CONGENITAL HEART DISEASE

AND

CARDIAC SURGERY:

PSYCHOLOGICAL IMPLICATIONS

FOR

CHILD AND FAMILY

BY

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ABSTRACT

In chronic physical illness in childhood, there is now an extensive literature on the role of psychological factors in determining the impact of illness on the child and family and their subsequent adjustment. Despite this, the psychological implications of congenital heart disease and cardiac surgery in particular have received relatively little attention. Some previous studies in this field have methodological problems, complicating interpretation of their findings. The present project, a prospective, controlled study, was designed to overcome some of the gaps in knowledge and deficiencies in methodology in the existing literature. A comprehensive battery of tests was administered to 75 children and their families before and 12 months after cardiac surgery. Two comparison groups of children and families were used - 75 children undergoing another form of stressful medical intervention (bone marrow transplantation) and 75 healthy children. Children were assessed on their developmental and cognitive functioning, academic attainments, behaviour at home and at school and their self perception. Parents completed measures assessing their own levels of psychological distress, marital relationship, locus of control and coping. Siblings' behaviour at home and at school was assessed. The results indicate that, in common with other chronic illnesses, children with congenital heart disease and their families are at increased risk of adjustment difficulties. Cardiac surgery is a potential stressor, but it does not have a uniform impact. Rather, its effects are mediated by a myriad of demographic, social, environmental and medical factors. Within a single disease category there may be more variability in psychological impact than there is between different disease categories (for example, cyanotic versus acyanotic presentations), and the effects can change with time/treatment. The results

support a model of psychological adjustment with both disease-specific and non-categorical elements. Implications of the findings for further research and clinical practice are outlined.

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Abbreviations and Symbols

Throughout the text a number of abbreviations and statistical symbols have been used, which are listed below:

ABBREVIATION	FULL MEANING
BMT	bone marrow transplantation
VSD	ventricular septal defect
ASD	atrial septal defect
PDA	patent ductus arteriosus
TOF	tetralogy of Fallot
TGA	transposition of the great arteries
ECG	electrocardiogram
ICU	intensive care unit
ALL	acute lymphoblastic leukaemia
AML	acute myeloid leukaemia
CGL	chronic granulocytic leukaemia
HLA	human leucocyte antigen
GVHD	graft versus host disease
IQ	intelligence quotient
CNS	central nervous system
CHD	congenital heart disease
CY	cyanotic
ACY	acyanotic
SR	spontaneous recovery
PH	physical handicap
SEIQoL	schedule for the evaluation of individual quality of life
pts	patients
CF	cystic fibrosis
BAS	British Ability Scales
BCL	behaviour checklist
GHQ	General Health Questionnaire
DAS	Dyadic Adjustment Scale
UCL	Utrecht Coping List
S.D.	standard deviation
SCBU	special care baby unit
ANOVA	analysis of variance
DQ	developmental quotient
CBCL	Child Behaviour Checklist
NYHA	New York Health Association

STATISTICAL SYMBOL	PROBABILITY VALUE
**	$p<.05$
*	$p<.01$
#	$p<.005$
~	$p<.001$

INTRODUCTION

Advances in medical treatment have resulted in significant changes in the pattern of childhood diseases. Some, such as tuberculosis and diphtheria, have been virtually eradicated whilst other previously fatal disorders, such as cystic fibrosis and leukaemia, have now taken on the status of chronic illnesses. It has been estimated that 10-18% of children suffer from chronic diseases (Pless & Douglas, 1971; Hobbs & Perrin, 1985; Cadman, Boyle, Szatmari & Offord, 1987), of which the most common is asthma. Duration of survival and quality of life for chronically ill children have improved dramatically and there is now a better prognosis for an ultimate disease free survival. However, the nature of the treatment regimens can also be intrusive and create long term psychological problems for these children and their families. With the prolongation of life in chronically sick children has come a resultant increase in psychological maladaption. It is now recognised that the care of such children is no longer a purely medical issue (Eiser, 1985) and that investigation and appropriate management are necessary of the potentially negative effects of intrusive treatment regimens on the child's cognitive, emotional and behavioural development, together with the effects on family adjustment.

The heart, more than any other organ in the body, has a deep-rooted symbolic significance and has many implications in a physical and an emotional sense (Toker, 1971; Bentovim, 1979). It has been suggested that diseases of the heart, with their chronicity and age-long association of death, are likely to have a greater impact on children and their families than other illnesses (Maxwell and Gane, 1962). Whilst some complex cardiac lesions still remain incurable, the advances in surgical and medical techniques have resulted in successful correction of many previously fatal congenital heart disorders. Paediatric open heart

surgery for palliation and correction of congenital lesions has been performed since the development of the pump oxygenator in the 1950's, and there has been a resultant increasing interest in the psychosocial implications of congenital heart disease itself, and subsequently in the impact of open heart surgery on psychological functioning. In the last decade, paediatric heart transplant programmes have been established, offering further treatment possibilities for the child with complex congenital heart disease. The relatively small number of operations performed means that there are very few data on the psychological aspects of transplantation. Paradoxically, whilst the 1970's and early 1980's saw a wealth of studies investigating congenital heart disease and the impact of open heart surgery on the paediatric patient and family, there has been very little more recent research on this topic (Wray & Yacoub, 1991). Many of the published studies are retrospective and cross-sectional. Furthermore, the majority have focused on a few factors in isolation, such as the impact of congenital heart disease on cognitive development or the relationship between adjustment in the child and maternal anxiety, rather than looking at the influence of variables within an integrated, multivariate framework. Despite some of these shortcomings of the existing literature, a substantial number of significant findings have emerged from the research that has been undertaken.

The current study was therefore conducted to investigate the psychosocial aspects of congenital heart disease and cardiac surgery for children and their families and to attempt to remedy some of the deficits in the existing literature. The assessment of cardiac surgery patients and their families was carried out in parallel with an evaluation of the impact of bone marrow transplantation (BMT) on children, enabling comparisons to be made between the effects of different chronic illnesses and subsequent treatment on psychological adjustment.

CHAPTER 1

BACKGROUND TO CONGENITAL HEART DISEASE, CARDIAC SURGERY AND BONE MARROW TRANSPLANTATION

1.1. CONGENITAL HEART DISEASE

1.1.1. Introduction

With the advances currently being made in medical technology there is now the possibility of a healthy future for the child born with a cardiac malformation (Glaser, Harrison & Lynn, 1964; O'Dougherty, Wright, Garmezy, Loewenson & Torres, 1983). Advances in the diagnosis and management of congenital heart defects have significantly increased survival rates of affected children. The survival rate amongst children undergoing open heart surgery is increasing and more complex disorders are becoming operable. Advances in diagnostic techniques and medical and surgical management have improved the prognosis of many cardiac disorders. By 1980 most patients with a cardiac malformation could expect to benefit from surgery (Taussig, 1982). The rate of development and perfection of new surgical techniques means that even for children who are at present deemed "inoperable", there is some hope for a new life-saving treatment in the near future. However, associated with this progress are certain ethical problems, such as the quality of treatment results and the relevance of financial and social burdens (Jonsen, 1987).

Information will be presented on the medical aspects of firstly congenital heart disease and cardiac surgery and secondly on bone marrow transplantation. The focus will be on those medical factors which could be influential in determining the psychological impact of the disease and its treatment.

1.1.2. Incidence and Aetiology of Congenital Heart Disease

Congenital heart disease has been defined as "...a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" (Mitchell, Korones & Berendes, 1971). Congenital heart disorders are a heterogeneous group of abnormalities and affect the heart and associated vessels. They may take an "almost infinite variety of forms" (Thomson, 1976). Congenital heart disease is the most common single group of congenital abnormalities, accounting for about 30% of the total (Jordan & Scott, 1989). The incidence of congenital heart disease has been cited as varying between 0.3% and 1% of all live births (Mitchell et al, 1971; Laursen, 1980; Dickinson, Arnold & Wilkinson, 1981; Nadas, 1984; Ferencz, Rubin, McCarter, Brenner, Neill, Perry, Hepner & Downing, 1985; Hoffman, 1990; Haworth & Bull, 1993). In approximately 1-2% of all patients with organic heart disease the condition is congenital (Thomson, 1976). The diversity of abnormalities is such that the defects may be so minor and inconsequential as to be only detectable in routine examinations as an abnormal murmur, causing no problems. In contrast, some defects are so major as to be incompatible with life and the child dies shortly after birth.

It has been estimated that in the first year of life three children per thousand live births (0.3%) become symptomatic because of heart disease and are hospitalised (Fyler, 1980), although there are no data regarding the remaining children born with a congenital heart defect. About 25% of affected children will die in the first year of life, the majority within the first month. Ten to 15% of children with congenital heart defects have more than one cardiac abnormality; 10-27% also have one or more associated non-cardiac congenital abnormalities

(Mitchell et al, 1971; Greenwood, Rosenthal, Parisi, Fyler & Nadas, 1975; Rowe, Freedom, Mehrizi & Bloom, 1981; Fyler, 1985).

1.1.3. Basic Physiology of the Heart

The heart is a pump consisting of four chambers - two atria and two ventricles. Blood returns from the body and passes into the right atrium, which acts as a reservoir to fill the right ventricle via a flap valve (the tricuspid valve). When the right ventricle contracts, the valve closes and the pressure in the ventricle rises above that in the pulmonary artery to the lungs. (The term "pulmonary" is the adjective to describe the lungs). The pulmonary valve opens and blood is pumped into the lungs, where it takes up oxygen. The blood then returns from the lungs in the pulmonary veins and enters the left atrium. This acts as a reservoir to fill the left ventricle through the mitral valve. When the ventricle contracts, the pressure rises, the aortic valve opens between the left ventricle and the aorta and blood is pumped into the aorta and then to the body in the systemic arterial circulation. The saturation of the blood refers to the amount of oxygen in the blood. Desaturated blood contains lower amounts of oxygen. When there is desaturated blood in the capillaries cyanosis, or a blue colouration of the skin and lips, results.

1.1.4. Classification of Congenital Heart Defects

Congenital heart disease can be broadly subdivided into two groups, based on changes in the circulation:

1. Acyanotic Defects - due to either a left to right shunt or to an obstructive lesion. There is no mixing of desaturated blood in the systemic arterial circulation.

2. Cyanotic Defects - with either increased or diminished pulmonary flow. Desaturated blood enters the systemic arterial circulation, regardless of whether cyanosis is clinically evident. Unsaturated venous blood by-passing the lungs can result in secondary polycythaemia (an increase in the number of red blood cells), which is a compensatory mechanism to carry more oxygen to the tissues. This causes increased viscosity, which in turn results in sluggish blood circulation and impeded blood flow, particularly in the capillaries. Poor peripheral blood flow and clubbing of the fingers and toes can result. Breathlessness and fatigue often result in a reduced exercise tolerance, and growth may be inhibited.

1.1.5. Complications of Congenital Heart Disease

When congenital heart disease causes reduced blood flow and low arterial blood oxygenation levels, complications may be encountered in any organ system. Conversely, cardiac lesions resulting in increased pulmonary blood flow and raised pulmonary arterial pressure may, over the course of time, produce a permanent vascular change that will itself limit life. Cyanosis causes an increased incidence of brain abscess and hypoxic spells produce a higher incidence of brain damage. Thrombosis and embolism are common complications of cyanotic congenital heart disease and can affect any part of the systemic circulation, although involvement of the cerebral circulation is the most common (Jordan & Scott, 1989). Abnormalities of the inner surface of the heart also lead to an increase in the incidence of bacterial endocarditis. Such complications can be ameliorated through early corrective surgery, although the exact effects of the timing of the surgical intervention and of the particular surgical procedure are far from clear.

There are eight congenital heart lesions which together make up approximately 80% of all cases (Table 1.1). The remaining 20% are comprised of numerous rarer, more complex anomalies (Jordan & Scott, 1989).

THE EIGHT MOST COMMON CONGENITAL HEART LESIONS AND APPROXIMATE INCIDENCE FIGURES	
ACYANOTIC	INCIDENCE
Ventricular septal defect (VSD)	33%
Atrial septal defect (ASD)	9%
Pulmonary stenosis	8%
Patent ductus arteriosus (PDA)	7%
Aortic stenosis	6%
Coarctation of the aorta	6%
CYANOTIC	
Tetralogy of Fallot (TOF)	5%
Transposition of the Great Arteries (TGA)	4%

Table 1.1

The aetiology of these defects is unknown, as with most congenital defects, but both genetic and environmental factors have been identified. Family studies suggest that the mode of inheritance is polygenic, although occasionally single gene mutations occur. The birth of a child with congenital heart disease triples the chances of a subsequent child being affected and there is a high degree of concordance - i.e. two affected siblings usually have the same lesion or one of its components. Family studies to determine empirical risks to first degree relatives (siblings and offspring) of patients with isolated congenital heart disease give risk

estimates of between 1% and 5% (Dennis, 1983). Ten to 25% of parents of children with a congenital heart defect report congenital heart disease among relatives (Boon, Farmer & Roberts, 1972; Rowe et al, 1981). The model of a genetic background predisposing to congenital heart disease is generally accepted (Campbell, 1959; Neill, 1972; Fyler, 1985) and the minimal variation in incidence of the different types of congenital heart lesions suggests unchanging factors that predispose to the development of congenital heart disease. The overall sex-ratio of cardiovascular malformations is of the order of 1:1 (Polani & Campbell, 1955) although the incidence by gender of specific lesions varies. Children with chromosomal abnormalities have a greatly increased incidence of congenital heart defects - almost 50% of all children born with Down's syndrome have a cardiac lesion. Environmental factors which are associated with an increased incidence of congenital heart defects include maternal rubella in the first three months of pregnancy, maternal diabetes (Rowland, Hubbell & Nadas, 1973) systemic lupus erythematosus (Scott, Maddison, Taylor, Esscher, Scott & Skinner, 1983) and drugs such as lithium, phenytoin and alcohol in pregnancy (Jordan & Scott, 1989). Premature birth is also associated with persistent patency of the ductus arteriosus. However, the majority of patients and parents provide no indication of an aetiologic factor for the child's congenital heart defect and at present the aetiology of congenital heart disease appears to be multi-factorial, with several largely uncertain factors simultaneously or consecutively involved.

1.1.6. Age: Diagnosis and Presentation

1.1.6.1. Age at Diagnosis

The variation in the nature and presentation of symptoms means that the relative frequency of the many diagnoses varies with age. In general, for the majority of children born with a cardiac malformation some, albeit tentative, diagnosis is made in infancy. Presentation of congenital heart disease usually occurs as a result of heart failure, the presence of cyanosis or through detection of a heart murmur during a routine examination in the neonatal period or during the first year. The remainder tend to be diagnosed as a result of school medicals or after some form of illness.

1.1.6.2. Age at Onset of Symptoms

Evidence indicates that most forms of congenital heart disease are compatible with survival of the unborn baby and surprising degrees of anatomic abnormality are tolerated before birth. At birth the circulation undergoes changes which enable separation of the pulmonary circulation from the systemic circulation. Normally each is pumped in equal volume by separate ventricles, but an abnormally small ventricle may not be able to manage. Depending on the degree of anatomic abnormality, the infant may have great difficulty in the first hours, days or weeks of life and the major life-threatening congenital cardiac defects cause death very early, often within a few hours of the manifestation of obvious symptoms. Generally, congenital lesions that obstruct the flow of blood from the heart are symptomatic at birth. The pathognomonic murmur of a patent ductus arteriosus or ventricular septal defect may not be present until a few days or weeks after birth and the murmur of an atrial septal defect is frequently not heard until the child reaches school-age. In children with Tetralogy of Fallot, the time of onset of cyanosis roughly correlates with the severity of the lesion

(Vanden Belt, Ronan & Bedynek, 1979). Early recognition and transfer of affected babies to specialist units has been recognised as vital for improved rates of survival. As attention is now being directed to newborn cardiac problems, the age of new patients with heart disease has been changing (Fyler, 1985). Of those children who will require hospitalisation, approximately 35% are admitted within the first year of life.

1.1.7. Survival Rates

The description of the natural history of many congenital heart defects is incomplete (Adams, 1977), resulting in incomplete survival statistics. The majority of studies have been initiated to assess the success of new surgical procedures and interest has focused on the pre- and post-surgical course of each cardiac lesion, rather than on the natural course.

1.1.8. Physiological Correlates of Congenital Heart Disease

In addition to the cardiac defect per se, other cardiac related effects may be present, including growth retardation (Ehlers, 1978; Levy, Rosenthal, Fyler & Nadas, 1978), chronic hypoxia (Silbert, Wolff, Mayer, Rosenthal & Nadas, 1969; Aisenberg, Rosenthal, Wolff & Nadas, 1977), and congestive heart failure (Jordan & Scott, 1989). As a result of his or her cardiac condition the child may also be more vulnerable to neurological complications, which can be life-threatening, further compromising the child's development (Tyler & Clark, 1957; Phornphutkul, Rosenthal, Nadas & Berenberg, 1973; Fishman & Parke, 1990).

1.1.9. Developmental and Neurological Factors

Developmental delay and failure to thrive are common in infants and children with congenital heart disease (Linde, Dunn, Schireson & Rasof, 1967; Linde, Rasof & Dunn, 1970;

Rosenthal & Castaneda, 1975; Poskitt, 1993) although growth retardation is not an inevitable accompaniment of congenital heart disease (Adams, Lund & Disenhouse, 1954). Poor growth has been cited as the most prominent complication of congenital heart disease, with 26% of 5 year old children who had survived symptomatic heart disease in infancy being found to be below the 5th percentile for height (Fyler, 1982). Growth retardation, more for weight than for height, has been found in all forms of congenital heart disease (Linde, 1975) but is more severe in children for whom heart disease is part of a chromosomal syndrome. It is usually more marked in males and in those children with a greater degree of cyanosis and physical incapacity (Campbell & Reynolds, 1949; Linde, Dunn, Schireson & Rasof, 1967), although no linear relationship has been found between these factors and delayed growth (Poskitt, 1993). The causes of growth retardation in children with congenital heart disease seem to be multiple and are poorly understood. Hypoxia (Naeye, 1967), malnutrition (Naeye, 1965(a); Gervasio & Buchanan, 1985; Thommessen, Heiberg & Kase, 1992; Unger, DeKleermaeker, Gidding & Christoffel, 1992), intrauterine growth factors (Naeye, 1965(b)), extra-cardiac anomalies, endocrine disorders (Linde, Dunn, Schireson & Rasof, 1967) and psychological factors (Adams et al, 1954; Krieger, 1970) have all been implicated in the mechanism of delayed growth and development.

The severity of growth inhibition is dependent on the type of cardiac lesion and its haemodynamic effects. Skeletal retardation is also associated with a retardation in height and weight but is not necessarily parallel to it (Bayer & Robinson, 1969). Whilst the presence of growth retardation in children with congenital heart disease is widely acknowledged, the effects of cardiac surgery on growth are far less clear, although weight gain is more evident than an increase in height after surgery (Bayer, 1976). For infants submitted to early

corrective surgery, good outcome with respect to catch-up growth has been reported (Sholler & Celermajer, 1986). Much conflicting and confusing data have been presented in the literature, due mainly to the lack of uniform criteria for defining growth failure and reporting results. Few studies have looked at parental/sibling growth or give accurate comparisons of pre- and post-operative growth patterns. The variability in growth in response to surgery has been suggested to be related to one or more of the following factors: genetic constitution, residual lesions, complications of surgery, associated cardiac and extra-cardiac anomalies, age at operation, chromosomal abnormality and intra-uterine growth retardation (Robinson & Bayer, 1969; Gidding & Rosenthal, 1984).

1.1.10. Treatment

The treatment of congenital heart disease is surgical (Nadas, 1984) and there is now emphatic agreement in the literature that early surgery should always be performed, with the majority of defects being corrected or at least palliated in the first year of life. Timing of the surgery depends on multiple factors, including the patient's size, complexity of the lesion and any secondary defects and psychosocial considerations, which include both patient and family factors (Graham, 1984). It is sometimes necessary for palliative surgery to be performed in infancy before corrective surgery is performed at a later age when the child is in a more favourable physical condition. One of the factors which needs to be considered is the effect of chronic hypoxemia on the brain and the consequences for future development, with earlier relief of hypoxemia reducing the risks of brain impairment.

Surgical correction of congenital heart defects may be closed heart (extracardiac) or open heart (intracardiac) procedures. Open heart surgery involves stopping the heart and

maintaining blood supply to the body with a heart-lung (cardiopulmonary bypass) machine, whereas for closed heart procedures the heart's action is not stopped. In general, closed heart procedures are considered to carry an operation risk of between 1% and 3% (Binet, 1985). The overall mortality rate due to open heart surgery is about 15%, with a greater risk of mortality in correction of cyanotic (30%) than acyanotic (3%) disorders (Hoffman, 1987). Whilst surgical repair is not possible for all children, the development of heart and heart-lung transplantation offers treatment for some previously inoperable defects - particularly in children with pulmonary hypertension secondary to a cardiac lesion.

1.1.11. Treatment Measures: Cardiopulmonary Bypass or Profound Hypothermia and Circulatory Arrest

With the refinement of surgical techniques, improvements have been achieved in mortality and morbidity and the correction of more complex congenital heart defects. Medical attention has now switched from performing reparative surgery using extracorporeal circulation (in which the circulation is maintained outside the body) to the debate of whether cardiopulmonary bypass or profound hypothermia and circulatory arrest produce the more positive results, and evidence has been cited in support of each approach.

Literature in the early 1970's focused on the possible neurological effects following cardiopulmonary bypass, mostly in adults. Work with children has been much more limited but demonstrated, in general, that intellectual functioning was not impaired by the procedure (Whitman, Drotar, Lambert, VanHeeckeren, Borkat, Ankeney & Liebman, 1973). With the development of the technique of profound hypothermia and circulatory arrest, attention is now being given to the merits of this procedure.

There is general agreement that profound hypothermia and circulatory arrest, in which oxygen consumption is drastically reduced at a temperature of 15°C, provides optimal conditions for surgery on neonates and infants (Subramanian, Vlad, Fischer & Cohen, 1976; Haka Ikse, Blackwood & Steward, 1978) and post-operative complications appear to be no more frequent than when bypass procedures are performed on children of the same age (Steward, Sloan & Johnston, 1974) - rather, post-operative problems may be less frequent in children who have hypothermic techniques as opposed to standard cardiopulmonary bypass (Gill & Page-Goertz, 1986). Furthermore, improved rates of survival and reduced morbidity have been quoted for infants undergoing surgery under hypothermic rather than conventional cardiopulmonary bypass conditions (Barratt-Boyes, Simpson & Neutze, 1971; Wagner & Subramanian, 1978). However, there is histopathological research indicating that there may be short- and long-term neurological sequelae after surgery under profound hypothermia (Egerton, Egerton & Kay, 1963; Brunberg, Reilly & Doty, 1974; Fisk, Wright, Hicks, Anderson, Turner, Baker, Lawrence, Stacey, Lawrie, Kalnins & Rose, 1976). Comparative studies of cardiopulmonary bypass and profound hypothermia and circulatory arrest have found a higher incidence of developmental abnormality in the group treated with deep hypothermic arrest (Wright, Hicks & Newman, 1979) and significantly lower IQ scores in the arrest group (Wells, Coghill, Caplan & Lincoln, 1983). In contrast, favourable results on the long-term effects of circulatory arrest on intellect and development have also been quoted (Stevenson, Stone, Dillard & Morgan, 1974; Messmer, Schallberger, Gattiker & Senning, 1976; Subramanian et al, 1976; Haka Ikse et al, 1978; Dickinson & Sambrooks, 1979; Clarkson, MacArthur, Barratt-Boyes, Whitlock & Neutze, 1980), although these studies tend to be retrospective. In one of the few prospective studies, no harmful effects on

development in a group of infants and young children were found after using circulatory arrest and profound hypothermia (Blackwood, Haka-Ikse & Steward, 1986). The literature seems to indicate, therefore, that for neonates and infants with complex intracardiac congenital heart defects the risks of hypothermia and circulatory arrest are far outweighed by the improved prognosis facilitated by the conditions under which surgery is performed. What is less clear is the use of deep hypothermia and circulatory arrest as opposed to cardiopulmonary bypass for older/bigger children.

1.1.12. Hospitalisation

1.1.12.1. Catheterisation

A child diagnosed as having a heart malformation by non-invasive techniques, such as echocardiography, X-ray and electrocardiogram, is usually admitted to hospital to undergo a cardiac catheterisation for further evaluation. Cardiac catheterisation is an invasive diagnostic procedure involving the introduction of a flexible catheter through an artery into the left or right heart, normally via needle puncture in the inguinal region. This measures intra-cardiac pressures and oxygen saturation and can detect defects in the atrial or ventricular septum, valves, coronary arteries or great vessels (Agamalian, 1986). Cardiac catheterisation also allows angiography - the injection of radio-opaque contrast medium into the heart - to be performed. The passage of the dye through the heart can then be monitored. The age at which cardiac catheterisation is performed on children depends on the type and severity of the cardiac defect. The use of general anaesthesia or sedation varies between centres, but infants and young children are normally given a general anaesthetic for the procedure, which lasts for 1-4 hours, whilst older children are more likely to remain conscious but sedated. The risks of the procedure are usually low (Gersony & Bierman,

1981) but major complications resulting from catheterisation have been reported to occur in approximately 3% of cases (Stanger, Heymann, Tarnoff, Hoffman & Rudolph, 1974). These include arrhythmias, atrioventricular blocks, thrombosis, anaphylactic reaction to the dye, anaesthesia reactions and, in extreme situations, death (Uzark, 1978: Finesilver, 1980). Minor complications - e.g. bleeding, loss of femoral pulses, drop in body temperature - are more frequent, occurring in about 12% of cases. Both morbidity and mortality associated with cardiac catheterisation have been greatly reduced when particular attention has been paid to the prevention of metabolic disorders and pulmonary difficulties, and small amounts of contrast agents and flexible catheters have been used (Ross & Peterson, 1977). The risks of the procedure are affected by the child's physical and emotional state (Uzark, 1978) and there is speculation that arrhythmias due to the manipulation of the catheter into the heart chambers in particular may be increased by the child's fear. An instance of death during catheterisation due to the patient's extreme distress has been reported (Engel, 1971). Following catheterisation patients normally have to endure a period of bed rest (usually up to 24 hours) to facilitate healing of the cannulated vessel.

1.1.12.2. Surgery

Prior to going down to theatre most children receive some form of premedication on the ward, often in the form of an intra- muscular injection. Once in the anaesthetic room anaesthesia is induced and the child is taken into the operating theatre. The surgical procedure usually lasts 2 or more hours, with the average time being 3-6 hours for bypass procedures and 2-4 hours for other operations. The incision is either a sternotomy (for open heart procedures) or a thoracotomy (for closed heart procedures) (see page 43). On returning to the intensive care unit (ICU) most children are sedated and ventilated, with

chest drains, pacemaker wires, urinary catheter, venous and arterial lines, naso-gastric tube and cardiac monitoring leads. Blood products are normally given to replace those lost during surgery and through chest drainage post-operatively, and drugs and fluids are administered intravenously or naso-gastrically. Patients are continuously monitored and nursed on a one to one basis until drains and lines are removed. For many children their stay in ICU may only last for 24-48 hours but the rate of progress varies, depending on the severity of the heart defect and the type of surgery performed. Some will remain in ICU for weeks, and sometimes months, and recovery can be very slow and traumatic. The risks of surgery are related to the condition itself and the ability of the surgeon to correct the lesion (Jordan & Scott, 1989). The overall mortality from open heart surgery remains significant because more complicated operations are now attempted at an earlier age and not all are corrective, although overall mortality rates are now less than 10% in most major units (Lansing, Girardet & Masri, 1984). In some cases it is necessary for the patient to return to theatre or to undergo further procedures on the ward. The most common complications are bleeding, arrhythmias, cardiogenic shock, wound breakdown, infection, paralysed diaphragm and pneumothorax. Acute kidney failure is sometimes a complication of cardiac surgery and peritoneal dialysis or haemofiltration can be required in the short term. Temporarily abnormal electroencephalograms (EEG's) in the early post-operative period have been reported for 39% of children (Finley, Calanchini & Riggs, 1976) but in the majority of cases the EEG returns to normal. However, a small number of patients suffer significant neurological damage (Ferry, 1987; 1990; Fallon, Aparicio, Elliot & Kirkham, 1995). This is more common in children with complex lesions and in those who have a traumatic post-operative recovery. A number of factors may be involved - including anaesthesia induction, difficulties with the bypass procedure and prolonged low cardiac output post-operatively.

Seizures may also occur, but frequently these are due to a biochemical imbalance and do not cause permanent damage. Long term seizures associated with cardiac surgery are rare (Ehyai, Fenichel & Bender, 1984).

Post-operatively the child will undergo physiotherapy to prevent chest infections and to help with mobilisation. The child will gradually be oriented back to normal activities. Most children are discharged home after a further stay of 5-7 days on the general ward. The majority are discharged with some oral medication to continue taking at home.

1.1.12.3. After Discharge

Children are normally seen in the outpatient clinic 6 weeks after surgery and if they are well are usually allowed back to school after that. Further out-patient follow-up depends on the individual child and the nature of his/her cardiac condition. Participation in normal school and extra-curricular activities is encouraged, but again varies depending on the individual's physical state.

1.1.13. Long-Term Impact

Although many patients with corrected lesions are functionally normal, there is a large incidence of residua, sequelae and coexisting lesions that can become important in time, such that clinical review should be maintained through adulthood (Maron, Rosing, Goldstein & Epstein, 1977; Garson, Nihill, McNamara & Cooley, 1979; Rowland, 1979; McNamara & Latson, 1982). It has been suggested that the only cardiac operation which should be regarded as completely curative is ligation of a patent ductus arteriosus (Somerville, 1983). The majority of children with heart disease require antibiotic prophylaxis for all dental

procedures for life to prevent bacterial endocarditis (Dube, 1990). Other long term consequences of congenital heart disease include difficulty obtaining life insurance, health insurance or insurance cover for mortgages (Truesdell, Skorton & Lauer, 1986; Shook, 1990; Mahoney & Skorton, 1991; Hart & Garson, 1993). For some patients with a long standing reduction in cardiac efficiency, heavy manual work may be impossible. Increasingly, discrimination is being reported against young adults with congenital heart disease (corrected or uncorrected) in the job market (Manning, 1981; Hamburgen, 1991). Risks to the children of patients with congenital heart disease are difficult to assess accurately, but a figure of a 3-10% chance of a patient having a child with a heart defect has been quoted, depending on the parental lesion (Czeizel, Pornoi, Peterffy & Tarcal, 1982; Emanuel, Somerville, Inns & Withers, 1983; Rose, Gold, Lindsay & Allen, 1985). For adults who have a syndrome and associated heart disease, the risks to their offspring are significantly greater (Nora & Wolf, 1976). The risks to the offspring are slightly higher if the mother, rather than the father, has the heart problem. Pregnancy is not contraindicated in patients with congenital heart disease unless they are particularly cyanosed, when the incidence of miscarriage is higher, or if they have significant pulmonary hypertension. However, women with congenital heart disease are at increased obstetric risk, regardless of the severity of their underlying defect (Gantt, 1992).

1.1.14. Drugs and Their Side Effects

Whilst many children will require drugs only for a short time, others will be on medication for life. Potential problems of long term drug use include electrolyte imbalance due to long term use of diuretics, nausea and vomiting - which can be side effects of anti-failure drugs - and allergic reactions to antibiotics.

1.2. BONE MARROW TRANSPLANTATION

1.2.1. Pathology

In contrast to cardiac surgery, which can take many forms, bone marrow transplantation (BMT) is a standard procedure involving the donation of bone marrow through an intravenous line in a manner similar to a blood transfusion. Within the bone marrow there are stem cells, which have the potential to develop into red or white blood cells. Any stem cell defect, in which the stem cell is either absent (as in aplastic anaemia), abnormally proliferating (as in acute leukaemia) or defective (as in severe combined immune deficiency and inborn errors of metabolism) is correctable by BMT through transplanting healthy cells. In practice, however, the life threatening nature of the procedure has meant that only conditions which would otherwise be fatal are at present considered suitable for BMT (Barrett & Gordon Smith, 1983).

The nature of paediatric conditions treated by BMT can be subdivided into acquired and genetic disorders (Table 1.2).

The timing of the BMT depends on the nature of the underlying disease and to a certain extent on the availability of a suitable donor. Children with an inherited enzyme deficiency, resulting in mental retardation, need to undergo transplantation before irreversible neurological damage occurs (Hugh-Jones, 1983). Similarly, patients with lysosomal storage diseases, such as Niemann-Pick, who also have early and severe liver disease die of hepatic impairment within a very short time and therefore require early BMT (Vellodi, Hobbs, O'Donnell, Coulter & Hugh-Jones, 1987). Patients with severe aplastic anaemia should undergo transplantation as soon as a suitable donor is found because transfusion of blood

products increases the chance of graft failure. Children with leukaemia are transplanted when in first (for myeloid leukaemias) or second (for lymphoblastic leukaemias) remission.

DISORDERS TREATED WITH BONE MARROW TRANSPLANTATION	
ACQUIRED DISORDERS	
Leukaemia: Acute lymphoblastic leukaemia (ALL)	
Acute myeloid leukaemia (AML)	
Chronic granulocytic leukaemia (CGL)	
Severe aplastic anaemia	
Lymphoma and other malignant tumours	
GENETIC DISORDERS	
Immunodeficiency disorders: Severe combined immune deficiency	
Wiskott Aldrich syndrome	
Metabolic disorders:	Hereditary anaemias
Fanconi's anaemia	
Thalassaemia B major	
Lysosomal storage diseases	
Mucopolysaccharidoses e.g. Hurler's, Hunter's, San Filippo syndrome	
Spingolipidoses e.g. Gaucher's disease	

Table 1.2

Donor selection involves several stages. Only one in four children will have a matched family donor. In the absence of a suitable family donor an unrelated HLA-matched donor is used whenever possible. Failing this either a partially matched family donor is used (this

last option has a lower chance of survival (Ash, Casper, Serwint, Coffey, Bruckman, Truitt, Greenwood, Geil, Romond, Camitta, McDonald, Thompson & Maruyama, 1987)) or BMT treatment is discontinued. With some specific malignancies autologous BMT can be used as an alternative (Buckner, Appelbaum, Clift, Doney, Sanders, Hill & Thomas, 1987; Phillips & Connors, 1987), in which the patient receives their own previously harvested marrow and no donor is involved.

BMT involves 3 phases - the pre-transplant conditioning regimen, the actual transplant and engraftment.

1.2.2. Conditioning Regimen

The preparation of the child prior to receiving the marrow differs substantially from the pre-operative preparation of the child undergoing cardiac surgery. The BMT patient is normally admitted 2 weeks before the day of the transplant to enable insertion of a Hickman catheter and the administration of the conditioning regimen, which is carried out to prepare the child for the marrow infusion. This involves treatment with chemotherapy and, for children with leukaemia or other malignancies, total body irradiation. The purpose of this regimen is to ablate the child's marrow cells and to immunosuppress the child in order to reduce the risk of graft rejection.

1.2.3. Marrow Donation

The donor is usually admitted to hospital the day before the marrow is due to be harvested. Marrow harvest takes place under general anaesthesia and usually takes between one and a half and two hours. The marrow is normally aspirated from multiple sites along the iliac

crests. The donor can normally leave hospital the following day, with no ill effects other than some soreness at the site of marrow collection. There are potential risks of the anaesthesia and of infection but complications are rare.

Following marrow harvest from the donor, the marrow is infused into the recipient in the second phase of the procedure.

1.2.4. Engraftment

Engraftment, in which the new marrow develops the capacity to produce mature blood cells, is normally seen during the second week after transplantation and haematologic reconstitution is usually achieved within 4 to 6 weeks after marrow infusion.

1.2.5. Nursing Care

Once the immunosuppression has begun, the child is nursed in conditions of strict reverse barrier nursing in order to prevent infection. This period of isolation lasts until the leukocyte count demonstrates moderate regeneration - usually between 28 and 42 days after transplantation (Barrett & Gordon Smith, 1983). During that time the child is restricted to the room but parents can visit provided they adhere to the special precautions. Such isolation procedures have psychological implications for both child and family (see page 92). Isolation procedures vary from centre to centre, but there is some evidence of a lower incidence of infection in patients treated in laminar air flow units (Storb, Prentice, Buckner, Clift, Appelbaum, Deeg, Doney, Hansen, Mason, Sanders, Singer, Sullivan, Witherspoon & Thomas, 1983). The intrusive daily treatment involves administration of blood products and medications, blood tests and decontamination of the mouth, skin and bowel by antiseptic and

antibiotic treatment. There are also side effects of the initial drug regimen, including nausea and vomiting, hair loss, skin breakdown and mucositis (deterioration of the mucous membranes in the mouth and gastrointestinal tract). There may also be effects associated with total body irradiation. Food restrictions are also necessary to reduce the risks of infection. Where oral intake is not possible due to gastrointestinal problems, the child's nutritional state is maintained by the use of total parenteral nutrition, where the necessary calories are given intravenously. Improvements in overall survival rates and disease free survival have been demonstrated in patients receiving total parenteral nutrition (Weisdorf, Lysne, Haake, Goldman, McGlave, Ramsay & Kersey, 1987).

1.2.6. Discharge

Discharge is normally after 3 to 4 months hospitalisation. During the first six months after transplantation regular hospital checkups are required, together with strict adherence to a drug regimen. Patients are not allowed to return to school or attend crowded public places during this time. It can take up to one year for full immunological reconstitution to occur. The major problems encountered during this period include chronic graft versus host disease (GVHD), graft rejection, infections and, in the case of leukaemia, a recurrence of the original disease. GVHD is caused by a reaction of the graft cells against the patient's own cells and is characterised by pyrexia, skin rash, jaundice and diarrhoea. It is the most significant cause of mortality and morbidity in patients treated with allogeneic BMT (Vega, Franco, Abdel-Mageed & Ragab, 1987). The acute form of GVHD starts within a few days of engraftment and primarily involves the skin, liver, bowel and lymphohaematologic system. Approximately 65% of paediatric patients suffer from acute GVHD and almost one third from chronic GVHD (Zwaan & Hermans, 1983; Kadota & Smithson, 1984).

The immunosuppressive regimen prior to BMT and the prolonged period of immunodeficiency afterwards make patients vulnerable to infections which can be fatal, particularly interstitial pneumonia (Weiner, 1987). Rejection of the graft - leading to a loss of the new bone marrow and a recurrence of the original disease - is also a problem, particularly in treatment of the leukaemias. Modification of the conditioning regimen to include higher doses of total body irradiation and additional chemotherapeutic drugs is being implemented to reduce the rate of relapse (Storb, 1984).

1.2.7. Long Term Implications

Children who have survived a BMT are at high risk for developing long term complications that can result in significant morbidity and mortality (van der Wal, Nims & Davies, 1988). The main problems encountered during the few years after transplantation are infections and recurrence of the original disease. The immune system remains incompetent for at least 2 years after the transplant, especially in patients with chronic GVHD. The risk of leukaemic relapse remains for 2-3 years (Barrett & Gordon-Smith, 1983). Secondary malignancies, infertility and gonadal dysfunction, cataracts, and growth and thyroid hormone deficiencies are other late effects linked to the preparatory regimen (Bender-Gotze, 1991; Sanders, 1991; Uderzo, Locasciulli, Rovelli, Rossi, Jankovic, Adamoli, Bonomi, Balduzzi, Biondi, Schiro, Rizzari, Valsecchi, Conter & Masera, 1992; Robertson, 1993; Schmidt, Niland, Forman, Fonbuena, Dagis, Grant, Ferrell, Barr, Stallbaum, Chao & Blume, 1993).

1.2.8. Side Effects of Drugs

In addition to the side effects of the total body irradiation and chemotherapy, patients can also experience effects related to other specific immunosuppressants, such as cyclosporin.

Cyclosporin is normally administered a day or so before marrow infusion and is continued on a twice daily basis for up to a year after transplantation. It is toxic to systems other than the bone marrow, with impaired kidney function being the most clinically significant side effect. Other side effects which can be distressing for the patient include hair growth, tremor, overgrowth of the gums and loss of appetite.

1.2.9. Clinical Results of BMT

BMT offers the chance of a cure for a number of acquired and inherited disorders. However, the risks of the procedure are high and survival and cure depend very much on donor compatibility and on the disease status at the time of transplant. Patients with acute myeloid leukaemia in first remission have a 40%-60% chance of a long-term, disease free survival (Gale, 1987) but the chance of curing the disease in second remission or relapse is smaller. Patients with acute lymphoblastic leukaemia have a greater than 80% chance of achieving long-term disease free survival if treated in first remission (Barrett & Gordon Smith, 1983); this drops to about 30% if treated in second or subsequent remissions (Johnson, Thomas, Clark, Chard, Hartmann & Storb, 1981). Comparison of patients with autologous and allogeneic grafts indicates earlier and more frequent relapses in autologous cases (Kersey, Weisdorf, Nesbit, Woods, LeBien, McGlave, Kim, Filipovich, Vallera, Haake, Bostrom, Hurd, Krivit, Goldman & Ramsay, 1987). Patients with chronic myeloid leukaemia treated during the chronic phase of their disease have a 60% chance of survival at 4 years (Goldman & Apperley, 1987) but this drops to 15-30% if treated in a later phase of the disease. Survival rates for children with severe aplastic anaemia are about 80% (Storb, 1984; Champlin, 1987). For children with genetic disorders, survival rates range from 50% for some of the inherited anaemias (Deeg, Storb, Thomas, Appelbaum, Buckner, Clift, Doney,

Johnson, Sanders, Stewart, Sullivan & Witherspoon, 1983) to 80%-90% for children with metabolic disorders treated with marrow from a matched donor.

CHAPTER 2

PSYCHOLOGICAL FACTORS FOR THE CHRONICALLY ILL CHILD

2.1. DEVELOPMENTAL STATUS AND COGNITIVE DEVELOPMENT

2.1.1. Introduction

Assessment of cognitive development is widely used to evaluate the impact of genetic and environmental factors on normal development. Cognitive development is assessed using standardised measures based on age-related norms. Whilst it is recognised that some illnesses do damage the developing brain and central nervous system, the majority of chronic illnesses in childhood are not known to cause any physiological damage. Consequently, it might be expected that the presence of a chronic illness would not lead to any cognitive impairment and that such children would not differ from healthy children in intelligence quotient (IQ) scores or on academic parameters. In practice, however, the restricted life style of the chronically sick child is often not conducive to maximising intellectual and academic potential. Schooling is often disrupted (Eiser, 1980(b)), with higher than average absenteeism (Cairns, Klopovich, Hearne & Lansky, 1982; Lansky, Cairns & Zwartjes, 1983; Fowler, Johnson & Atkinson, 1985). The frequent, albeit short, absences from schooling that many of these children experience are known to be potentially more disruptive than a single, extended period of absence (Rutter, 1975). Girls are more likely than boys to have increased absence (Charlton, Larcombe, Meller, Morris Jones, Mott, Potton, Tranmer & Walker, 1991). There is a wealth of documented evidence concluding that chronically sick children have IQ scores within the normal range but that their academic achievements are significantly lower than expected from age-related norms (Rutter, Tizard & Whitmore, 1970; Olch, 1971; Pless & Pinkerton, 1975; Eiser, 1980(a); Gath, Smith & Baum, 1980; Mearig, 1985; Peckham, Meadows, Bartel & Marrero, 1988). Furthermore, children with mild

reading disability have also been found to be at risk of emotional difficulties (Casey, Levy, Brown & Brooks-Gunn, 1992).

Following the diagnosis of a chronic illness, schooling for young children is inevitably disrupted, with a resulting negative effect on academic attainments. This is not only relevant for the child's current academic performance but also has implications for the child's management of stress throughout his/her schooling. Work by Dweck and colleagues (Dweck & Reppucci, 1973; Dweck & Licht, 1980; Dweck & Wortman, 1982) indicates that for some children early school failure leads to a cycle of helplessness and poor achievement. They suggest that children differ in their responses to school failure, distinguishing between "mastery-oriented" children who are effective copers and "helpless" children who are inefficient copers. Differences between the two groups are primarily focused on the cognitive coping strategies employed in response to failure and the types of attributions made with respect to failure (Dweck & Reppucci, 1973; Dweck & Licht, 1980; Dweck & Wortman, 1982). Such possible school failure among children with physical illness must be seen not only in the context of the disease but in a wider framework in which the cognitive coping strategies of the child are considered, which has important implications for effective intervention. A recent follow-up study of former paediatric renal patients found that lower self esteem in adulthood was linked to reduced educational achievement (Morton, Reynolds, Garralda, Postlethwaite & Goh, 1994), indicating the importance of early educational intervention to minimise disruption caused by chronic illness and maximise academic achievement.

Intellectual development is likely to be compromised among those children who develop chronic illness early in life (Ack, Miller & Weil, 1961; Dikmen, Matthews & Harley, 1975; Eiser & Lansdown, 1977; Eiser, 1981; Stewart, Uauy, Kennard, Waller, Benser & Andrews, 1988). Conversely, it has been suggested that the more intelligent child is more adaptive in dealing with a chronic illness (Vignos, Thompson, Katz, Moskowitz, Fink & Svec, 1972). IQ tests have been criticised as not sufficiently sensitive to detect changes that might be attributable to illness or drug effects (St. James Roberts, 1979). However, some measure of cognitive performance as part of a detailed psychological assessment is widely considered to be not only valid but also valuable in the study of chronically sick children.

2.1.2. Cognitive Development and Congenital Heart Disease

Impaired central nervous system functioning (Aisenberg, Rosenthal, Wolff & Nadas, 1974) and developmental and cognitive impairment have been reported in children with congenital heart disease, particularly those with cyanotic lesions (Bret & Kohler, 1956; Feldt, Ewert, Stickler & Weidman, 1969; Silbert et al, 1969; Gonzalez-Pardo, Miles, Taylor & Mattioli, 1981; O'Dougherty et al, 1983; Newburger, Silbert, Buckley & Fyler, 1984; Aram, Ekelman, Ben-Shachar & Levinsohn, 1985; DeMaso, Beardslee, Silbert & Fyler, 1990). Developmental delay has been recognised in infants with congenital heart lesions as early as two months of age (Aisenberg, Rosenthal, Nadas & Wolff, 1982). The general finding that children, who as infants were undernourished and failing to thrive, have developmental and cognitive scores below the normal range (Stoch & Smythe, 1963; Singer & Fagan, 1984) is of particular relevance for a number of children with congenital heart disease. In a five year longitudinal study Linde et al (1970) found that children with either acyanotic or cyanotic heart disease obtained scores below those of healthy children on various measures

of intelligence. Children with cyanosis achieve significantly lower scores than children with acyanotic lesions, which has been attributed to the presence of chronic hypoxia (O'Dougherty, Wright, Loewenson & Torres, 1985) but it is questionable whether this is a simple effect of chronic hypoxia leading to organic brain damage and an intelligence deficit. Instead, a number of additional risk factors have been postulated, such as the extent of physical incapacity and degree of parental over-protection (Linde, Rasof & Dunn 1967). A strong relationship has also been found between the presence of congestive heart failure and developmental delay (Aisenberg et al, 1982). Whilst it seems to be widely accepted that the presence of a congenital heart defect can have some deleterious effect on intelligence, mean scores for cardiac patients have been found to be within the normal range, albeit at the lower end (Chazan, Harris, O'Neill & Campbell 1951; Kramer, Awiszus, Sterzel, van Halteren & Classen, 1989).

In contrast to the majority of studies of congenital heart disease and cognitive function, one study of a heterogeneous group of 126 children found no significant differences in IQ scores between cyanotic and acyanotic patients (Fyler, Silbert & Rothman, 1976). However, it was unclear which children had undergone repair of the lesion and which had not, thereby making it impossible to draw any firm conclusions from this study.

Studies suggest that perceptual motor function is more profoundly impaired than other cognitive functions in both cyanotic and acyanotic heart disease in children (Honzik, Buse, Fitzgerald & Collart, 1976; Newburger et al, 1984). It has also been found that these differences in IQ diminish with age (Rasof, Linde & Dunn, 1967), a finding which has been attributed to the nature of intellectual functions tapped by intelligence tests at different age

levels. IQ tests in the early years rely more heavily on sensory-motor function, whilst those at a later age emphasise verbal and memory abilities. The curtailment of physical activity in the child with a cyanotic lesion affects the development of sensory-motor processes and limitations in responsiveness and energy during tasks relying on gross motor functioning may produce proportionately lower scores in these infants, scores which bear little relation to their IQ after the age of 3 years.

Children with congenital heart disease are at risk of cardiac arrest and a study of children who had survived cardiac arrest found that they had globally lowered IQ, visual, perceptual-motor, achievement, adaptive and fine and gross motor abilities (Morris, Krawiecki, Wright & Walter, 1993). The population in this study included children with cyanotic and acyanotic heart disease and children with no heart disease. However, the authors did not distinguish between these different groups in the analyses, so it is possible that the effects described are at least partly attributable to the presence of congenital heart disease and the recognised cognitive deficits associated with cyanotic heart disease in particular, rather than being purely due to the cardiac arrest itself. However, increased duration of cardiac arrest was associated with decreased functioning, so it would appear that cardiac arrest is an additional risk factor for children with congenital heart disease.

2.1.2.1. Treatment of Congenital Heart Disease

2.1.2.1.1. Nature of the Cardiac Lesion

Surgical intervention has been found to lead to a significant increase in IQ scores (Meyendorf, Jansch, Trondle, Takke, Buhlmeyer & Sebening, 1980) and in particular in cyanotic patients (Linde et al, 1970), attributed in part to improved oxygenation levels.

Children with acyanotic defects tend to exhibit less deficit pre-operatively and tend to be functioning at a level closer to their true potential. They therefore show less change in IQ after surgery, which also explains why acyanotic patients who do not undergo surgical intervention do not differ significantly from those children with corrected acyanotic lesions. In a small study of Tetralogy of Fallot patients, 4 out of 7 children showed post-operative increases in IQ (Bret & Kohler, 1956). A retrospective study of a small group of patients with surgically corrected transposition of the great arteries (TGA) who were compared with a group of children who had had a ventricular septal defect (VSD) repaired indicated that the TGA patients had a greater impairment of cognitive skills than the VSD patients (Hesz & Clark, 1988), further supporting the theory of impeded cognitive development in patients with cyanotic heart disease, even after surgical repair of the lesion. A further follow-up study of 24 children who had corrective surgery for TGA in infancy also found significant morbidity in the form of neurodevelopmental problems (Mendoza, Wilkerson & Reese, 1991). In contrast, 17 children assessed after repair of Tetralogy of Fallot before two years of age were found to be indistinguishable in terms of intellectual and social development from a group of randomly selected healthy children (Sunderland, Matarazzo, Lees, Menashe, Bonchek, Rosenberg & Starr, 1973). However, no information was given on the matching of this healthy group with the study group. One of the few prospective, long-term follow-up studies found a high correlation between pre-operative child IQ scores and post-operative adult IQ scores in Tetralogy of Fallot patients (Shampaine, Nadelman, Rosenthal, Behrendt & Sloan, 1990). This latter study highlights the importance of prospective evaluations.

Despite poor school performance being increasingly recognised as a concomitant of chronic illness, academic functioning in the child with congenital heart disease has received little

attention. Most studies have focused on IQ, but a recent study of school performance in a group of children with surgically corrected cyanotic lesions found them to have significantly poorer performance in all areas of academic functioning compared with a group of children who had had previously diagnosed innocent murmurs or a spontaneously closing ventricular septal defect (Wright & Nolan, 1994). Despite having had corrective surgery before two and a half years of age, children with cyanotic heart disease are at "great risk" of having difficulties at school (Wright & Nolan, 1994). Poor concentration, particularly at school, has also been identified in a significantly greater number of children with surgically corrected cyanotic, rather than acyanotic, defects (Kallfelz, Kaemmerer, Luhmer, Lacher, Anacker & Wietzke, 1992). In a Belgian study of children with corrected cyanotic or acyanotic lesions, 41% of the sample had "school retardation", compared with 13% of the normal population (Dhont, De Wit, Verhaaren & Matthys, 1992). A further follow-up study of children with surgically corrected TGA found that 13% went to special schools, indicating some degree of intellectual impairment (Turina, Pasic, Fry & Von Segesser, 1992).

2.1.2.1.2. Age at Treatment

Greater impairment of gross motor function (Newburger, Tucker, Silbert & Fyler, 1983) and of general cognitive function have been found to be associated with older age at time of corrective surgery for children with TGA (O'Dougherty et al, 1983; Newburger et al, 1984) but age at time of repair was found to correlate poorly with cognitive function in children with a VSD - an acyanotic defect. The data suggested that postponing the repair of a cyanotic lesion - but not an acyanotic defect - is associated with a progressive impairment of cognitive function. Several hypotheses have been postulated to explain why an increased duration of hypoxemia has a harmful effect on intellectual functioning. Suggestions include

effects of hypoxemia on central nervous system (CNS) functioning and development and time dependent effects of chronic hypoxemia on the brain, together with the increasing cumulative risk of a cerebrovascular accident with time. However, no clear mechanism has yet emerged to explain the findings.

Retarded developmental outcome following corrective surgery for TGA in infancy was found to be positively correlated with various medical and family stress factors, such as prolonged hypoxia, growth retardation, congestive heart failure and socioeconomic status (O'Dougherty et al, 1983). Overall outcome was compromised if multiple risk factors occurred, indicating that the timing of surgery is not the only relevant factor in post-operative intellectual development.

Palliative surgery has also been found to be beneficial for subsequent intellectual development (Finley, Buse, Popper, Honzik, Collart & Riggs, 1974). Pre-operative IQ scores were higher in children undergoing corrective surgery for Tetralogy of Fallot who had had a previous palliative shunt compared with those who had not. Post-operative IQ score increases were also greater in the shunt group.

2.1.2.1.3. Type of Treatment

Most research has focused on cardiopulmonary bypass as opposed to deep hypothermia and circulatory arrest. Both positive (Messmer et al, 1976) and negative (Wells et al, 1983) results of the impact of hypothermia and circulatory arrest on intellectual development have been reported. No conclusions about the implications for cognitive development can be drawn at present, although it may be significant that the authors of these studies regard lack

of cognitive impairment as a positive factor, rather than looking for an improvement in post-operative functioning. In addition to the duration of circulatory arrest as a factor in post-operative cognitive performance, the duration of cooling before circulatory arrest is a predictor of cognitive development. The data from a retrospective study of 28 children who underwent repair for TGA in infancy suggest that it is possible to define an optimal period of cooling which will allow a safe maximum duration of circulatory arrest (Bellinger, Wernovsky, Rappaport, Mayer, Castaneda, Farrell, Wessel, Lang, Hickey, Jonas & Newburger, 1991).

2.1.2.2. Gender Differences in Cognitive Function

A few studies have investigated characteristics of the child - in particular the sex of the child - as determinants of intellectual functioning. There is a lack of detail in reporting the data and the results are inconclusive. A reversal of the normal IQ findings was found in a group of North American children with congenital heart disease, in which girls had lower pre- and post- operative verbal IQ's than those of boys and also than their own performance IQ scores (Honzik, Collart, Robinson & Finley, 1969). In a replication of the study using children from a cultural background in which boys are valued more highly than girls, Cravioto, Lindoro & Birch (1971) found a reversal of the previous results and they suggest that the presence of a congenital heart defect results in excessive experiential constriction for boys due to overprotection in this culture, with deleterious effects on various aspects of intelligence, and they suggest that the results of Honzik et al can be explained in terms of child rearing differences.

2.1.2.3. Cardiac "Non-Disease" : Psychological Effects

Innocent or functional heart murmurs exist in a large percentage of the childhood population, the actual prevalence varying with the age of the child and the experience of the examiner (Bergman & Stamm, 1967). Figures for the number of normal children with innocent murmurs have been quoted to be as high as 44% (Quinn & Campbell, 1962) and the prevalence of such murmurs together with the fact that they are often misdiagnosed as being due to cardiac defects results in many infants and children being incorrectly considered to have heart disease (Bergman & Stamm, 1967; Cayler & Warren, 1970; Kupst, Blatterbauer, Westman, Schulman & Paul, 1977). It has been estimated that the amount of disability from cardiac non-disease in children is greater than that due to actual heart disease (Bergman & Stamm, 1967). In a study to look at intellectual and perceptual-motor development, intelligence scores were found to be lowest in a group of restricted children with cardiac non-disease (Cayler, Lynn & Stein, 1973). Non-restricted children with cardiac non-disease, whilst performing better than the restricted group, demonstrated a lower level of intellectual functioning than a group of normal children. The lack of significant differences between the three groups on a test of perceptual motor development excludes organic brain disease as a factor in the impaired performances of the cardiac non-disease groups and it is concluded that the accumulative emotional factors of the diagnosis of heart disease, especially if accompanied by physical restrictions, are detrimental to intellectual development. This fits in with findings with children who do have a cardiac malformation that it is the perception of the disease, rather than the severity of the condition per se, that has a greater influence on developmental and intellectual outcome (Linde, Rasof, Dunn & Rabb, 1966).

2.1.3. Bone Marrow Transplantation

2.1.3.1. Effects of Initial Disease

Among children with leukaemia, there is a comprehensive literature detailing the impact of treatment regimens on cognitive function. The emphasis has been on the effects of prophylactic central nervous system (CNS) treatment by irradiation and the wealth of retrospective and prospective studies conclusively highlight the potentially deleterious effects of CNS irradiation, particularly for young children (Eiser & Lansdown, 1977; Moss & Nannis, 1980; Meadows, Gordon, Massari, Littman, Fergusson & Moss, 1981; Moss, Nannis & Poplack, 1981; Jannoun, 1983; Stehbens, Kisker & Wilson, 1983; Twaddle, Britton, Craft, Noble & Kernahan, 1983). A meta-analysis of all research published between 1975 and 1985 in which children with acute lymphatic leukaemia (ALL) who were treated with cranial irradiation were compared with patients with a similar disease profile who did not receive irradiation revealed a significant IQ deficit in the irradiation group (Cousens, Waters, Said & Stevens, 1988). However, this did not yield information about whether the patients had a generalised IQ deficit or whether specific skills were affected, nor was any account taken of potential mediating variables such as gender or social class (Eiser, 1991). Further retrospective research found that CNS prophylaxis for ALL affected four cognitive processes - namely short term memory, speed of processing, visuomotor coordination and sequencing ability (Cousens, Ungerer, Crawford & Stevens, 1991). Specific auditory learning deficits, independent of IQ, have also been reported (Jannoun & Chessells, 1987). Whilst most of the published research is retrospective, a prospective study of ALL patients confirmed the reduction in IQ scores previously reported and further identified age and initial IQ as associated with a drop in IQ score (Meadows et al, 1981).

The effects on intelligence of the other diseases for which BMT is a treatment regimen are not well documented with the exception of some of the mucopolysaccharidoses, such as Hurler's and Hunter's syndromes, for which a degree of mental handicap and retarded development are recognised consequences of the disease (Hull & Johnston, 1981; Churchill's Medical Dictionary, 1989; Kurihara, Kumagai, Goto & Yagishita, 1992).

2.1.3.2. Treatment of BMT

There is little documented evidence on the effects of BMT on intellectual functioning in children. One prospective neurodevelopmental study of two infants with ALL who received a BMT found no significant late neurodevelopmental sequelae - contradicting published research on the deleterious effects of irradiation (Kaleita, Tesler & Feig, 1987). After five year follow-up of the two children the authors suggested that intensive chemotherapy and TBI, without intrathecal therapy (injecting drugs directly into the spinal column), may not cause permanent injury during a period of rapid brain development. This small study further highlights the importance of the age of the child at the time of the treatment and would seem to suggest that there is a period during infancy when TBI administration will not result in impeded neuropsychological development. This paper, however, would appear to say more about the effects of irradiation, rather than of BMT, on cognitive function.

A recent retrospective study of neuropsychological functioning in 32 children who had undergone BMT 1-6 years previously found that age at the time of BMT was a critical factor (Smedler, Ringden, Bergman & Bolme, 1990). Children who were 12 years or older at the time of BMT had no observable deficits, whereas neuropsychological impairment was clearly present in those children with an original diagnosis of leukaemia who were between the ages

of 1-3 years at the time of BMT. Children between 3 and 11 years showed some subtle deficits in tasks of perceptual and fine motor speed. However, in view of the fact that 25 children had an original diagnosis of leukaemia and had received irradiation, the results of this study could also be attributed in part to the effects of irradiation, rather than BMT, on neuropsychological functioning. A small number of the group had non-leukaemic diseases and had not received irradiation as part of the conditioning regimen. Although the number of these children was too small for statistical analyses, their performance was more similar to that of the sibling comparison group. The clear indication from these data is that it is the conditioning regimen, rather than the BMT itself, which has a significant effect on neuropsychological functioning, particularly in those children treated before the age of 3 years (Smedler & Bolme, 1995). One further study reports favourable psychological development after BMT for Gaucher disease, with an "excellent IQ" in one patient ten years after BMT (Ringden, Groth, Erikson, Granqvist, Mansson & Sparrelid, 1995). However, few details of the psychometric testing are given by the researchers.

For children with Hurler's syndrome, stabilization of intellectual status has been demonstrated following BMT (Hobbs, Barrett, Chambers, James, Hugh-Jones, Byrom, Henry, Lucas, Rogers, Benson, Tansley, Patrick, Mossman & Young, 1981; Hugh-Jones, 1986), but there is no evidence that the brain damage already present at the time of treatment can be reversed.

2.2. ADJUSTMENT OF CHRONICALLY ILL CHILDREN

The psychological consequences of chronic disease in childhood have been the subject of prolific research since the 1970's, with research assuming an "increasingly important position in medical psychology and behavioural medicine" (Watson & Kendall, 1983). However,

unnecessary and superfluous replication and revision of previous research is now occurring (Pless & Nolan, 1991). The traditional focus on maladjustment as a consequence of chronic illness is now being replaced with models which take into account coping strategies and individual competence (Eiser, 1990(a)) and there is now a need to identify specific determinants or modifiers of the risks associated with chronic illness.

Most of the published research supports the view that chronically ill children are at increased risk of psychosocial maladjustment compared with healthy children (Gortmaker, Walker, Weitzman & Sobol, 1990; Pless & Nolan, 1991; Lavigne & Faier-Routman, 1992) and children with fatal illnesses have been found to have a higher prevalence of psychiatric disorder than those with non-fatal chronic illnesses (Howarth, 1972). However, there is now increasing acknowledgement of the methodological difficulties of much of this research, and resulting confusion surrounding the findings. Furthermore, because the course of childhood chronic disease has been altered through changes in medical management, early reports about psychological sequelae may now be of limited relevance (Breslau, 1990).

2.2.1. Ambiguities in Terminology

Throughout the literature the terms "coping", "adaptation", "adjustment" and "competence" are used interchangeably (Rutter, 1981; Compas, 1987) but the focus is shifting away from defining maladjustment and negative concepts associated with chronic illness towards identifying individual and family coping strategies and skills (Varni & Wallander, 1988). A number of studies have also assessed the prevalence of "depression" in chronically ill children (e.g. Kashani & Hakami, 1982; Worchel, Nolan, Wilson, Purser, Copeland & Pfefferbaum, 1988). The view that "children with chronic physical disease are invariably depressed"

(Battle, 1975) was not borne out by a meta-analysis, which found that children with chronic medical problems are at a slightly elevated risk for depressive symptoms but most are not clinically depressed (Bennett, 1994).

2.2.2. Methodological Concerns

Relatively few population-based studies, based on large representative samples, have been carried out despite the fact that they are considered preferable to clinic based studies (Starfield, 1985). Instead, much of the reported data have been collected in small, often specifically selected clinic samples. Specific characteristics of such populations and policies of medical management are infrequently described and the inevitable differences in such factors make comparisons across studies and between clinic samples difficult. This is borne out by the considerable intra-order variation in outcome for those disorders where multiple studies are available (Lavigne & Faier-Routman, 1992). Furthermore, within single disease categories there is a need to identify subgroups that differ on relevant descriptive characteristics and evaluate the significance of these subgroups for diagnosis and treatment (Drotar, 1994). The use of control groups is variable, with some studies relying on normative comparisons. In those studies where control groups are employed, insufficient attention is frequently given to relevant variables (Lemanek, Lytle Moore, Gresham, Williamson & Kelley, 1986). Although within-group differences are now considered more pertinent (Lemanek, 1994), there is still an emphasis on comparisons to normative data and study controls (Kazak, 1993). The lack of a comprehensive theoretical framework has resulted in a wide range of outcome measures and variability in the source of the information (parent, child, teacher etc.) although the importance of basing investigations on theoretical models is now acknowledged (Harper, 1991).

2.2.3. Risk Factors

More specific attributes of the psychosocial dysfunction in chronic illness are being sought in order to further understand the impact of chronic illness and to plan appropriate interventions and target resources. Such risk factors may be associated with demographic, medical, social, environmental or individual personality characteristics (Pless & Nolan, 1991).

2.2.3.1. Demographic Factors

The risk of emotional problems increases with age for children with chronic illness (Pless, Roghmann & Haggerty, 1972), mirroring the findings for the general population (Rutter et al, 1970). Chronic illness in general has a more detrimental impact on the emotional wellbeing of boys compared with girls (Hurtig & White, 1986) , although very few gender related effects of specific illnesses have been documented. Family size influences the amount of information parents give their children about their illness and treatment, with children from larger families tending to be told more (Chesler, Paris & Barbarin, 1986). Social class appears to be a relatively unimportant factor, with no clear pattern of cause and effect. Within the general population, children from inner city areas have been found to be at greater risk for behaviour problems and psychiatric disorder compared with children from rural locations (Rutter, Cox, Tupling, Berger & Yule, 1975). Psychosocial differences in cultural background are also influential in determining a child's and family's response to chronic illness (Spinetta, 1984).

2.2.3.2. Specific Diseases

Much research supports the idea that specific disease characteristics are not risk factors for psychosocial problems (Stein & Jessop, 1982(a); Breslau, 1985). However, CNS

involvement has been found to be a significant predictor of behaviour problems (Rutter et al, 1970; Breslau, 1985; Breslau & Marshall, 1985; Howe, Feinstein, Reiss, Molock & Berger, 1993; Fletcher, Brookshire, Landry, Bohan, Davidson, Francis, Thompson and Miner, 1995) although it is unclear whether the higher prevalence of adjustment difficulties is a direct or secondary result of brain dysfunction. There is also evidence that children with sensory impairments are at increased risk (Pless & Satterwhite, 1975(b); Pless, 1984(a)).

2.2.3.3. Severity

Much of the evidence on the effects of illness severity on adjustment is confusing, in particular due to the lack of standardised measures of severity that are valid and reliable across different diseases (Stein, Gortmaker, Perrin, Perrin, Pless, Walker & Weitzman, 1987). Some studies have found a direct relationship between severity and maladjustment (e.g. McNichol, Williams, Allan & McAndrew, 1973; Heller, Rafman, Zvagulis & Pless, 1985) whilst others have found a curvilinear relationship (McAnarney, Pless, Satterwhite & Friedman, 1974; Pless & Satterwhite, 1975(b)) and yet others have found no linear relationship (Harper, 1983; Garralda, Jameson, Reynolds & Postlethwaite, 1988; Hurtig, Koepke & Park, 1989; Pless & Nolan, 1991). Parental rather than clinicians' or other objective assessments of severity have been suggested to be better predictors of adjustment (Perrin, MacLean & Perrin, 1989; Bradford, 1994). The strength of the relationship between "severity" and psychological adjustment may differ within demographic subgroups, which has been suggested as an explanation for the controversies over the relationship between the child's chronic condition and psychological adjustment (Stein & Jessop, 1984).

2.2.3.4. Visibility

Better psychosocial adjustment has been found in some patients with visible physical characteristics revealing the presence of an illness or disability (Zahn, 1973). Chronically ill children with an essentially normal appearance were found to have poorer adjustment (Jessop & Stein, 1985). However, a comparison of children with congenital heart disease (invisible) and facial burns (visible) indicated better adjustment in those with the invisible defect (Goldberg, 1974). A study of young adult survivors of end-stage renal disease found that those with more visible handicaps were at greater risk of maladjustment (Beck, Nethercut, Crittenden & Hewins, 1986).

2.2.3.5. Age at Onset and Duration

Adverse consequences of chronic illness have a potentially greater impact the younger the child at diagnosis. It has been suggested that there are qualitatively different consequences of illness depending on age at diagnosis (Maddison & Raphael, 1971). It is still unclear how duration of illness affects adjustment. There is support for the view that adjustment difficulties are not stable over time, but tend to vary according to disease state or at times of crisis (Steinhausen, 1976) and as a function of the child's age (Ungerer, Horgan, Chaitow and Champion, 1988; Nash, 1990).

2.2.3.6. Individual Characteristics

A major flaw with research in this area is the lack of data on personality and individual attributes before the onset of chronic disease. An external health locus of control has been found to be far more prevalent in ill children than in healthy children (Perrin & Shapiro,

1985), but the conclusion that this is due to the effects of the disorder is premature (Pless & Nolan, 1991).

2.2.3.7. Social Environment

The effects of family, teachers and peers on adjustment is receiving growing attention. There is considerable evidence that family dysfunction is associated with emotional problems among children with chronic illness (e.g. Steinhausen, Schindler & Stephan, 1983; Sabbeth, 1984). However, the lack of data on family functioning prior to the onset of illness and the fact that the illness itself may well disrupt family functioning make the direction of the relationship unclear. There are also indications that chronically ill children experiencing higher levels of life stress have poorer self-concepts and more illness episodes than those with lower levels of life stress (Bedell, Giordani, Amour, Tavormina & Boll, 1977). The role of perceived social support from parents, teachers and peers has been shown to be an important predictor of adjustment for children with chronic conditions (Varni & Setoguchi, 1991). The degree of social skills displayed has also been found to predict adjustment (Kapp-Simon, Simon & Kristovich, 1992) and it has been suggested that social skills training may be beneficial for increasing perceived social support and reducing behaviour problems (Varni, Katz, Colegrove & Dolgin, 1993).

2.2.3.8. Medical Environment

Medical treatment itself can have adverse effects on psychological functioning (Pless, 1984(b)). Enforced dependency on others, the violation of the body by medical treatment, and restrictions on voluntary actions and behaviour due to care and treatment regimens result in a loss of control which can be overwhelming for the child (Van Dongen-Melman &

Sanders Woudstra, 1986(b)). The medical setting and the pattern of care, despite having been poorly studied, are also potentially influential factors. For example, out-patient regimens, particularly where follow-up visits are scheduled according to protocol rather than individual medical wellbeing, can give conflicting messages of psychological significance as they emphasise illness and hospitalisation yet the child is being told that they are well and encouraged to lead a "normal life".

2.2.4. Adjustment From a Developmental Perspective

Age affects adjustment in terms of the types of difficulties experienced. Developmental level influences a child's interpretation of illness, although the traditional focus on the cognitive approach as a means of understanding the impact of illness (e.g. Bibace & Walsh, 1981; Perrin & Gerrity, 1984) has been criticised for failing to incorporate the influences that social and cultural factors have on a child's understanding of illness. More recent studies suggest that conceptual changes in development occur when an individual child constructs their own theories and understandings based on the information available to them from their own experience (Charman & Chandiramani, 1995). What is clear is that illness challenges the accomplishment of specific tasks at different ages. Chronic illness in infancy can affect attachment behaviour (Fischer-Fay, Goldberg, Simmons & Levison, 1988). Security of attachment has been found to be predictive of the quality of a child's other relationships (Park & Waters, 1989) as well as reflecting the quality of the mother-child relationship (Goldberg, Perrotta, Minde & Corter, 1986).

Schooling is important for promoting independence and autonomy. Chronic illness in middle childhood and resultant absenteeism from school can not only lead to poorer academic

achievement but can influence the development of peer relationships and integration within the school environment (Nash, 1990). Social isolation (Spirito, Stark, Cobiella, Drigan, Androkites & Hewett, 1990; Blum, Resnick, Nelson & Germaine, 1991) and lower levels of social competence (Uzark, Sauer, Lawrence, Miller, Addonizio & Crowley, 1992) have been found in some groups of chronically ill children, but other ill groups do not differ from physically healthy children on measures of social competence and peer relationships (Graetz & Shute, 1995; Nassau & Drotar, 1995). As well as the potentially negative impact of chronic illness on social development, the experience of illness may also have more positive effects in terms of increased empathy towards, and awareness of, other people. Although this has been postulated with respect to the experience of minor illnesses (Parmelee, 1986) there has been little systematic investigation of this with respect to the experience of chronic illness (Eiser, 1993).

For the adolescent the challenge is to achieve the desired independence and autonomy whilst also maintaining close and supportive links with the nuclear family (Eiser, 1993). Illness at this time exaggerates the challenge. Increased dependence, for example, may be unavoidable, thereby curtailing the move towards autonomy (Coupey & Cohen, 1984). The social consequences of chronic illness may be more significant in adolescence than earlier (Ungerer et al, 1988) as physical and practical limitations become more apparent and the presence of disability increases vulnerability in specific areas of psychosocial functioning (Orr, Weller, Satterwhite & Pless, 1984). Chronically ill adolescents may be particularly vulnerable to social isolation and delayed development of peer- support networks (Melzer, Leadbeater, Reisman, Jaffe & Lieberman, 1989; Noll, Bukowski, Davies, Koontz & Kulkarni, 1993), although peers have been found to be an important source of emotional, companionship and

disease management support for the ill adolescent (La Greca, 1992). Body image and self perception become particularly important in adolescence, although the presence of chronic illness does not necessarily result in lower self esteem (Sullivan, 1978; Kellerman, Zeltzer, Ellenberg, Dash & Rigler, 1980; Kovacs, Iyengar, Goldston, Stewart, Obrosky & Marsh, 1990). Certain diseases, such as cystic fibrosis, do, however, seem to be associated with low self-image (Boyle, di Sant'Agnese, Sack, Millican & Kulczycki, 1976; Offer, Ostrov & Howard, 1984).

2.2.5. Hospitalisation

Hospitalisation is a frequent experience for many children with chronic disease. It is well documented that repeated and long-term hospital stays can cause a negative pattern of emotional reactions (Bowlby, 1952; Douglas, 1975; Quinton & Rutter, 1976). Repeated hospital admissions are also associated with an increased risk of psychosocial problems in later childhood and adolescence, particularly in young children already experiencing chronic family adversity (Rutter, 1982). The period 6 months to 4 years is particularly critical in terms of adverse psychological consequences of hospital admission (Prugh, Staub, Sands, Kirsschbaum & Lenihan, 1953; Vernon, Schulman & Foley, 1966; Goslin, 1978). The recognised trauma associated with hospitalisation has resulted in greater attention being given to the child's psychological needs in hospital (Platt Committee, 1959). It is now accepted that parents should be able to stay with their child in hospital, that they should have unlimited visiting and that children should be treated on paediatric wards by specially trained nurses, and with access to appropriate educational and play services. More recent studies now suggest that in a modern paediatric setting there is little evidence to indicate that hospital admission has any significant effect on a child's subsequent behavioural pattern (Shannon,

Fergusson & Dimond, 1984). It has also been acknowledged that separation from the family, rather than just the mother, is stressful for the child (Rutter, 1979). Although changes in hospital policy mean that a parent usually stays with the child, there can still be lengthy separations from other family members (Hamlett, Walker, Evans & Weise, 1994).

The importance of understanding children's anxiety responses to hospital and being sensitive to their fantasies regarding hospitalisation is being increasingly recognised (Willis, Elliott & Jay, 1982) and a second point of focus has been on trying to reduce anxiety and stress in hospitalised children. A number of techniques for preparing children have been devised, such as puppet therapy (Cassell, 1965), rehearsal (Visintainer & Wolfer, 1975) and filmed modelling (Vernon & Bailey, 1974; Melamed & Siegel, 1975; Robinson & Kobayashi, 1991). Relaxation training and stress inoculation techniques (Meng & Zastowny, 1982; Zastowny, Kirschenbaum & Meng, 1986) have also been used for anxiety reduction. The importance of targeting preparation techniques at a level appropriate for the child's conceptual abilities has also been highlighted (Rasnake & Linscheid, 1989). However, not all children require preparation and some may become even more upset by it (Melamed & Ridley-Johnson, 1988). Research into the efficacy of such practices has highlighted a number of difficulties, such as the unsuitability of some of these techniques for the most vulnerable children and the discrepancy between research findings and clinical practice (Peterson & Mori, 1988; Saille, Burgmeier & Schmidt, 1988). The clinical use of individual preparation techniques does not appear to be based on research data on their efficiency and some of the most frequently used techniques have below average effectiveness. Furthermore, most of the research on preparation for hospital has focused on children requiring short-stay hospitalisation for minor, low risk surgery (Eiser, 1984).

Research on adjustment to hospitalisation has indicated the importance of the number of physical stressors experienced by the child during their hospital stay and factors such as personality style in determining adjustment (Saylor, Pallmeyer, Finch, Eason, Trieber & Folger, 1987). Children's coping with specific hospital stressors has been found to be related to characteristics of the situation itself and the actions of professionals in the situation (Ellerton, Ritchie & Caty, 1994). The behaviour of parents is also widely acknowledged to influence children's coping and distress during acute painful events (Blount, Davis, Powers & Roberts, 1991; Frank, Blount, Smith, Manimala & Martin, 1995). A history of previous surgery has been found to predict presurgical anxiety, with elevated presurgical anxiety predicting later behaviour problems in children hospitalised after surgery (Lumley, Melamed & Abeles, 1993).

2.2.6. Theoretical Approaches To Coping with Chronic Illness

There are two main theoretical perspectives. One view follows the prevailing medical approach that all diseases differ. Treatment of, and adjustment to, each disease is dependent on its specific characteristics and the focus of research should be to attempt to identify and describe these. The alternative approach is the non-categorical approach (Pless & Pinkerton, 1975; Stein & Jessop, 1982(a,b)) which suggests that children with chronic physical disorders "face common life experiences and problems based on generic dimensions of their conditions rather than on idiosyncratic characteristics of any specific disease entity" (Stein & Jessop, 1982(a)). A problem with this approach is that it is difficult to evaluate the relative impact of any one dimension on adjustment or to isolate diseases that only vary across a single dimension.

Current research is unable to distinguish the relative merits of these two theoretical approaches, due to the lack of comparative, properly controlled research on different diseases and the bias towards studying a few conditions such as cancer in depth whilst the psychological impact of other conditions remains virtually unknown. However, in terms of intervention and provision of practical assistance, the non-categorical approach has many advantages and allows policies of care and liaison for the ill child to be established irrespective of the specific disease entity.

2.2.7. Coping

The vast majority of chronically ill children appear to cope with their disease and treatment. Understanding the ways in which children cope and how illness impacts on this will allow the development of appropriate interventions to improve adaptation.

Most of the current research utilises some aspects of the adult model of coping proposed by Lazarus and Folkman (1984). Four approaches to the study of coping in children and adolescents have been identified (Compas, Worsham & Ey, 1992), all of which emphasize a basic distinction between two fundamental types of coping. Coping can be achieved either by attempting to control or change some aspect of the situation (problem-focused) or by managing negative emotions associated with the stressor (emotion-focused). As with all other aspects of children's responses to chronic illness, use of coping strategies is also influenced by developmental changes. Research is still at an early stage but there are indications of differences in the type and number of coping strategies utilised by boys and girls and the fact that medical (e.g. number of hospital admissions) and demographic (e.g. age at diagnosis) factors can influence the type of strategy used (Spirito, Stark & Tyc, 1989).

Emotion-focused coping in medical situations appears to increase with age (Band & Weisz, 1988; Bull & Drotar, 1991) although problem-focused coping has been found to lead to better adaptation in diabetic children (Band, 1990) and those with sickle cell disease (Gil, Williams, Thompson & Kinney, 1991). Time since diagnosis is also influential in determining changes in coping strategies in response to illness (Phipps, Fairclough & Mulhern, 1995).

Assessment of the effects of intervention strategies on adjustment have tended to look at the effects of increasing disease-related knowledge (Rubin, Leventhal, Sadock, Letovsky, Schottland, Clemente & McCarthy, 1986), improving self-care skills (Johnson, 1988) and developing social skills to enable a child to cope with their disease and treatment. Many issues still wait to be resolved: How should efficacy of intervention strategies be measured?; Who should facilitate the strategies? The issues already mentioned in the context of assessing adjustment to chronic illness are pertinent for assessing intervention strategies too. What is clear is that for children with chronic illness concern about maladjustment should focus on the introduction, improvement, and strengthening of adaptive processes and coping skills within a developmental framework encompassing medical, social and cultural influences, with the aim of preventing the emergence of adverse psychological sequelae.

2.2.8. Congenital Heart Disease

Congenital heart disease, perhaps more than any other individual chronic illness, has the potential for creating psychological maladjustment entirely out of proportion to the severity of the lesion. The reasons for this centre on the emotional and psychological significance attached to the heart. Parents and children are inclined to exaggerate the danger of any symptoms related to the heart and to respond to their misgivings (Pless & Pinkerton, 1975).

The literature on the adaptation of children with congenital heart disease is limited and often contradictory, although this would appear to be due largely to methodological problems such as variability in outcome measures, lack of consideration for age-related developmental differences and wide variation in the type of lesion studied. Table 2.1 summarises the main studies, their patient groups and the major findings.

ADJUSTMENT IN CHILDREN WITH CONGENITAL HEART DISEASE

AUTHORS	PATIENT POPULATION	AGE RANGE	MAJOR FINDINGS
Freed et al (1953)	71 CHD pts and 71 age matched healthy controls		Projective sentence completion task to investigate fear. CHD pts. had a fear of their handicap but had less fear of inter-personal relationships than healthy group. Specific situations feared by both groups were essentially similar.
Landtman et al (1959)	42 CHD pts assessed pre- and post-op	5-15 years	Pre-op, 21 had severe maladjustment problems at home. Post-op improvement occurred in 17 of these, particularly in sleep and eating problems.
Green et al (1962)	25 CHD pts. 3 control grps.	8-16 years	CHD grp had constricted body image compared with normal children.
Linde et al (1966)	98 CY pts. 100 ACY pts. 2 control grps.		Poorer adjustment and anxiety in cardiac pts. related more highly to maternal anxiety and pampering than to degree of medical incapacity.
Linde et al (1970)	As above		Following surgery, CY grp but not ACY grp were significantly more confident and showed better adjustment after surgery compared with pre-op. Non-operated cyanotics showed a significant deterioration in general adjustment. Neither acyanotic grp showed any significant changes in adjustment.
Auer et al (1971)	28 VSD pts- 14 had surgery; 14 did not	6-11 years	No significant difference in incidence of emotional disturbance between 2 grps. VSD grp overall showed rates of emotional disturbance higher than in a "control group".
Danilowicz et al (1971)	68 CHD pts under-going surgery	0.5-15 years	63 of 67 survivors showed reactions consistent with expectations for adults (anxiety, anger, cooperation, compliance). In 4 children grossly abnormal responses were seen.
Barnes et al (1972)	11 pts under-going surgery	5-14 years	Study of neuroendocrine and behavioural response to stress. Neuroendocrine activity higher on day before surgery and day of return from ITU, but no correlation between such activity and overt anxiety levels.
Offord et al (1972)	20 CHD pts and their mothers	9-17 years	Child's perception of CHD on his life was influenced by over-estimation of disease severity by mothers and previous cardiac surgery.
Landtman (1973)	50 pts under-going surgery	5-12 years	Prior to discharge pts asked to draw a picture of their heart on admission and now (after surgery). Common feature was that the heart image had changed after surgery, generally in a positive direction.
Goldberg (1974)	26 pts with CHD 26 pts with burns	11-15 years	CHD pts ("invisible handicap") better psychological adjustment than burns ("visible handicap") pts.
Gabriel et al (1978)	50 CHD pts studied after surgery	2-19 years	No initial psychoses or delirium responses. 1 pt. had long-term "reaction". Pts of 8-11 years were the most emotionally stable, but in general responses in adequately prepared pts are relatively mild and of short duration.
Myers-Vando et al (1978)	12 CHD pts; 12 age matched healthy controls	8-16 years	Tests of cognitive development, illness causality and vulnerability to illness. Cardiac grp were functioning at lower levels of cognitive development than healthy grp. No differences between grps on illness causality. Vulnerability was less clear cut.

Zeltzer et al (1980)	345 healthy adolescents; 168 ill adolescents (including cardiac disease)		Cardiac adolescents reported more illness-related school disruption than healthy adolescents. They also reported less treatment related problems than other ill adolescents. Cardiac adolescents were more concerned about their sexuality than other ill adolescents.
Donovan et al (1983)	4 male CHD pts		Pts evaluated before and after completing one year cardiac and fitness programme. After programme pts reported improvement in feelings of adequacy and self esteem. Drawings indicated better self image, more positive investment in their body and an increase in positive feelings.
Baer et al (1984)	48 pts with corrected TOF	20-29 years	Pts corrected after 9 years of age were less venturesome and more insecure than those corrected earlier.
Heller et al (1985)	140 children with cardiovascular disorders, cleft lip and palate or hearing defects	4-13 years	Type of disorder and severity were associated with remaining or becoming maladjusted. Boys with cardiac problems and deaf girls were most likely to be affected. A direct relationship was found with severity.
Kong et al (1986)	29 CHD pts; 21 controls	6-16 years	CHD grp had a higher % of behaviour and social adjustment problems. Adjustment and behaviour problems in the child significantly related to maternal anxiety and guilt.
Fowler et al (1987)	62 CHD pts; 62 controls	6-18 years	School absenteeism in CHD pts predominantly linked to medical factors. Absence not related to any measures of social adaptation, self-esteem or academic achievement in either grp.
Alpern et al (1989)	30 CHD pts with pace-makers (PM) 30 CHD without pace-makers 30 controls	7-19 years	Tests of locus of control (LOCS), anxiety, IQ. No differences in IQ. PM grp had significantly more external LOCS than healthy but not non-PM grp. PM grp had greater knowledge of pacemakers, facilitating use of intellectualisation as a coping mechanism. Non-PM and healthy grp perceived significant differences (social and emotional) between PM grp and their peers, but PM grp did not view themselves as different from peers.
Kramer et al (1989)	128 CHD pts (77 with PH; 51 without PH). 89 controls	4-14.9 years	No differences in behaviour between 2 cardiac grps. Pts with PH had increased feelings of inferiority and anxiety and more impetuous than pts without PH and controls. PH grp not more dependent.
DeMaso et al (1990)	63 TGA pts; 77 TOF pts; 36 SR pts	5.5-6.3 years	TGA and TOF pts had poorer overall psychological functioning than SR grp. TGA and TOF pts had a higher % of CNS and IQ impairment. Strong association between CNS and IQ impairment and psychological functioning; Controlling for effects of impairment resulted in no differences between TGA/TOF and SR grps.
DeMaso et al (1991)	99 CHD pts and their mothers	4-10 years	Maternal perceptions of parenting skills and interaction with children were strong predictors of emotional adjustment in child. Illness severity less influential in adaptation than quality of mother-child relationship.
Brandhagen et al (1991)	168 adult CHD pts	24-42 years	Long term follow-up study. Psychological stress higher than for a normal population. Unrelated to severity of original cardiac defect. Psychological stress occurred even when educational achievement and occupational level indicated "success".
Weiss (1992)	24 infants with CHD	0-6 months	Investigated effects of verbal and tactile stimulation on arousal. Touch conducive to neural excitation produced significantly greater arousal than verbal stimuli or touch not conducive to neural excitation. Some infants showed physiological and behavioural distress. Such arousal may have life-threatening consequences for the health status of infants with CHD.
Attie-Aceves et al (1992)	Pts with CHD and controls		Investigation of differences in response to frustration and disintegration of body image. No differences between grps found; children with CHD have special "capabilities" that enable them to respond to illness in adaptive ways.
Utens et al (1992) (1993)	499 pts operated on for CHD	9-35 years	Follow-up study to assess quality of life after surgical correction of CHD in childhood. CHD pts had significantly more behaviour problems than healthy reference grps.
Wright & Nolan (1994)	29 pts with corrected TGA or TOF; 36 pts with cardiac murmurs	6-12 years	Study of school performance. Parental assessment of adjustment: TGA/TOF grp had significantly more cases of maladjustment than control grp. No significant differences on teacher ratings.
Casey et al (1994)	26 CY pts 26 normal controls		Physical data on quality of life after surgical palliation. Only 2 pts not at school. Parents under-estimated child's exercise tolerance in 80% of cases.
Casey et al (in press)	26 CY pts 26 normal controls	6-12 years	Study of behavioural adjustment indicated that CY pts were rated as being more withdrawn than controls by both parents and teachers. Family strain and exercise tolerance were strong predictors of teacher-rated school adjustment.

Table 2.1

Abbreviations
CHD = congenital heart disease
CY = cyanotic
ACY = acyanotic
ITU = intensive care unit
TGA = transposition of the great arteries
TOF = Tetralogy of Fallot
SR = spontaneous recovery
VSD = ventricular septal defect
PH = physical handicap

The infant with congenital heart disease, in common with other chronically sick babies, has difficulty in achieving normal emotional development (Garson & Baer, 1990). In the specific case of congenital heart disease, feeding difficulties in particular can disrupt the parent-child relationship (Loeffel, 1985; Lobo, 1992). Although studies of "adjustment" of infants with congenital heart disease are scarce, there are several descriptive accounts of care-giving problems and the difficulties parents have in establishing appropriate relationships with their infants (Gillon, 1972; Gudermuth, 1975; Pinelli, 1981). As the child gets older, emotional development can be hampered by parental attitudes of pampering and over-protection and by maternal anxiety (Cooper, 1959; Linde, Adams & Rozansky, 1971; Offord, Cross, Andrews & Aponte, 1972; Donovan, Mathews, Nixon, Stephenson, Robertson, Dean, Fricker, Beerman & Fischer, 1983; Bowen, 1985; Kong, Tay, Yip & Chay, 1986). Maladjustment in the child is related more to maternal variables than to disease severity (Linde, 1982; DeMaso, Campis, Wypij, Bertram, Lipshitz & Freed, 1991). A study of self-image found that children with congenital heart disease had a constricted body image compared with healthy children (Green & Levitt, 1962) but a more recent study did not find such differences and concluded that children with congenital heart disease have "special

capabilities" that enable them to respond to illness in adaptive ways (Attie-Aceves & Cardenas, 1992). As well as negative psychological outcome (Landtman & Valanne, 1959; Auer, Senturia, Shopper & Biddy, 1971; Garson, Williams & Reckless, 1974; Donovan, Fricker, Neches, Park, Mathews, Lenox & Zuberbuhler, 1979; Doucet, 1981; Kashani, Lababidi & Jones, 1982; Heller et al, 1985) a number of studies have reported more positive outcomes for the child with congenital heart disease (Chazan et al, 1951; Freed, Hastings & Cruickshank, 1953; Reed, 1959; Linde et al, 1966; Goldberg, 1974; O'Dougherty, 1981; Pelcovitz, DeMaso, Russo & Freed, 1984; Wright, Jarvis, Wannamaker & Cook, 1985). Children and adolescents with cyanotic heart disease have been reported to have poorer emotional adjustment (Garson et al, 1974; Donovan et al, 1979; Spurkland, Bjornstad, Lindberg & Seem, 1993; Wright & Nolan, 1994; Casey, Craig, Sykes, Power & Mulholland (in press)). In cyanotic children in whom the defect remains uncorrected a significant deterioration in adjustment over time has been reported (Linde et al, 1970). More recently, poor overall psychological functioning has been found to be linked not to the presence of a cyanotic lesion but rather to the presence of CNS impairment (DeMaso et al, 1990), an increased prevalence of which is found in children with cyanotic lesions (Silbert et al, 1969). The presence of physical handicap has also been identified as a risk factor for the development of "abnormal" personality characteristics (Steinhausen & Bruhn, 1980) and for increased feelings of anxiety and inferiority in patients with congenital heart disease (Kramer et al, 1989). Some children and adolescents have been reported to experience loneliness and social isolation because they feel more mature than their peers as a result of their experiences, whilst others have the same difficulties because of their childish behaviour, resulting from being over-protected and pampered (Utens, 1992). Adolescents with congenital heart disease are significantly more concerned about sexuality than are other ill adolescents and fears of

death during intercourse are particularly prevalent among teenage boys (Zeltzer, Kellerman, Ellenberg, Dash & Rigler, 1980). A study of 90 adolescents with congenital heart disease found that whilst they did not differ from their healthy peers in terms of overall self concept, those with more severe disease had lower self concepts (Uzark, VonBargen-Mazza & Messiter, 1989). Patients with congenital heart disease also reported greater perceived stress in comparison to peers. Adolescents with congenital heart disease have been found to lack knowledge about their medical condition (Ferencz, Wiegmann & Dunning, 1980) and about preventive health care and life-style implications of the cardiac diagnosis (Uzark, 1992). They also have a constant fear of sudden and early death (Donovan, 1985).

Long-term follow-up studies indicate that levels of psychological stress (Brandhagen, Feldt & Williams, 1991) difficulties in relationships with others (Utens, Verhulst, Meijboom, Duivenvoorden, Erdman, Bos, Roelandt & Hess, 1993) and dependency (Kokkonen & Paavilainen, 1992) are higher in patients with congenital heart disease compared with the normal population. Low self-esteem, insecurity and feelings of vulnerability have also been reported (Perloff & Marelli, 1992). Adults who had lesions corrected after 9 years of age were less venturesome and more insecure than those corrected at a younger age (Baer, Freedman & Garson, 1984). Adolescent and young adult males, in particular, have psychosocial difficulties (Donovan et al, 1979; Kokkonen & Paavilainen, 1992).

2.2.8.1. Cardiac Catheterisation

Cardiac catheterisation can have a detrimental effect on emotional adjustment (Sohni, Geiger & Schmidt-Redemann, 1987), particularly in pre-school and young school-aged patients (Aisenberg, Wolff, Rosenthal & Nadas, 1973). Different techniques of preparation, such as

puppet therapy (Cassell, 1965; Cassell & Paul, 1967), rehearsal (Naylor, Coates & Kan, 1984), stress management training (Campbell, Clark & Kirkpatrick, 1986) and filmed modelling (Bradlyn, Christoff, Sikora, O'Dell & Harris, 1986) have all been shown to have significant benefits, although no one technique has been shown to be significantly more beneficial than any other (Bradlyn et al, 1986). School-aged children undergoing echocardiography and cardiac catheterisation were found to predominantly exhibit orienting behaviours during the procedures, whereby they used verbal, visual, tactile and body movement to familiarise themselves with their surroundings, circumstances and what was happening to them (Youssef, 1981). The child's adjustment to hospital and the stress of catheterisation can have an important impact on the child's future (Uzark, 1978), particularly if further surgery is indicated.

2.2.8.2. Cardiac Surgery

Both children and adults have been reported to show grossly abnormal responses after heart surgery, particularly after procedures requiring the heart-lung machine (Egerton & Kay, 1964; Abram, 1971; Danilowicz & Gabriel, 1971; Kaplan, Achtel & Callison, 1974). In the immediate post-operative period, however, the incidence of extreme psychological reactions in children is low compared with psychotic reactions seen after cardiac surgery in adults (Kornfeld, Zimberg & Malm, 1965). Behavioural disturbance has been reported in approximately 10% of children in the immediate post-operative period (Danilowicz & Gabriel, 1971; Kaplan et al, 1974) but for the majority of children abnormal responses are mild and of short duration (Weinstein & Fitzgerald, 1976). As well as negative responses to hospitalisation and surgical intervention, positive responses have also been observed, such as an increase in self-confidence and social skills. Evaluation of reactions by age indicated

that children of 8-11 years of age tended to have the least number of negative reactions and the highest number of positive gains (Gabriel & Danilowicz, 1978). Children between 2 and 7 years of age demonstrated the highest number of adverse reactions and long term problems. A more recent study of pre-school aged children undergoing cardiac surgery demonstrated the benefits of using therapeutic play in the preparation of these children (Abbott, 1990) and the provision of coping skills training for pre-school and school-aged children prior to cardiac surgery has been found to result in better adjustment in hospital, at home and at school (Campbell, Kirkpatrick, Berry & Lambert, 1995). A further study of neuroendocrine and behavioural responses to psychological stress found increased neuroendocrine activity at times of particular stress (e.g. day before surgery), suggesting that children have insufficient psychological defenses to prevent such an increase in response to the threat of major surgery (Barnes, Kenny, Call & Reinhart, 1972).

In an assessment of coping behaviours of children hospitalised for cardiac surgery (Corbo-Richert, Caty & Barnes, 1993), direct action and information seeking were the predominant coping strategies utilised by the children. Younger children tended to seek comfort and emotional support more than school-aged children, whilst school-aged patients asked more questions. These findings were similar to those of other groups of hospitalised children (Caty, Ellerton & Ritchie, 1984) and the authors suggest that developmental level, rather than the specific medical intervention, is more influential in determining children's coping patterns.

A difficulty in interpreting a number of the studies on adjustment to congenital heart disease is that the samples are often not homogeneous with respect to whether the children have undergone cardiac surgery and if so, whether it was corrective. The few prospective studies

comparing pre- and post-operative psychological functioning have generally indicated improvement in adjustment after surgery (Landtman & Valanne, 1959; Landtman, 1973), particularly for children with cyanotic lesions (Linde et al, 1970).

In summary, children with congenital heart disease are at risk for emotional and behavioural difficulties, even when the lesion has been repaired. Follow-up of adults suggests that problems persist and highlights the importance of not only identifying risk factors for maladjustment but also of identifying adaptive responses. As surgery continues to offer greater medical benefits, prospective studies are needed to assess the psychological implications. Research on congenital heart disease and cardiac surgery is still focusing on the traditional approach of assessing maladjustment. However, in common with children with other chronic illnesses, the vast majority of children with congenital heart disease appear to cope with their disease and treatment and studies now need to look at how they cope.

2.2.9. Bone Marrow Transplantation

The procedure of BMT involves prolonged medical treatment over several months (see page 53) and a number of psychological stressors associated with the treatment have been identified. At each stage of the transplant different psychological demands are placed on the child and family.

2.2.9.1. Isolation

Isolation in a germ-free environment is a major stressor for BMT patients. The process of isolation involves elimination of skin-to-skin contact with other individuals, restriction of movement due to confinement to a relatively small area and generally increases the possibility

of both sensory deprivation and social isolation (Kellerman, Siegel & Rigler, 1980). Reported psychological reactions include anxiety, depression, sleep disturbance, inhibition of motor activity and the development of behaviours such as rhythmic rocking, and extreme dependency (Kohle, Simons, Weidlich, Dietrich & Durner, 1971; Simons, Kohle, Genscher & Dietrich, 1973; Kellerman, Rigler, McCue, Pospisil & Uno, 1976). Studies of children have found that feelings of entrapment (Patenaude, Szymanski & Rappeport, 1979) and fears of abandonment (Artinian, 1982) are heightened by the isolation process. In contrast, other researchers have concluded that long-term isolation does not necessarily result in adverse psychological consequences (Gordon, 1975; Drotar, Stern & Polmar, 1976; Freedman, Montgomery, Wilson, Bealmear & South, 1976; Lesko, 1994).

2.2.9.2. Physical Stressors

In common with other forms of hospital treatment, children are subjected to a number of unpleasant and distressing procedures during the work-up period and BMT itself. The experience of total body irradiation has been found to be particularly frightening, not only because of the requirement to spend several hours immobilised in a shielded room, but also because of the physical effects of the treatment (Patenaude et al, 1979). Children treated with cranial irradiation have been found to be at higher risk of subsequent behaviour problems compared with non-irradiated children (Deasy-Spinetta, Spinetta & Oxman, 1988). Anticipation of medical procedures intensifies feelings of anger, depression and anxiety (Gardner, August & Githens, 1977). Hair loss, weight loss, skin lesions and other physical effects engender negative feelings about appearance and intensify fears of dying, although it has also been suggested that, whilst children find their change in appearance distressing, they focus primarily on the associated discomfort and pain (Gardner et al, 1977). The

insertion of a Hickman catheter also alters body image, although self esteem does not appear to be adversely affected (Newman, Schnaper, Reed, deJongh & Schimpff, 1984).

2.2.9.3. Psychological Adjustment During the BMT Process

Prior to transplantation, children describe fears of life-threatening disease, anxiety about illness and death and feelings of helplessness and vulnerability (Gardner et al, 1977). Concerns about being a burden to their family are also expressed.

The majority of early papers on adjustment of children and adults to BMT itself were descriptive and tended to focus on the emotional and behavioural responses during the hospitalisation. Depression and anxiety were widely reported, together with increased dependency and regressive behaviour (Brown & Kelly, 1976; Gardner et al, 1977; Popkin, Moldow, Hall, Branda & Yarchoan, 1977; Gluckman, Alby, Devergie, Marty & Bernard, 1979; Kamphuis, 1979; Patenaude et al, 1979). The loss of autonomy, together with the isolation and uncertainty about the future, combine to engender feelings of anger, depression and fear (Gardner et al, 1977; Patenaude et al, 1979). Confining the child to a sterile room provides few constructive outlets for such negative feelings (Linn, Beardslee & Patenaude, 1986). Side effects of medication can result in gastro-intestinal problems and children may therefore be unwilling to eat or drink. The prevalence of eating problems is higher in chronically ill children (Garralda & Palanca, 1994) and it has also been suggested that children undergoing BMT refuse food in an attempt to maintain some control and mastery over their situation (Gardner et al, 1977; Patenaude et al, 1979). Although reactions vary according to developmental level, feelings of loss of control bridge the developmental spectrum (Abramovitz & Senner, 1995). In a study of compliance with the strict medical

regimen, 52% of patients exhibited significant adherence difficulties (Phipps & DeCuir-Whalley, 1990). The highest rates of non-compliance were found in pre-school and school-age children, with adolescents showing a lower frequency. These findings for adolescents contradict studies of other chronically ill adolescent groups (Korsch, Fine & Negrete, 1978; Tebbi, Cummings, Zevon, Smith, Richards & Mallon, 1986; Jacobson, Hauser, Lavori, Wolsdorf, Herskowitz, Milley, Bliss, Gelfand, Wertlieb & Stein, 1990; Johnson, Kelly, Henretta, Cunningham, Tomer & Silverstein, 1992). Phipps and DeCuir-Whalley suggested that, for adolescents, BMT totally denied them any control and autonomy, so they gave up altogether, exhibiting patterns of depression and withdrawal. In one of the few interventional approaches to helping young children cope with BMT, Linn et al (1986) demonstrated the efficacy of puppet therapy using mastery, expression and modelling techniques.

Children were found to identify closely with other patients on the ward, which could have both positive and negative repercussions (Patenaude & Rappeport, 1982). When both patients were treated successfully, identification was found to help reduce feelings of anxiety and isolation. However, the death of another patient resulted in increased fear and anxiety in the survivor about their own mortality and, in some instances, guilt about being the survivor.

A number of factors other than the treatment itself have been identified as influencing the way in which children react to BMT. Developmental level (Pot-Mees & Zeitlin, 1987; McConville, Steichen-Asch, Harris, Neudorf, Sambrano, Lampkin, Bailey, Fredrick, Hoffman & Woodman, 1990; Nespoli, Verri, Locatelli, Bertuggia, Taibi, & Burgio, 1995), medical state (Kamphuis, 1979; McConville et al, 1990), duration of the hospitalisation (Patenaude

et al, 1979), adequate parental support (Artinian, 1982) and trust in the staff (Popkin et al, 1977) are important considerations.

Despite the reported emotional reactions of children undergoing BMT, the incidence of frank psychiatric disorder is lower and less severe than might be expected (Gardner et al, 1977; Rait, Jacobsen, Lederberg & Holland, 1988). Adjustment disorders are often found to correlate with specific medical stressors and improve as the child's medical condition improves (McConville et al, 1990). Whilst psychopathology in the child has been reported as being mild to moderate, children who died or had unexpectedly high complication rates have been found to show more psychosocial distress patterns than those surviving up to one year after transplantation (McConville et al, 1990). It has, however, been suggested that symptoms such as depression, withdrawal and anorexia should be considered to be normal reactions to bone marrow transplantation (Gluckman et al, 1979).

2.2.9.4. Adjustment after Discharge

Discharge from hospital has been found to provoke feelings of ambivalence about leaving the safe environment of the transplant unit (Freund & Siegel, 1986). Children may have unrealistic expectations about their immediate return to normality (Atkins & Patenaude, 1987) and are often unprepared for any setbacks (Freund & Siegel, 1986; Patenaude et al, 1979). A study of BMT recipients 12 months post-transplant found that they exhibited symptoms of post-traumatic stress-disorder, with denial and avoidance particularly prevalent (Stuber, Nader, Yasuda, Pynoos & Cohen, 1991). The authors also suggest that the symptoms observed are unlikely to be unique to children undergoing BMT - rather, it is probable that children experiencing similar medical trauma will also demonstrate a similar

pattern of symptoms. Longer-term adjustment to BMT in children has not been systematically studied. However, a study of adult survivors found that, whilst 75% of recipients were doing well from a psychosocial perspective, 15-25% reported significant emotional distress, low self-esteem and less than optimal life satisfaction (Wolcott, Wellisch, Fawzy & Landsverk, 1986(a)).

2.3. QUALITY OF LIFE

2.3.1. What is Quality of Life?

Quality of life is a term to which reference is often made, but its multiple definitions and meanings and subsequent multiple applications have resulted in controversy and confusion in the literature.

As a general concept, quality of life has been applied to the healthy as well as to the physically and/or mentally ill. Measurement of quality of life has been undertaken with respect to the general population, particularly in America, and a number of studies have been published attempting to identify the major factors affecting the quality of life of adult Americans (Flanagan, 1982). "Happiness" and "satisfaction" have been equated with quality of life in a number of studies (Campbell, Converse & Rodgers, 1975).

2.3.2. Quality of Life Applied to Illness and Health

A World Health Organisation publication in 1947 introduced a broadened definition of health as, " a state of complete physical, mental and social wellbeing and not merely the absence of disease" (World Health Organisation Constitution, 1947), thereby equating health and quality of life. Contrary to popular belief, health is not always of prime importance to the individual,

even for those with a "health problem". In studies evaluating the impact of a specific disease or treatment, the relevance of health parameters to the quality of life of the individual must therefore be determined. In the literature on chronic illness quality of life is frequently, but incorrectly, deemed to be synonymous with health status, which refers to social, physical and psychological wellbeing. Measurement of "health status" is often a more accurate description of the content of some tools used to purportedly assess quality of life.

Quality of life is a subjective, multi-dimensional concept. There is no consensus regarding its definition. Quality of life has been defined as, "the degree of satisfaction with perceived present life circumstances" (Young & Longman, 1983) - a nebulous concept but one which is nevertheless fundamental in everybody's life, irrespective of their individual circumstances. There is an increasing recognition that in evaluating the impact of illness or treatment, single outcome measures are inadequate to quantify such a multi-dimensional concept. In addition to objective measures, the importance of an individual's perceptions is also now being acknowledged. Despite the extensive range of instruments currently employed to assess quality of life, there are similarities in the content of the measures (Fitzpatrick, Fletcher, Gore, Jones, Spiegelhalter & Cox, 1992). The facets most commonly assessed are shown in Table 2.2.

The shared aim of all quality of life measures is to try to quantify the impact of illness and/or treatment on (some aspect of) an individual's daily life. Applications range from measuring the impact of specific illnesses and interventions generally (Goodinson & Singleton, 1989) to detailed health economic assessments which aim to determine the relative costs of illnesses and interventions e.g. quality of life adjusted years (Kind, Rosser & Williams, 1982).

FACETS OF QUALITY OF LIFE IN THOSE WITH ILLNESS
Physical function - e.g. mobility, self-care
Emotional function - e.g. anxiety, depression
Social function - e.g. social support, intimacy
Role performance - e.g. job, housework, school
Pain
Other symptoms - e.g. disease specific symptoms, fatigue, nausea

Table 2.2

2.3.3. Measurement of Quality of Life

2.3.3.1. Assessment Tools

A wide range of objective and subjective criteria have been used to assess quality of life and in a review of 69 empirically- based studies from 1980 to 1984, 83 different measures of quality of life parameters were found to have been utilised (Hollandsworth, 1988). An earlier review of studies carried out between 1975 and 1979 found there to be an emphasis on objective indicators (Najman and Levine, 1981).

The vast majority of quality of life measures are either questionnaires - e.g. personality inventories, self-esteem scales and measures of anxiety and depression - or analogue scales representing a continuum on which the patient rates themselves or is rated by an informant on a particular parameter. One disadvantage of a scoring system using a linear numerical range is the definition of the end points - e.g. on a health status scale the criteria of "death" and "perfect health" may be selected to define the poles, thereby making the basic

assumption that death is the worst possible outcome. Such scales take no account of the fact that severe disability, coma etc. may be regarded by some as worse than death. In many of the early quality of life studies survival, or recurrence-free survival, and death were the limited end points commonly used. In addition, early quality of life measures tended to rely on a single score but latterly instruments have reflected the multi-dimensional nature of "quality of life".

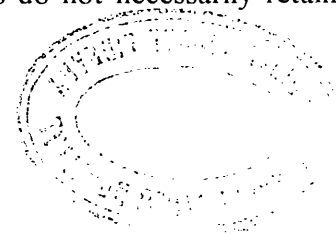
There are two basic categories of assessment tool - disease specific and generic. Disease specific instruments are those which have been specifically designed for one disease or narrow range of diseases (e.g. Guyatt, Berman, Townsend, Pugsley & Chambers, 1987; Jacobson, Barofsky, Cleary & Rand, 1988; Parfrey, Vavasour, Bullock, Henry, Harnett & Gault, 1989). They have the advantage of often being more acceptable to patients because they only deal with relevant factors, but they do not allow comparisons to be made between different disease groups. Generic tools are applicable to a wide range of health problems - e.g. the Nottingham Health Profile (Hunt, McEwen & McKenna, 1986) - and facilitate comparisons among different disease groups. Another form of generic instrument is health indices, in which an individual is rated on a continuum and a single score is given - e.g. the Quality of Well-Being Scale (Kaplan, Bush & Berry, 1976; Kaplan & Bush, 1982). Use of a single score has limitations in terms of clinical applicability, unless the score can be broken down into components. Health profiles, on the other hand, provide individual scores for different dimensions and where appropriate these can be summed to give a single aggregate score. A compromise between disease specific and generic instruments is to use relevant dimension specific instruments (Fletcher, Gore, Jones, Fitzpatrick, Spiegelhalter & Cox,

1992), such as the Profile of Moods States (McNair, Lorr & Droppleman, 1971), which evaluate a specific area such as psychological wellbeing.

A further problem with quality of life scoring systems is that they are limited by the fact that they describe the health of the individual at one point in time. If the test is reapplied after a short time, reliability may be over-rated because of memory effects. If a longer time elapses reliability may be under-rated because of real changes in actual health status (Nelson, Landgraf, Hays, Wasson & Kirk, 1990).

The design of studies and methods of sample selection are also important, with many studies being hampered by poor design and inadequate methods of assessment (O'Young & McPeek, 1987). The majority of studies tend to be based on single assessments with no control groups, with samples being selected either on the basis of consecutive patients accepted for treatment or in terms of all those who have survived following a particular treatment. However, there is an increasing awareness of the importance of randomised clinical trials for assessing the impact of specific medical treatments on quality of life, although the ethical implications of randomly assigning treatments - particularly those which are potentially life-saving - are complex.

A recent criticism of quality of life indicators has focused on the fact that they impose an external value system on patients by virtue of their extrapolation from group data (O'Boyle, 1992). The relevance of such measures to the individual is questioned - not only because the assessment tools may be too broad and not represent specific goals important to the individual, but also because similar behaviours/goals do not necessarily retain the same



significance for an individual over time. To meet these perceived short-comings, a schedule for the evaluation of individual quality of life was devised (SEIQoL), in which patients elicited their own individual cues which they judged to be most important to their overall quality of life and assigned scores representative of their current status on each of these constructs (McGee, O'Boyle, Hickey, O'Malley & Joyce, 1991; O'Boyle, 1994). O'Boyle and colleagues used this technique prospectively, in conjunction with traditional measures of health status, with patients undergoing hip replacement (O'Boyle, McGee, Hickey, O'Malley & Joyce, 1992). They found that the SEIQoL demonstrated the very individual nature of quality of life, reflected in the variety of areas nominated as important by individual patients, the differences in significance attached to them and the changes that occurred post-operatively. Importantly, they also found that standardised measures were insensitive to change over time. They also emphasised the importance of recognising that level of functioning in a particular area and the significance attached to that area can be independent of one another and can change independently. Whilst administration of such a measure is more time consuming than the traditional questionnaire or standardised visual analogue scale, it is applicable to all patients and is not disease or culture specific.

2.3.3.2. Who Should Assess Quality of Life?

A number of studies purporting to assess the quality of life of adults have focused exclusively on medical and physiological parameters (e.g. Bentdal, Fauchald, Brekke, Holdaas & Hartmann, 1991), but increasingly researchers are becoming aware of the importance of patients' own perceptions and feelings as a factor in evaluating therapeutic success. It is now also recognised that the most appropriate source of information on quality of life is the patient themselves (Slevin, Plant, Lynch, Drinkwater & Gregory, 1988; Aaronson, 1991),

although a number of studies have utilised medical and nursing staff to evaluate quality of life parameters in their patients (e.g. McClellan, Anson, Birkeli & Tuttle, 1991) with some measures being specifically designed for completion by physicians (Spitzer, Dobson, Hall, Chesterman, Levi, Shepherd, Battista & Catchlove, 1981). Whilst assessment by professionals is often very efficient in terms of time, low levels of agreement have been found between doctors' and patients' ratings of the patient's emotional and social functioning (Pearlman & Uhlmann, 1988; Slevin et al, 1988). Other studies have used parents, spouses or other significant people, but it has been shown that patient-informant agreement on measures of functional status and social activity depends on the degree of involvement of the informant with the patient (Epstein, Hall, Tognetti, Son & Conant, 1989). In some instances quality of life has been assessed without the patient's involvement - particularly if their physical and/or mental state precludes their cooperation.

2.3.4. Quality of Life of Children

The adult literature on quality of life has generated considerable debate and controversy but there are still relatively few studies of quality of life in children. However, quality of life is being used with increasing frequency as an indicator of the success of a particular intervention in childhood and researchers are now recognising its importance in the total care of sick children (Glaser & Walker, 1994). Despite this, many studies have tended to focus on long-term implications of childhood illness from an adult perspective (e.g. Makipernaa, 1989; Lannering, Marky, Lundberg & Olsson, 1990; Almond, Morel, Matas, Gillingham, Chau, Brown, Kashtan, Mauer, Chavers, Nevins, Dunn, Sutherland, Payne & Najarian, 1991; Morel, Almond, Matas, Gillingham, Chau, Brown, Kashtan, Mauer, Chavers,

Nevins, Dunn, Sutherland, Payne & Najarian, 1991), with very few attempting to look at quality of life parameters for the patient during childhood.

The lack of comprehensive standardised measures for the evaluation of quality of life in children makes it necessary to use parameters such as development and cognition, behaviour, schooling, and self esteem as indicators, and to use a composite set of variables rather than single factors in isolation. Use of this type of global assessment does address the issue of quality of life and the outcome for the child as a whole and ideally should be used prospectively (Lancet, 1992). Few studies are prospective, however - rather the tendency is to try and evaluate quality of life in a group of children with a particular disease who have undergone a particular treatment regimen or to make a single assessment at one point in time. There are even fewer controlled studies in which the effects of different treatments and/or diseases are compared. Whilst a number of studies set out to evaluate quality of life, few authors attempt to define their interpretation of the concept, thereby making it difficult to adequately assess the implications of their results and to make comparisons between published data from different sources.

Whilst the importance of evaluating individual quality of life of adults is rapidly gaining credence, it is debatable whether this is as crucial for children. Throughout childhood values and life-styles tend to be more standardised and uniform. Children are not yet necessarily evaluating different attributes of everyday life and assigning their own values of significance to them, so it is probable that well validated and reliable measures will be applicable and meaningful in a far more general way for a paediatric, rather than an adult, population. In contrast, developmental change makes it difficult to apply any single measure to all age

groups and a unitary concept of childhood quality of life is probably not possible (Rosenbaum, Cadman & Kirpalani, 1990). This is particularly true for the pre-school age group because of the rapid rate of change in normal children. In studies of sick children, their understanding of health and illness concepts are also necessary considerations. The difficulty of developing appropriate standardised tools for children in a broad age spectrum means that specific measures need to be designed for specific developmental levels, a method currently being adopted for children of 7 to 12 years with asthma (Christie, French, Weatherstone & West, 1991) and chronic illness in general (Neff & Dale, 1990). This approach, however, makes it difficult to assess changes over time for a particular individual or group of patients, especially those crossing developmental level boundaries.

Who should provide information concerning quality of life of the paediatric patient is open to debate - at what age do children become a reliable source of information and less likely to be influenced by their parents' perceptions? It has been suggested that children themselves may not be reliable informants (Lehmann, Bendebba & De Angelis, 1990; Falconer, Oldman & Helms, 1993; Schwab-Stone, Fallon, Briggs & Crowther, 1994) and in some studies practical problems have prohibited direct questioning of the child themselves (Gerard, 1990). A number of studies have therefore used parents, physicians and/or teachers as principal informants (e.g. Ferreira, 1993). However, the lack of agreement between different informants is evident in paediatric studies as in those of adults (Cadman, Goldsmith & Bashim, 1984) and in areas such as the reporting of life events, uneven agreement between child and parent reporting has been found (Bailey & Garralda, 1990). It has been suggested that response bias may be a particular problem when parents are the source of information about their children, due to their tendency to respond in a socially desirable manner (Eisen,

Ware, Donald & Brook, 1979). Discrepancies also exist between what the informant and child perceive to be important in the context of quality of life (Neff & Dale, 1990). Whilst correlations of children's self-reports of functional status and those of parents may be good for some aspects of quality of life, such as impact of illness on independence (Pantell & Lewis, 1987), other areas have been found to be poorly correlated (Achenbach, McConaughy & Howell, 1987; Pantell & Lewis, 1987), demonstrating the need for multiple sources of information in children's quality of life measures. Studies in which there are no data provided by the child themselves must be questionable as to the accuracy of their interpretation of the child's quality of life.

2.3.4.1. Paediatric and Adolescent Quality of Life Studies

A review of the brief literature in this area indicates a predominance of studies with paediatric oncology patients (e.g. Kun, Mulhern & Crisco, 1983; Lannering et al, 1990; Chang, 1991; Bradlyn, Harris, Warner, Ritchey & Zaboy, 1993; Olson, Boyle, Evans & Zug, 1993). Whilst some investigations have looked exclusively at medical parameters (e.g. Sanders, Sullivan, Witherspoon, Doney, Anasetti, Beatty & Petersen, 1989; Lozano, Rovirosa, Reig & Salva, 1990) researchers are increasingly incorporating psychological and social factors into their methodology (e.g. Poznanski, Miller, Salguero & Kelsh, 1978; Lansky, List, Lansky, Cohen & Sinks, 1985; Lindstrom & Kohler, 1991; Townsend, Feeny, Guyatt, Furlong, Seip & Dolovich, 1991; Feeny, Furlong, Barr, Torrance, Rosenbaum & Weitzman, 1992). There have been some attempts to design specific measures of quality of life and these tend to be disease specific rather than generic (e.g. Ditesheim & Templeton, 1987). There is also a tendency for researchers to modify existing adult measures for use with children and adolescents (e.g. Ingersoll & Marrero, 1991; Orenstein & Kaplan, 1991), although this

approach rarely results in measures suitable for younger age groups. Due to the difficulties inherent in measuring a multi-dimensional concept such as quality of life in children, and the argument, put forward by Schipper, that there is a reluctance to recognise the potential significance of "soft", as well as "hard", data (Schipper, 1983), IQ has been frequently used as an indicator of quality of life, particularly in retrospective studies of brain tumours or the effects of cranial irradiation (e.g. Balestrini, Zanette, Micheli, Fornari, Solero & Broggi, 1990). Whilst these studies provide data about cognitive function following treatment, they cannot really be said to provide information about "quality of life" - a criticism which can be levelled at a number of paediatric studies. Other researchers have used standardised measures of behaviour, in conjunction with parameters such as survival and complication rates, as quality of life tools (e.g. Chin, Shepherd, Cleghorn, Patrick, Javorsky, Frangoulis, Ong, Balderson, Koido, Matsunami, Lynch & Strong, 1991), although these too are often not eliciting global measures of quality of life. Another focus has been to assess "health outcomes", utilising a broad definition of health encompassing domains such as longevity, disease, comfort, perceived well-being, activity, achievement and resilience (Vivier, Bernier & Starfield, 1994). In their review of recent studies, Vivier et al (1994) found that most researchers focused on a small subset of health concerns, although increasingly the importance of multiple health measures and multi-dimensional instruments is being recognised and such tools used (e.g. Barr, Furlong, Dawson, Whitton, Strautmanis, Pai, Feeny & Torrance, 1993; McCormick, Brooks-Gunn, Workman-Daniels & Peckham, 1993; Starfield, Bergner, Ensminger, Riley, Ryan, Green, McGauhey, Skinner & Kim, 1993).

2.3.5. Quality of Life and the Paediatric Cardiology Patient

There are very few relevant studies in this area. A study purporting to look at the quality of life of the adolescent cardiac patient (Ferencz, 1974) actually studied males of 23 - 27 years and assessed variables such as marital status, smoking, criminal involvement and employment, together with their recollections of cardiac catheterisation and/or cardiac surgery. There was no control group, making it difficult to assess the possible effects of the heart disease on adult functioning, and it would seem that the study did not really measure quality of life in adolescent cardiac patients.

Whilst other researchers have looked at variables such as IQ, personality and emotional development in the congenital heart disease patient, no systematic attempts to evaluate quality of life in this group appear to be forthcoming. A recent study of "quality of life" in patients undergoing palliative surgery for complex cardiac lesions reported physical parameters (Casey, Craig & Mulholland, 1994) and parent- and teacher-rated behavioural adjustment (Casey et al (in press)), but no data were provided by the patients themselves. With the increasing use of heart transplantation as a treatment for children with end-stage heart disease, quality of life is now being recognised as an important consideration for the paediatric patient (Parness & Nadas, 1988; Radley-Smith, 1989; Fricker, Trento & Griffith, 1990) although the studies tend to be retrospective, based on small sample sizes and lacking control groups. Small patient numbers have largely precluded looking at transplant patients according to their initial diagnosis, but a recent study suggested that children with an initial diagnosis of congenital heart disease performed less well on cognitive and behavioural parameters 12 months after transplantation compared with children undergoing transplantation for acquired heart disease or cystic fibrosis (Wray, Radley-Smith & Yacoub,

1993). Other retrospective studies have suggested that transplant patients demonstrate positive adjustment to the surgical intervention and an ability to return to age-appropriate activities in most cases (Pennington, Sarafian & Swartz, 1985; Fricker, Griffith, Hardesty, Trento, Gold, Schmeltz, Beerman, Fischer, Mathews, Neches, Park, Zuberbuhler, Lenox & Bahnson, 1987; Lawrence & Fricker, 1987; Pahl, Fricker, Trento, Griffith, Hardesty, Gold, Lawrence, Beerman, Fischer & Neches, 1988) and a prospective study indicated early post-operative improvement in behaviour following transplantation (Wray, Radley-Smith & Yacoub, 1992).

2.3.6. Definition of Quality of Life in This Study

From the literature, global measures comprising assessments of cognitive, emotional and sensory functioning, in which individual factors are looked at in combination, are the most appropriate method of assessing the concept of quality of life in children, within the paradigm of a prospective, controlled study. A composite set of variables, including measures of development and cognition, behaviour at home and at school and various parameters related to the disease and its treatment will be used to give a global assessment of quality of life. Two further indicators, in which the child provides their own "standard", will also be used. The first is achievement on academic parameters, measuring school performance relative to a child's own IQ. The second is a global self perception measure, which evaluates children's assessments of themselves on a number of constructs relative to their perception of their ideal self on the same constructs.

CHAPTER 3

ADJUSTMENT OF PARENTS AND SIBLINGS

Research on the adjustment of parents and siblings will be presented in the context of the general literature on chronic illness and then specifically in the areas of congenital heart disease and bone marrow transplantation.

3.1. PARENTS

3.1.1. Introduction

One of the most stressful experiences for any family is caring for a child with a chronic illness (Bouma & Schweitzer, 1990). Illness of a child has been shown to disturb the family equilibrium (Gillon, 1972) and the effects of chronic illness on the family can be manifested in many spheres - such as financial, social and behavioural (Sabbeth, 1984). There is a huge literature on parents' - particularly mothers' - reactions to chronic illness and increasingly the role of fathers is being recognised, together with the importance of considering the father-child relationship and the mother-father-child triad (Bristol & Gallagher, 1986; Parke, 1986). Much of the early work took a deficit-centred approach. Increasingly, however, researchers are acknowledging the adaptive responses of parents, rather than assuming that chronic illness is inevitably associated with maladjustment. The focus now is on identifying factors which are influential in determining parents' responses (positive as well as negative) to chronic illness. Table 3.1 outlines the main areas which have been studied.

3.1.2. Mental Health

Mothers of children with chronic disease have been found to be more depressed and anxious than mothers of healthy children (McCrae, Cull, Burton & Dodge 1973; MacCarthy, 1975;

Breslau, Staruch & Mortimer, 1982; Cadman, Rosenbaum, Boyle & Offord, 1991; Fisman & Wolf, 1991), although variability in the degree of reported problems suggests that other factors may also be influential (Wallander, Varni, Babani, DeHaan, Wilcox & Banis, 1989).

PARENTAL RESPONSE TO CHRONIC ILLNESS: MAIN AREAS OF INVESTIGATION
Mental health
Adjustment
Marital relationship
Coping styles
Perception of stresses associated with the illness
Effect of diagnosis
Time since diagnosis
Characteristics of the disease
Age of the child
Perception of illness severity
Social support
Financial/employment factors
Care-taking responsibilities
Impact of illness on parenting and the parent-child relationship
Compliance with treatment
Personality
Locus of control

Table 3.1

Fathers have lower levels of anxiety and depression than mothers (Walker, Thomas & Russell, 1971; Cadman et al, 1991) but when mothers report high levels of disease related stress, fathers' mental health is worse (Nagy & Ungerer, 1990).

3.1.3. Adjustment

A number of variables have been identified which influence maternal adjustment (Varni & Wallander, 1988; Wallander et al, 1989). Varni and Wallander (1988) classified these into risk and resistance factors, covering areas such as temperament and coping of the mother, family environment, maternal perception of stress, disease characteristics and temperament of the child. This multi-dimensional, dynamic approach assumes that the variables are inter-dependent and it emphasises adaptation rather than maladjustment. Although the spectrum of variables incorporated into such an approach is fairly comprehensive, it is still not clear how the variables influence adaptation or the mechanisms of the underlying processes.

3.1.4. Marital Relationship

The stresses of caring for a sick child can have significant consequences for the parents' own relationship. A number of ways in which this relationship can be undermined have been identified, including the physical and mental demands of care-taking and striving to maintain family integrity. It has frequently been suggested that divorce rates are higher for such families compared with the normal population, although reported results have been inconsistent. Whilst increased divorce rates have been cited in families of sick children (e.g. Tew, Payne & Laurence, 1974) other studies have not found elevated rates of divorce (e.g. Lansky, Cairns, Hassanein, Wehr & Lowman, 1978; Finley, Putherbough, Cook, Netley & Rowe, 1979; Silbert, Newburger & Fyler, 1982; Perrin & MacLean, 1988). Pre-existing

marital problems may be accentuated by the presence of chronic illness (Allan, Townley & Phelan, 1974).

Levels of marital distress have also been studied, with far more consistent results indicating significantly higher distress levels than in healthy control families, and in mothers compared with fathers (e.g. Crain, Sussman & Weil, 1966; Tew et al, 1974; Gath, 1977; Lansky et al, 1978; Cairns & Lansky, 1980; Gordon Walker, Manion, Cloutier & Johnson, 1992; Dahlquist, Czyzewski, Copeland, Jones, Taub & Vaughan, 1993). In contrast, some studies have shown that coping with disease can have positive effects and actually bring parents closer together (Pless & Satterwhite, 1975(a); Vance, Fazan, Satterwhite & Pless, 1980; Barbarin, Hughes & Chesler, 1985). The relationship between marital distress and divorce has not been studied systematically and a number of pertinent areas - such as marital communication - have been largely neglected. Research has been characterised by the lack of a theoretical framework (Sabbeth & Leventhal, 1984).

A study of parents of children with cancer reported a higher degree of marital satisfaction in those whose children had been diagnosed for less than three years compared with those parents whose children had been diagnosed for more than three years (Barbarin et al, 1985), indicating not only the importance of time since diagnosis, but also the dynamic aspects of parents' response to illness.

3.1.5. Coping Styles

Different parental strategies for coping with illness have been identified (McCubbin, McCubbin, Patterson, Cauble, Wilson & Warwick, 1983; Shapiro, 1983) and the implications

of these have been studied, not only for the parents but also for the children (Sanger, Copeland & Davidson, 1991). In this latter study, children with most difficulties came from families in which parents reported fewer coping strategies. Persistently poor maternal adjustment has been found to be associated with more use of palliative, rather than adaptive, coping mechanisms (Thompson, Gil, Gustafson, George, Keith, Spock & Kinney, 1994). The extent to which different coping strategies are helpful has been found to be dependent on the child's specific disease (Eiser & Havermans, 1992). These authors also found that perception of disease related problems influenced the degree of use of different coping mechanisms and that there were differences between mothers and fathers. Mothers who perceived more difficulties utilised social support more extensively, whilst fathers endorsed strategies related to autonomy.

Parental perception of the stresses associated with the illness has been suggested to explain some of the contradictory findings of adaptation to the illness (Wallander et al, 1989). Consistently, parents of ill children report more stresses than parents of healthy children (Holroyd & Guthrie, 1986; Hauenstein, Marvin, Snyder & Clarke, 1989; Goldberg, Morris, Simmons, Fowler & Levison, 1990; Phipps & Drotar, 1990; Singhi, Goyal, Pershad, Singhi & Walia, 1990), with mothers responding in a more emotional way than fathers.

3.1.6. Effects of Diagnosis

Diagnosis of chronic illness is commonly associated with psychological distress in the parents. With the birth of an infant with a congenital anomaly, attention has focused on parental mourning for the loss of a perfect child (Solnit & Stark, 1961) and the resulting reduction in self esteem (Gordeuk, 1976) and the reactions of shock, denial, guilt, anger, inadequacy,

sorrow and depression (Gordeuk, 1976; Waechter, 1977; Fost, 1981; McKeever, 1981; Collins-Moore, 1984). The same reactions have also been reported in parents of children diagnosed at a later age (e.g. Koski, 1969). Following these reactions, gradual adaptation - characterised by a lessening of intense anxiety and emotional reactions - has been reported (Drotar, Baskiewicz, Irvin, Kennell & Klaus 1975). Recognition is now being given to the differences in reaction to terminal illness and chronic illness. Whilst families facing terminal illness go through fairly predictable stages of grief, those dealing with chronic illness go through recurrent and cyclical stages (Worthington, 1989), often with no apparent end-point (Collins-Moore, 1984).

3.1.7. Time Since Diagnosis

Responses to illness have been found to alter with time (Venters, 1981; Ungerer et al, 1988). The time of diagnosis (Kupst & Schulman, 1980; Kupst, Schulman, Honig, Maurer, Morgan & Fochtman, 1982; Kovacs, Finkelstein, Feinberg, Crouse-Novak, Paulauskas & Pollock, 1985) and episodes of deterioration in the child's health (Kupst, 1992) are particularly stressful. Increased time since diagnosis was found to be associated with more distorted maternal perceptions of illness severity (Offord & Aponte, 1967).

3.1.8. Disease Characteristics

A number of disease characteristics and their effects on parents' response to illness and parenting strategies have been studied, such as visibility of the condition, disease severity, involvement of the central nervous system and prognosis (e.g. Steinhauer, Mushin & Rae-Grant, 1974; Dolgin, Phipps, Harow & Zeltzer, 1990; Saddler, Hillman & Benjamins, 1993). There are relatively few comparative studies assessing the impact on parents of different

chronic illnesses, but CNS involvement has been linked to more adjustment difficulties. No consistent relationship between disease severity and adjustment has been found.

3.1.9. Age of the Child

Care of pre-schoolers and adolescents can be associated with particular difficulties, which may be exacerbated by illness. A study of mothers of children with cystic fibrosis found that mothers of children in these two age groups had higher levels of depression than mothers of healthy children, but for parents of children in other age groups depression levels did not differ from those of parents of healthy children (Walker, Ford & Donald, 1987).

3.1.10. Perception of Disease Severity

Whilst there is little evidence that objective measures of medical severity are predictive of parents' adjustment (Wallander et al, 1989; Mullins, Olson, Reyes, Bernardy, Huszti & Volk, 1991) parents' perceptions of disease severity have been found to be influential in determining parental anxiety, coping and their behaviour towards their sick child (Bergman & Stamm, 1967; Linde et al, 1970; Cayler et al, 1973; Parcel, Gilman, Nader & Bunce, 1979; Berenbaum & Hatcher, 1992). However, the relationship between perception of disease severity and adjustment is not always direct - in some instances parents who perceived their child's illness to be less severe than it actually was had greater problems than those parents who accurately perceived the severity of their child's disease (Frydman, 1980). Furthermore, mothers and fathers differ in their perception of disease severity (Eiser, Eiser, Town & Tripp, 1991(b)). Parental perception of child health is also a major predictor of the impact of a child's condition on the family (McCormick, Charney & Stemmler, 1986).

3.1.11. Social Support

There are many definitions of social support, ranging from the actual size or density of the social networks to parents' perceptions of the support they receive. Larger, less dense networks are, in general, associated with less stress (Hirsch, 1980; Kazak & Wilcox, 1984; Kazak, Reber & Carter, 1988) but in families with a chronically ill child, extended family members are reported to be less helpful and to provide less emotional support than in families with healthy children. Satisfaction with received social support is also related to maternal adjustment (Barakat & Linney, 1992). Lack of perceived social support is associated with a risk for psychological distress in parents (Quittner, 1992; Speechley & Noh, 1992) and adjustment problems in the sick child (Hamlett, Pellegrini & Katz, 1992). Mothers receiving significant support from their husbands report better mental health. However, it is not the amount of help offered by fathers but rather the value they attach to child rearing activities that is important to mothers' mental health (Nagy & Ungerer, 1990). Mothers value social support more highly than fathers (Eiser & Havermans, 1992) whilst fathers tend to withdraw socially from others (Hobfoll, 1991).

3.1.12. Employment and Financial Concerns

Chronic illness can result in significant extra expenses through the need for special equipment, trips to hospital etc. and cause financial worries for parents (Lansky, Cairns, Clark, Lowman, Miller & Trueworthy, 1979). Mothers' opportunities to take on employment are decreased due to the demands of caring for a sick child (Breslau et al, 1982; Cowen, Corey, Keenan, Simmons, Arndt & Levison, 1985), although for those who do work, levels of depression are lower than for those at home (Walker, Ortiz-Valdes & Newbrough, 1989). Fathers may be limited in promotion opportunities, particularly if they are unwilling to go

away from the family (Tiller, Ekert & Rickards, 1977) or the family is unable to move due to the child's illness (McKeever, 1981).

3.1.13. Care-Taking Responsibilities

Mothers have been found to take more responsibility for caring for the sick child (Barbarin et al, 1985; Cowen et al, 1985; Nagy & Ungerer, 1990; Eiser, Eiser, Town & Tripp, 1991(a); Havermans & Eiser, 1991), although such a pattern of care is also more prevalent in families of healthy children. The burden of caring for an ill child can result in feelings of isolation and loneliness for the mothers (Walker et al, 1971; Cook, 1984). Fathers, who often take on more general responsibilities (Klein & Simmons, 1979), also feel isolated - but from the ill child (Cook, 1984).

3.1.14. Parenting

Early work focused on the observations of restrictive and overprotective parenting in response to the child's illness and a number of studies reported that parents had difficulties with discipline (e.g. Wasserman, Thompson, Wilimas & Fairclough, 1987). However, the view that chronic childhood illness compromises normal parenting practices is not supported by empirical work (Eiser et al, 1991(a); Eiser, 1993), although rearing a child with a life-threatening illness is acknowledged to make extra demands on parenting skills (Van Dongen-Melman & Sanders-Woudstra, 1986(a)). Empirical work has identified few significant differences between parents of sick and healthy children. Areas of difference which have been found include the greater concern expressed by parents of sick children about their child's physical health (Davies, Noll, DeStefano, Bukowski & Kulkarni, 1991; Eiser et al, 1991(a)), mothers' greater anxiety about being too involved with their sick child (Davies et

al, 1991) and less limit setting by mothers of chronically ill compared with healthy children (Ievers, Drotar, Dahms, Doershuk & Stern, 1994). Furthermore, children's misbehaviour is more likely to be excused by parents when children exhibit symptoms of physical illness, particularly if associated with a medical diagnosis (Whitt, 1984; Walker, Garber & Van Slyke, 1995). Many of the general findings about parenting - such as depressed mothers reporting more behaviour problems in their child (Lancaster, Prior & Adler, 1989) and mothers experiencing a large number of life events reporting higher rates of child rearing problems (Beautrais, Fergusson & Shannon, 1982) - are equally applicable to the parents of chronically ill children. One contradictory finding was that for children with a chronic illness (cystic fibrosis) there was a positive correlation between poor psychosocial functioning in the child and parental over-protection, whereas for healthy children poor psychosocial functioning was associated with a lack of parental care (Capelli, McGrath, MacDonald, Katsanis & Lascelles, 1989).

Much of this research has focused exclusively on two parent families, but single parents have reported more stress than those from two parent families (DeMaso et al, 1991). Although some studies have found chronically ill children from one parent families to be at some disadvantage (Christiaanse, Lavigne & Lerner, 1989), others have found good adjustment in children from one parent families and better adherence to treatment regimens compared with children from two parent families (Hanson, Henggeler, Rodrigue, Burghen & Murphy, 1988).

The effects of personality characteristics and perceived locus of control on parenting have also been studied. Mothers with a more internal locus of control have reported better

adjustment in their children (DeMaso et al, 1991). Personality has been found to influence attitudes to parenting, irrespective of the health status of the child (Boll, Dimino & Mattsson, 1978). Some medical aspects of the child's care are also related to parenting. Non-compliance in paediatric (rather than adolescent) populations is almost totally attributable to parental behaviour (Eiser, 1993). For older children and adolescents, inconsistent discipline practices, lack of organisation, inadequate family functioning, family conflict and lack of agreement between parents and children have been found to contribute to non-compliance (Tebbi, Richards, Cummings, Zevon & Mallon, 1988; Hauser, Jacobson, Lavori, Wolfsdorf, Herskowitz, Milley, Bliss, Wertlieb & Stein, 1990; Hanson, DeGuire, Schinkel, Henggeler & Burghen, 1992). In contrast, supportive parenting style has been linked to better adherence (Manne, Jacobsen, Gorfinkle, Gerstein & Redd, 1993).

3.2. PARENTS OF CARDIAC CHILDREN

3.2.1. Introduction

There is a substantial literature on parents of cardiac children, much of it descriptive, but little reporting of parental variables, such as coping strategies or levels of psychological distress. Mothers in particular have been studied in terms of the relationship between their attitudes and functioning of the cardiac child, but fathers are rarely mentioned. The major studies and their main findings are given in Table 3.2.

3.2.2. Time of Diagnosis

The diagnosis of congenital heart disease in a child creates a crisis for the parents (Higgins & Kashani, 1986) and has been widely reported to elicit feelings in the parents of grief, anxiety, fear, anger, resentment, guilt and shock (Cooper, 1959; Glaser et al, 1964; Pidgeon,

1967; Garson, Benson, Ivler & Patton, 1978; Gottesfeld, 1979; Giboney, 1983). Interviews with parents indicate that the initial mention of cardiac disease is particularly stressful, particularly if the lesion is not found until some time after birth (Furgal, 1981). It has been suggested that families with congenital heart deformities have more difficulties and stress than those with other congenital problems (Emery, 1989), which has been attributed in part to the hidden nature of the cardiac lesions. There is also a greater dependency on medical personnel than for many other congenital abnormalities.

ADJUSTMENT OF PARENTS OF CHILDREN WITH CONGENITAL HEART DISEASE: MAIN STUDIES

AUTHORS	STUDY GROUP	MAJOR FINDINGS
Landtman et al 1960	84 CHD pts and their families	25 of 70 mothers were overprotective before surgery; 8 mothers were "rejective". 1 year after surgery 11 of the overprotective mothers and 4 of the rejective mothers treated their child normally. 33 of 74 mothers were "normally" distressed at time of operation; 28 were extremely distressed, 5 of whom sought medical help. 8 were classed as "indifferent" and 5 as particularly negative, such that they took their child out of the hospital.
Apley et al 1967	Families of 88 children with CHD	In 60 families (68%), mothers were felt to have an unsatisfactory relationship with the child. A disturbed relationship was significantly correlated with maternal immaturity, a disrupted family balance, maternal anxiety and dissatisfaction with consultations. 42% of fathers had a disturbed relationship with the child.
Linde et al 1970	Families of 198 CHD pts	Following surgery there was a significant decrease in maternal anxiety, pampering and overprotectiveness of the cardiac child. Compared with mothers of children who did not undergo surgery, mothers of operated children were less anxious, overprotective and pampering.
Hackett 1976	25 mothers of children undergoing surgery	Pre-operative maternal counselling resulted in greater discipline with their cardiac child. No differences in maternal anxiety or protectiveness between those who received counselling and those who did not.
D'Antonio 1976	20 mothers of CHD pts	Study of mothers' responses to rearing their child. Five maternal responses elicited from interview data - watchful, avoidance of conflict, preventive, adjustive and restrictive. Mothers were anxious, exhibited feelings of hopelessness and did not view their child positively. Family life appeared to revolve around the cardiac child.
Boll et al 1978	20 mothers of children with ACY CHD and 20 mothers of normal children	No overall differences in personality or parenting attitudes between the 2 groups. In both groups, different personality styles were significantly linked to different parenting attitudes. Knowledge of parents' personality style may help in predicting the need for intervention in coping with a child's chronic illness.
Finley et al 1979	Families of 40 children who had TOF corrected and 40 families of children who had appendectomies.	In 6 of the 40 TOF families parents had divorced since the birth of affected child (mean 9.8 years). Rate not significantly different to that in appendectomy group. Within Tetralogy of Fallot group there were no differences between families with a child with moderate compared with less severe disability.

Pinelli 1981	Mothers of 10 infants with CHD	Interviewed during hospitalisation and 1 month after discharge about their concerns regarding care-taking tasks of their infants. Area of greatest concern to mothers was how to anticipate and recognise the needs of the baby. There was an overall increase of 61% in the number of concerns reported at the second interview.
Silbert et al 1982	Families of 438 CHD pts and 2 control groups of 25 families where the child's heart disease had cured spontaneously and 26 families whose child was catheterised in infancy but who had no cardiac disease	No statistical differences at the time of testing (parents interviewed when affected child 5.5 years old) in divorce or separation rates for the 3 groups (12.1% for CHD group and 4% and 11.6% for the 2 reference groups).
Donovan et al 1983	4 CHD pts and their mothers	After completion by the patients of a comprehensive cardiac fitness programme, mothers were less restrictive, less anxious about their child dying and better understood their child's condition.
Naylor et al 1984	40 CHD pts undergoing cardiac catheterisation and their parents	Rehearsed parents had significantly lower anxiety scores before and after cardiac catheterisation than non-rehearsed group. Their anxiety reduced after the procedure. Anxiety level of control parents was the same before and after cardiac catheterisation.
Kaden et al 1985	Mothers of 285 infants with CHD	36% of mothers demonstrated poor comprehension of their child's diagnosis. No relationship between understanding and stress. Suggested that ignorance of the cardiovascular system may be so important as to minimise the impact of other variables. Distorted perceptions may result in unnecessary anxiety, restrictions on the child and impairment of the child's self perception. There are significant implications for the accurate understanding of the diagnosis.
Campbell et al 1986	26 children undergoing cardiac catheterisation and their parents	Stress management training prior to cardiac catheterisation resulted in significantly lower levels of parental stress responses both during hospitalisation and afterwards compared with parents who received standard information.
Goldberg et al 1990	Parents of infants with CF or CHD and of healthy babies	Parents of CF and CHD pts reported more stress than parents of healthy babies. Parents in the CHD group reported the highest levels of stress.
Goldberg et al 1991	42 infants with CHD and their mothers and 46 healthy infants and their mothers	Significantly fewer infants with CHD had a secure relationship with their mother. The quality of this relationship in the CHD group was not related to parents' reports of their own stress or psychological wellbeing or to illness severity. Securely attached infants showed more subsequent improvement in health status compared with insecurely attached infants. Focusing on this relationship may result in improved social and physical development of babies with CHD.
Lobo 1992	10 infants with CHD and their mothers; 10 healthy infants and mothers	Study of feeding interactions indicated that CHD infants scored significantly lower on measures of responsiveness to their mother and their ability to give signals to the mother. CHD mothers scored significantly lower on measures of social interaction with their infant.

CHD = congenital heart disease
pts = patients
ACY = acyanotic heart disease
CF = cystic fibrosis
TOF = Tetralogy of Fallot

Table 3.2

It has been suggested that parents commonly view their child's condition and their own responsibilities from within a moral framework (Silverman, 1987), thereby assuming responsibility for the birth of a congenitally malformed child. Parents typically find

unsatisfying and unacceptable the clinical explanation that their child's cardiac defect is unrelated to their own actions and is a random event. Within this moral framework, parents also need to demonstrate that they are caring and responsible. Involvement of hospital staff may be perceived as a threat to their perceived responsibilities to care for their child (Silverman, 1987). In situations where a late diagnosis is made, parents frequently have stories of incompetence on the part of the medical staff and refusal of doctors to listen to their anxieties about their child. To outsiders, the discrepancy between the presence of congenital heart disease at birth and its delayed discovery could reflect on the moral status of the parents, so parents produce these accounts which indicate that they are caring and responsible parents whose anxieties were dismissed and who then accepted the reassurances of the professionals.

3.2.3. Specific Problems of a Diagnosis in Infancy

The diagnosis of congenital heart disease in the neonatal period, necessitating urgent transfer of the baby to a specialist centre, can disrupt the usual pattern of parent-infant bonding (Shor, 1978; Bentovim, 1983). The impact of the diagnosis at this time may be heightened by other stressors (such as the birth itself) and may be exacerbated by the need to make difficult treatment decisions urgently (Fisk, 1986).

Infants with congenital heart disease have frequently been described as difficult to feed (Gillon, 1973; Gudermuth, 1975; Cloutier & Measel, 1982; Clare, 1985) and this can result in feelings of frustration and in difficulties in the development of the mother-child bond (Garson & Baer, 1990). A small study of 8 infants with congenital heart disease and their mothers reported that all mothers had experienced some degree of difficulty in bonding with

their infants and in 3 cases the feeding process was implicated as a source of difficulty (Gudermuth, 1975). The difficulties experienced in feeding such babies, together with their poor growth patterns, can be experienced by the mother as rejection of her and an indication of her own inadequacy and failure as a competent mother (Gudermuth, 1975; D'Antonio, 1976; Shor, 1978). When timing of surgery is contingent on sufficient weight gain, the situation can be even more frustrating for mothers (Glaser & Bentovim, 1987). A study of the mother-infant interaction during feeding found that both mothers and infants differed significantly from healthy infants and their mothers in their interactions. Infants with congenital heart disease were less responsive to their mothers and less able to give appropriate signals and mothers were less able to engage in social interactions with their babies (Lobo, 1992). Pinelli (1981) investigated mothers' concerns about caring for their infants with congenital heart disease and found a significant increase in their number of concerns once they were caring for their child at home, indicating their feelings of vulnerability and inadequacy.

Significantly fewer infants with congenital heart disease, compared with healthy babies, have secure relationships with their mothers (Goldberg, Simmons, Newman, Campbell & Fowler, 1991). The quality of the relationship was not found to be influenced by parental stress levels or the severity of the diagnosis, but it did have a bearing on later social and physical development. In a comparative study of parents of infants with congenital heart disease or cystic fibrosis and parents of healthy infants, parents of chronically ill infants consistently experienced more stress than those of healthy infants. Congenital heart disease in infancy was more stressful for parents than cystic fibrosis and mothers tended to report more stress than fathers (Goldberg, Morris, Simmons, Fowler & Levison, 1990). However, whilst in the

healthy group the most securely attached infants had the most positive mother-child interaction at two years of age, this was not the case in either the congenital heart disease or cystic fibrosis groups (Goldberg, Washington, Morris, Fischer-Fay & Simmons, 1990). The authors suggest that the influence of the early parent-child relationship may be altered by a child's health status and that parent-child attachments are less consistent in the presence of chronic illness. A number of babies with congenital heart disease are born prematurely and, due to this and their heart lesion, require more prolonged hospitalisation. Preterm infants hospitalised for longer than a month at birth have been found to have a significantly different pattern of attachment (more anxious-resistant) compared with healthy preterm infants (Plunkett, Meisels, Stiefel, Pasick & Roloff, 1986).

3.2.4. Impact of Congenital Heart Disease

Apley, Barbour & Westmacott (1967) found that 68% of mothers and 42% of fathers had an unsatisfactory or disturbed relationship with their child. A disturbed mother-child relationship was significantly related to maternal immaturity and anxiety. A study of the parents of 260 children with congenital heart disease highlighted the importance of mourning the loss of a normal, healthy child in order that parents could accept the defective child (Garson et al, 1978; Garson & Baer, 1990). Denial is the most frequently reported coping mechanism utilised by parents (Rozansky & Linde, 1971), particularly among parents of asymptomatic children (Garson et al, 1978). Parents of younger cardiac patients are more willing to discuss their feelings whereas parents of older children attempt to deny them (Rozansky & Linde, 1971). This study also highlighted the difficulties experienced by parents in discussing death. Parents avoided "deep feelings" about death and when discussion

approached this topic they generally stopped communicating and became mildly euphoric or stopped attending group sessions.

Parental anger and resentment - often displaced on to God, medical personnel or themselves, rather than being overtly displayed towards the child - have also been reported (Linde & Linde, 1973; Gidding & Rosenthal, 1984). It has been suggested that the use of denial and guilt as defense mechanisms can serve to keep parents from becoming too overwhelmed (Bowen, 1985). In one of the few studies to focus on both parents, fathers, in contrast to mothers, did not tend to express feelings of guilt but a loss of self esteem was more evident in the fathers. This loss of self esteem was often related to the child's retarded development or cyanosis (Rozansky & Linde, 1971; Linde, 1982). The manifestation of denial and anger may be seen as overprotective, infantilising behaviour towards the sick child (Rozansky & Linde, 1971; Garson et al, 1978) and overprotective behaviour by mothers in particular has been reported (Landtman & Valanne, 1959; Linde et al, 1966; Apley et al, 1967; Kimball, 1973; Rausch de Traubenberg, 1973). The degree of maternal overprotection is often unrelated to the actual severity of a child's cardiac lesion (Landtman, Valanne, Pentti & Aukee, 1960; Offord et al, 1972; D'Antonio, 1976) and a number of early studies highlighted the importance of maternal perceptions of disease severity (Bergmann & Stamm, 1967; Offord & Aponte, 1967; Offord et al, 1972; Cayler et al, 1973). It is suggested that the presence of a heart condition, rather than its severity, is a main determinant of maternal anxiety (Linde et al, 1966; Kitchen, 1978). A recent study of children who had undergone palliative surgery found that 80% of parents underestimated their child's exercise tolerance (Casey et al, 1994). Another recent study, however, found that mothers had accurate and realistic perceptions of the severity of their child's heart lesion and that their perceptions of

severity were not correlated with adjustment of their child. However, maternal perceptions of their parenting skills and interaction with the cardiac child were significantly correlated with the child's adjustment (DeMaso et al, 1991). Poor adjustment in the cardiac child has been found to be related more to maternal anxiety, guilt and pampering than to the degree of medical incapacity (Linde et al, 1966; 1976; Kong et al, 1986). The influence of maternal perceptions on parenting was also demonstrated in a study of children with Down's syndrome, with and without heart defects (Barrera, Watson & Adelstein, 1987). Parents perceived children with cardiac lesions as less active, less skilful and needing less environmental stimulation than those without congenital heart disease, resulting in poorer caretaking.

The presence in a family of a child with congenital heart disease can place constraints and stressors on all aspects of family life. Family recreational activities may be restricted and additional financial burdens imposed. In a study of 100 patients and their families there was a tendency for mothers to postpone subsequent pregnancies (Boon, 1972), although parents have been shown to have little knowledge about recurrence risks for future children (Reiss & Menashe, 1972).

3.2.5. Impact of Cardiac Surgery

For families whose child requires immediate surgery, the crisis is prolonged and intensified beyond the time of diagnosis. For the majority of children, however, surgical correction is not undertaken for several months or years after diagnosis. Parents of these children have identified uncertainty about their child's future as a major stressor in their lives (Fisk, 1986). Such stress may engender feelings of helplessness and frustration and can influence parental

expectations of the sick child. The time of the surgery can precipitate extreme distress in parents and in a study of 74 mothers, five had to seek medical advice for a nervous breakdown (Landtman et al, 1960). Indifference and negative reactions to the proposed surgery, resulting in the child being removed from hospital, were also reported. In the perioperative period parents have been found to be highly stressed in the intensive care unit by the alteration in their parental role and by their child's behavioural and emotional reactions (Miles, Carter, Hennessey, Riddle & Eberly, 1989). Change or loss of parental role during hospitalisation and the resulting increase in parental anxiety have also been reported for other groups of parents (Kasper & Nyamathi, 1988). Maternal anxiety is also influenced by parental beliefs regarding their hospitalised children and their own role (Melnyk, 1995). Following cardiac surgery, parental anxiety reduces, with a resulting decrease in overprotective and pampering behaviour towards the children (Landtman et al, 1960; Linde et al, 1970).

A seemingly contradictory finding is the second mourning phase that has been identified in parents after successful corrective surgery (Garson et al, 1978). For parents whose child had significant cardiac symptoms pre-operatively, surgical correction had produced a perceptibly different child who could be accepted only after mourning the loss of the previously symptomatic one. For children who were asymptomatic but overprotected pre-operatively, the authors identified difficulties for their parents with this second mourning phase. Parents were unable to alter their behaviour towards their child after surgery and continued to be overprotective. A follow-up study of young adults with Tetralogy of Fallot also found that patients with the least physical symptomatology pre-operatively were more dependent and protected post-operatively (Garson et al, 1974).

3.2.6. Parents' Marital Relationship

Two studies investigating the impact of congenital heart disease on the marital relationship found that the divorce rate was not significantly higher than for parents of children who had undergone appendectomy (Finley et al, 1979) or for parents of children with heart defects which had spontaneously recovered or who had had previous cardiac catheterisation but had no heart disease (Silbert et al, 1982). Finley et al also reported a high degree of family stability in the families of children with congenital heart disease, and neither divorce rates nor family stability were related to the severity of disability of the children. The physical demands of caring for the child can be a major drain on parents' resources and leave them with little time and energy for the pursuit of other relationships or responsibilities (Fisk, 1986).

3.2.7. Parental Characteristics

Studies of parental personality characteristics and their relationship to parenting attitudes have found similar findings for parents of children with congenital heart disease and parents of healthy children. A study of school absenteeism found that children from families with high external locus of control beliefs tended to have increased school absence compared with children from families with internal health locus of control beliefs. This finding was applicable to children with congenital heart disease and healthy children (Fowler, Johnson, Welshimer, Atkinson & Loda, 1987). A comparison of parenting attitudes of mothers of children with acyanotic heart disease with mothers of healthy children found no overall differences between the two groups. However, parental personality style was significantly correlated with parenting attitude, irrespective of the child's health status (Boll et al, 1978). Little attention has been given to coping strategies, although cardiac parents find trust in

medical care and the use of autonomy more helpful than social or family support (Eiser & Havermans, 1992).

3.2.8. Provision of Information

A number of studies have described the importance of and/or assessed the impact of providing specific information/counselling at the time of diagnosis (Kupst, Blatterbauer, Westman, Schulman & Paul, 1977; Kashani & Higgins, 1986), prior to undergoing cardiac catheterisation (Naylor et al, 1984; Campbell et al, 1986; Campbell, Kirkpatrick, Berry, Penn, Waldman & Mathewson, 1992) or cardiac surgery (Peay, 1960; Peterson, 1979; Bavin, 1983; Rushton, 1983) and on discharge from hospital following cardiac surgery (Stinson & McKeever, 1995). Parents often have to administer life-saving, but potentially lethal, toxic drugs to their child at home, and the need for accurate information and instruction has also been emphasized (Jackson, 1979).

There is general agreement that the provision of detailed information, both from a medical and a psychological perspective, is of benefit to parents. An intervention study in which parents were taught techniques of stress management and given supportive counselling as well as information prior to their child undergoing cardiac catheterisation demonstrated the positive effects on the way in which parents experienced the hospitalisation (Campbell et al, 1986). The authors suggest that such an approach may have given the parents a sense of control beyond that gained from just being given information. Negative reactions in children after catheterisation were found to correlate significantly with parental anxiety (Naylor et al, 1984), but parental anxiety was reduced after catheterisation when parents and their children had been involved in a rehearsal programme prior to catheterisation. A small study of four

cardiac patients, who participated in a comprehensive cardiac fitness programme for one year, and their mothers found that after the programme mothers were less restrictive, less anxious about their child dying and had a better understanding of their child's condition (Donovan et al, 1983).

3.2.9. Communication

Attention has increasingly been focused on the communication between parents and physicians and parents' understanding of their child's heart defect. Kaden, McCarter, Johnson & Ferencz (1985) found that 36% of mothers demonstrated poor comprehension of the lesion but the authors could only explain a small proportion of the variability of maternal knowledge by factors such as sociodemographic variables and maternal stress levels. The authors suggest that fundamental ignorance of the cardiovascular system may be of such importance as to minimize the impact of other variables. The potential harm of distorted perceptions of congenital heart disease in causing unnecessary anxieties, inappropriate restrictions and impairment of the child's self perception is also highlighted.

In initial consultations parents do not distinguish between different conditions and medical severity and in situations where symptoms are not obvious, parents rely on their lay understanding of the heart for understanding the condition (Silverman, 1987). Parents tend to focus their attention on areas where they are directly responsible, such as instructions about medication and physical activity, and many feel that the academic, clinical information is not very important to them (Kupst, Dresser, Schulman & Paul, 1976). A high percentage of parents report dissatisfaction with consultations (Apley et al, 1967), particularly at the early stages (Korsch & Negrete, 1972; Silverman, 1983,1987). Parental participation in

outpatient clinics is greatest at times where specific knowledge is available, such as after catheterisation, or where active intervention is contemplated - e.g. surgery, but does not appear to be related to traditional variables of parental social class and educational level or consulting style of the doctor (Silverman, Hilliard, Baruch & Shinebourne, 1984).

Parents' groups have been suggested as useful reference points for parents (Silverman, 1987) and the sharing of mutual experiences can resolve feelings of isolation. Group discussion permits negative expression regarding the medical profession and can add status to the role of the parent in the parent-physician relationship (Linder, 1970). However, there may also be pitfalls of such groups, such as undermining the confidence of some parents in their child's physician, which can potentially interfere with optimal medical care (Rowland & Armstrong, 1983). For parents of children with uncomplicated congenital heart disease, fears may be expanded through discussion with parents of children with more complex lesions.

3.3. PARENTS OF BMT RECIPIENTS

3.3.1. Introduction

Despite the increasing numbers of paediatric bone marrow transplants being performed, there is still relatively little detailed systematic information about the effects on parents and descriptive accounts still predominate. The focus of these accounts has tended to be on parental adjustment to childhood malignancy. Parents coping with other diseases for which BMT is offered as a treatment have been largely ignored. However, the differences in the underlying diseases and the varying impact of BMT on parents is now being acknowledged.

3.3.2. Nature of the Disease

For parents of children with leukaemia, BMT represents the final stage in treatment. Parents have usually begun to accept and integrate the illness into their lives. They are already used to uncertainty about the future, focusing on the sick child and disruption to usual family routines. However, their sense of desperation at this "last ditch" attempt can be extreme. In contrast, children with aplastic anaemia have usually only been recently diagnosed and for their parents there is little time to deal with their feelings about the diagnosis or begin to integrate the illness into their lives before they have to cope with the stresses of BMT. Parents of children with genetic diseases often experience significant feelings of guilt. They may have already lost one or more children to the disease and this may heighten their anxieties through the transplant stages.

3.3.3. Parental Consent for the BMT

The issue of parental consent for a child's bone marrow transplant has received some attention in the literature. The experimental nature of the treatment and the significant morbidity and mortality associated with it means that giving consent is potentially highly stressful. An ethical study on the use of BMT and an evaluation of the risk/cure trade-off found that most parents were unwilling to accept a 15% mortality and 15% morbidity risk for their child to be cured. These findings reflected the views of an ethics committee, suggesting not only that the use of BMT should be restricted but also raising the issue of who should be able to make treatment decisions for children (Kodish, Lantos, Siegler, Kohrman & Johnson, 1990). A study of 46 mothers and 15 fathers consenting to BMT for their child found that 60% of mothers and 47% of fathers exhibited significant psychological distress. Use of emotion-focused coping, such as avoidance and suppression, was positively correlated

with psychological distress, suggesting that such coping mechanisms are ineffective in alleviating parental distress at the time of consent. Parental perception of the quality of the communication between physician and parent was the strongest predictor of parental level of distress (Dermatis & Lesko, 1990). A further issue concerns the conflict of interest that can occur when parents are asked to give consent for both donor and recipient. A judicial procedure has been proposed to deal with the medical and legal interests of all involved (Serota, August, O'Shea, Woodward & Koch, 1981).

3.3.4. Marrow Donation

Due to the low incidence of physiological morbidity associated with donation of bone marrow, the role of donors has tended to be de-emphasized (Folsom & Popkin, 1987), even though some adverse psychological effects have been reported for other groups of donors (e.g. Simmons, Klein & Simmons, 1977). A strong bond between the child and resident parent is often forged as a result of the experiences they have shared and the dependency of each on the other for emotional and physical closeness (Freund & Siegel, 1986). This may continue after discharge and can cause the other parent to feel jealous and excluded. In instances where a parent is the marrow donor, the non-donor parent may feel estranged and inadequate (Wiley, Lindamood & Pfefferbaum-Levine, 1984) and there may also be significant repercussions for the parent-child relationship. When the donor is a sibling, parents have to divide their concern and attention between two children (Pfefferbaum, Lindamood & Wiley, 1978).

3.3.5. Impact of the Treatment

With the relatively small numbers of centres undertaking BMT, resulting in some children being treated a considerable distance from home, and the prolonged hospitalisation required, separation of the family is a significant issue. One parent tends to stay at the hospital, leaving the other parent and children at home, and this can affect the parents' own relationship (Patenaude et al, 1979; Pot-Mees & Zeitlin, 1987). In some instances parents alternate spending time with the ill child, but pre-existing distinctions between parental roles are significant in determining how they adapt to this. If parents have had traditionally differentiated roles, fathers may find it difficult to assume a more nurturing "maternal" role (Patenaude et al, 1979).

Parents resident in hospital have been found to have high rates of anxiety and distress and feelings of helplessness in relation to the isolation procedures (Pfefferbaum et al, 1978) and may feel guilt at submitting their child to the treatment (Gardner et al, 1977). Sleep disturbance, feelings of claustrophobia and increased mental and physical fatigue have also been reported (Pot-Mees & Zeitlin, 1987). Fathers of children with more severe clinical conditions were found to show greater distress than fathers of children with fewer medical problems, but mothers of the sicker children were more supportive towards their children (McConville et al, 1990). In relatively uncomplicated cases with good outcome many of the parents coped well (McConville et al, 1990).

3.3.6. Support

In some cases parents have difficulties in maintaining effective support for their child through the treatment process. A fear of the child dying, anxiety about giving the child an infection

through touching or being with them and parents' own physical state (e.g. exhaustion) have been suggested as reasons for this (Artinian, 1982). Pfefferbaum, Lindamood & Wiley (1977) retrospectively studied the impact of family support and the parent-child interaction on outcome and although there was no direct correlation, the authors felt that these factors were critical to patient wellbeing. Throughout the hospitalisation parents have been found to rely on support from other parents in the hospital (Gardner et al, 1977) and from staff (Popkin & Moldow, 1977), rather than on their own family and social network. This has also been found to be true of families who are local to the treating hospital (Patenaude et al, 1979).

3.3.7. Discharge from Hospital

Discharge from hospital is a time of ambivalence for parents and can evoke apprehension, particularly in mothers who are now responsible for daily care. Parents are often physically and mentally exhausted and their continuing anxiety about the child's health, together with the child's increased dependency, can influence family reintegration. Financial and employment concerns may also be of greater significance once the family are back at home.

3.4. SIBLINGS

3.4.1. Introduction

The significance of the sibling experience in the development of personality has been increasingly recognised (Sutton Smith & Rosenberg, 1970; Dunn, 1983). Sibling relationships have also been acknowledged as important precursors for childhood and adult relationships formed outside the family (Abramovitch, Pepler & Corter, 1982) although the importance of not considering the sibling relationship in isolation from other family

relationships has been emphasized (Dunn, 1988). The sibling relationship plays an important role in the process of learning how to cooperate, compete, negotiate, support and reward one another (Minuchin, 1974). At an emotional level the relationship is constantly changing (Walker, 1990). By the end of the first year of life children are spending as much time interacting with their siblings as they do with their mothers and more time than with their fathers (Lawson & Ingleby, 1974). Whilst many relationships within society may be short-lived and transient, that with a sibling is unique because it usually spans 60 or more years (McKeever, 1983), a rather greater overlap of life-span than that of parents with their children. Furthermore, most people have sibling relationships, with estimates that more than 80% of children in the U.S.A. have siblings (Brown Miller & Cantwell, 1976).

Early research on sibling relationships tended to focus on the effects of constellation variables such as gender, birth order and sibship size on intelligence, personality and academic functioning (Zajonc & Markus, 1975; Rosenberg, 1982; Sutton-Smith, 1982). More recent research has looked at dynamic interactions within the family, and a wide variety of variables, including the role of parental behaviour, have been studied in normal sibling pairs (Dunn & McGuire, 1992). The influence of contextual and ecological variables in the development of sibling relationships has also been studied, such as the physical setting in which the relationship occurs and the presence or absence of specific other people (Brody & Stoneman, 1986). It is now acknowledged that "outcome", in terms of child development and sibling interactions, is "the evolving result of an interacting system of child, family, situational and cultural variables" (Lobato, Faust & Spirito, 1988).

Difficulties, particularly aggression, in sibling relationships have been linked to difficulty with peer relationships later on (Dishion, 1986), although other research into childrens' relationships with other children found links between their relationships with siblings and close friends but no other association between sibling and peer relationships (Stocker & Dunn, 1990). Such research highlights the need to clarify the processes linking children's relationships within the family to those formed outside the family, whilst at the same time emphasising the importance of the sibling relationship in the formation of other extra-familial relationships.

3.4.2. Chronic Illness and Its Effect on Siblings

In view of the significance of sibling relationships it is reasonable to expect that "trauma", such as chronic illness, affecting a child will affect other siblings in a family. It has been suggested that the presence of a chronically sick child in a family is potentially one of the most stressful experiences for healthy children (Coddington, 1972). Chronic illness threatens the integrity of the sibling relationship both directly and indirectly (Eiser, 1993). Directly, concern and anxiety influences the relationship emotionally and at a practical level opportunities may be reduced for joint activities and even direct contact between siblings. Healthy siblings loose their equal relationship with their sick brother or sister (Trahd, 1986). Indirectly, siblings may be restricted in interactions with their parents as parental attention is increasingly focused on the sick child. Such differential treatment of children by parents is known to result in increased conflict between the siblings (Handel, 1986; Boer, 1990). Relationships between parents and their well children are also affected (Gallo, 1988) and healthy siblings can feel "displaced and unimportant" (Martinson, Gilliss, Colaizzo, Freeman & Bossert, 1990). Parents may be too busy and/or exhausted to spend time with their well

children and may endeavour to "protect" their healthy children from knowledge about the disease. In a study of 36 families of chronically ill children, none of the parents talked openly to their well children about how they felt (Canam, 1987). Chronic illness alters the quality and quantity of intra- and extra-familial communication (McKeever, 1983), often with serious consequences for the healthy siblings. A number of studies have found that children are poorly informed about their sibling's illness (Burton, 1975; Spinetta, 1981), although others report that siblings are well informed but totally dependent on their parents for information (Marky, 1982). A study of cystic fibrosis patients and families found that siblings were poorly informed about certain aspects of the disease, such as genetics and reproductive risks, and that a high percentage expressed a need for "a great deal " more information on certain medical and psychological aspects of the disease (Henley & Hill, 1990 (a,b)). However, children who are aware of their sibling's diagnosis are a significant source of stress for their parents (Meyerowitz & Kaplan, 1967) due to the additional anxiety parents have about the possible traumatic effects of such knowledge on the well children. Hospitalisation of the sick child can result in increased problems for the well siblings with difficulties sleeping, increased anger and impaired concentration at school (Craft & Wyatt, 1986). Siblings have also been found to be ill prepared for hospital visiting, with 75% reporting feelings of fear before the first visit and seeing the visit as a source of stress (Steiner, 1984). Healthy siblings may also become isolated from the outside world, either as an attempt to protect the ill child from infection or because of the perceived stigma associated with the condition. Feelings of jealousy and resentment towards the sick child are commonly reported and subsequently guilt that their resentment has precipitated a deterioration in the child's medical condition, or even their death. It is therefore perhaps not surprising that some researchers have concluded that, within the family, healthy siblings bear the greatest burden of stress (Spinetta, 1981).

Whilst the literature on the psychosocial effects of chronic illness for the ill child and their parents is quite extensive, it is only comparatively recently that research attention has focused on the healthy siblings. Early reporting of sibling adjustment tended to be anecdotal and based on unstructured interviews and/or projective test measures (Ferrari, 1984; Drotar & Crawford, 1985). Three early studies highlighted the consequences for children affected by the death of a sibling and it was concluded that they were at significant risk of developing severe psychological problems (Cobb, 1956; Cain, Fast & Erickson, 1964; Binger, Ablin, Feuerstein, Kushner, Zoger & Mikkelsen, 1969). These included low self-worth, poor school performance, guilt and feelings of responsibility for their sibling's death, and distorted concepts of illness and death.

Later researchers have examined sibling reactions during the illness, with many studies focusing on the impact of childhood cancer. Overall, there is no simple conclusion to be drawn from this literature. The commonly held belief that siblings of chronically ill and disabled children have more adjustment difficulties than siblings of healthy children has not been consistently supported. Some adverse psychological effects have been reported at home and at school, such as difficulties with peers, academic problems, reduction in school and social activities, lowered self concept, anxiety, depression, irritability, somatization and increased aggressive behaviour (Binger et al, 1969; Gath, 1973; Tew & Laurence, 1973; Burton, 1975; Lavigne & Ryan, 1979; Peck, 1979; Vance et al, 1980; Breslau, Weitzman & Messenger, 1981; Harvey & Greenway, 1984; Cadman, Boyle & Offord, 1988; Reynolds, Garralda, Jameson & Postlethwaite, 1988; Tritt & Esses, 1988; Williams, Lorenzo & Borja, 1993). Siblings of children with cancer were found to experience severe stress and in some areas they showed even more distress than the patients, such as perceived social isolation

(Cairns, Clark, Smith & Lansky, 1979). Low self esteem in children whose sibling died was also found to be common (Pettle Michael & Lansdown, 1986). Family relationships can be disrupted and siblings of sick children have been found to express more negative emotion in relation to their parents - particularly their fathers - than siblings of healthy children (Stewart, Stein, Forrest & Clark, 1992). A study in which children were looked at in terms of Piagetian levels of cognitive development found that children - particularly adolescents who were at the formal operational stage of cognitive development - with ill siblings had lower levels of illness conceptualisation than would be expected from their level of cognitive development. Their level of illness conceptualisation was also lower than that of children with healthy siblings (Carandang, Folkins, Hines & Steward, 1979). However, other studies have not found evidence of a uniformly negative psychological impact on the healthy siblings (Gayton, Friedman, Tavormina & Tucker, 1977; Ferrari, 1984; Fielding, Moore, Dewey, Ashley, McKendrick & Pinkerton, 1985; Gallo, Breitmayer, Knafl & Zoeller, 1992; Noll, Yosua, Vannatta, Kalinyak, Bukowski & Davies, 1995) and some have found positive psychological effects, including less sibling aggression and teasing, increased social competence, socialisation skills, tolerance, empathy and sensitivity (Iles, 1979; Taylor, 1980; Ferrari, 1984; Horwitz & Kazak, 1990; Gallo, Breitmayer, Knafl & Zoeller, 1991).

A number of mediating variables have been linked to sibling adjustment, including gender, birth order, socioeconomic status, parental reaction to the illness and time since diagnosis. The importance of considering and controlling for characteristics, such as gender and age, of the sibling pairs, rather than just of the sibling, has also been highlighted (Brody & Stoneman, 1986). The source of the data collected has also been found to be an important factor, with mothers providing a more negative perspective on sibling functioning than

siblings themselves (Lobato, Barbour, Hall & Miller, 1987), although this may also reflect maternal depression and anxiety about the sick child (Lancaster et al, 1989). Other discrepancies between siblings' and parents' views of sibling difficulties have also been found (Menke, 1987). Siblings reported worry and concern about their sick sibling as being hardest for them, while parents thought that the major impact on their well children was the loss of parental attention (Breyer, Kunin, Kalish & Patenaude, 1993).

A few studies have looked at areas other than adjustment and behaviour problems, such as caring for the ill child and the performance of home based chores by healthy siblings. Age and sex of the well sibling were found to be particularly important (Gath, 1974; Burton, 1975; Schwirian, 1976; Lobato et al, 1987) with sisters bearing the brunt of such duties. More recently, attention has focused on the effects of disability on the quality of sibling relationships and whether characteristics such as age and birth order of the siblings influence the nature of the interaction (Dallas, Stevenson & McGurk, 1993 (a,b)). A few studies have also attempted to look at coping strategies in siblings (Iles, 1979; Koch-Hattem, 1986; Breyer et al, 1993). Obtaining information about the illness, expression of emotions, seeking support from others and involvement with the patient and participation in their care were particularly identified as helping siblings cope (Kramer, 1981), although the importance has been acknowledged of identifying specific stressors and coping strategies for the individual child (Walker, 1990). Support groups have been found to provide siblings with the opportunity to decrease their sense of isolation, express negative feelings and learn from each other (Heiney, Goon-Johnson, Ettinger & Ettinger, 1990).

Findings in the general literature on after-school care when parents are out at work indicate that children who are on their own or just with younger siblings are more isolated socially than children in adult care, whilst children in the care of older siblings are at greater risk for negative effects on self-esteem and social development (Berman, Winkleby, Chesterman & Boyce, 1992). For the healthy sibling of a sick child, one might expect that they will spend more time alone or with other siblings, particularly when the ill child is hospitalised, further adding to the stress associated with the illness.

The research reviewed above demonstrates the lack of a direct relationship between childhood illness and psychopathology in the well siblings. It has been suggested that the impact of illness or disability on siblings can be best conceptualised as a risk factor, the effects of which are mediated by other individual and family characteristics and resources (Lobato et al, 1988).

3.4.3. Factors Influencing Psychological Adjustment Of Siblings

A number of individual and family characteristics may influence psychological adjustment of healthy siblings to chronic illness, although findings are inconclusive. Birth order (Gath, 1972, 1974) and the interaction of birth order and gender have been closely studied, with illness/handicap having a greater impact on younger male and older female siblings (Breslau et al, 1981). However, the opposite has also been found, with younger female and older male siblings being most severely affected (Lavigne & Ryan, 1979). The consensus for older siblings does seem to be that it is girls who experience greater psychological distress (Gath, 1974; Breslau et al, 1981; Simeonsson & McHale, 1981), which has been attributed to their greater care responsibilities (Burton, 1975; Lobato et al, 1987). In terms of the age of the

siblings, older siblings are more likely to report more positive effects of the illness than younger siblings, suggesting that level of maturity can moderate the stress of an ill child within the family (Sargent, Sahler, Roghmann, Mulhern, Barbarian, Carpenter, Copeland, Dolgin & Zeltzer, 1995). Other variables which have been studied include size of the sibship (Gath, 1974), socioeconomic status (Gath, 1974; Breslau & Prabucki, 1987) and level of sibling knowledge about the illness (Evans, Stevens, Cushway & Houghton, 1992). Family variables which have been found to affect sibling adjustment include mother's adjustment (Tew & Laurence, 1973; Fisman & Wolf, 1991), parental stress (Dyson, Edgar & Crnic, 1989) and communication within the family and family cohesion (Daniels, Miller, Billings & Moos, 1986; Daniels, Moos, Billings & Miller, 1987). Influential disease variables include severity (Tew & Laurence, 1973), maternal perception of disease severity (Klein & Simmons, 1977), visibility (Lavigne & Ryan, 1979) and time since diagnosis (Ferrari, 1984). However, there is little evidence of consistent relationships between single factors and adjustment - rather, it is the interaction of variables that is significant. Furthermore, siblings appear to demonstrate similar personal and social reactions, regardless of the specific chronic and/or handicapping condition. Simeonsson and Bailey (1986), in discussing the commonality of sibling reactions across different conditions, propose that there are child variables influencing the sibling reaction which transcend the specific characteristics of the illness or disability - i.e. sibling reactions are likely to be mediated by individual characteristics of the sick child as well as by the actual condition. Such an approach accounts for differences in sibling reaction to children with the same condition.

3.4.4. Methodological Difficulties In Research On Siblings

One of the fundamental problems with the majority of studies of sibling adaptation is that they are based on parental perceptions; very few have data obtained directly from the siblings themselves. It may be difficult for researchers to see the siblings, particularly in hospital based studies where siblings are infrequent visitors and when they do visit it tends to be in evenings or at weekends. A further difficulty with gaining access to the siblings involves obtaining parental consent. In families where open communication and opportunities for family discussion are common, parents are normally prepared for siblings to participate in research. In those families who are unwilling to involve healthy siblings in the patient's illness or do not want information to be given to the well children, parents are likely to refuse permission for their healthy children to be involved in the research, thereby leading to a skewed sample (Eiser, 1993). A further problem is that the cross-sectional design of many studies ignores the developmental aspects inherent in any relationship (McKeever, 1983). Focusing on one member of a dyad also ignores the bidirectional interactions between the ill and well siblings. The way in which chronic illness modifies the sibling relationship has been largely overlooked, particularly aspects such as the effects on the healthy siblings of the patient's anger at them for being well (Sourkes, 1980). One recent study does suggest, however, that qualitative aspects of sibling relationships can influence both illness-specific and general psychosocial adaptation of chronically ill adolescents (Hanson, Henggeler, Harris, Cigrang, Schinkel, Rodrigue & Klesges, 1992). In order to achieve sufficiently large sample sizes studies have tended to involve siblings across a wide age-range, thereby ignoring developmental and age-related considerations such as sibling understanding of the illness and the child's role within the family. The tendency has been to look at a narrow range of variables, with the emphasis on a deficit-centred approach of

detecting "maladjustment" and vulnerability in the siblings, rather than focusing on coping strategies, adaptive responses and resilience (Leonard, 1991) to chronic illness. Furthermore, many studies have not controlled for factors such as birth order, gender and family size, disease characteristics such as time since diagnosis, or looked at parental coping and the effects on sibling adjustment. It seems clear that sibling adaptation is multifactorially determined and that in order to understand more adequately the impact of chronic illness on siblings, direct study of sibling interactions and experiences needs to be carried out, within the context of the functioning of the family as a whole.

3.4.5. Congenital Heart Disease and Siblings

There is very little information on the impact of congenital heart disease on the healthy siblings and studies reporting quantifiable data are particularly scarce. Table 3.3 summarises the main studies, their subject groups and the major findings.

Siblings of cardiac patients have been found to be jealous, resentful and hostile towards the patient, as well as expressing anxiety about their sick brother or sister (Maxwell & Gane, 1962; Boon, 1972). An early study of 155 siblings of cardiac patients from 70 families found that siblings in 64% of the families had behaviour problems and/or psychosomatic disorders (Apley et al, 1967). Comparison of cardiac siblings with siblings of haematology and plastic surgery patients and with siblings of healthy children indicated that, although siblings in all 3 illness groups were more likely to experience adjustment or behaviour problems than siblings of healthy children, cardiac and haematology siblings were less socially isolated and had lower levels of overall disturbance than the siblings of the plastic surgery patients (Lavigne & Ryan, 1979). A more recent study reported positive findings for cardiac siblings

AUTHORS	PATIENT POPULATION	MAJOR FINDINGS
Maxwell & Gane (1962)		38% of siblings of CHD patients reported to have been "affected" - anxiety, hostility, jealousy and feelings of deprivation reported.
Linde et al (1966)	81 cardiac siblings, 40 siblings of healthy children and 2 groups of CHD patients	Cardiac siblings received different parental treatment than siblings of healthy children. They received less attention and specifically were pampered less.
Apley et al (1967)	155 siblings of CHD patients	Siblings in 64% of families had behaviour and/or psychosomatic problems. Siblings of most severely affected patients were more disturbed.
Boon (1972)		Siblings in 17 of 58 families reported to show jealousy and resentment.
Lavigne & Ryan (1979)	57 CHD siblings, 62 haematology siblings, 37 plastic surgery siblings and 40 siblings of healthy children.	Collectively, siblings of ill children more likely to show irritability and social withdrawal than healthy group siblings. Overall levels of psychopathology higher in siblings of children with visible, compared with invisible, defects.
Henry (1982)	33 siblings of CHD patients who had died.	4 siblings had significant problems, including significant depression, deterioration in school performance and severe behaviour problems. One adolescent was admitted to a psychiatric hospital.
Faux (1991)	20 siblings of CHD patients; 22 siblings of children with craniofacial anomalies and 25 siblings of healthy children	Cardiac siblings were kinder, more empathetic and accepting of their siblings than in the 2 reference groups. Both groups of siblings of ill children displayed less hostility and anger towards their sick sibling than siblings in the healthy group.

Table 3.3

compared with siblings of children with craniofacial anomalies and siblings of healthy children. Siblings of cardiac children were kinder towards their ill sibling than siblings in the other two groups. In both illness groups siblings were reported to show less anger and hostility towards their impaired sibling than was shown by the children in the healthy group towards their siblings (Faux, 1991). Mothers' caretaking of well siblings of children with cardiac problems has been found to be significantly reduced following illness onset to a greater extent than for siblings of children with neurological illness (Williams et al, 1993), with possible implications for adjustment of the healthy cardiac sibling. In a study of families of children who had died from congenital heart disease, 12% of siblings had significant problems, including depression, deterioration in school performance and behaviour problems (Henry & Taylor, 1982).

These small number of studies on sibling adjustment to congenital heart disease are inconsistent in their findings. There are indications that cardiac siblings can be adversely affected but it is also clear that the majority of siblings cope and, furthermore, that some show positive responses to their sick sibling. The lack of consistency in the results is consistent with the findings for siblings in other illness groups and further investigation is clearly needed.

3.4.6. BMT and Siblings

The focus of research on the siblings of BMT patients has been on the issues of donors and non-donors. However, all siblings can experience a sense of abandonment due to their separation from one or both parents and this can result in feelings of anger, jealousy and resentment, coupled with guilt about having these feelings and being the healthy one (Patenaude et al, 1979; Freund & Siegel, 1986). Older siblings frequently take on parenting and domestic roles at home (Patenaude et al, 1979).

In the majority of allogeneic transplants donors are siblings. However, selection of the donor affects the entire family and a family's willingness to allow a child to donate can be a reflection of the relative value placed on a particular child (Levine, Camitta, Nathan & Curran, 1975; Patenaude et al, 1979). Competition among siblings (Freund & Siegel, 1986) and feelings of ambivalence in donor and non-donor siblings (Patenaude et al, 1979; Wiley et al, 1984) have been noted at the time of donor selection. The relationship between donor and recipient frequently changes as a result of transplantation (Patenaude et al, 1979; Freund & Siegel, 1986) and both positive and negative aspects of the sibling relationship may be accentuated (Wiley et al, 1984). A follow-up study of young adult donors several years after

transplant found that the quality of the donor-recipient relationship was highly correlated with health status variables in the patient (Wolcott, Wellisch, Fawzy & Landsverk, 1986(b)). A comparison of renal and bone marrow donors found that donors of bone marrow have a more positive attitude, due to the fact that donation impinges less on their own life (Gardner et al, 1977) but that feelings of irrational guilt seem to develop more easily in bone marrow donors. Donors can have unrealistic feelings of responsibility for the success or failure of treatment (Gardner et al, 1977) and risks of psychopathology in donor siblings increase if the patient dies or develops serious medical complications (Wiley et al, 1984). Psychological problems also seem to be more common when twins or opposite sex siblings are involved in transplantation (Wiley et al, 1984). Paediatric donors have been found to have sleeping difficulties and problems at school and also to experience a threat to their body integrity (Kinrade, 1987).

The implications of the transplant for each child depend on his or her developmental stage (Wiley et al, 1984). There is increasing recognition of the importance of providing age-appropriate information for siblings and the need to support the emotional well-being of the donor and non-donor siblings (Lenarsky & Feig, 1983).

CHAPTER 4

METHODOLOGY OF THE STUDY

4.1. RATIONALE FOR PERFORMING THE STUDY

The review presented above of the psychological effects of heart surgery for the child and family indicates a variety of potential adverse effects of the treatment as well as of the disease. Studies have looked at individual factors such as changes in cognitive performance following surgery and the effects of specific medical interventions but there have been few controlled, systematic, prospective evaluations of children undergoing cardiac surgery for correction or palliation of congenital lesions, and their families. Whilst researchers and reviewers have acknowledged the importance of a multifactorial approach, there is still a lack of research in which disease, treatment, environmental and family factors are looked at together. This study was therefore set up to look at the treatment of heart surgery within a multifactorial framework in order to elaborate on areas previously investigated and to elucidate areas so far uninvestigated. Whilst there are some hypotheses based on the results of previous studies, this study is essentially exploratory with the core question being:

What are the long term effects of a congenital heart defect and subsequent cardiac surgery on the psychosocial functioning of the child and family?

To give further insight into this, attention was focused on three more specific questions:

1. Does cardiac surgery share psychosocial effects with other types of potentially stressful hospital treatment?

2. How do children who have undergone cardiac surgery differ in psychosocial status from children who have not undergone hospitalisation and who do not suffer from any chronic medical condition?

3. If a high psychosocial morbidity is found following treatment, is there any indication that this is related to any treatment, environmental or family characteristics?

4.2. HYPOTHESES

Based on the results of previous studies, several hypotheses were tested, looking more specifically at differences in cognitive functioning in relation to diagnosis:

1. Children with cyanotic lesions will perform at a lower level of intellectual functioning pre-operatively compared with children with acyanotic defects.
2. Corrective surgery will result in a greater increase in cognitive function for children with cyanotic defects compared with those with acyanotic lesions.
3. Age at time of repair of the cardiac defect will be positively correlated with a greater degree of post-operative cognitive impairment in children with cyanotic defects, but for children with acyanotic lesions there will be no association between age at repair and degree of cognitive impairment.

In addition, a specific set of hypotheses relating to psychosocial functioning were tested:

CHILDREN

4. There will be a higher prevalence of behaviour problems both before and at 12 months after surgery among those patients with a cyanotic lesion compared with those with an acyanotic lesion, and in the group with acyanotic defects the prevalence

of behaviour problems will be higher in those with symptomatic, compared with asymptomatic, lesions.

5. Positive self perception, both before and at 12 months after treatment, will be correlated with Rutter A scores at 12 months.

6. For the older age group (5-17 years) those who score as poorly adjusted on the Rutter A and B scales at 12 months will have a longer mean time after surgery before returning to school compared with those whose Rutter A or B scores are below the cut-off points.

7. Time in hospital since discharge will be correlated with Rutter A and B scores at 12 months.

8. Pre-operative adjustment of the child will be significantly associated with post-operative adjustment.

CHILD-MOTHER

9. There will be no relationship between mothers' and clinicians' perception of disease severity.

10. Maternal levels of psychological distress at 12 months will be correlated with Rutter A scores of the child at 12 months.

PARENTS

11. Pre-operatively, those mothers showing psychological distress will perceive their child's disease as more severe compared with those mothers showing no psychological distress, regardless of clinicians' assessment of actual urgency of treatment.

12. Pre-operative levels of parental distress will be significantly associated with post-operative distress.

FAMILY

13. Psychosocial functioning of the child, siblings and parents will be more compromised in the cardiac and BMT groups compared with the healthy group, but the cardiac and BMT groups will not differ significantly from each other.

4.3. EXPERIMENTAL DESIGN

Psychological adjustment of children and their families was evaluated by comparing the psychosocial functioning of the child and family before and after cardiac surgery using:

1. a longitudinal assessment of the cardiac surgery patients and their families
2. a cross-sectional comparison between cardiac surgery patients and their families and two reference groups.

4.3.1. Longitudinal Assessment

Psychosocial functioning of the child and family was assessed at two times - just prior to surgery and one year after surgery. The pre-operative assessment was performed in the hospital during the few days immediately before surgery. Ideally this would have been carried out in the child's home but this option was precluded for practical reasons, such as the distance children lived from the hospital and the short notice given for surgery. The pre-operative assessment therefore focused retrospectively on functioning prior to the hospital admission. The post-operative assessment was carried out during a routine out-patient follow-up at either the treating or referring hospital.

4.3.2. Cross-sectional Comparison with Reference Groups

Two reference groups were available with which to compare the cardiac surgery group:

1. a sample of children undergoing bone marrow transplantation (BMT) and their families
2. a sample of normal healthy children with no medical problems and their families

Both cardiac surgery and BMT were seen as potentially life- threatening interventions for potentially fatal or chronic conditions, necessitating hospitalisation and causing a degree of physical discomfort to the child.

The healthy group had not experienced any of the stressors related to the presence of a chronic illness and they served to provide a "baseline", against which the effects of the illness and hospitalisation could be compared. Both reference groups were seen twice in 12 months, the BMT group, like the cardiac surgery group, being seen first before treatment.

4.4. SELECTION OF CASES

4.4.1. Cardiac Group

4.4.1.1. Sample

Consecutive admissions for surgery were seen from three London cardiac units during the period July 1984 until June 1986.

4.4.1.2. Inclusion/Exclusion Criteria

Selection was according to the following criteria:

1. The presence of a congenital heart defect for which cardiac surgery was seen as a suitable treatment.
2. Age range: 0 - 17 years.
3. Patients and families were resident in the U.K. and were potentially able to attend the 12 month follow-up appointments.

Patients were excluded on the following criteria:

1. The patient was too ill to participate.
2. The family did not speak English sufficiently well to enable adequate communication.
3. Families were not willing to cooperate with the study.

One in 20 children born with a heart defect has Down's syndrome (Kenna, Smithells & Fielding, 1975) and 40% of children born with Down's syndrome have a defect of the heart and/or great vessels (Rowe & Uchida, 1961), with atrioventricular septal defects being the most commonly occurring lesions (Rowe & Uchida, 1961; Park, Mathews, Zuberbuhler, Rowe, Neches & Lenox, 1977). A degree of mental retardation is invariably associated with Down's syndrome, with the mean intelligence quotient being about 50 (Burn, 1987). To include such patients in the main experimental group would cause distortion of its homogeneity on average level of cognitive function, but to exclude them would mean that the sample was not fully representative of children awaiting cardiac surgery as a whole. It was therefore decided to include patients with Down's Syndrome but to look at mental retardation as a separate variable.

4.4.2. The BMT Group

4.4.2.1. Sample

Selection of the BMT patients was based on consecutive admissions to four London units during the period September 1983 to June 1986.

4.4.2.2. Inclusion/exclusion Criteria

Selection criteria were the same as for the cardiac cases. Some patients awaiting BMT had Hurlers or Hunters syndrome and were therefore intellectually subnormal but, as with the cardiac children with Down's syndrome, they were included as a subgroup of the BMT group.

4.4.3. Healthy Group

4.4.3.1. Sample

As part of another study (Pot-Mees, 1989), each child in the healthy group was individually matched with a BMT patient, and selection was from local health centres, the dental department at Westminster Children's Hospital and local schools.

4.4.3.2. Inclusion/exclusion Criteria

Selection was according to the following criteria:

1. The BMT and healthy child were of the same sex.
2. The BMT and healthy child were of the same age, using an age margin of +/- 3 months for children under the age of 5 years and +/- 6 months for children over 5 years of age.

3. The BMT and healthy child were at the same level of cognitive function, in terms of normal or subnormal. Criteria for normal cognitive function were a) being in normal schooling, where appropriate, and b) parents/caretakers reporting the child to be of normal intellectual abilities. Subnormal cognitive function was defined as having the diagnosis of a mental disorder such as Down's syndrome, with a mild (IQ: 50-80) degree of mental handicap.

4. The BMT and healthy child fell into the same broad category of socioeconomic status, according to the profession of the main breadwinner in the house. The Standard Occupational Classification (1966) was used and for the purpose of the study grouped into:

i) Non-manual (professional, intermediate, skilled non-manual)

ii) Manual (skilled manual, partially skilled, unskilled/unemployed)

After identification of suitable cases, exclusion was still possible according to the following criteria:

1. occurrence of serious medical problems;
2. permission not given by parents/caretakers;
3. parents and/or child did not speak English sufficiently well;
4. inability to attend for follow-up.

4.5. INSTRUMENTS

This study focuses on patients, siblings and parents. Pertinent factors from the literature review are investigated with respect to these groups, according to the model proposed below (Fig. 1).

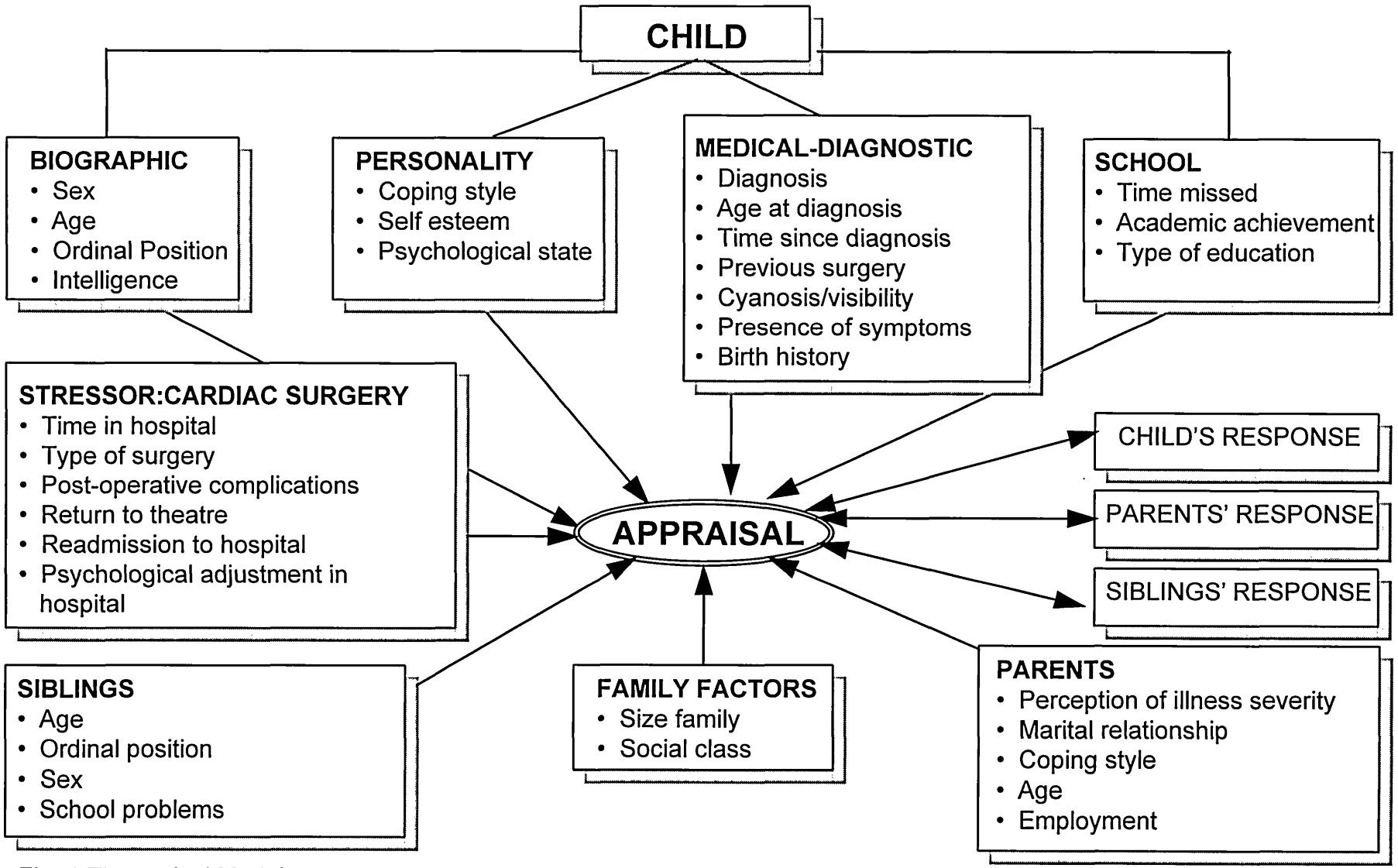


Fig. 1 Theoretical Model

4.5.1. Child Measures

4.5.1.1. Cognitive Ability

4.5.1.1.1. The Ruth Griffiths Mental Development Scales

The Ruth Griffiths Mental Development Scales (Griffiths, 1954, 1970) were used to assess the developmental status of children below three and a half years of age. The scales involve testing of the child but also rely on mothers' reports, elicited by specific questioning - e.g. about eating habits, toilet training etc. For children younger than two years there are five subscales: locomotor, personal-social, hearing-speech, eye-hand coordination and performance. A sixth scale, the practical reasoning scale, is added from the third year onwards. They are widely used scales which have been standardised on British children (Griffiths, 1954, 1970) although some concerns have been expressed over the use of mothers' reports, with greater inter-rater reliability being reported on those scales assessing coordination and performance than on the others (Aldridge Smith, Bidder, Gardner & Gray, 1980; Hanson, Aldridge Smith & Hume, 1984). The scales have been used with chronically ill or handicapped children (Bidder, Bryant & Gray, 1975; Balestrini et al, 1990; Ludman, Spitz & Lansdown, 1993) and are the most widely used infant scale in Britain (Tyler & Miller, 1986).

4.5.1.1.2. The British Ability Scales

4.5.1.1.2.1. IQ Assessment

For children older than 3.5 years the British Ability Scales (BAS)* were used to assess intellectual status. A short form IQ estimate of the BAS was used (Elliott, 1983), comprising measures of verbal and non-verbal reasoning ability, short term memory, speed of information

processing and retrieval of knowledge skills. The scales have been validated on a British population and are used in a wide range of diagnostic contexts as an aid in identification, classification and selection of children with learning difficulties (Elliott, 1983, 1986). They have also been used in the study of children with chronic illness (Jannoun, 1983; Twaddle et al, 1983). The BAS was chosen in preference to the Wechsler scales (Wechsler, 1974) because of its standardisation on a British, rather than an American, population and also because it has the facility to be used with a large age-range. Adequate correlation has been reported between the BAS and Wechsler scales (Elliott, 1983; Jannoun, 1983).

*Footnote: Whilst the term "British Ability Scales" is used to refer to the scales in the plural in the manual for the tests, the abbreviation "BAS" is used in the singular and will therefore be used in that way in this thesis.

4.5.1.1.2.2. School Attainments

To assess academic performance, children of 3.5 to 14.5 years completed the basic number skills (BAS) attainment test to assess arithmetic, and children of 5 to 14.5 years completed the word reading subtest (BAS) and the Schonell graded spelling test (Schonell & Schonell, 1949). The measures of academic attainment have been previously used in studies of chronically ill children (Jannoun & Chessels, 1987).

4.5.1.2. Behaviour

There are numerous methods of assessing behaviour and adjustment of children, including assessments by parents and/or teachers, and direct psychiatric assessments. In this study a direct psychiatric assessment was not carried out for a number of reasons. First, there was

apprehension at the treatment centres that a direct interview of the child on the day prior to surgery or at the onset of BMT could have harmful effects and possibly affect the child's attitude to the treatment procedure. Secondly, for the cardiac patients in particular, there was insufficient time to carry out a lengthy pre-operative psychiatric interview. Thirdly, such an interview just before surgery would probably have doubtful validity. Fourthly, it has been suggested elsewhere that information obtained from the child in such an interview contributes little to the overall clinical judgement based on information obtained from parents (Verhulst, Althaus & Berden, 1987). Information was, however, collected from parents and also from teachers, recognised as an objective source of information regarding the child's functioning (Rutter et al, 1970).

4.5.1.2.1. Standardised Measures

4.5.1.2.1.1. Measures Completed by Parents

Due to the lack of suitable measures no behaviour instrument was used for those children under three years of age.

4.5.1.2.1.1.1. Richman Behaviour Checklist

For children of 3 to 5 years behaviour at home was assessed with the Richman Behaviour Checklist (Richman & Graham, 1971; Richman, 1979), a 21-item questionnaire. Reliability and validity have been reported to be satisfactory (Richman & Graham, 1971) and the recommended scoring methods were used. A score of 9 or more was used to identify those children with a significant degree of problem behaviour. The Richman BCL has been previously used in British studies of chronically ill children (Reynolds, Garralda, Postlethwaite & Goh, 1991; Bradford, 1994).

4.5.1.2.1.1.2. Rutter A Scale

Behaviour of children of 5 to 17 years was assessed with the Rutter A Scale (Rutter et al, 1970), a 31-item questionnaire. Reliability and validity have been reported to be satisfactory (Rutter et al, 1970) and the recommended scoring methods were used. A criterion score of 13 was used to identify those children with a significant degree of problem behaviour at home and behaviour disturbance was classified into antisocial or neurotic behaviour disorders, based on the total scores on the items of each of these dimensions (Rutter et al, 1970), or, if the neurotic and antisocial scores were equal, a classification of "undifferentiated" was given.

4.5.1.2.1.2. Measures Completed by Teachers

Behaviour at school of children of 5 to 17 years was assessed with the Rutter B Scale (Rutter, 1967; Rutter et al, 1970), a 26-item questionnaire. Using the recommended scoring methods a criterion score of 9 was used to identify those children with a significant degree of problem behaviour at school. As with the Rutter A Scale, behavioural disturbance was classified into the two diagnostic categories of antisocial and neurotic behaviour.

The Rutter Scales have been extensively used in British studies, including studies of physical illness and mental handicap in childhood (Stevenson, Hawcroft, Lobascher, Smith, Wolff & Graham 1979; Eiser, 1980(a); Jannoun, 1983; Sawyer, Crettenden & Toogood, 1986; Udwin, Yule & Martin, 1987; Garralda et al, 1988; Chisholm, Bloomfield & Atkinson, 1994). The correlation between the Rutter A and B scales is low due to the differences between parental and teachers' perceptions and to the situation-specificity of children's behaviour (Graham, 1967; Rutter et al, 1970).

4.5.1.3. Observation

The research worker also spent a session with the child during which they did house, tree and person drawings (Buck, 1948) and had the opportunity to talk about their illness and impending treatment.

4.5.1.4. Semi-Structured Interview

In order to collect further demographic, medical and social information on the child and family an interview was held with the parents. Due to the lack of appropriate interview schedules, the existing ones of which tended to be too lengthy and therefore impractical for use in the limited time available for interviewing parents before surgery, a semi-structured interview was designed specifically for use in the study. It was used with parents of children of 3 years and older and covered eight areas of behaviour, which were selected on the basis of the effects of chronic illness documented in the literature (Korsch, Negrete, Gardner, Weinstock, Mercer, Grushkin & Fine, 1973; Koocher & O'Malley, 1981; Eiser, 1985). The areas investigated were: reaction in a new situation; temper tantrums; disobedience; anxieties; relationships with peers; relationships with adults; degree of dependency and level of activity. Parents' descriptions of their child's behaviour in each of these areas were rated on a 3 point scale of 1 (no problems), 2 (some problems), or 3 (definite problems). The interview data were recorded on a printed schedule in order to gain further insight into the behaviour of the children. A further general question, asking parents whether they considered their child to have behaviour problems, was scored in the same way.

Parents were further asked how their child generally reacted to stress, in order to provide information on the child's coping style. Parents were asked to describe their child's

behaviour and the researcher then categorised their response into one of the following options: aggression, withdrawal, calmness, excitement and emotional/tearful behaviour. These categories were selected on the basis of work done on the coping styles of adults and children (Folkman & Lazarus, 1980; Caty et al, 1984). If parents felt that their child did not exhibit any of these coping modes, their response was categorised as undifferentiated.

4.5.1.5. Self Perception

A visual analogue scale, devised specifically for this research in the absence of any more suitable measures, was used to assess self-perception in the children of 5 years and over. Utilising constructs and elements - based on the technique of semantic differentials (Osgood, Suci & Tannenbaum, 1957) - the instrument was designed to provide information about how the child perceived him/herself on a series of different dimensions. Eight constructs considered to be of particular salience for the self-perception of chronically ill children (Burns & Zweig, 1980) were used and were represented on individual cards with a cartoon drawing and descriptive words at each end to depict the two extremes of the construct (Fig. 13; Appendix A). The constructs were body image (weak-strong), mood (feels like crying-cheerful), self image (likes self-doesn't like self), sociability (feels lonely-has friends), anxiety (safe-frightened), aggression (angry-calm), self esteem (bad-good) and physical health (ill-well). Each of these was rated for two elements, "self" and "ideal self". Each construct was rated on a 5 point scale, with the child placing the picture cards representing the elements in the appropriate box. In order to prevent the child automatically choosing the same pole each time, four of the cards were reversed so that half of the cards had the negative pole at the extreme left hand side and half had the positive pole at the left hand end. In addition to the individual construct scores, a total self perception score was computed (by

scoring all of the eight constructs from 1 (most negative) to 5 (most positive)). Differences between ratings of "self" and "ideal self" on the eight constructs were also analysed. A small study utilising this technique with 15 hospitalised children and 15 control children has shown satisfactory test-retest reliability, but further assessment of the reliability and validity of the scale is required. For the purposes of this study, the internal consistency between the items of the scale was used as a measure of reliability and was found to be satisfactory ($\alpha = .84$).

4.5.1.6. Parental Assessment of Child's Self Perception

Previous researchers have found poor agreement between parents' and children's reports of children's adjustment to and experience of chronic illness (Weissman, Orvaschel & Padian, 1980; Nelms, 1986; Ennett, DeVellis, Earp, Kredich, Warren & Wilhelm, 1991; Engstrom, 1992). The importance of cross-validating children's and parents' reports has been emphasised by these authors. Utilising the same visual analogue scale, parents were therefore asked to rate how they perceived their child on the 8 constructs. This enabled comparisons to be made between the child's perception of themself and parental perception of the child. Parents were also asked to rate their child on a 5 point scale of introversion/extroversion.

4.5.2. Sibling Measures

4.5.2.1. Behaviour at Home

The same structured questionnaires were completed on the siblings as for the patients - namely the Richman BCL (Richman & Graham, 1971; Richman, 1979) for children of 3 - 5 years and the Rutter A scale (Rutter et al, 1970) for children of 5 - 17 years.

During the interview with the parents, they were asked a general question about the occurrence of behaviour problems in the sibling(s) and this was scored in the same way as for the patient.

4.5.2.2. Behaviour at School

Behaviour of the siblings of 5 - 17 years was assessed with the Rutter B Scale (Rutter, 1967; Rutter et al, 1970), completed by teachers.

During the interview with the parents, they were asked whether or not the sibling had academic and/or adjustment problems at school. These were individually scored as a yes/no response.

4.5.3. Measures of Parental Functioning

In addition to the semi-structured interview and the questionnaires for completion on their child's behaviour, parents were asked to complete a series of questionnaires relating directly to themselves. Separate questionnaires were given to each parent. Research on adjustment in children has found that a number of parental factors are likely to be salient. Parents' - and in particular mothers' - mental state (Cox, 1988; Caplan, Coghill, Alexandra, Robson, Katz & Kumar 1989; Dumas, Gibson & Albin, 1989), the presence of marital discord (Porter & O'Leary, 1980), parents' perceived locus of control (Campis, Lyman & Prentice-Dunn, 1986; DeMaso et al, 1991) and parental coping (Kupst et al, 1982) have all been found to influence the adjustment of the child. Marital satisfaction and parental depression have also been found to influence parenting practices (Stoneman, Brody & Burke, 1989). Furthermore, studies have shown that the chronic illness of a child can affect parental

adjustment, with findings of poorer mental health in the mothers of chronically sick children compared with the mothers of healthy children (Breslau et al, 1982; Wallander et al, 1989; Miller, Gordon, Daniele & Diller, 1992; Quittner, DiGirolamo, Michel & Eigen, 1992). It has been suggested that the stress of having a sick child can have a significant and negative impact on the parental relationship (Lansky et al, 1978; Tew et al, 1974) although, contrary to popular belief, higher than normal levels of divorce have not been found (Sabbeth & Leventhal, 1984). Questionnaires were therefore selected to measure these parental variables.

4.5.3.1. Mental State

The mental and emotional state of parents was assessed with the 30 item version of the General Health Questionnaire (GHQ) (Goldberg, 1972, 1978) which consists of questions concerned with psychological distress or altered behaviour. For each item the respondent is asked to compare their recent state with their normal state, with a four point response scale for each item. The GHQ focuses on two major classes of phenomena: inability to continue to carry out one's normal "healthy" functions and the appearance of new phenomena of a distressing nature (Goldberg, 1978). Satisfactory reliability and validity have been reported and the GHQ has been used in a number of studies in Britain (Goldberg, 1978; Tarnopolsky, Hand, McLean, Roberts & Wiggins, 1979). It has also been used in studies of parents of chronically sick children (Reynolds et al, 1988; Vandvik & Eckblad, 1991).

4.5.3.2. Marital Satisfaction

The Dyadic Adjustment Scale (DAS) (Spanier, 1976), the most widely used marital/partner adjustment scale (Kronenberger & Thompson, 1992), was chosen to assess parental

satisfaction within their married/unmarried dyadic relationship. To save time, the "Dyadic Satisfaction" subscale was used, rather than the full 32-item questionnaire, a practice justified by the author of the scale (Spanier, 1976). This 10 item scale rates overall happiness and satisfaction with the relationship. Satisfactory reliability and validity have been reported (Spanier, 1976; Spanier & Thompson, 1982; Antill and Cotton, 1982). In addition, responses to one global item assessing overall happiness within the relationship were looked at, which has been found to be a sufficient means of classifying respondents into high and low adjustment groups (Sharpley & Cross, 1982). The DAS has been previously used in studies of parents of chronically sick and disabled children (Spaulding & Morgan, 1986; Kazak, Reber & Snitzer, 1988; Gordon Walker et al, 1992; Kronenberger & Thompson, 1992).

4.5.3.3. Locus of Control

The most widely used locus of control scale (Rotter, 1966) was felt to be too lengthy and too non-specific for this study, so a questionnaire was devised specifically for this research. The six item scale comprised those five items from the Rotter scale which had a correlation of .30 or higher with the total test score (Rotter, 1966) and one question from a similar questionnaire by Van der Ploeg (1983). Parents were asked to rate the six statements on a scale of 1 - 4, from the most internal to the most external, although the polarity was reversed in three questions to safeguard against parents habitually selecting the same "box". The internal consistency between the items (α=.56) was taken as a measure of reliability.

4.5.3.4. Coping Style

The Utrecht Coping List (UCL) (Schreurs, Tellegen & Van de Willige, 1984) was used to assess the coping strategies of the parents. This Dutch scale was translated for the study and

consists of 47 statements of different coping behaviours which are rated by the parents on a 4 point scale according to how often they react in that way. The item scores are clustered into 7 coping styles - actively solving, palliation, avoidance, social support, depression, expression of emotions and comforting thoughts. The instrument has been reported to have satisfactory reliability and validity (Schreurs, Van de Willige, Tellegen & Brosschot, 1988).

4.5.3.5. Parental Opinions About Services Offered and Concerns for the Future

Parents were asked a series of questions at the post-operative follow-up about whether or not they were satisfied with a number of aspects of care received at the time of treatment - namely, communication with staff, accommodation, facilities, support and medical management. They were also asked whether they had any concerns relating to their child's health and the future.

4.6. STATISTICAL ANALYSIS

4.6.1. Child Measures

Developmental and cognitive measures at each test occasion were compared by one way analysis of variance (Table 4.1). Scheffe's multiple comparison tests were used to identify the source of any significant differences ($p<.05$) between the cardiac, BMT and healthy groups, and between the cyanotic, acyanotic, BMT and healthy groups. Performance of the cyanotic and acyanotic subgroups was compared using independent t-tests. Within group changes over time in developmental and cognitive function were compared using paired t-tests. Between group comparisons over time were assessed using repeated measures analysis of variance.

STATISTICAL TESTS USED IN STUDY		
CHILD MEASURES		
SCALE	COMPARISON	TEST USED
Ruth Griffiths and British Ability Scales	Cardiac and reference groups	1-way analysis of variance
	Cyanotic and acyanotic groups	Independent t-test
	Within group changes over time	Paired t-test
	Between group changes over time	Repeated measures analysis of variance
	Correlation of age and performance	Pearson correlation coefficient
	Effects of age on performance	Analysis of variance
Academic parameters	Incidence of underachievement in cardiac and reference groups	Chi-squared test*
Richman and Rutter Scales	Cardiac and reference groups	Chi-squared test
	Association between pre- and post-operative behaviour problems within each group	Chi-squared test
	Correlation between time in hospital/time before returning to school and adjustment	Spearman correlation coefficient
Parental interview, self-perception and parental perception	Cardiac and reference groups	Mann-Whitney test
	Within group changes over time	Wilcoxon test
Incidence of behaviour problems on parental interview	Cardiac and reference groups	Chi-squared test

PARENT MEASURES		
SCALE	COMPARISON	TEST USED
General Health Questionnaire	Cardiac and reference groups	Chi-squared test
	Association between pre- and post-operative distress levels	Chi-squared test
Marital relationship and locus of control scales	Cardiac and reference groups	1-way analysis of variance
	Cyanotic and acyanotic groups	Independent t-test
	Within group changes over time	Paired t-test
	Between group changes over time	Repeated measures analysis of variance
Disease severity	Mothers' and physicians' perceptions	Kruskal-Wallis analysis of variance
Maternal GHQ scores and child Rutter A scores	Within group	Spearman correlation coefficient
Mothers' GHQ scores and perception of disease severity	Cardiac group	Kruskal-Wallis analysis of variance
SIBLING MEASURES		
Richman and Rutter scales	Cardiac and reference groups	Chi-squared test
	Association between pre- and post-operative behaviour problems within each group	Chi-squared test

* Where appropriate, the Fisher Exact test was substituted for the chi-squared test.

Table 4.1

Correlations of age and performance were measured using Pearson correlation coefficients and the effects of age were assessed with analysis of variance with age as a covariate. The differences between the groups in the incidence of underachievement on academic parameters and reading problems were assessed using chi-squared tests.

The prevalence of behaviour problems measured on the Richman and Rutter scales was compared using chi-squared tests. The degree of association between the prevalence of pre- and post-operative behaviour problems within each group was measured with a chi-squared test. Correlations between time spent in hospital or time before returning to school and adjustment were measured using Spearman correlation coefficients.

Between group comparisons of parental interview, self perception and parental perception data were performed by Mann-Whitney tests. Within group changes over time were assessed using a Wilcoxon test. Between group comparisons of the prevalence of behaviour problems on the parental interview were measured by chi-squared tests.

4.6.2. Parent Measures

The GHQ scores were analysed using non-parametric tests. Between group comparisons of levels of psychological distress were performed by chi-squared tests (Table 4.1). The association between pre- and post-operative psychological distress levels was also assessed by chi-squared tests and the direction of change between the pre- and post-operative ratings was assessed by a Wilcoxon test.

The marital relationship scale and locus of control scale results were compared by one way analysis of variance and Scheffe's multiple comparison tests were used to identify the source of any differences between the groups. Scores in the cyanotic and acyanotic subgroups were compared by independent t-tests. Within group changes over time were analysed by paired t-tests. Between group changes over time were assessed by repeated measures analysis of variance.

The relationship between mothers' and physicians' perceptions of disease severity was assessed using a Kruskal-Wallis analysis of variance. Spearman correlation coefficients were calculated to determine whether or not there was a correlation between the presence of maternal psychological distress and behaviour problems in the patient. To see whether or not there was a relationship between maternal psychological distress and maternal perception of disease severity, irrespective of clinicians' assessment of the urgency of treatment, a new variable was computed to reflect the possible combinations of psychological distress and urgency of surgery, and a Kruskal-Wallis analysis of variance was then performed with maternal perception of illness severity and this new variable as the grouping variable.

4.6.3. Sibling Measures

Sibling measures, all of which were non-parametric, were analysed in the same way as the patient non-parametric measures (Table 4.1).

The SPSS statistical package was used for the analyses (SPSS for Windows, 1992).

A number of symbols have been used throughout the presentation of the results to represent specific probability values, as listed below:

SYMBOL	PROBABILITY VALUE
**	$p<.05$
*	$p<.01$
#	$p<.005$
~	$p<.001$

4.7. DATA COLLECTION

4.7.1. Assessment of the Cardiac Group

In the majority of cases, admission for cardiac surgery only occurred one or two days before the operation. Each week the research worker contacted the cardiac centres to determine the planned admissions for the week and then visited the family after their admission to the ward. The research worker discussed the project with the family and gave them an information sheet and a consent form to sign to indicate their agreement to participate in the study. Due to the short time between admission and surgery, assessments were carried out the day before surgery. In most instances this was in a room away from the ward. Parents' and children's wishes were respected as to whether or not parents were present during the assessment of the child, although for all of the younger age group parents were present due to the positive influence of parental presence during testing (Magrab & Lehr, 1982). The assessments all followed the same format, starting with the developmental or cognitive testing. For school-age children this was followed by the assessment of school attainments, the semantic differential technique and finally some drawings and an informal conversation. Assessment of the children usually lasted between 45 and 90 minutes (with the exception of some of the developmental testing of the young babies, which took less time).

Parents were seen for an interview after the child was seen but before surgery, and in the first instance this was intended to facilitate the development of the relationship between the research worker and the parents. Initially the interview focused on medical information and experiences prior to admission and was in essence to put the family experience of the patient's illness into context. The second part of the interview was used to collect information on family and demographic factors and on the development, behaviour and personality of the child. In some cases there was insufficient time to complete the second part of the interview in a single session and it was necessary for the research worker to return at a later date. Parents were given their questionnaires to complete and asked to return them in the pre-stamped envelope provided. With parental consent, Rutter B questionnaires, together with a covering letter, were also sent to the teachers of each of the school-age children in the family.

At the initial assessment children and families were also told that they would be seen again 12 months after surgery. Whilst the majority of patients were not due to be seen at the treating hospital at this time, they were invariably seen at their referring hospital approximately 12 months after treatment. In order to standardise the testing environment arrangements were made with the families and their local hospitals to see them at a routine out-patient visit and, with the exception of one child who was seen at home, all of the children were seen in a hospital outpatient department. The assessment of the child followed a format identical to that of the initial test occasion and parents were additionally seen for an interview during which information was collected on the adjustment of the child and family and on events of the past year. Parents were given another set of questionnaires to be

completed at home and questionnaires were again sent to the teachers of the patients and siblings.

4.7.2. Assessment of the Reference Groups

4.7.2.1. The BMT Group

In the majority of cases data collection took place during the first two weeks after admission, before the BMT treatment had started. The same format of informed consent, testing and interviews was used as for the cardiac group. The date of the 12 month follow-up assessment was arranged so that it coincided with an out-patient visit to the hospital. In contrast to the cardiac group, patients continued to be followed-up at the treating, rather than the referring, centre, so that the initial and follow-up assessments could be carried out at the same hospital.

4.7.2.2. The Healthy Group

On both test occasions the family were seen at home, where the assessment of the child and parental interviews were conducted using the same format as for the cardiac and BMT groups. Parents were asked to complete the same questionnaires. Due to the individual matching of the healthy and BMT groups, follow-up of the healthy group was primarily of those children matched with a BMT survivor.

CHAPTER 5

STUDY SAMPLE

5.1. SAMPLE SELECTION

5.1.1. Experimental Group: Sample Selection

Initially cardiac cases were selected from the paediatric cardiac units at Westminster and Hammersmith hospitals. During 1985 Westminster Hospital stopped operating on paediatric patients and instead the children underwent surgery at the Brompton Hospital, from which further patients were recruited. During the period July 1984 until June 1986, 75 cardiac surgery patients were seen from the cardiac units of the Brompton (n=38; 51%), Hammersmith (n=31; 41%) and Westminster (n=6; 8%) Hospitals. Ninety-eight patients were initially eligible for inclusion, but 23 (23.5%) had to be excluded because they were too ill at the time of admission for surgery (n=12), unavailable for follow-up (n=3), not sufficiently able to communicate in English (n=7) or declined to participate (n=1).

5.1.2. Reference Groups: Sample Selection

BMT patients were selected from 4 units in the London and Greater London areas. During the period September 1983 to June 1986 seventy-five patients were enrolled into the study from the BMT units at Westminster Children's Hospital (n=44; 59%), Great Ormond Street (n=17; 23%), Hammersmith Hospital (n=10; 13%) and the Royal Marsden Hospital (n=4; 5%). Ninety-six patients were initially eligible for inclusion but 21 (22%) had to be excluded because they were too ill at the time of entering treatment (n=10), unavailable for follow-up (n=4), not sufficiently able to communicate in English (n=6) or declined to participate (n=1).

The BMT group formed part of another study and the healthy group were initially collected as matched controls for the BMT group. Children were selected for the healthy reference group from local health centres (n=25; 33%), the dental department register at Westminster Children's Hospital (n=35; 47%) and local schools (n=15; 20%). After identification of a suitable child, parents were approached by letter which was followed up by a telephone call. From September 1984 until June 1986, 94 families were initially approached, 19 (20%) of whom were excluded due to the occurrence of serious medical problems (n=5), permission not given by parents/caretakers (n=7), not sufficiently able to communicate in English (n=4) and unavailability for follow-up (n=3).

5.2. DATA COLLECTION

5.2.1. Assessment of the Cardiac Group

The rate of completion of the parental questionnaires at the initial assessment was satisfactory (mothers: 73%; fathers: 70%). Of the 75 patients tested pre-operatively, 54 children (72%) were seen 12 months after treatment. Twenty-one patients could not be seen for follow-up because they died during or after the operation (n=11), they did not want to participate in another assessment (n=6) or they could not be traced (n=4).

5.2.2. Assessment of the Reference Groups

5.2.2.1. The BMT Group

Four of the 75 BMT patients (5.3%) could not be tested because they had already started the isolation procedures. Completion rates for the parental questionnaires at the initial assessment were satisfactory (mothers: 85%; fathers: 71%). The forty-three patients alive one year after BMT were followed-up.

5.2.2.2. The Healthy Group

The completion rates for the parental questionnaires at the initial assessment were again satisfactory (mothers: 88%; fathers: 88%). Forty-nine healthy children were followed-up after one year.

5.3. DESCRIPTION OF THE SAMPLE

The sample is described in terms of: 1). sociodemographic; 2). educational; 3). developmental history prior to assessment; 4). medical history; 5). medical intervention and 6). family data

5.3.1. Sociodemographic Data

The mean age of the cardiac group at the time of the initial assessment was 5.2 years (range: 0.2 - 14.8 years) and there were 45 boys and 30 girls. Distribution of the sample according to socioeconomic status indicated that slightly more children came from the non-manual classes. Only 3 of the 75 children had a degree of mild mental handicap, the majority functioning in the normal range.

Comparison with the reference groups (Table 5.1) indicated that on these matching criteria the only significant difference between the three groups was the mean age. The cardiac patients were significantly younger than the BMT and healthy groups ($p<0.05$) and this is attributable to the fact that all of the cardiac group had congenital heart disease, which was generally diagnosed at birth or soon after. Corrective surgery was frequently performed within the first five years, which has been shown to be more beneficial for later medical and psychological functioning (O'Dougherty et al, 1983; Nadas, 1984; Newburger et al, 1984).

SOCIODEMOGRAPHIC DATA			
	CARDIAC (n=75)	BMT (n=75)	HEALTHY (n=75)
AGE (YEARS):			
RANGE	0.2-14.8 **	0.3-16.7	0.3-16.7 **
MEAN	5.2**	6.6**	6.7
S.D.	3.8	4.2	4.2
SEX:			
BOYS	45 (60%)	40 (53%)	40 (53%)
GIRLS	30 (40%)	35 (47%)	35 (47%)
COGNITIVE LEVEL:			
NORMAL	72 (96%)	66 (88%)	66 (88%)
SUBNORMAL	3 (4%)	9 (12%)	9 (12%)
SOCIOECONOMIC STATUS:			
NON-MANUAL	40 (53%)	32 (43%)	36 (48%)
MANUAL	35 (47%)	43 (57%)	39 (52%)
RESIDENCY:			
< 30 MILES FROM LONDON	40 (53%)	19 (25%)	72 (96%)
30-100 MILES FROM LONDON	20 (27%)	21 (28%)	3 (4%)
> 100 MILES FROM LONDON	15 (20%)	35 (47%)	0 (100%)

** $p < .05$

Table 5.1

The majority of the patients were of Caucasian background (88%), the remainder being of Asian (9%) or Afro-Caribbean (3%) origin. A similar ethnic distribution was found in the reference groups. Forty (53%) children were resident within 30 miles of London, with the remainder coming from further afield in Wales and England. For those children living outside London there tended to be a clustering in specific geographical locations due to the fact that some of the London based cardiologists also held local clinics elsewhere in the U.K. and sent patients seen in the joint local clinics to their London cardiac unit for treatment. In the BMT

group only 25% (n=19) children lived in the London area whereas 96% (n=72) of the healthy children were resident within 30 miles of London.

5.3.2. Education

Information on the education received by children in the 3 groups is given in Table 5.2.

In the cardiac group none of the 3 children with mental handicap were attending school. Of the children with mental handicap in the BMT group, 2 were attending special schools and 1 was attending a normal playgroup. In the healthy group 2 children with mental handicap were attending special schools.

EDUCATION OF CHILDREN WITHIN THE THREE GROUPS (EXCLUDING MENTALLY HANDICAPPED SUBGROUPS)

	CARDIAC (n=72)	BMT (n=66)	HEALTHY (n=66)
< 3 years	23 (32%)	12 (18%)	13 (20%)
> 3 years and eligible for education	49 (68%)	54 (82%)	53 (80%)
Not receiving any education:			
3-5 years	2 (4%)	2 (4%)	0 (0%)
5-17 years	0 (0%)	3 (6%)	0 (0%)
Special education	2 (4%)	1 (2%)	0 (0%)
Normal education	45 (92%)	48 (89%)	53 (100%)

Table 5.2

Whilst the majority of the cardiac patients attended school up until the time of their admission for surgery - with, on average, 5 days (S.D. 6 days) elapsing between their last school attendance and date of admission - the BMT patients had missed a significantly greater amount of time from school, with an average period of 99 days (S.D. 88 days) between their last school attendance and admission for BMT ($t=6.32$; $p<0.001$).

5.3.3. Developmental History Prior to Assessment

Thirty-one (44%) children in the cardiac group were known to have experienced problems at birth, generally due to prematurity or related to the cardiac lesion. (Information was not available on 5 patients). In 25 (35%) cases this necessitated admission of the child to a special care baby unit (SCBU). Many parents reported problems with their child during infancy, primarily related to poor feeding and failure to thrive, and 11 (21%) children were known to have exhibited developmental delay. In the majority of cases this was delayed locomotor development, where the child was not walking unaided by the age of 18 months. This profile did not differ significantly from that of the BMT group. In this group, 19 (25%) children had problems at birth, necessitating SCBU treatment in 14 cases, and 10 (15%) children were reported as exhibiting developmental delay, 7 of whom had genetic disorders.

5.3.3.1. Previous Psychological/Emotional Problems

Five of the cardiac surgery patients were known to have been referred to a psychologist for problems which were predominantly of an emotional nature. This was similar to the BMT and healthy groups.

5.3.3.2. Coping Style

The following table shows the coping styles utilised by the children of 3 years and older when confronted with a stressful situation (Table 5.3).

REACTION IN A STRESSFUL SITUATION			
	CARDIAC (n=44)	BMT (n=55)	HEALTHY (n=54)
Withdrawal	5 (11.4%)	16 (29.1%)	14 (25.9%)
Anger	4 (9.1%)	21 (38.2%)	4 (7.4%)
Emotional	19 (43.2%)	9 (16.4%)	22 (40.7%)
Calm	3 (6.8%)	2 (3.6%)	4 (7.4%)
Excited	7 (15.9%)	4 (7.3%)	7 (13.0%)
Undifferentiated	6 (13.6%)	3 (5.4%)	3 (5.6%)

Table 5.3

The cardiac group showed similar patterns of reaction to the healthy group, with tearfulness being the most prevalent response. In contrast, the BMT group more frequently tended to react with aggression or withdrawal.

5.3.4. Medical History

5.3.4.1. Diagnosis

Within the cardiac group, the heart defects (all congenital) were divided into cyanotic and

acyanotic defects. Twenty-nine (39%) children had cyanotic disorders and 46 (61%) had acyanotic disorders (Table 5.4).

DIAGNOSES IN THE CARDIAC GROUP	
DIAGNOSIS	NUMBER
Cyanotic	29
Fallot's Tetralogy	11
Pulmonary Atresia	6
Single Ventricle	6
Atrioventricular Canal	3
Transposition of the Great Arteries	2
Truncus Arteriosus	1
Acyanotic	46
Ventricular Septal Defect	15
Atrial Septal Defect	12
Pulmonary Stenosis	4
Patent Ductus Arteriosus	4
Coarctation of the Aorta	3
Aortic Valve Stenosis	3
Ventricular/Atrial Septal Defect	2
Partial Atrioventricular Canal	2
Double Outlet Right Ventricle	1

Table 5.4

In the BMT group 37(49%) children had genetic disorders and 38 (51%) had acquired disorders (Table 5.5).

DIAGNOSES IN THE BMT GROUP	
DIAGNOSIS	NUMBER
Acquired Disorders	38
Leukaemia: Acute Lymphoblastic Leukaemia	20
Acute Myeloid Leukaemia	6
Chronic Granulocytic Leukaemia	2
Aplastic Anaemia	5
Neuroblastoma	2
B Cell Lymphoma	2
Solid Tumour	1
Genetic Disorders	37
Hurler's Syndrome	9
B Thalassemia Major	8
Fanconi's Anaemia	7
Gaucher's Disease	6
Hunter's Syndrome	3
Severe Combined Immune Deficiency	3
Niemann-Pick Disease	1

Table 5.5

5.3.4.2. Age at Diagnosis

For the cardiac group the mean age at diagnosis was 0.6 years (S.D. 1.59 years), with the cyanotic patients being significantly younger than the acyanotic patients (0.08 vs 0.92 yrs, $t=-2.94$; $p<0.005$). The cardiac group, as expected, were significantly younger at diagnosis than the BMT patients, whose mean age at diagnosis was 4.6 years (S.D. 4.0 years). Those children with genetic disorders were diagnosed at a younger age than those with acquired conditions.

5.3.4.3. Previous Hospitalisation

Seventy (93%) of the children in the cardiac surgery group had had at least one previous hospital admission, 25 (36%) of whom had had previous surgery. In the remaining 45 cases hospitalisation had usually been for cardiac catheterisation and other diagnostic tests and was therefore of short duration. However, 17 children who had not had previous surgery had required hospital admission for chest infections, heart failure etc. and in some instances admissions had been frequent and/or for several weeks duration. In the BMT group 73 of the 75 (97%) children had previously been in hospital, although those with genetic disorders had spent less time in hospital than those with acquired disorders. For the majority of children with acquired disorders some form of treatment was started on diagnosis. Of those with acquired disorders, 63.2% had spent the period leading up to the BMT in hospital, compared with 13.5% of those with genetic diseases.

5.3.5. Medical Interventions and Outcome

5.3.5.1. Age at Treatment

The mean age of the cardiac patients at treatment was 5.2 years, with no significant differences between the age at treatment of the cyanotic (5.02 years, S.D. 4.03 years) and the acyanotic (5.31 years, S.D. 3.74 years) groups. As previously mentioned, the cardiac patients were significantly younger at the time of treatment than the BMT patients ($p<0.05$) but when the BMT group were subdivided according to the type of disorder, it was found that the age at treatment for the genetic group was similar to that for the cardiac group (4.9 years, S.D. 2.9 years) but that the group of patients with acquired disorders were significantly older at the time of treatment (8.3 years, S.D. 6.5 years).

5.3.5.2. Type of Treatment

Seventy (93%) of the 75 cardiac surgery patients had corrective surgery, while in 4 cases (5%) surgery was palliative. One child with truncus arteriosus was taken to theatre and her chest opened, but the lesion was found to be inoperable and no further surgical procedures were performed.

In 63 cases (85%) treatment involved open-heart surgery, the remainder having closed heart procedures.

In the BMT group one child did not receive a transplant because of social and behavioural problems. Of the 74 children who did undergo BMT, 60 (81.1%) had an allogeneic transplant, 12 (16.2%) had an autologous transplant and 2 (2.7%) had a syngeneic*

transplant. Forty-two of the 74 patients (56.8%) were treated with total body irradiation as part of their conditioning regimen, most of whom (n=35; 83.3%) had acquired disorders.

* Syngeneic refers to transplants in which the donor and recipient are genetically identical; allogeneic transplants involve genetically dissimilar donors and recipients.

5.3.5.3. Time in Hospital

The average time spent in hospital by the cardiac group was 20 days (Range:3-355; median:14). (Those children who spent less than 7 days in hospital all died in the perioperative period). The cyanotic patients spent longer in hospital (mean: 30 days) than the acyanotic patients (mean: 13 days) but this difference did not reach significance. The BMT patients spent significantly longer in hospital than the cardiac group (t= 8.26; p=.000), with a mean hospital stay of 67 days (Range: 25-176; median:60). Those children with genetic disorders tended to spend longer in hospital than the children with acquired disorders (means of 76 and 57 days respectively).

5.3.5.4. Complications of Medical Intervention

Thirteen children in the cardiac group had unforseen problems - as defined by the operating team - during surgery, such as bleeding, difficulties coming off bypass etc. In a further 4 cases post-operative complications necessitated returning to theatre. Nine children had problems both during surgery and during the post-operative period. There was also a high incidence of complications in the BMT group, with 38 children experiencing acute and/or chronic graft versus host disease and 25 having problems with Cyclosporin A toxicity. Seven children (9.5%) needed a second BMT due to initial graft failure.

5.3.5.5. Cancellation of Treatment

Nine patients (12%) in the cardiac surgery group had their planned surgery cancelled having once been admitted to hospital, which was reported by the parents of those children as being an additional source of stress.

5.3.5.6. Survival

At the time of discharge 64 cardiac patients (85%) were alive. Of the 11 children who died, 8 patients had cyanotic defects and 3 patients had acyanotic defects, which represents 28% of the cyanotic group and 7% of the acyanotic group. There were no late deaths in the cardiac group, with all of the patients who were alive at discharge still alive 12 months after surgery. At the time of discharge 54 of the 74 BMT children (73%) were alive, 20 children having died during the hospitalisation. During the period between discharge and the 12 month follow-up a further 11 children died, primarily due to relapse of their initial disease.

5.3.6. The Families

5.3.6.1. Parents

The majority of children in all three groups came from intact two-parent families with two children. Table 5.6 shows the family composition for the three groups, together with mean parental ages.

The size of the family in the cardiac group was significantly smaller than that of the BMT group (chi-squared = 21.51019; p=.0006) and of the healthy group (chi-squared = 12.56746; p=.0278).

FAMILY DEMOGRAPHIC DATA			
	Cardiac	BMT	Healthy
Two parents	70 (93%)	67 (89%)	64 (85%)
One parent	5 (7%)	8 (11%)	11 (15%)
Ordinal position of patient:			
only child	19 (25%)	3 (4%)	9 (12%)
eldest	15 (20%)	24 (32%)	20 (27%)
youngest	33 (44%)	34 (45%)	31 (41%)
middle	8 (11%)	14 (19%)	15 (20%)
Number of children under 18 years:			
1	19 (25%)	4 (5%)	11 (15%)
2	28 (37%)	46 (61%)	41 (55%)
3	22 (29%)	14 (19%)	10 (13%)
4+	6 (8%)	11 (15%)	13 (17%)
Mean Age: Mothers (Years)	33.30**	34.05	35.97**
S.D. (Years)	5.95	6.83	6.37
Mean Age: Fathers (Years)	36.55	35.76**	39.71**
S.D. (Years)	7.09	7.33	8.35

** $p<.05$

N.B. In one BMT family and 2 families of healthy children, the patient was one of 2 or more children but the other child(ren) were older than 18 years of age.

Table 5.6

Seven of the 70 (10%) fathers in the cardiac group were unemployed. The rate of unemployment in the BMT group was 24% but in the healthy group none of the 64 fathers were unemployed. The rate of unemployment was significantly higher in the cardiac and BMT groups compared with the healthy group (chi-squared = 4.88399; p=.0271; chi-squared = 14.99816; p=.0001 for the cardiac and BMT groups respectively).

Within the cardiac group 5 (7%) mothers and 4 (6%) fathers had received treatment at some time for psychiatric symptoms. The incidence of parental psychiatric problems was similar for all three groups.

5.3.6.2. Siblings

The following table (Table 5.7) shows the age and sex distribution of the siblings under 18 years of age in the three groups, together with the prevalence of chronic and/or life threatening illnesses.

SIBLINGS: AGE AND SEX DISTRIBUTION, ORDINAL POSITION AND PREVALENCE OF CHRONIC ILLNESS

	CARDIAC (n=86)	BMT (n=108)	HEALTHY (n=105)
Mean age (yrs):	7.95	7.82	7.80
S.D.:	4.44	4.94	4.80
Females:	43 (50%)	52 (48%)	60 (57%)
Males:	43 (50%)	56 (52%)	45 (43%)
Ordinal position: eldest:	37 (43%)	43 (40%)	38 (36%)
middle or other:	29 (34%)	32 (30%)	35 (33%)
youngest:	20 (23%)	33 (30%)	32 (31%)
Medical Problems:			
None/minor:	74 (86%)	90 (83%)	94 (90%)
Chronic/life threatening:	6 (7%)	18 (17%)	11 (10%)
Not known:	6 (7%)	0 (0%)	0 (0%)

Table 5.7

Educational status of the siblings in each of the 3 groups is given in Table 5.8.

EDUCATION OF SIBLINGS WITHIN THE THREE GROUPS			
	CARDIAC (n=86)	BMT (n=108)	HEALTHY (n=105)
< 3 years	12 (14%)	27 (25%)	20 (19%)
> 3 years and eligible for education	74 (86%)	81 (75%)	85 (81%)
Not receiving any education: 3-5 years 5-17 years	 1 (1%) 1 (1%)	 4 (5%) 1 (1%)	 1 (1%) 0 (0%)
> 16 years and left school	2 (3%)	2 (2%)	2 (2%)
Special education	2 (3%)	1 (1%)	2 (2%)
Normal education	68 (92%)	73 (90%)	80 (94%)

Table 5.8

CHAPTER 6

PRE-OPERATIVE COGNITIVE AND BEHAVIOURAL FUNCTIONING

The results will be presented in turn of each area of functioning tested - developmental and cognitive, behaviour at home, behaviour at school, parental interview, self perception and parental perception. For each area, results for the younger children will be presented first, followed by those for the older group. In each area results for the whole cardiac group will be presented first, looking specifically at any differences between the cyanotic and acyanotic patients and the cardiac group will then be compared with the reference groups. In areas where there is a significant difference between the cyanotic and acyanotic patients, comparisons will also be made between these two subgroups and the 2 reference groups. The influence, (if any), of the child's age on development and cognition will be described for the cardiac group. The pre-operative results of those patients who were not seen after surgery will be compared where appropriate with those who were followed up. Finally, the results will be briefly discussed in terms of the existing literature on children with chronic illness and congenital heart disease in particular.

6.1. DEVELOPMENTAL FUNCTION: 0 - 3.5 YEARS

6.1.1. Cardiac Group

In the cardiac group there were 24 children with no known mental handicap who were assessed with the Ruth Griffiths developmental scales. One patient who was eligible for assessment was not well enough to be assessed. Those with cyanotic heart disease (n=10) and those with acyanotic lesions (n=14) did not differ on any of the developmental parameters (Table 6.1).

PRE-OPERATIVE DEVELOPMENTAL FUNCTIONING						
SCALE	CYANOTIC (n=10)	ACYANOTIC (n=14)	TOTAL CARDIAC (n=24)	BMT (n=14)	HEALTHY (n=15)	COMMENTS
LOCOMOTOR	#		~	*	#~*	* p<.01
MEAN D.Q.	92.00	96.50#	94.63	99.57	124.00 #	# p<.005
S.D.	27.18	13.46	19.91	20.49	12.08	~ p<.001
PERSONAL-SOCIAL	**	#	~	~	** # ~ ~	**p<.05 # p<.005
MEAN D.Q.	105.80	103.50	104.46	95.07	121.87	~ p<.001
S.D.	14.98	9.40	11.79	6.64	14.88	
SPEECH/HEARING			*		*	* p<.01
MEAN D.Q.	98.60	102.36	100.79	96.69 *	120.27 *	
S.D.	16.50	12.83	14.25	18.24	23.34	
EYE/HAND COORDINATION						
MEAN D.Q.	103.40	101.64	102.38	103.28	112.27	
S.D.	10.75	6.91	8.55	13.54	16.90	
PERFORMANCE						
MEAN D.Q.	112.80	106.64	109.21	104.93	118.60	
S.D.	13.70	15.05	14.53	17.21	15.03	
PRACTICAL REASONING	(n=4)	(n=2)	(n=6)	(n=3)	(n=7)	
MEAN D.Q.	99.50	106.50	101.83	92.67	118.57	
S.D.	15.52	6.36	12.88	4.04	20.90	
	#		~		# ~	~ p<.001
OVERALL I.Q.	102.30	102.14	102.21	99.93 ~	119.27 ~	# p<.005
S.D.	9.74	7.21~	8.16	10.18	12.33 ~	

DQ = Developmental Quotient

Table 6.1

6.1.2. Comparison with the Reference Groups

Fourteen BMT patients and 15 healthy children were also assessed using the same measures, with one BMT patient not being assessed due to poor health. Pre-operative testing indicated that all of the mean subtest scores and overall IQ score were within the normal range for all three groups (Griffiths, 1970). However, both the cardiac and BMT groups obtained significantly lower scores on the locomotor, personal-social and speech and hearing scales, together with the overall IQ score, compared with the healthy group (Table 6.1). There were no significant differences between the two groups of hospitalised patients. Comparison of the cyanotic and acyanotic subgroups with the reference groups indicated no significant

differences between either subgroup and the BMT group, but both the cyanotic and acyanotic subgroups differed significantly from the healthy group on the locomotor and personal-social subscales and on the overall IQ.

6.1.3. Significance of Age

The mean age of the two cardiac subgroups was not significantly different. (Cyanotic group: mean age 1.68 years; S.D .0.7857 years; acyanotic group: mean age 1.40 years; S.D. 0.8814 years; p=.431). In the cyanotic group performance on the eye-hand coordination subscale was negatively correlated with age (r=-.7171; p=.010). In the acyanotic group performance on the locomotor subscale was positively correlated with age (r=.6324; p=.008) (Table 6.2). Using analysis of variance to compare the 2 subgroups, with age as a covariate (see page 172), age made a significant contribution to the locomotor and eye-hand coordination scores. However, when the effects of age were controlled for there were still no significant differences in performance between the cyanotic and acyanotic subgroups.

PRE-OPERATIVE DEVELOPMENTAL FUNCTIONING: 0 - 3.5 YEARS: CORRELATION OF AGE WITH PERFORMANCE FOR THE CYANOTIC AND ACYANOTIC SUBGROUPS, EFFECT OF AGE ON THE DIFFERENCE IN PERFORMANCE BETWEEN THE TWO SUBGROUPS, AND PROBABILITY VALUES FOR DIFFERENCES IN PERFORMANCE BETWEEN THE TWO SUBGROUPS CONTROLLING FOR AGE

	CORRELATION OF AGE AND PERFORMANCE		COMPARISON OF PERFORMANCE BETWEEN CYANOTIC AND ACYANOTIC SUBGROUPS WITH AGE AS A COVARIATE	
	CYANOTIC (n=10)	ACYANOTIC (n=14)	EFFECTS OF AGE	CONTROL-LING FOR AGE
SCALE	CORRELATION: AGE & SCORE	CORRELATION: AGE & SCORE		
LOCOMOTOR	r=- .4740 p= .083	r= .6324 p= .008	t= 2.663 p= .015	p=.307
PERSONAL-SOCIAL	r=- .0296 p= .468	r= .0604 p= .419	t= .069 p= .946	p=.669
SPEECH/HEARING	r=- .2972 p= .202	r= -.0864 p= .384	t= - .823 p= .420	p=.639
EYE/HAND COORDINATION	r=- .7171 p= .010	r= -.1603 p= .292	t=-2.101 p= .048	p=.393
PERFORMANCE	r= .3422 p= .167	r= .3238 p= .125	t= 1.622 p= .120	p=.448
PRACTICAL REASONING	r=-1.000 p= .000	r= .6765 p= .162	t= .894 p= .437	p=.705
OVERALL IQ	r= .0749 p= .419	r= .3377 p= .119	t= 1.002 p= .328	p=.903

Table 6.2

6.1.4. Mentally Handicapped Subgroups

IQ scores of the children with known mental retardation indicated that these children in each of the three groups functioned in the range of mild to moderate mental handicap (Table 6.3). The cardiac group performed at a lower level on all of the subtests compared with the reference groups, particularly on the locomotor scale, but these differences failed to reach statistical significance.

PRE-OPERATIVE DEVELOPMENTAL FUNCTIONING: 0 - 3.5 YEARS :
MENTAL HANDICAP SUBGROUPS:
COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS

SCALE	CARDIAC (n=3)	BMT (n=9)	HEALTHY (n=9)
LOCOMOTOR			
MEAN DQ:	39.33	65.67	69.78
S.D.	27.23	17.75	14.90
PERSONAL-SOCIAL			
MEAN DQ:	56.33	66.67	69.89
S.D.	18.72	12.66	11.36
SPEECH/HEARING			
MEAN DQ:	53.33	55.44	52.67
S.D.	20.55	22.45	14.38
EYE/HAND COORD			
MEAN DQ:	63.33	71.22	66.00
S.D.	22.75	15.68	9.51
PERFORMANCE			
MEAN DQ:	53.33	80.33	66.89
S.D.	17.56	18.51	13.01
PRACTICAL REASONING	(n=0)	(n=1)	(n=3)
MEAN DQ:	-	61.00	61.00
S.D.		.00	8.54
OVERALL IQ:	53.33	70.33	65.00
S.D.	18.56	14.83	10.52

Table 6.3

6.1.5. Children not Followed-Up

Of the 24 cardiac children assessed pre-operatively, 18 were followed up at one year after surgery. Of the six who were not followed, 5 died and 1 was lost to follow-up. Comparison of the follow-up sample and those who died indicated that the two samples only differed significantly on the locomotor scale, with those children not followed-up obtaining a lower score ($t=3.06$; $p=.006$).

6.2. COGNITIVE ABILITY: 3.5 - 17 YEARS

6.2.1. Cardiac Group

All children in this age range ($n=47$) were assessed with the British Ability Scales. Differences were found between the 17 children with cyanotic heart disease and the 30 with acyanotic heart disease. The cyanotic group showed poorer performance than the acyanotic group on all of the subtests. The differences reached significance on the matrices ($p=0.001$) and speed of information processing ($p=0.007$) subtests (Table 6.4), with the difference on the similarities subtest almost reaching significance ($p=.055$).

The overall IQ scores were within the normal range for each subgroup, as were the arithmetic and reading attainments. However, the cyanotic patients achieved a significantly lower IQ score than the acyanotic group ($p<0.001$) and obtained significantly lower scores on all of the school attainments compared with the acyanotic patients (Table 6.4).

PRE-OPERATIVE COGNITIVE FUNCTIONING: 3.5 - 17 YEARS						
SCALE	CYANOTIC	ACYANOTIC	TOTAL CARDIAC	BMT	HEALTHY	COMMENTS
COGNITIVE						
RECALL OF DIGITS MEAN T SCORE S.D.	(n=16) 49.31 9.74	(n=29) 52.93 9.26	(n=45) 51.64 9.48	(n=47) 49.11 9.44	(n=51) 53.18 9.68	
VISUAL RECOGNITION MEAN T SCORE S.D.	(n= 5) 51.80 10.09	(n= 5) 63.20 13.50	(n=10) 57.50 12.74	(n= 5) 63.40 11.41	(n= 7) 58.29 10.19	
NAMING VOCABULARY MEAN T SCORE S.D.	(n=10) 50.70 10.19	(n=21) 56.76 9.85	(n=31) 54.81 10.20	(n=21) 49.95 11.12	(n=23) 53.87 8.81	
VERBAL REASONING MEAN T SCORE S.D.	(n= 5) 49.40 8.30	(n= 5) 54.40 2.61	(n=10) 51.90 6.37	(n= 6) 54.67 7.03	(n= 7) 56.29 4.57	
MATRICES MEAN T SCORE S.D.	(n=12)~** 47.00 13.71	(n=24)~# 61.83 9.27	(n=36) 56.89 12.88	(n=41)# 52.76 8.77	(n=44)** 55.66 8.47	~ p=.001 ** p=.05 # p .005
SIMILARITIES MEAN T SCORE S.D.	(n=12) + 48.85 12.22	(n=24) + 55.40 8.39	(n=36) 53.28 10.07	(n=41) 51.39 9.19	(n=44) 53.50 7.92	+ p=.055
SPEED OF INFORMATION PROCESSING MEAN T SCORE S.D.	(n= 7)+~ ** 40.86 11.05	(n= 9) + 59.89 12.38	(n=16) ** 51.56 15.02	(n=25) ** 55.84 9.52	(n=28)**~ 60.82 9.40	+ p=.007 ** p=.05 ~ p=.001
ACADEMIC						
OVERALL BAS: MEAN IQ S.D.	(n=17)## # 96.06 19.83	(n=30)## ** 114.93 14.19	(n=47) 108.11 18.65	(n=47) ** 104.57 14.18	(n=51) # 111.00 12.52	## p .0001 # p=.005 ** p .05
ARITHMETIC: MEAN IQ S.D.	(n=16)+ 98.19 20.81	(n=29)+* 111.59 17.51	(n=45)** 106.82 19.62	(n=41)** * 97.63 13.56	(n=46) 105.24 15.97	+ p=.027 ** p .05 * p .01
READING: MEAN IQ S.D.	(n=12)+ 92.92 18.30	(n=24)+* 107.42 17.52	(n=36)** 102.58 18.84	(n=35)**#* 92.66 16.05	(n=39)# 105.41 12.81	+ p=.027 ** p=.05 # p .005 * p=.01
SPELLING: MEAN IQ S.D.	(n=12)+ ** 81.00 20.08	(n=24)+ 98.42 15.37	(n=36) 92.61 18.74	(n=29)** 87.48 17.17	(n=39)** ** 98.21 14.06	+ p=.007 ** p=.05

Table 6.4

To assess overall academic levels of achievement, a score was calculated for each child to measure achievement on their arithmetic and reading performance relative to overall IQ (Thomson 1982; Elliott, 1983). The incidence of reading problems was also investigated

using as a criterion a T score of more than one standard deviation below the norm (Table 6.5). Underachievement was more prevalent in the acyanotic group. Compared with the acyanotic group, the cyanotic group was performing at a comparable level on school attainments and overall IQ, but they had a higher prevalence of reading problems (although this failed to reach statistical significance).

PRE-OPERATIVE ACADEMIC FUNCTIONING UNDERACHIEVEMENT ON ARITHMETIC AND READING AND THE PREVALENCE OF READING PROBLEMS					
	CYANOTIC	ACYANOTIC	TOTAL CARDIAC	BMT	HEALTHY
NUMBER UNDER-ACHIEVING ON ARITHMETIC	1 (8%)	3 (13%)	4 (11%)	9 (22%)	11 (28%)
NUMBER UNDER-ACHIEVING ON READING	2 (17%)	7 (29%)	9 (25%)	13 (37%)	7 (18%)
INCIDENCE OF READING PROBLEMS	4 (33%)	2 (8%)	6 (17%)	12 (34%)	3 (8%)

CY = CYANOTIC
ACY = ACYANOTIC

Table 6.5

6.2.2. Comparison with the Reference Groups

Four of the 51 BMT patients eligible for testing were not well enough to be assessed, so that 47 completed the IQ testing, and 51 normal children were also assessed with the same measures. Performance on all of the subtests and the overall IQ of each of the three groups

fell within the normal range. However, the cardiac group performed at a significantly lower level than the healthy group on the speed of information processing subtest (Table 6.4).

Comparison of the cyanotic and acyanotic subgroups with the reference groups indicated that the cyanotic patients performed at a significantly lower level than the BMT group on speed of information processing. The cyanotic group also performed at a significantly lower level than the healthy group on matrices and speed of information processing. The acyanotic patients did significantly better than the BMT patients on the matrices subtest.

Performance on the school attainments was also within the normal range for the three groups. However, the BMT patients attained significantly lower scores than the cardiac patients on the arithmetic and reading tests. There were no significant differences between the total cardiac group and the healthy controls. (Table 6.4). However, the cyanotic group obtained significantly lower scores than the healthy group on the overall IQ and on the spelling attainment. The acyanotic patients performed at a significantly higher level than the BMT group on the overall IQ, arithmetic and reading tests.

The achievement status and incidence of reading difficulties are given in Table 6.5. There were no significant differences in the prevalence of underachievement on reading and arithmetic or in the prevalence of reading problems between the cardiac group and either of the 2 reference groups, but the BMT group had a significantly higher prevalence of reading problems than the healthy group (Chi-squared = 6.51015; p=.0107).

6.2.3. Effects of Age

As the number of children completing each test was not always the same, and because the test results could be influenced by age, the mean ages of the cyanotic and acyanotic groups were compared for each test. No significant differences were found between the cyanotic and acyanotic children who completed each of the subtests.(See Appendix B; Table 1).

Age was significantly correlated with performance on the matrices and similarities subtests in both the cyanotic and acyanotic subgroups. (In both of the reference groups there was also a significant correlation between age and performance on the matrices subtest: r=-4.115; p=.01 and r=-.3827; p=.01 for the BMT and healthy groups respectively). Age was also significantly correlated with performance on recall of digits in the cyanotic group and with performance on speed of information processing in the acyanotic group. The effect of age on the difference in performance between the two subgroups was significant on the matrices and similarities subtests (Table 6.6). However, the significant differences remained between the two subgroups on the matrices and speed of information processing subtests even when the effects of age were controlled for.

There were again no significant differences in mean age between the two subgroups assessed on overall IQ and school attainments. Performance on overall IQ, arithmetic and spelling was significantly correlated with age in the cyanotic group. In the acyanotic group age was significantly correlated with arithmetic. (In the healthy group there was also a significant correlation between age and performance on arithmetic: r=-.5785; p=.001). The effect of age on the difference in performance between the two subgroups was significant on the overall IQ, arithmetic and spelling (Table 6.6).

PRE-OPERATIVE COGNITIVE FUNCTIONING: 3.5 - 17 YEARS: CORRELATION OF AGE WITH PERFORMANCE FOR THE CYANOTIC AND ACYANOTIC SUBGROUPS, EFFECT OF AGE ON THE DIFFERENCE IN PERFORMANCE BETWEEN THE TWO SUBGROUPS, AND PROBABILITY VALUES FOR DIFFERENCES IN PERFORMANCE BETWEEN THE TWO SUBGROUPS CONTROLLING FOR AGE				
	CORRELATION OF AGE AND PERFORMANCE		COMPARISON OF PERFORMANCE BETWEEN CYANOTIC AND ACYANOTIC SUBGROUPS WITH AGE AS A COVARIATE	
	CYANOTIC	ACYANOTIC	EFFECTS OF AGE	CONTROLLING FOR AGE
SCALE	CORRELATION: AGE & SCORE	CORRELATION: AGE & SCORE		
RECALL OF DIGITS	r=-.4830 p= .029	r=-.0261 p= .446	t=-1.394 p= .171	p=.227
VISUAL RECOGNITION	r=-.5111 p= .189	r=-.0584 p= .463	t=- .658 p= .532	p=.156
NAMING VOCABULARY	r=-.2764 p= .220	r= .2549 p= .132	t= .345 p= .732	p=.171
VERBAL REASONING	r=-.2383 p= .350	r=-.1495 p= .405	t=- .503 p= .630	p=.229
MATRICES	r=-.7669 p= .002	r=-.6118 p= .001	t=-5.282 p= .000	p=.001
SIMILARITIES	r=-.8123 p= .001	r=-.3589 p= .039	t=-3.585 p= .001	p=.130
SPEED OF INFORMATION PROCESSING	r= .1851 p= .346	r= .6618 p= .026	t= 2.041 p= .062	p=.004
BAS IQ	r=-.6067 p= .005	r=-.1168 p= .269	t=-2.442 p= .019	p=.000
ARITHMETIC IQ	r=-.7660 p= .000	r=-.3846 p= .020	t=-4.315 p= .000	p=.031
READING IQ	r=-.4548 p= .069	r=-.0603 p= .390	t=-1.202 p= .238	p=.056
SPELLING IQ	r=-.6543 p= .010	r=-.2333 p= .136	t=-2.580 p= .015	p=.020

Table 6.6

Controlling for age resulted in the significant differences remaining between the cyanotic and acyanotic groups on overall IQ, arithmetic and spelling, but on the reading test the difference was no longer significant, with the probability value changing from .027 (see Table 6.4) to .056.

6.2.4. Children not Followed-up

Of the 47 cardiac children assessed pre-operatively, 35 were followed-up one year after surgery. Of the 12 not seen at one year, 5 died perioperatively and 7 were lost to follow-up. Comparisons were made between the 35 followed up, 5 who died and 7 who were alive but not followed-up.

In general, performance of those children who were lost to follow-up resembled that of the follow-up sample on the majority of tests. In contrast, the children who died performed at a significantly lower level than the follow-up sample on the majority of parameters. Performance of those children who died was significantly worse than the follow-up sample on matrices ($p<.0005$), similarities ($p=.004$) and recall of digits ($p=.010$) and on overall IQ ($p<.0005$), arithmetic ($p=.004$) and spelling ($p=.022$). On the naming vocabulary and recall of digits subtests the follow-up sample obtained significantly higher scores than the non-followed up survivors ($p=.015$ and $p=.032$ respectively).

6.3. BEHAVIOUR AT HOME: 3 - 5 YEARS

6.3.1. Cardiac Group

Thirteen children in the cardiac group were in the age range of 3-5 years. Completed Richman BCL questionnaires were received on 8 (62%), of whom 3 (37%) obtained a score

of 9 or more, indicating behaviour problems at home. Two of the 8 children had cyanotic lesions and 6 had acyanotic defects. Comparison of the 2 subgroups indicated no significant differences in the prevalence of problem behaviour at home (Table 6.7) although the small numbers involved are likely to have introduced a type II statistical error.

PRE-OPERATIVE SCORES ON THE RICHMAN BEHAVIOUR CHECKLIST		
	NUMBER SCORING < 9	NUMBER SCORING ≥ 9
CYANOTIC (n=2)	1 (50%)	1 (50%)
ACYANOTIC (n=6)	4 (67%)	2 (33%)
TOTAL CARDIAC (n=8)	5 (63%)	3 (37%)
BMT (n=5)	4 (80%)	1 (20%)
HEALTHY (n=8)	7 (88%)	1 (12%)

Table 6.7

6.3.2. Comparison with the Reference Groups

Ten BMT patients and 9 healthy children were eligible for completion of the Richman BCL and completed questionnaires were received on 5 BMT patients and 8 healthy children. There were no significant differences in the prevalence of problem behaviour between the three groups (Table 6.7).

6.3.3. Children not Followed-up

Nine of the 13 cardiac children seen pre-operatively were followed up. Of the 4 not followed-up, 3 died and 1 was lost to follow-up. Completed questionnaires were not received on any of these children.

6.4. BEHAVIOUR AT HOME: 5 - 17 YEARS

6.4.1. Cardiac Group

Of the 37 children aged 5-17 years, Rutter A Scale questionnaires were returned on 29 (78%). Five obtained a score of 13 or more, indicating a significant degree of problem behaviour at home. Three of these had neurotic problems and 2 had antisocial problems. Eight of the 29 had cyanotic lesions and 21 had acyanotic defects. Comparison of the 2 subgroups indicated no significant differences in the prevalence of problem behaviour at home or in the nature of the problems (Table 6.8). Within the acyanotic group 5 were symptomatic and 16 were asymptomatic. There were no significant differences in the prevalence of problem behaviour between these two groups.

PRE-OPERATIVE SCORES ON THE RUTTER A SCALES		
	NUMBER SCORING < 13	NUMBER SCORING ≥ 13
CYANOTIC (n= 8)	6 (75%)	2 (25%)
ACYANOTIC (n=21)	18 (86%)	3 (14%)
TOTAL CARDIAC (n=29)	24 (83%)	5 (17%)
BMT (n=40)	35 (88%)	5 (12%)
HEALTHY (n=42)	36 (86%)	6 (14%)

Table 6.8

6.4.2. Comparison with the Reference Groups

Of the 44 children in each of the BMT and healthy groups eligible for the Rutter A questionnaire, completed questionnaires were received on 40 BMT patients and 42 healthy

children. Comparison of these with the cardiac group indicated no significant differences in the prevalence of problem behaviours at home (Table 6.8).

A comparable proportion of children had neurotic and antisocial patterns of behaviour in each of the three groups.

6.4.3. Children not Followed-up

Of the 37 cardiac children in the age range 5 - 17 years seen pre-operatively, 3 died during surgery or soon after and a further 6 were lost to follow-up. Completed questionnaires were received on 3 of the 6 lost to follow-up but on none of those who died. In those cases where the children died it was not considered appropriate to remind parents to complete the forms after the child's death. Of the 3 lost to follow-up on whom Rutter A questionnaires were completed, one child was rated as showing antisocial behaviour at home.

6.5. BEHAVIOUR AT SCHOOL

6.5.1. Cardiac Group

Questionnaires were sent to the teachers of the 37 children who were aged 5 years or older and were attending school. Completed Rutter B questionnaires were received on 33 (89%). Seven (21%) obtained a score of 9 or more, indicative of a significant degree of problem behaviour at school. Four children had neurotic problems, 1 had antisocial problems and in 2 cases the problems were undifferentiated.

Nine of the 33 had cyanotic lesions and 24 acyanotic defects and comparison of the 2 subgroups indicated no significant differences (Table 6.9).

PRE-OPERATIVE SCORES ON THE RUTTER B SCALES		
	NUMBER SCORING < 9	NUMBER SCORING ≥ 9
CYANOTIC (n= 9)	6 (67%)	3 (33%)
ACYANOTIC (n=24)	20 (83%)	4 (17%)
TOTAL CARDIAC (n=33)	26 (79%)	7 (21%)
BMT (n=30)	23 (77%)	7 (23%)
HEALTHY (n=44)	36 (82%)	8 (18%)

Table 6.9

6.5.2. Comparison with the Reference Groups

Completed Rutter B questionnaires were returned on 30 of the 41 BMT patients aged 5 or older and attending school and on all 44 healthy children of 5 years or older. Comparison of the 3 groups indicated no significant differences in the prevalence of problem behaviour at school (Table 6.9).

In the cardiac and BMT groups the majority of children had neurotic type behaviour problems (57% and 86% respectively) but only 37% of children in the healthy group had problems of a neurotic nature, with the majority (50%) having antisocial type behaviour problems.

6.5.3. Children not Followed-up

Of the 37 cardiac children seen pre-operatively, 9 were not followed-up. The 6 children lost to follow-up all had completed Rutter B forms, 1 of whom had behaviour problems at school

of a neurotic nature. Completed questionnaires were not obtained for any of the 3 children who died, even though they were at school prior to surgery and questionnaires were sent.

6.6. PARENTAL INTERVIEW: 3 - 17 YEARS

6.6.1. Cardiac Group

Fifty children were 3 years or older and parental interview data concerning the child's behaviour were collected on 44 (88%), 12 of whom had cyanotic lesions and 32 of whom had acyanotic defects. Cyanotic children were rated as being significantly more dependent (U=122.5; p=.0291) and also less active (U=129.0; p=.0133) than acyanotic children (See Appendix B: Table 2).

Four children (9%) were judged by their parents to have behaviour problems, 3 of whom had cyanotic lesions and 1 had an acyanotic defect. There was a significant difference in the prevalence of behaviour problems between the 2 subgroups (chi squared =6.31354; p=.0426).

For the 21 acyanotic patients there was a significant association between the presence of behaviour problems on the Rutter A and the presence of behaviour problems at home according to the interview (Chi-squared = 6.3000; p=.0121). However, this association was not significant for the 8 cyanotic patients on whom both measures were available (Fisher's exact test: p=.53571) or for the total cardiac group (chi-squared = .0000; p=1.0000).

The 6 children on whom data were not collected all died. Practical reasons - primarily that the children were admitted such a short time before surgery - precluded collecting the data pre-operatively.

6.6.2. Comparison with the Reference Groups

Fifty-seven BMT patients and 55 healthy children were 3 years or older and parental interview data were collected on 54 children in each group. The cardiac and BMT groups did not differ on any of the parental interview items. The cardiac group were significantly more anxious (U=725.5; p=.0002), more dependent (U=817.0; p= .0004) and less active (U=974.0; p=.0183) than the healthy group (See Appendix B: Table 3). The BMT group were significantly more anxious (U=1023.5; p=.0096) and dependent (U=956.5; p=.0036) and had more difficulties in relationships with other children (U= 1087.0; p=.0389) than the healthy group.

Comparison of the cyanotic and BMT groups indicated that the cyanotic children were significantly more disobedient (U=228.0; p=.0179), more dependent (U=180.0; p=.0216) and less active (U=191.5; p=.0089). Relative to the healthy group the cyanotic group were also judged to be more disobedient (U=232.0; p=.0163), anxious (U= 159.5; p=.0004), dependent (U=145.5; p=.0000) and less active (U=190.0; p=.0001) (See Appendix B: Table 4). The acyanotic group did not differ from the BMT group on any of the items but they were rated as more anxious (U=565.5; p=.0017) and more dependent (U=671.5; p=.0116) than the healthy group (See Appendix B: Table 5).

There were no significant differences in parental ratings of behaviour problems between the cardiac, BMT and healthy groups (9%, 6% and 11% for the 3 groups respectively) or between the cyanotic or acyanotic subgroups and the 2 reference groups.

Comparison of Rutter A and parental interview data indicated a significant association between the presence of behaviour problems on the two measures for the 42 healthy children (chi-squared = 4.28588; p=.0384) but not for the 40 BMT patients (chi-squared = .05148; p=.8205).

6.6.3. Children not Followed-up

All of the 7 children seen pre-operatively but not after surgery were lost to follow-up; none died. They did not differ from the follow-up sample on any of the behavioural parameters.

6.7. SELF PERCEPTION

6.7.1. Cardiac Group

Thirty-one of the 37 children (84%) aged between 5 and 17 years completed the self-perception test. Two children refused to complete the task and in 4 cases it was considered that the children had not understood what was required of them, so the test was not completed. Eleven of the 31 children performing the test had cyanotic lesions and 20 had acyanotic defects. There were no significant differences between the two subgroups on any of the constructs (See Appendix B: Table 6). In order to examine whether self perception varied in younger and older children, each subgroup was divided into two according to the median age of the total cardiac sample. The median age was 7.3 years. In the younger age group the cyanotic children rated themselves as stronger than the acyanotic children (U=5.5; p=.0453) whereas in the older age group the cyanotic children rated themselves as weaker (U=16.0; p=.0441).

As well as rating themselves on the 8 constructs, children were also asked to rate their "ideal self". There were no significant differences between the cyanotic and acyanotic groups (See Appendix B: Table 7).

Finally, a score was computed for each construct which reflected the difference between "self" and "ideal self" ratings, but again there were no significant differences between the two subgroups (See Appendix B: Table 8).

6.7.2. Comparison with the Reference Groups

Forty-one of the 44 eligible BMT patients and all 44 of the eligible children in the healthy group completed the self-perception test. The cardiac group rated themselves as significantly weaker, but less angry than the BMT group. Comparison with the healthy group indicated that the cardiac group rated themselves as weaker, more frightened and more ill (Table 6.10). The total self perception score in the cardiac group was lower (more negative) than in the healthy group. The BMT group rated themselves as significantly more frightened (U=634.0; p=.0063) and more ill (U=486.0; p=.0002) than the healthy group. Overall, the total self perception score in the BMT group was lower (more negative) than that of the healthy group (U=567.0; p=.0291).

PRE-OPERATIVE SELF PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS

25, 50 and 75 centiles.

	C (n=31)	BMT (n= 41)	H (n= 44)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	1.00 3.00 5.00	3.00 5.00 5.00	3.00 4.00 5.00	368.5	.0013	458.5	.0124
CRYING-CHEERFUL	3.00 5.00 5.00	3.00 5.00 5.00	4.00 5.00 5.00	610.5	.5871	646.5	.6341
DOESN'T LIKE SELF-LIKES SELF	4.00 5.00 5.00	3.00 5.00 5.00	3.00 5.00 5.00	569.0	.7757	676.0	.9405
LONELY-HAS FRIENDS	4.00 5.00 5.00	3.00 5.00 5.00	3.25 5.00 5.00	613.5	.6104	661.0	.7815
FRIGHT-ENED-SAFE	2.00 5.00 5.00	1.00 5.00 5.00	5.00 5.00 5.00	557.5	.5297	409.0	.0002
ANGRY-CALM	4.00 5.00 5.00	1.00 5.00 5.00	3.00 5.00 5.00	441.0	.0393	605.5	.3251
BAD-GOOD	3.00 5.00 5.00	3.00 5.00 5.00	3.00 4.00 5.00	475.0	.2366	639.0	.6176
ILL-WELL	1.00 3.00 5.00	1.00 4.00 5.00	5.00 5.00 5.00	523.0	.5091	335.5	.0000
TOTAL SELF-PERCEPTION SCORE	26.00 31.00 35.00	25.25 31.00 36.00	32.00 34.50 38.75	523.0	.6952	427.5	.0060

C = Cardiac group
H = Healthy group

Table 6.10

In order to examine whether self perception varied in younger and older children, each group was divided into two according to the median age of the whole sample (all 3 groups). The median age was 8.7 years. For the younger age group, the cardiac group rated themselves as weaker (U=93.5; p=.0359) but less frightened (U = 80.5; p=.0019) than the BMT group. Comparison with the healthy group indicated that the cardiac group rated themselves as weaker (U=117.5; p=.0552) and more ill (U=98.0; p=.0104). For the group of 8.7 years and older, the cardiac patients perceived themselves as weaker (U=80.5; p=.0078), more bad (U=79.0; p=.0196) but less lonely (U=48.0; p=.0081) than the BMT group. Comparison with the healthy group indicated that the cardiac children rated themselves as weaker (U=87.5; p=.0078), more miserable (U=113.0; p=.0486), more frightened (U=74.5; p=.0004) and more ill (U=65.5; p=.0002).

The 3 groups did not differ on ratings of "ideal self" on any of the constructs (See Appendix B: Table 9). There were also no significant differences between the groups when the sample was divided according to the mean age.

A score was computed to reflect the difference between self and ideal self (Table 6.11). The difference between self and ideal self ratings on the weak-strong construct was significantly larger in the cardiac group compared with the BMT group. Comparison of the cardiac and healthy groups indicated that on the frightened-safe and ill-well constructs the difference between self and ideal self ratings was significantly larger in the cardiac group. The difference in total scores between self and ideal self was also significantly larger in the cardiac group.

PRE-OPERATIVE DATA: DIFFERENCE BETWEEN CHILD'S SELF AND IDEAL SELF PERCEPTION SCORES: COMPARISON BETWEEN CARDIAC, BMT AND HEALTHY GROUPS

25, 50 and 75 centiles.

	C (n=31)	BMT (n=38)	H (n= 44)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	-4.00 -2.00 .00	-2.00 .00 .00	-2.00 -1.00 .00	366.0	.0082	520.5	.0751
CRYING-CHEERFUL	-2.00 .00 .00	-1.00 .00 .00	-1.00 .00 .00	588.0	.9893	625.0	.6049
DOESN'T LIKE SELF-LIKES SELF	-1.00 .00 .00	-1.00 .00 .00	-1.00 .00 .00	539.0	.6645	660.0	.9360
LONELY-HAS FRIENDS	-1.00 .00 .00	-2.00 .00 .00	-1.00 .00 .00	572.0	.8188	661.0	.9442
FRIGHT-ENED-SAFE	-3.00 .00 .00	-3.25 .00 .00	.00 .00 .00	525.0	.4047	434.0	.0022
ANGRY-CALM	.00 .00 .00	-2.00 .00 .00	-1.00 .00 .00	465.5	.1392	628.0	.6342
BAD-GOOD	-2.00 .00 .00	-1.00 .00 .00	-2.00 .00 .00	492.5	.4847	659.5	.9350
ILL-WELL	-4.00 -2.00 .00	-3.75 -1.00 .00	.00 .00 .00	532.5	.7398	394.0	.0010
TOTAL SCORE	-13.00 -8.00 .00	-8.00 -6.00 .00	-6.00 -3.00 .00	496.5	.5535	476.0	.0365

C = Cardiac Group
H = Healthy Group

Table 6.11

6.7.3. Children not Followed-up

Six of the 31 children assessed pre-operatively were not followed-up, 3 of whom died and 3 were lost to follow-up. Those children who died rated themselves as being weaker (U=9.0; p=.0284) and more miserable (U=16.5; p=.0493) than those who were followed-up. There were no significant differences between those lost to follow-up and those seen after surgery.

6.8. PARENTAL PERCEPTION

6.8.1. Cardiac Group

Data on 42 of the 50 eligible children (84%) were collected, where parents were asked to rate their children on the same 8 constructs as the children had rated themselves. In addition, a rating of personality was also given (see page 165). Six of the 8 children on whom data were not collected died (see page 209). Parents of the other 2 children felt unable to rate them on the constructs. There were no significant differences in parental ratings on any of the constructs between the cyanotic (n=12) and acyanotic (n=30) groups (See Appendix B: Table 10). When the sample was divided into younger and older children according to the median age of the total cardiac sample (6.2 years), there were no differences in parental perception in the younger age group. Parents of older cyanotic children felt that they were weaker (U=22.0; p=.0452).

Comparison of parents' and children's ratings indicated a lack of agreement. There were no significant correlations in the cyanotic group. In the acyanotic group there were significant correlations on the lonely-has friends and ill-well constructs (Table 6.12).

PRE-OPERATIVE DATA: CHILD AND PARENTAL PERCEPTION: CORRELATION: CYANOTIC AND ACYANOTIC SUBGROUPS, TOTAL CARDIAC, BMT AND HEALTHY GROUPS

	CYANOTIC (n=8)	ACYANOTIC (n=20)	CARDIAC (n=28)	BMT (n=34)	HEALTHY (n=43)
WEAK-STRONG	r= .4428 p= .136	r=-.0823 p= .365	r= .0244 p= .451	r= .1990 p= .130	r= .2842 p= .032
CRYING-CHEERFUL	r=-.3217 p= .219	r= .1920 p= .209	r= .0324 p= .435	r= .4288 p= .006	r= .1563 p= .158
DOESN'T LIKE SELF-LIKES SELF	r= .0726 p= .432	r= .1541 p= .258	r= .1049 p= .298	r= .0786 p= .332	r=-.0716 p= .324
LONELY-HAS FRIENDS	r= .0357 p= .467	r= .5310 p= .008	r= .3206 p= .048	r= .0461 p= .398	r=-.0083 p= .479
FRIGHTENED-SAFE	r= .4083 p= .158	r= .2200 p= .176	r= .3214 p= .048	r= .3714 p= .017	r= .0529 p= .368
ANGRY-CALM	r=-.5029 p= .102	r= .3421 p= .070	r= .1896 p= .167	r= .1042 p= .282	r= .2868 p= .031
BAD-GOOD	r=-.3198 p= .220	r=-.3041 p= .096	r=-.3048 p= .057	r= .0059 p= .487	r= .0915 p= .280
ILL-WELL	r=-.0579 p= .446	r= .3931 p= .043	r= .3438 p= .037	r=-.1297 p= .240	r= .5402 p= .000
TOTAL SCORE	r= .2033 p= .315	r= .2577 p= .136	r= .2721 p= .081	r= .3518 p= .031	r= .3089 p .022

Table 6.12

6.8.2. Comparison with the Reference Groups

Comparison of the cardiac, BMT (n=44) and healthy (n=54) groups indicated that parents in the cardiac group felt that their children were more cheerful, less lonely and less frightened than parents of BMT children. The total score was higher (more positive) in the cardiac group. Cardiac parents felt that their children were more ill than parents of the healthy group (Table 6.13) and BMT parents felt that their children were more lonely (U=718.0; p=.0016), more frightened (U=704.5; p=.0015) and more ill (U=656.5; p=.0001) compared with ratings

PRE-OPERATIVE PARENTAL PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS

25, 50 and 75 centiles

	C (n = 42)	BMT (n = 44)	H (n = 54)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	3.00 4.00 5.00	3.00 3.00 5.00	3.00 4.00 5.00	822.5	.4596	941.0	.2242
CRYING-CHEERFUL	4.00 5.00 5.00	3.00 5.00 5.00	4.00 5.00 5.00	702.5	.0421	937.5	.1784
DOESN'T LIKE SELF-LIKES SELF	4.00 5.00 5.00	4.00 5.00 5.00	3.00 5.00 5.00	838.5	.6635	1003.5	.4600
LONELY-HAS FRIENDS	3.75 5.00 5.00	1.00 3.00 5.00	3.00 5.00 5.00	566.5	.0018	1065.0	.8213
FRIGHT-ENED-SAFE	3.75 5.00 5.00	2.00 3.00 5.00	3.00 5.00 5.00	597.5	.0073	1021.0	.5432
ANGRY-CALM	3.00 3.00 4.00	3.00 4.00 5.00	3.00 3.00 5.00	744.0	.1979	1022.0	.5685
BAD-GOOD	3.00 4.00 5.00	4.00 4.00 5.00	3.00 4.00 5.00	722.5	.1280	1084.0	.9486
ILL-WELL	3.00 4.00 5.00	1.00 3.00 5.00	4.00 5.00 5.00	827.5	.6108	561.5	.0000
TOTAL SCORE	29.00 33.00 35.00	26.00 31.00 34.00	30.00 33.00 36.00	567.5	.0170	955.0	.2957
PERSON-ALITY	3.00 3.00 4.00	2.75 3.00 4.00	3.00 4.00 4.00	954.5	.6365	1109.5	.9469

Table 6.13

of the healthy group by their parents. The total score was significantly lower in the BMT group compared with the healthy group (U=639.0; p=.0026).

When the sample was divided into younger and older children according to the median age (7.3 years), parents of younger cardiac children felt that they were less frightened (U=136.5; p=.0327) than BMT parents rated their children and more miserable (U=185.0; p=.0244) and more ill (U=157.5; p=.0068) than parents in the healthy group rated their children. In the older age group, parents of the cardiac children felt that they were less lonely (U=106.5; p=.0042) than parents in the BMT group rated their children. Comparison of the cardiac and healthy groups indicated that the cardiac children were rated as being more frightened (p=130.5; p=.0039) and more ill (U=105.0; p=.0003).

There was no particular pattern either across or within the groups of correlations between children's and parents' perceptions. There were significant correlations for the cardiac group as a whole on the lonely-has friends, frightened-safe and ill-well constructs; in the BMT group on the crying-cheerful and frightened-safe constructs and total score and in the healthy group on the weak-strong, angry-calm and ill-well constructs and total score (Table 6.12).

6.8.3. Children not Followed-up

The 5 children who were not seen after surgery were all lost to follow-up and did not differ on any of the parental ratings from the follow-up sample.

6.9. DISCUSSION OF THE RESULTS

6.9.1. Developmental Testing: 0 - 3.5 Years

The results of the developmental testing indicated that there were no significant differences between the 2 cardiac subgroups. Other research which has found differences between cyanotic and acyanotic children has tended to focus on older children, with little attention being given to children under three years old. Results from this study, although based on small samples, suggest that the potentially deleterious effects associated with a cyanotic lesion are not as yet evident in the youngest children when compared with children of a similar age with acyanotic defects. For the cardiac group as a whole, and for each of the two subgroups, locomotor development scores were lower than those of the other subtests, supporting previous findings (Newburger et al, 1984).

In terms of developmental and cognitive ability, findings in the literature indicate that cardiac children perform at a lower level compared with healthy children Although most of the research has been conducted with older children (e.g. Linde et al, 1970), developmental delay has been reported in infants with cardiac defects too (Aisenberg et al, 1982). Results from the current study support the findings of other researchers, with the cardiac group performing at a significantly lower level than the healthy group on developmental parameters, particularly on the locomotor scale.

There are four particular reasons for the results. Firstly, children with cardiac disease are often physically less able to interact with their environment due to the limiting nature of their condition. For children in this younger age group, physical activities constitutes a significant part of the tests used. Lack of physical abilities also hinders the development of other skills,

such as exploratory behaviour. Secondly, maternal overprotectiveness of cardiac children, which has been previously reported (Landtman et al, 1960; Linde et al, 1966; Offord et al, 1972). A number of mothers in this study admitted keeping their children away from others through fear of infection etc., thus limiting their social interactions and potentially limiting the development of speech and socialisation skills. In this study, the cardiac children performed significantly less well on the personal-social and speech and hearing scales. Thirdly, the effects of hospitalisation and illness. A number of children in the cardiac group had spent prolonged periods of time in hospital, resulting in environmental inconsistency not only in terms of the physical environment but also in terms of the large number of people involved with the child, which could further have compromised their development. Fourthly, feeding difficulties, particularly in the infants, may also have made parents feel inadequate and caused them to withdraw emotional support from their infant, which has been reported by others (Gudermuth, 1975; D'Antonio, 1976; Shor, 1978), thus further impacting on developmental function.

The cardiac group did not differ from the BMT group on any of the developmental parameters. The comparison of the BMT and healthy groups indicated that the two groups differed significantly on the same parameters as the cardiac and healthy groups. This suggests that chronic illness in infancy does compromise development, supporting the views of other researchers (Ack et al, 1961; Dikmen et al, 1975; Eiser & Lansdown, 1977; Eiser, 1981; So, Chang, Najarian, Mauer, Simmons & Nevins, 1987; Stewart, Uauy, Waller, Kennard & Andrews, 1987) and that gross motor and language development in particular are affected (Polinsky, Kaiser, Stover, Frankenfield & Baluarte, 1987). However, the physical nature of the illness does not differentiate different diagnostic groups at this stage.

6.9.2. Cognitive Testing: 3.5 - 17 Years

Within the older age group there were significant differences on a number of the cognitive parameters (section 6.2.1.) between the cyanotic and acyanotic subgroups, supporting the work of previous researchers (O'Dougherty et al, 1985; DeMaso et al, 1990). However, performance in the cyanotic group as well as in the acyanotic group was within the normal range, as found previously (Chazan, 1951; Kramer et al, 1989). Compared with findings for the younger age group, there were significant differences between the cyanotic and acyanotic groups, which contradicts suggestions that IQ differences diminish with age due to the change in focus of the tests (Rasof et al, 1967). These results support the view that impairment of cognitive function increases with age in cyanotic patients (O'Dougherty et al, 1983; Newburger et al, 1984). In addition, a greater number of the cyanotic group had been previously hospitalised, particularly in infancy, and it has been suggested that stresses associated with hospitalisation and surgery at an early age may affect subsequent mental and intellectual development (Kato, Kanto, Yoshino, Hebiguchi, Koyama, Arakawa & Hishikawa, 1993).

The results on the attainment tests (section 6.2.1.) show that although cyanotic children were performing at a lower level on measures of cognitive ability compared with the acyanotic group, their academic performance was in line with their overall IQ. In contrast, the acyanotic group had a higher overall IQ but were performing below their potential on arithmetic and reading. These findings corroborate those of other researchers that chronically ill children have IQ scores within the normal range but perform at lower levels than expected from age-related norms on measures of academic functioning (Olch, 1971; Pless & Pinkerton, 1975; Eiser, 1980(a); Gath et al, 1980; Mearig, 1985; Peckham et al, 1988).

Whilst the cyanotic children performed at a significantly lower level than the healthy group on a number of the cognitive measures, the acyanotic patients did not differ from the healthy group. This lack of difference between the acyanotic and healthy groups is contrary to the findings of Linde et al (1970), who found that both cyanotic and acyanotic patients obtained lower scores than healthy children on various measures of intelligence. The majority of acyanotic children in this study were leading relatively normal lives, with minimal effects of symptoms. One explanation for the discrepancy in findings is that in the 1980's parents were encouraged to treat their children more normally than in the 1960's. The advances in medicine mean that for many children with acyanotic lesions a normal life is quite possible and the outlook after surgery is excellent. As well as changes in parental attitudes, other organisations such as schools are now more accustomed to dealing with children with congenital heart disease, thereby further encouraging normal development. For the cyanotic patients, as well as the potentially harmful effects of hypoxia etc., they tended to be more symptomatic and to be affected in terms of appearance, thus resulting in being treated differently - all of which can potentially influence their cognitive performance.

The poorer developmental and cognitive functioning of the cardiac children who died compared with the follow-up sample is likely to be largely attributable to their more serious medical condition and greater degree of physical impairment.

The cyanotic and BMT groups were broadly comparable in terms of cognitive performance, with both demonstrating similar differences with the healthy group. The differences between both of these groups and the acyanotic group suggest that, for older children at least, the nature of the disease is a significant factor for cognitive development. Their poorer

performance on academic parameters supports the view that schooling is affected, despite IQ being within the normal range. Although the performance of the cyanotic and BMT groups was within the normal range, the higher scores of the acyanotic and healthy groups also needs interpretation. One view is that the standardised norms for IQ tests are no longer accurate. A study of healthy children tested with the BAS found a high mean IQ score (IQ: 112; S.D. 13.4) (Fulton, Raab, Thomson, Laxen, Hunter & Hepburn, 1987), which is consistent with the results of this study. Other authors have also found higher than expected scores on subtests of the BAS (Cockburn & Ounsted, 1983). A similar finding has also been reported using the Wechsler scales (Fuggle, Tokar, Grant & Smith, 1992). If, as Fuggle et al (1992) suggest, IQ scores are rising, then the results of the cyanotic and BMT groups need to be interpreted with a greater degree of caution before it is categorically stated that their performance is within the normal range.

6.9.3. Behaviour at Home: 3 - 5 Years

In terms of behaviour at home, the small number of completed questionnaires obtained on pre-school-aged children makes interpretation of the results difficult. On individual items, sleeping difficulties, eating problems and temper tantrums were specific areas of concern mentioned by parents (See Appendix D: Table 23). There is very little information in the literature about behaviour of children with congenital heart disease in this age range. The majority of patients in this study had been diagnosed in infancy and it has been suggested that the younger the child at diagnosis, the poorer the outcome in terms of behaviour (Eiser, 1985). For the pre-school age group, mastery of new skills and the development of autonomy and initiative, together with separation from parents and involvement in peer relationships, become the focus of development. For the ill child, these tasks may be

compromised (Garrison & McQuiston, 1989). For the child with congenital heart disease, where limitations on physical activities and social interactions (the latter either through the illness itself or due to parental attitudes) are particularly evident, the achievement of such normal developmental tasks may be severely hindered.

The prevalence of problem behaviours in pre-school children was similar in the cardiac and reference groups, but again the small numbers are likely to have influenced the findings. The prevalence of behaviour problems in the healthy group was similar to that found in a study of healthy 3 year olds using the same measure (Richman, Stevenson & Graham, 1982). Eating and sleeping problems were two of the areas of greatest concern to parents in this latter study. Although not significant, in our study a higher percentage of parents of cardiac and BMT children compared with parents of healthy children reported these as areas of difficulty. A further study of healthy children found that poor appetite, sleeping problems and temper tantrums were more evident at times of minor illness, demonstrating the frequent coexistence of health and behaviour problems in pre-school aged children (Hart, Bax & Jenkins, 1984). The results highlight the susceptibility of pre-school aged children with chronic illness and also suggest that the presence of chronic illness, rather than its underlying nature, is the relevant factor for potential adjustment difficulties.

6.9.4. Behaviour at Home: 5 - 17 Years

The literature indicates that older children with congenital heart disease are at risk for emotional and behavioural difficulties (Landtman et al, 1959; Green et al, 1962; Linde et al, 1966; Auer et al, 1971; Kong et al, 1986; Kramer et al, 1989; DeMaso et al, 1990), particularly those children with cyanotic lesions. Although there were no significant

differences in the prevalence of problem behaviours at home between the cyanotic and acyanotic subgroups, a higher proportion of children with cyanotic lesions had behaviour problems (25% vs 14%). Similarly elevated rates of behaviour problems have been reported in other studies of chronically ill children (Heller et al, 1985; Garralda et al, 1988). There are a number of factors which are likely to have contributed to the higher prevalence of behaviour problems in the cyanotic group compared with the acyanotic group. The majority of children in both subgroups were diagnosed in infancy, increasing the likelihood of disturbed emotional development during childhood (Linde et al, 1971; Linde, 1982). However, compared with the acyanotic children, the cyanotic children had undergone more hospitalisation and were more affected in terms of symptoms and their physical appearance (tending to have delayed puberty, be "blue", smaller and thinner). Other studies of chronically ill children and adolescents have found that altered physical appearance increases the prevalence of adjustment difficulties (Boyle et al, 1976; Sinnema, Van Der Laag & Stoop, 1991). Short stature in particular can result in other people relating inappropriately to the child as a much younger child (Mattsson, 1972; Law, 1987) and lead to restrictions in social life (Henning, Tomlinson, Rigden, Haycock & Chantler, 1988) and lower levels of social competence (Sandberg, Brook & Campos, 1994). Overprotective behaviour by mothers, recognised as a significant factor in poor adjustment of the cardiac child (Linde et al, 1966), may also be exacerbated by the way in which others treat the child and the influence of the child's appearance on her own perceptions.

From the behaviour questionnaire, areas most often mentioned by parents included the occurrence of headaches and stomachaches, temper tantrums, disobedience, eating difficulties, restlessness, anxieties, irritability, fearfulness and difficulty settling to anything

(See Appendix D; Table 24). Parents of cyanotic children reported a lower, although not significant, prevalence of temper tantrums, disobedience and irritability and a higher prevalence of solitary behaviour and anxiety than parents of the acyanotic children. For the cyanotic group, the more apparent effects of their condition on their appearance and the greater severity of their symptoms are likely to have contributed to their feelings of social isolation and anxiety. In contrast, many of the acyanotic children were well, with no overt signs of their condition, yet they were having to undergo hospitalisation and surgery. It has been suggested that for some children with chronic illness the lack of any indicators of their condition can result in more adjustment difficulties (McAnarney et al, 1974) and a possible manifestation of this could be more attention-seeking and antisocial type behaviour patterns. Such children are neither "normal" nor do they elicit the support and allowances accorded more readily to the more disabled child (Heisler & Friedman, 1981).

The prevalence of problem behaviours was the same in the acyanotic and healthy groups (14%) and was similar to that of the normal population (Rutter et al, 1970). As for the acyanotic group, temper tantrums and disobedience were two particular areas of concern for parents of the healthy children. These results suggest that the acyanotic group were more similar to the healthy group than to the cyanotic group, both in the nature and prevalence of problem behaviours, although with small sample sizes the differences with the cyanotic group were not significant.

Neither the cardiac nor BMT group showed any extreme deviant behaviour prior to treatment, although within the cardiac group the prevalence of problem behaviour in the cyanotic group was twice that of the BMT group. For the cyanotic group, the congenital

nature of the disorder and early diagnosis, together with their more overt symptoms, are likely to be contributory factors to such differences.

6.9.5. Behaviour at School: 5 - 17 Years

Little work has been carried out on the adjustment of children with congenital heart disease in the school environment - rather, the focus has been on performance at school. Assessment of other groups of chronically ill children has largely investigated return to school and adjustment at various times after the diagnosis of a chronic condition in childhood, with most research to date on children diagnosed with cancer (Lansky et al, 1983; Sawyer et al, 1986; Katz, Rubinstein, Hubert & Blew, 1988; Sawyer, Toogood, Rice, Haskell & Baghurst, 1989). Among children with congenital diseases those with phenylketonuria show some evidence of higher rates of behaviour disturbance in the school environment (Stevenson et al, 1979), with neurotic behaviour in particular being identified by teachers. However, a study of cystic fibrosis patients found that teachers' ratings of overall adjustment did not differ from ratings of healthy children or other chronically ill groups (Drotar, Doershuk, Stern, Boat, Boyer & Matthews, 1981). The results of our study indicated that 21% of the cardiac group had problem behaviours at school, predominantly of a neurotic type, which is higher than the reported rate of 10% for the normal population (Rutter et al, 1970). Although not significantly different, the prevalence of problem behaviour was twice as high in the cyanotic group as in the acyanotic group. Physical problems, such as tiredness and lack of mobility, have been found to create difficulties for children at school (Larcombe, Walker, Charlton, Meller, Morris Jones & Mott, 1990) and these are likely to be a contributory factor.

On individual items, fearfulness and anxiety were specific areas of concern for teachers of the cardiac group overall (see Appendix D; Table 25). For teachers of the cyanotic children solitary behaviour, difficulty settling, fussiness and unresponsive or apathetic behaviour were particular concerns. Withdrawn and anxious behaviour patterns have been found in other children with cognitive deficits (Anderson, Smibert, Ekert & Godber, 1994) and, as previously mentioned, IQ deficits and academic difficulties were more prevalent in the cyanotic group. The more obvious symptoms and physical effects of the disease in the cyanotic group are also likely to have influenced their treatment by, and the attitudes of, teachers and peers. Teachers have been found to have minimal knowledge of, and a number of misconceptions about, several chronic conditions (Eiser & Town, 1987) and fears about illness associated with the heart are likely to be a particular problem. Although, in contrast to some other illnesses, prolonged and frequent periods of absence were less of a factor in the cardiac group, it is highly likely that teachers were reacting to the child's physical appearance and treating and judging them differently from their peer group. For the acyanotic children this is likely to have had less impact because they were frequently physically indistinguishable from their peers.

Teachers of the healthy children reported more concerns about lying, theft, bullying and aggressive/resentful behaviour than teachers of the cardiac group, and this was borne out by the greater number of children with antisocial, rather than neurotic, behaviour problems in the healthy group. Children with other chronic diseases have also been found to exhibit more neurotic behaviours than normal controls (Stevenson et al, 1979), but the differences could also be due in part to different perceptions and expectations of the teachers. What may be

considered resentful behaviour in a healthy child may be justified, and ignored, in a sick child because of their illness.

The prevalence of problem behaviour was similar in the cardiac and BMT groups, and the pattern of predominantly neurotic behaviour was also similar in the two groups. Both the cardiac and BMT groups were felt to be more anxious, miserable and fearful than healthy children.

6.9.6. Parental Interview

On the parental interview, the findings of significantly greater dependency and less active behaviour in the cyanotic group compared with the acyanotic group is not unexpected in view of the more limiting and severe nature of their cardiac lesions. However, it is well documented that development can be influenced by parental attitudes of pampering, over-protection and anxiety (Cooper, 1959; Linde et al, 1971; Offord et al, 1972; Donovan, 1983; Bowen, 1985; Kong et al, 1986). Such parental attitudes are more likely to be precipitated by having a more symptomatic and frail child, so it is difficult to separate out how much of the dependent and less active behaviour is due to the physically limiting nature of the lesion and how much is due to the restricting and over-anxious behaviour of the parents.

There was a significant association between the presence of behaviour problems on the parental interview and Rutter A scales for the acyanotic group, but the association was not significant for the cyanotic group. This suggests that for the acyanotic group, the majority of whom were asymptomatic and leading relatively normal lives, problem behaviour as measured on the Rutter A scale was also considered to be problem behaviour by the parents

of these children. In contrast, in the cyanotic group children who were rated as having a significant degree of problem behaviour on the Rutter A scale were not necessarily considered to have behaviour problems by their parents. On the other hand, parents reported that other aspects of behaviour not measured on the Rutter A scale were "problems". The implication is, therefore, that for at least some groups of chronically ill children, the Rutter A scale is not an adequate measure for assessing behaviour problems as perceived by the parents.

With surgery imminent, greater anxiety and dependent behaviour would not be unexpected in the cardiac group compared with the healthy group, although this may also be a reflection of maternal anxiety. Increased dependency has been reported in other groups of chronically sick children (Zitelli, Miller, Gartner, Malatack, Urbach, Belle, Williams, Kirkpatrick & Starzl, 1988; Eiser, Havermans, Pancer & Eiser, 1992; Daud, Garralda & David, 1993) and it has been suggested that for children diagnosed before 3 years of age there may be an increase in maternal control and a resultant rise in more passive and helpless behaviour (Maddison & Raphael, 1971). Furthermore, the cardiac group were younger and therefore might be expected to be more dependent. The less active behaviour reported for the cyanotic, but not the acyanotic, group compared with the healthy group is, as previously mentioned, likely to be a combination of physical limitations and maternal over-protection. It also supports the view that the acyanotic children were not markedly impaired compared with the healthy group.

The significant association between the presence of behaviour problems on the Rutter A scales and parental interview for the healthy group further supports the idea that the

acyanotic group, for whom the association was also significant, were more similar to the healthy children than to another group of chronically sick children. The finding also suggests that, for healthy children, the Rutter A scale does assess those areas of behaviour which are of importance to parents, and that it is an accurate reflection of whether parents perceive their child to have behaviour problems.

The presence of a congenital lesion, diagnosed at birth and with overt, limiting symptoms, is likely to be a contributory factor to the greater dependency and less active behaviour in the cyanotic group compared with the BMT group. The BMT group were also more dependent than the healthy group, so all of the illness groups had a more dependent pattern of behaviour than well children. The BMT children, like the cardiac children, were also considered by their parents to be more anxious than the healthy group and this is again likely to be a reflection of impending treatment and the parents' own anxieties. The finding of more difficulty with peers in the BMT group compared with the healthy group could be due to a number of factors, including prolonged absence from school (not a factor in the cardiac group) and resulting social isolation, and altered appearance due to previous chemotherapy.

As for the cyanotic group, the lack of association between the presence of behaviour problems on the Rutter A and parental interview in the BMT group further questions the validity of the Rutter A scales for assessing the prevalence of behaviour problems in chronically ill children. The inclusion of somatic elements on the Rutter A may explain some of the discrepancy - nausea and eating problems, for example, are acknowledged side effects of chemotherapy and are not seen as behaviour problems per se by parents. However, for many of the BMT children in particular, they scored highly on these questions on the Rutter

A. For three of the four BMT children whose parents felt that they did not have behaviour problems according to the parental interview but who achieved Rutter A scores above the criterion, a large proportion of their score was comprised of somatic elements.

6.9.7. Self Perception

There is little information on the self perception of children with congenital heart disease, but the consistent findings of poorer emotional adjustment in patients with cyanotic compared with acyanotic lesions, the actual differences in symptomatology, and effects on physical appearance would suggest that self perception may also be more negative in children with cyanotic heart disease. The results of this study did not support such a view - there were no differences on any of the constructs between the cyanotic and acyanotic groups. However, when the sample was divided into two age bands, the younger cyanotic children rated themselves as stronger than the acyanotic group, but the older cyanotic patients rated themselves as weaker. For the older group this difference is likely to reflect an actual physical difference between the cyanotic and acyanotic children. Within the younger group it is more likely that all of the children had polarised responses - i.e. picked either the weak or the strong pole, rather than the interim points on the scale. Within the cyanotic group it is possible that the children were using denial as a means of coping with their condition and were therefore perceiving themselves as strong. Children who died perceived themselves as more miserable and weaker than those who were followed-up, reflecting their poorer physical condition at the time of surgery and perhaps a greater insight, resulting in more misery, into what lay ahead.

The cardiac group perceived themselves as weaker, more frightened and more ill than the healthy group. In view of their actual physical condition and their impending surgery, these differences would be expected. Poor body image (measured in this study by the weak-strong construct) has been reported in other groups of chronically ill children (Kaufman & Hersher, 1971; Rovet, Ehrlich & Hoppe, 1987). Within the younger age group the finding that the cardiac children felt that they were weaker and more ill indicates that younger children tend to focus on the physical aspects of their condition. In the older age group, the cardiac children additionally rated themselves as more miserable and frightened, indicating that they were also focusing (appropriately, in view of the surgery) on their emotions. The greater discrepancy in the cardiac group between self and ideal self on the frightened-safe and ill-well constructs clearly indicates that the children recognised their greater anxiety and poorer health as being related to their condition and treatment and that this was not how they wished to feel.

The cardiac group rated themselves as weaker, but less angry, than the BMT group. The perception of being weaker is again likely to be a reflection of genuine physical differences, particularly as many of the cardiac group had been ill for a longer time and were physically more impaired. The greater discrepancy in the cardiac group on the weak-strong construct between self and ideal self indicates that these children were less satisfied with their physical abilities. The reported anger in the BMT group may be attributable to the older age of the BMT patients. Anger has been reported in other groups of children with acquired, rather than congenital illnesses - particularly related to intrusive treatment regimens (Kashani & Hakami, 1982; Wray et al, 1993) - and the finding in this study may also be a reflection of the differences between those with acquired and congenital disorders. Younger cardiac

children rated themselves as less frightened than BMT children, probably reflecting the fact that many of the BMT patients had already experienced stressful hospital procedures, such as chemotherapy. Anxiety related to illness has been reported previously for BMT patients prior to treatment (Gardner et al, 1977). In the older age group cardiac patients perceived themselves to be less lonely but more bad than BMT patients. The cardiac children had, in general, experienced less disruption to schooling and many had led a "normal" life up until surgery. In contrast, many BMT children had spent time in hospital and had prolonged periods of absence from school. The findings that cardiac children rated themselves as less good may be due to the younger age of these patients and the fact that in some younger children punishment for being bad is given by them as an explanation for illness (Perrin & Gerrity, 1981; Springer, 1994).

6.9.8. Parental Perception

The lack of a clear and consistent relationship between actual disease severity and parental - particularly maternal - perception of the child with congenital heart disease has been well documented in the literature (Landtman et al, 1960; Offord et al, 1972; D'Antonio, 1976). A number of studies have highlighted the importance of maternal perceptions of disease severity (Bergman & Stamm, 1967; Offord et al, 1972; Casey et al, 1994). It has been suggested that it is the presence, rather than the actual severity, of a heart lesion which is significant in determining maternal anxiety (Linde et al, 1966; Kitchen, 1978) and the resulting maternal attitude towards the ill child. Although there is no other information about how parents perceive their child with congenital heart disease in terms of the constructs used in this study, the lack of difference between parents' perceptions of children with cyanotic or acyanotic lesions supports the findings in much of the literature that actual disease severity

has little impact on maternal perception of the child. (In this study the vast majority of the ratings were given by the mothers).

Parents' and children's ratings showed a lack of agreement, particularly in the cyanotic group, with children tending to have more polarised perceptions than parents. In studies of other illness groups, children's and mothers' reports of children's adjustment and experience of the disease showed poor agreement (Weissman et al, 1980; Nelms, 1986; Ennett et al, 1991; Engstrom, 1992; Wachtel, Rodrigue, Geffken, Graham-Pole & Turner, 1994) and the authors emphasised the importance of cross-validating parental reports with children's self-reports.

The finding that parents of cardiac children perceived their child to be more ill than parents of the healthy children is expected. Parents rated older, but not younger, cardiac children as being more frightened than the healthy children, which agrees with the children's own perceptions. With surgery imminent, this is an expected finding but it may also be a reflection of parents' own fears and worries. The lack of agreement between parents' and children's perceptions on five of the eight constructs suggests that, for healthy and ill children, parental reporting of their children is not an accurate assessment of how the child feels, particularly in areas dealing with emotional, rather than physical, attributes.

Cardiac children were rated as being more cheerful, less lonely and less frightened than BMT children. The majority of cardiac patients had been leading a relatively normal life, with school, up until the time of surgery. In contrast, many BMT children were already undergoing traumatic and intrusive treatment, with disruption to schooling, and so might be expected to feel more miserable, lonely and frightened. In neither the cardiac nor the BMT

group was there a significant association between children's and parents' perceptions on the ill-well construct; however, the association was significant for the healthy group. Within the BMT group the poor correlation between parents' and children's ratings on the ill-well construct may also be due in part to the fact that a number of BMT patients were not overtly ill - e.g. those with aplastic anaemia - and so the children themselves may have underestimated the severity of their illness. However, overall, the results indicate the importance of accurately assessing how the child feels about and interprets his/her illness rather than purely using parents as their advocates.

CHAPTER 7

POST-OPERATIVE COGNITIVE AND BEHAVIOURAL FUNCTIONING

The results will be presented in turn of each area of functioning tested - developmental and cognitive, behaviour at home, behaviour at school, parental interview, self perception and parental perception. For each area results for the younger children will be presented first, followed by those for the older group. In each instance the results at follow-up for the whole cardiac group will be presented, looking specifically at any differences between cyanotic and acyanotic patients. To simplify presentation children will still be referred to in terms of cyanotic and acyanotic heart disease, (and, for the acyanotic group, as symptomatic or asymptomatic), even though they have had surgery and in most cases have had the lesion repaired. Post-operative functioning in the cardiac group will then be compared with that of the two reference groups. Changes over time will be looked at, firstly for the cyanotic (n=18) and acyanotic (n=36) patients and secondly for the total cardiac (n=54), BMT (n=43) and healthy (n=49) groups. The influence (if any) of the child's age at the time of surgery on developmental and cognitive function will be described for the cardiac group. Finally, the results will be briefly discussed in terms of the existing literature on children with chronic illness and congenital heart disease in particular.

7.1. POST-OPERATIVE DEVELOPMENTAL FUNCTION: 0 - 4.5 YEARS

7.1.1. Cardiac Group

There were 19 children who were eligible for and were assessed with the Ruth Griffiths developmental scales, 7 of whom had cyanotic lesions and 12 who had acyanotic heart disease. Complete pre- and post-operative assessments were available for 6 cyanotic children

DEVELOPMENTAL FUNCTIONING

SCALE	CYANOTIC (n=6)			ACYANOTIC (n=12)			TOTAL CARDIAC (n=18)			BMT (n=7)			HEALTHY (n=11)		
	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE
LOCOMOTOR	** +	>>+ ** ~	abd	~	>>	ace	~				**	bc	~ ** ~	~	de
MEAN D.Q.	103.83	79.33	- 24.50	99.25	106.92	7.67	100.78	97.72	- 3.06	105.86	107.29	1.43	125.45	118.55	- 6.91
S.D.	13.14	15.57	14.96	11.75	11.70	12.46	12.04	18.41	20.24	14.77	23.94	15.76	13.28	11.47	12.13
PERSONAL-SOCIAL	> **	< > *	fg	*	<	g	~					f	~ * **	*	
MEAN D.Q.	103.17	88.17	- 15.00	104.92	105.58	0.67	104.33	99.78	- 4.56	96.43	99.86	3.43	125.45	116.27	- 9.18
S.D.	18.23	17.07	9.94	9.19	17.81	16.05	12.37	19.03	15.92	5.38	13.16	12.97	15.29	10.12	17.21
SPEECH/HEARING							**						**		
MEAN D.Q.	97.33	93.00	- 4.33	102.75	103.67	0.92	100.94	100.11	- 0.83	104.67	101.00	- 3.67	121.00	119.36	- 1.64
S.D.	16.57	25.12	23.36	13.90	23.32	15.58	14.59	23.75	18.00	7.31	17.03	21.29	23.11	21.29	28.07
EYE/HAND COORDINATION															
MEAN D.Q.	99.67	95.83	- 3.83	101.92	102.08	0.17	101.17	100.00	- 1.17	105.86	102.57	- 3.29	113.18	114.82	1.64
S.D.	12.45	16.56	25.70	7.48	10.57	10.32	9.11	12.73	16.34	16.99	14.57	16.42	19.48	14.68	28.07
PERFORMANCE															
MEAN D.Q.	117.33	102.83	- 14.50	109.42	114.75	5.33	112.06	110.78	- 1.28	112.86	111.57	- 1.29	118.36	120.00	1.64
S.D.	15.36	19.14	25.77	13.81	17.44	15.96	14.40	18.38	21.28	19.60	15.84	21.16	16.29	14.24	18.80
PRACTICAL REASONING			(n=3)			(n=2)			(n=5)			(n=1)			(n=5)
MEAN D.Q.	97.00	94.67	- 2.33	106.50	101.50	5.00	100.80	97.40	- 3.40	95.00	92.00	- 3.00	117.00	116.00	- 1.00
S.D.	18.00	21.46	9.07	6.36	14.85	8.49	14.11	17.30	7.83	.00	.00	.00	19.79	13.51	15.31
	**	++ **	hij	#	++	j	# ~					I	** # ~ #	**	h
OVERALL I.Q.	104.00	91.67	- 12.33	103.67	106.50	2.83	103.78	101.56	- 2.22	103.86	104.29	0.43	120.45	116.91	- 3.55
S.D.	10.88	16.10	14.61	6.26	14.17	10.32	7.76	16.06	13.63	9.62	15.70	13.54	12.34	12.09	13.89

Table 7.1

PAIRED T-TEST: CYANOTIC GROUP

+	p = .010
>	p = .014

INDEPENDENT T-TEST: CYANOTIC VS ACYANOTIC GROUP

>>	p = .001
<	p = .065
++	p = .062

MULTIPLE COMPARISONS: CARDIAC/CYANOTIC/ACYANOTIC GROUPS VS BMT/HEALTHY GROUPS

**	p<.05
*	p<.01
#	p<.005
~	p<.001

CHANGES OVER TIME: REPEATED MEASURES ANOVA

a	p = .00018
b	p = .0002
c	p = .002
d	p = .0002
e	p = .0009
f	p = .017
g	p = .045
h	p = .042
I	p = .025
j	p = .021

and 12 acyanotic children. The seventh cyanotic child was not seen pre-operatively, but his post-operative results did not differ significantly from the overall sample.

The 2 subgroups differed significantly on the locomotor subscale and the differences almost reached significance on the personal-social subscale and on the overall IQ (Table 7.1).

7.1.2. Comparison with the Reference Groups

Eight BMT patients and 11 healthy children were also assessed with the same measures. In the cardiac group performance on all of the subtests was within the normal range (Griffiths, 1970) but the cardiac group obtained significantly lower scores on the locomotor, personal-social and eye-hand coordination subscales and on the overall IQ compared with the healthy group ($p<0.05$). The BMT group did not differ from the cardiac or healthy groups on any of the parameters (Table 7.1).

Comparison of the cyanotic and acyanotic subgroups with the 2 reference groups indicated that the cyanotic group obtained significantly lower scores than the BMT group on the locomotor subscale ($p<.05$) and than the healthy group on the locomotor ($p<.001$) and personal-social ($p<.01$) subscales and on the overall IQ ($p<.05$). There were no significant differences between the acyanotic patients and the BMT or healthy groups.

7.1.3. Changes Over Time: Cardiac Group

Eighteen children were assessed both pre- and post-operatively, 6 of whom had cyanotic lesions and 12 had acyanotic heart disease. There were no significant changes over time in the acyanotic group, but there was a significant deterioration in performance on the locomotor and personal-social subscales in the cyanotic group. The degree of change on these 2 subscales and on the overall IQ differed significantly between the cyanotic and acyanotic patients (Table 7.1).

7.1.4. Changes Over Time: Comparison with the Reference Groups

Seven BMT patients and 11 healthy children were assessed pre- and post-operatively. There were no significant changes over time on any of the parameters in either of the reference groups or for the total cardiac group (Table 7.1). There was a significant group x time interaction on the locomotor subscale ($p<.0001$).

7.1.5. Significance of Age

The mean age of the two cardiac subgroups was not significantly different at follow-up (cyanotic group: mean age 2.64 years; S.D. 0.99 years; acyanotic group: mean age 2.66 years, S.D. 0.86 years; $p=.972$). In the cyanotic group performance on the eye-hand coordination and performance subscales was positively correlated with age at repair. In the acyanotic group there were no significant correlations between subscale scores and age. When the effects of age were controlled for, the 2 subgroups still differed significantly on the locomotor and personal-social subscales and on the overall IQ (Table 7.2).

POST-OPERATIVE DEVELOPMENTAL FUNCTIONING: 0 - 4.5 YEARS: CORRELATION OF AGE AT REPAIR WITH PERFORMANCE FOR THE CYANOTIC AND ACYANOTIC SUBGROUPS, EFFECT OF AGE ON THE DIFFERENCE IN PERFORMANCE BETWEEN THE TWO SUBGROUPS, AND PROBABILITY VALUES FOR DIFFERENCES IN PERFORMANCE BETWEEN THE TWO SUBGROUPS CONTROLLING FOR AGE

	CORRELATION OF AGE AND PERFORMANCE		COMPARISON OF PERFORMANCE BETWEEN CYANOTIC AND ACYANOTIC SUBGROUPS WITH AGE AS A COVARIATE	
	CYANOTIC	ACYANOTIC	EFFECTS OF AGE	CONTROL-LING FOR AGE
SCALE	CORRELATION: AGE & SCORE	CORRELATION: AGE & SCORE		
LOCOMOTOR	r= .3435 p= .225	r= -.0577 p= .429	t= .571 p= .576	p= .000
PERSONAL-SOCIAL	r= .1683 p= .359	r= .1879 p= .279	t= .727 p= .477	p= .050
SPEECH/ HEARING	r= .4503 p= .155	r= .1344 p= .339	t=1.060 p= .305	p= .333
EYE/HAND CO-ORDINATION	r= .7662 p= .022	r= .0488 p= .440	t=1.557 p= .139	p= .136
PERFORMANCE	r= .8873 p= .004	r= .1514 p= .319	t=1.633 p= .122	p= .053
PRACTICAL REASONING	r= .4725 p= .264	r= -.2683 p= .260	t= .435 p= .674	p= .279
OVERALL IQ	r= .6661 p= .051	r= .0252 p= .469	t=1.274 p= .221	p= .021

Table 7.2

7.2. POST-OPERATIVE COGNITIVE ABILITY: 4.5 - 17 YEARS

7.2.1. Cardiac Group

All children in this age range (n=35) were assessed with the British Ability Scales. Differences were found between the 11 children with cyanotic heart disease and 24 with acyanotic lesions. The cyanotic group showed poorer performance than the acyanotic group on all of the subtests, with the differences reaching significance on the recall of digits, naming vocabulary, similarities and speed of information processing subtests (Table 7.3).

The overall IQ scores were within the normal range for each subgroup but the acyanotic patients achieved a significantly higher score than the cyanotic group ($p<.001$). The cyanotic patients obtained significantly lower scores on the reading and spelling attainments compared with the acyanotic patients (Table 7.4).

Underachievement on arithmetic was more prevalent in the acyanotic group, but not significantly so. The incidence of reading problems was significantly higher in the cyanotic group (chi-squared = 5.11913; $p=.0237$) (Table 7.5).

COGNITIVE FUNCTIONING

SCALE	CYANOTIC			ACYANOTIC			TOTAL CARDIAC			BMT			HEALTHY		
	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE
RECALL OF DIGITS		>>	(n=10) ab	++	++ >>	(n=23)	**		(n=33)	**		(n=27) b			(n=35) a
MEAN T SCORE	53.90	51.00	- 2.90	54.52	58.48	3.96	54.33	56.21	1.88	48.37	52.30	3.93	54.09	55.29	1.20
S.D.	8.50	11.94	12.21	8.16	8.67	8.39	8.13	10.19	10.02	9.74	10.57	9.68	9.41	10.16	7.68
VISUAL RECOGNITION			(n=3)			(n=1)			(n=4)			(n=1)			(n=1)
MEAN T SCORE	58.00	55.67	- 2.33	60.00	66.00	6.00	58.50	58.25	- 0.25	71.00	66.00	- 5.00	63.00	52.00	-11.00
S.D.	6.25	15.37	9.24	0.00	0.00	0.00	5.20	13.57	8.62	.00	.00	.00	.00	.00	.00
NAMING VOCABULARY		~~	(n=8)		~~	(n=13)			(n=21)			(n=9)			(n=12)
MEAN T SCORE	51.13	47.13	- 4.00	59.46	57.62	- 1.85	56.29	53.62	- 2.67	47.89	52.33	4.44	53.75	51.58	- 2.17
S.D.	11.19	11.98	17.25	8.31	7.52	7.90	10.13	10.56	11.95	12.75	14.56	11.25	6.47	6.43	6.01
VERBAL REASONING			(n=3)			(n=1)			(n=4)			(n=1)			(n=1)
MEAN T SCORE	51.00	45.33	- 5.67	56.00	60.00	4.00	52.25	49.00	- 3.25	60.00	58.00	- 2.00	62.00	58.00	- 4.00
S.D.	10.15	13.01	3.79	0.00	0.00	0.00	8.66	12.91	5.74	.00	.00	.00	.00	.00	.00
MATRICES	+		(n=8)	+ ** #		(n=19) ce	< *	<	(n=27)	* #		(n=23) de	**		(n=30) cd
MEAN T SCORE	53.13	48.38	- 4.75	62.42	55.53	- 6.89	59.67	53.41	- 6.26	50.87	51.57	0.70	54.80	55.47	0.67
S.D.	12.61	13.81	10.54	9.06	7.76	6.06	10.88	10.21	7.51	8.00	7.44	4.97	8.83	9.21	6.02
SIMILARITIES		<<	(n=7) fgh		<<	(n=20) h			(n=27)			(n=23) g			(n=30) f
MEAN T SCORE	53.43	46.29	- 7.14	55.30	56.85	1.55	54.81	54.11	- 0.70	52.00	52.48	0.48	52.63	52.40	0.23
S.D.	12.53	10.01	10.90	8.76	8.24	7.94	9.64	9.74	9.41	9.12	8.83	7.62	8.26	7.65	5.22
SPEED OF INFORMATION PROCESSING	** >	** #	(n=3)	>		(n=8)			(n=11)		**	(n=16)	**	#	(n=22)
MEAN T SCORE	39.67	34.00	- 5.67	61.23	61.00	- 0.63	55.64	53.64	- 2.00	54.44	52.19	- 2.25	60.27	59.91	- 0.36
S.D.	12.66	7.21	8.15	12.01	9.40	3.46	15.44	15.21	5.22	8.12	9.73	6.34	9.14	11.49	8.20

Table 7.3

INDEPENDENT T-TEST: CYANOTIC VS ACYANOTIC

+	p = .040
>	p =. 026
>>	p = .045
~~	p =. 023
<<	p = .007
^	p = .000

PAIRED T-TEST: ACYANOTIC GROUP

++	p = .034
^^	p = .000

PAIRED T-TEST: CARDIAC GROUP

<	p = .000

MULTIPLE COMPARISON TESTS: CARDIAC/CYANOTIC/ACYANOTIC VS BMT AND HEALTHY GROUPS

**	p < .05
*	p < .01
#	p < .005

CHANGES OVER TIME: REPEATED MEASURES ANOVA

a	p = .052
b	p = .046
c	p = .001
d	p = .009
e	p = .019
f	p = .009
g	p = .019
h	p = .033

FUNCTIONING ON ACADEMIC PARAMETERS

SCALE	CYANOTIC			ACYANOTIC			TOTAL CARDIAC			BMT			HEALTHY		
	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE	PRE	POST	CHANGE
OVERALL BAS	+	** <	(n=11) ab	** +	<	(n=24)			(n=35)	**		(n=27) b		**	(n=35) a
MEAN I.Q.	103.82	95.91	- 7.91	117.42	116.25	- 1.17	113.14	109.86	- 3.29	104.15	105.78	1.63	110.83	111.23	0.40
S.D.	19.36	21.66	18.76	12.74	12.02	8.26	16.16	18.10	12.64	13.95	15.74	10.35	12.35	14.78	8.31
ARITHMETIC			(n=10)	**		(n=21)	**		(n=31)	** **		(n=22)			(n=30)
MEAN I.Q.	105.90	97.70	- 8.20	112.17	109.33	- 2.84	111.03	105.58	- 5.45	97.27	97.86	0.59	103.60	102.90	- 0.70
S.D.	20.77	19.70	15.66	15.94	14.11	14.21	17.23	16.72	14.55	15.16	15.25	9.02	17.76	18.42	11.12
READING		** >	(n=8) ceg	**	>	(n=17) dfg	**		(n=25)	** **	**	(n=19) ef		** **	(n=25) cd
MEAN I.Q.	97.25	85.50	-11.75	107.26	109.25	1.99	105.28	102.64	- 2.64	93.21	92.16	- 1.05	106.88	107.52	0.64
S.D.	17.26	17.35	21.99	15.71	16.14	7.23	16.78	20.32	14.72	17.86	19.61	6.99	12.28	13.97	7.24
SPELLING		** >>	(n=8) h		>>	(n=17)			(n=25)		**	(n=18)		** **	(n=25) h
MEAN I.Q.	86.25	78.38	- 7.88	97.37	100.90	3.53	95.32	95.32	- 1.08	85.83	85.22	- 0.61	98.84	100.36	1.52
S.D.	18.86	15.81	18.77	15.08	17.65	10.16	16.75	16.75	13.93	18.39	19.00	11.47	15.48	16.74	9.44

Table 7.4

INDEPENDENT T-TEST: CYANOTIC VS ACYANOTIC

+	p = .018
<	p = .001
>	p = .002
>>	p = .004

MULTIPLE COMPARISONS: CARDIAC/CYANOTIC/ACYANOTIC VS BMT AND HEALTHY GROUPS

**	p < .05

CHANGES OVER TIME: REPEATED MEASURES ANOVA

a	p = .017
b	p = .038
c	p = .002
d	p = .050
e	p = .009
f	p = .030
g	p = .030
h	p = .045

7.2.2. Comparison with the Reference Groups

Twenty-eight of the 31 eligible BMT patients and all 35 healthy children were assessed with the BAS. Performance on all of the subtests and the overall IQ of each of the 3 groups fell within the normal range. There were no significant differences between the 3 groups on any of the parameters (Table 7.3).

Comparison of the cyanotic and acyanotic subgroups with the 2 reference groups indicated that the cyanotic group performed at a significantly lower level on the speed of information processing subtest than both the BMT ($p<.05$) and healthy ($p<.005$) groups. On the overall IQ the cyanotic group obtained a lower score than the healthy group ($p<.05$). The acyanotic group did not differ from either of the reference groups on any of the parameters.

POST-OPERATIVE ACADEMIC FUNCTIONING UNDERACHIEVEMENT ON ARITHMETIC AND READING AND THE PREVALENCE OF READING PROBLEMS					
	CY	ACY	TOTAL CARDIAC GROUP	BMT	HEALTHY
NUMBER UNDER-ACHIEVING ON ARITHMETIC	1 (10%)	7 (33%)	8 (26%)	8 (35%)	8 (27%)
NUMBER UNDER-ACHIEVING ON READING	2 (25%)	5 (25%)	7 (25%)	9 (39%)	3 (10%)
INCIDENCE OF READING PROBLEMS	4 (50%)	1 (5%)	5 (18%)	9 (39%)	3 (10%)

CY = CYANOTIC
ACY = ACYANOTIC

Table 7.5

Performance on the school attainments was also within the normal range for the 3 groups. The cardiac group did not differ significantly from either of the reference groups on any of the attainments (Table 7.4). Comparison of the cyanotic and acyanotic subgroups with the reference groups indicated that the cyanotic group had lower scores than the healthy group on the reading ($p<.05$) and spelling ($p<.05$) attainments.

The achievement status and incidence of reading difficulties were also assessed for each group, the results of which are given in Table 7.5. There were no significant differences in the prevalence of underachievement on reading and arithmetic between the total cardiac group and the reference groups or between the cyanotic and acyanotic subgroups and the

reference groups. The cardiac group also did not differ from either of the reference groups in the prevalence of reading problems. However, the cyanotic group had a significantly higher prevalence of reading problems than the healthy group (chi-squared = 4.10269; p=.0428) and the acyanotic group had a significantly lower prevalence of reading problems than the BMT group (chi-squared = 5.20086; p=.0226). The BMT group had a significantly higher prevalence of underachievement on the reading test (chi-squared = 4.47561; p=.0344) and of reading problems (chi-squared = 4.47561; p=.0344) compared with the healthy group.

7.2.3. Changes Over Time: Cardiac Group

There were no significant changes over time on any of the subtests in the cyanotic group, although they fared worse after surgery on every subtest and on overall IQ. The results for the acyanotic group were mixed. There was a significant improvement in performance on the recall of digits subtest but a significant deterioration in performance on the matrices subtest (Table 7.3).

On the school attainments there were no significant changes over time within either subgroup, although performance was worse after surgery in the cyanotic group. In the acyanotic group the results were again mixed. The degree of change on the reading test differed significantly between the 2 subgroups (Table 7.4).

7.2.4. Changes Over Time: Comparison with the Reference Groups

For the cardiac group as a whole, the only significant change over time on the subtests and overall IQ was on the matrices subtest. In the BMT group there was a significant improvement in the recall of digits subtest. There were no changes in the healthy group

(Table 7.3). There was a significant group x time interaction on the matrices subtest ($p<.0001$).

On the school attainments there was a significant deterioration in performance on the arithmetic test in the cardiac group. There were no significant changes in either of the reference groups (Table 7.4). There was a significant group x time interaction on the reading test ($p=.014$).

7.2.5. Effects of Age

There were no significant differences in the mean ages of the cyanotic and acyanotic children completing each of the subtests. Age was not significantly correlated with performance on any of the subtests in either the cyanotic or acyanotic groups. With regard to academic attainments, there was a significant negative correlation between arithmetic and age in both subgroups. (In the healthy group there was also a significant correlation between age and arithmetic: $r=-.6300$; $p=.001$). Using analysis of variance to compare the 2 subgroups with age as a covariate, age made a significant contribution to arithmetic IQ and, even allowing for age, the differences between the 2 subgroups remained significant. When the effects of age were controlled for, the two groups differed significantly on the recall of digits, naming vocabulary, matrices, similarities and speed of information subtests. They also differed significantly on the overall IQ and on all 3 attainment tasks (Table 7.6).

POST-OPERATIVE COGNITIVE FUNCTIONING: 4.5 - 17 YEARS: CORRELATION OF AGE AT REPAIR WITH PERFORMANCE FOR THE CYANOTIC AND ACYANOTIC SUBGROUPS, EFFECT OF AGE ON THE DIFFERENCE IN PERFORMANCE BETWEEN THE TWO SUBGROUPS, AND PROBABILITY VALUES FOR DIFFERENCES IN PERFORMANCE BETWEEN THE TWO SUBGROUPS CONTROLLING FOR AGE

	CORRELATION OF AGE AND PERFORMANCE		COMPARISON OF PERFORMANCE BETWEEN CYANOTIC AND ACYANOTIC SUBGROUPS WITH AGE AS A COVARIATE	
	CYANOTIC	ACYANOTIC	EFFECTS OF AGE	CONTROL-LING FOR AGE
SCALE	CORRELATION: AGE & SCORE	CORRELATION: AGE & SCORE		
RECALL OF DIGITS	r=-.1786 p= .311	r= .0531 p= .403	t=- .094 p= .926	p=.050
VISUAL RECOGNITION	r= .5821 p= .302	r= - p= -	t= .716 p= .604	p=.560
NAMING VOCABULARY	r=-.1587 p= .354	r= .3160 p= .146	t= .143 p= .888	p=.027
VERBAL REASONING	r= .8430 p= .181	r= - p= -	t= 1.567 p= .362	p=.301
MATRICES	r=-.3415 p= .204	r=-.2717 p= .105	t=-1.428 P= .164	p=.049
SIMILARITIES	r=-.3927 p= .192	r=-.2217 p= .155	t=-1.319 p= .198	p=.006
SPEED OF INFORMATION PROCESSING	r=-.4327 p= .358	r= .4513 p= .082	t= 1.434 p= .179	p=.000
BAS IQ	r=-.3565 p= .141	r=-.0511 p= .406	t=- .864 p= .394	p=.001
ARITHMETIC IQ	r=-.6890 p= .014	r=-.4867 p= .013	t=-3.585 p= .001	p=.031
READING IQ	r=-.0721 p= .433	r= .3022 p= .098	t= 1.036 p= .310	p=.002
SPELLING IQ	r=-.2966 p= .238	r= .1062 p= .328	t= .068 p= .946	p=.005

Table 7.6

7.3. POST-OPERATIVE BEHAVIOUR AT HOME: 3 - 5 YEARS

7.3.1. Cardiac Group

Ten children in the cardiac group were in the age range of 3-5 years and completed Richman BCL questionnaires were received on 8. Five of the 8 children had cyanotic lesions and 3 had acyanotic defects. One child (13%) obtained a score of 9 or more, indicating behaviour problems at home. Comparison of the 2 subgroups indicated no significant differences in the prevalence of problem behaviour at home, although again the small numbers involved are likely to have introduced a Type II statistical error (Table 7.7).

POST-OPERATIVE SCORES ON THE RICHMAN BEHAVIOUR CHECKLIST		
	NUMBER SCORING < 9	NUMBER SCORING ≥ 9
CYANOTIC (n=5)	4 (80%)	1 (20%)
ACYANOTIC (n=3)	3 (100%)	0 (0%)
TOTAL CARDIAC (n=8)	7 (88%)	1 (12%)
BMT (n=4)	4 (100%)	0 (0%)
HEALTHY (n=7)	6 (86%)	1 (14%)

Table 7.7

7.3.2. Comparison with the Reference Groups

Five BMT patients and 7 healthy children were eligible for completion of the Richman BCL and completed questionnaires were received on 4 BMT patients and 7 healthy children. There were no significant differences between the 3 groups in the prevalence of problem behaviour (Table 7.7).

7.3.3. Changes Over Time: Cardiac Group

Richman BCL data were collected on 5 children before and after surgery, 2 of whom had cyanotic disorders and 3 had acyanotic lesions. There was no significant association between pre- and post-operative scores in either subgroup.

7.3.4. Changes Over Time: Comparison with the Reference Groups

Only 2 children in each of the reference groups had Richman BCL questionnaires completed before and after surgery, so the numbers were too small for any statistical analyses.

7.4. POST-OPERATIVE BEHAVIOUR AT HOME: 5 - 17 YEARS

7.4.1. Cardiac Group

Of the 30 children aged 5-17 years, Rutter A scale questionnaires were returned on all 30. Five (17%) obtained a score of 13 or more, indicating a significant degree of problem behaviour at home. Four of them had neurotic problems and 1 had an undifferentiated pattern of behaviour problems. Eight of the 30 had cyanotic lesions and 22 had acyanotic defects. Comparison of the 2 subgroups indicated no significant differences in the prevalence of problem behaviour at home or in the nature of the problems (Table 7.8). Within the acyanotic group, 6 were symptomatic and 16 were asymptomatic. Those children with symptoms had a significantly higher prevalence of problem behaviour than those who were asymptomatic (chi-squared = 5.50402; p=.0190). One child in the cardiac group had developed psychological problems since surgery, necessitating professional intervention. This child was rated as having a significant degree of problem behaviour on the Rutter A scale.

BEHAVIOUR AT HOME: RUTTER A SCALE			
		POST-OPERATIVE	
	PRE-OPERATIVE	< 13	≥ 13
CYANOTIC (n= 7)	< 13	5 (100%)	
	≥ 13		2 (100%)
ACYANOTIC (n=19)	< 13	15 (88%)	2 (12%)
	≥ 13	1 (50%)	1 (50%)
TOTAL CARDIAC (n=26)	< 13	20 (91%)	2 (9%)
	≥ 13	1 (25%)	3 (75%)
BMT (n=21)	< 13	14 (74%)	5 (26%)
	≥ 13	1 (50%)	1 (50%)
HEALTHY (n=26)	< 13	22 (92%)	2 (8%)
	≥ 13	2 (100%)	

Table 7.8

7.4.1.1. Readmission to Hospital

Thirteen of the children had to be readmitted to hospital, 10 of whom had cyanotic lesions and 3 of whom had acyanotic lesions. There was a significant correlation between time spent in hospital after the initial surgery and the presence of behaviour problems on the Rutter A scale (r=.2992; p=.047).

7.4.2. Comparison with the Reference Groups

Completed questionnaires were received on 24 of the 30 eligible BMT patients and on 31 of the 34 eligible children in the healthy group. Comparison of these with the cardiac group indicated no significant differences in the prevalence of problem behaviours at home (Table

7.8) but the BMT group had a higher prevalence of problem behaviours at home compared with the healthy group (chi-squared = 8.76748; p=.0125).

A higher proportion of children in both reference groups had antisocial patterns of behaviour (50% in each) compared with the cardiac group.

No children in either of the 2 reference groups developed psychological problems which necessitated professional intervention.

7.4.3. Changes Over Time: Cardiac Group

For the 7 children in the cyanotic group on whom data were available at both test occasions, the presence of behaviour problems before surgery was a significant predictor of behaviour problems after surgery (Fisher's Exact Test: p=.04762), but this was not true for the acyanotic patients (Fisher's Exact Test: p=.29825) (Table 7.8).

7.4.4. Changes Over Time: Comparison with the Reference Groups

For the 26 children in the cardiac group on whom Rutter A data were collected before and after surgery, there was a significant association between pre- and post-operative scores (chi-squared = 5.698; p=.0170) but this was not found in either of the 2 reference groups (Table 7.8).

7.5. POST-OPERATIVE BEHAVIOUR AT SCHOOL

7.5.1. Cardiac Group

Forty cardiac children were aged 3 years or older and were therefore eligible for some form of education. Three of the 10 children aged 3-5 years were not attending nursery or playgroup. Two children aged 5 years or older were attending special school; the remainder were in full-time, normal school. The mean time after surgery before returning to school was 2.9 months. Questionnaires were sent to the teachers of the 30 children aged 5 years or older and completed Rutter B questionnaires were received on 29 (97%). Six (21%) obtained a score of 9 or more, indicative of a significant degree of problem behaviour at school. Four had neurotic problems and in 2 cases the problems were undifferentiated.

BEHAVIOUR AT SCHOOL: RUTTER B SCALE			
		POST-OPERATIVE	
	PRE-OPERATIVE	< 9	≥ 9
CYANOTIC	< 9	4 (80%)	1 (20%)
(n= 8)	≥ 9	2 (67%)	1 (33%)
ACYANOTIC	< 9	15 (94%)	1 (6%)
(n=19)	≥ 9		3 (100%)
TOTAL CARDIAC	< 9	19 (90%)	2 (10%)
(n=27)	≥ 9	2 (33%)	4 (67%)
BMT	< 9	10 (83%)	2 (17%)
(n=17)	≥ 9	3 (60%)	2 (40%)
HEALTHY	< 9	21 (95%)	1 (5%)
(n=27)	>/ 9	4 (80%)	1 (20%)

Table 7.9

Eight of the 29 had cyanotic lesions and 21 had acyanotic defects. Comparison of the 2 subgroups indicated no significant differences (Table 7.9).

There was no significant correlation between time spent in hospital after the initial surgery and the presence of adjustment problems on the Rutter B scale. There was also no significant correlation between mean time after surgery before returning to school and adjustment problems on the Rutter B scale.

7.5.2. Comparison with the Reference Groups

Thirty-five BMT patients and 41 healthy children were eligible for some form of education. Two of the 5 BMT children aged 3-5 years were not attending playgroup or nursery and 5 children in the older age group were not attending school. Two children in this latter age group were going to school part-time and one other child was at a special school. The remainder were at normal full-time school. The mean time after BMT before returning to school was 7 months. All children in the healthy group were at normal full-time school. Completed Rutter B questionnaires were received back on 22 of the 25 BMT patients aged 5 years or older and attending school and on 31 of the 34 healthy children of 5 years or older. Comparison of the 3 groups indicated no significant differences in the prevalence of problem behaviour at school (Table 7.9). In the BMT group there was no significant correlation between mean time after BMT before returning to school and adjustment problems on the Rutter B scale.

There was a higher proportion of antisocial patterns of behaviour in both reference groups (50% and 100% for the BMT and healthy groups respectively) compared with the cardiac group .

7.5.3. Changes Over Time: Cardiac Group

Rutter B questionnaires were completed on 27 children before and after surgery. In the cyanotic group pre-operative scores were not significantly associated with post-operative scores (Fisher's Exact Test: p=.64286) but in the acyanotic group there was a significant association between pre- and post-operative scores (Fisher's Exact Test: p=.00413) (Table 7.9).

7.5.4. Changes Over Time: Comparison with the Reference Groups

For the cardiac group overall, pre-operative Rutter B score was a significant predictor of post-operative score (chi-squared = 5.82015; p=.0158) but there was no significant association between pre- and post-operative scores in either the BMT or healthy groups (Table 7.9).

7.6. POST-OPERATIVE PARENTAL INTERVIEW: 3 - 17 YEARS

7.6.1. Cardiac Group

Forty children were 3 years or older and parental interview data concerning the child's behaviour were collected on 39 (98%), 14 of whom had cyanotic lesions and 25 had acyanotic defects. Parents were asked to rate whether or not their child had problems in 8 different areas of behaviour and were also asked a general question regarding the occurrence of "behaviour problems". Cyanotic children were rated as having significantly more temper

tantrums (U= 132.0; p=.0298) and being significantly more anxious (U=97.5; p=.0197) and less active (U=137.5; p=.0175) than acyanotic children (See Appendix C: Table 11).

Eleven children (21%) were felt by their parents to have behaviour problems, 7 of whom had cyanotic lesions and 4 had acyanotic defects. The prevalence of behaviour problems was significantly higher in the cyanotic group (chi squared = 3.90813; p=.0481).

7.6.2. Comparison with the Reference Groups

Thirty-five BMT patients and 41 healthy children were 3 years or older and parental interview data were collected on 32 BMT patients and on all 41 children in the healthy group. Relative to the BMT group the cardiac group were rated as having significantly more problems in new situations (U = 411.5; p=.0192), being more disobedient (U=487.0; p=.0181), and having more difficulties in relationships with adults (U = 544.0; p=.0371). Comparison of the cardiac and healthy groups indicated that cardiac children were significantly more disobedient (U = 607.5; p=.0039), more anxious (U = 577.0; p=.0068), more dependent (U=663.5; p=.0477) and had more problems in relationships with other children (U = 673.5; p=.0357) (See Appendix C: Table 12). Children in the BMT group were significantly more anxious (U=442.0; p=.0016) and had more difficulties in relationships with other children (U=485.5; p=.0030) compared with the healthy group.

Comparison of the cyanotic and acyanotic subgroups with the 2 reference groups indicated that the cyanotic group had more problems with new situations (U=119.5; p=.0068), were more disobedient (U=174.0; p=.0408), had more problems with relationships with adults (U=176.0; p=.0074), were more dependent (U=155.0; p=.0218) and less active (U=182.0;

p=.0402) than the BMT group (see Appendix C: Table 13). Compared with the healthy group, the cyanotic group had more problems in new situations (U=196.0; p=.0324), more temper tantrums (U=219.0; p=.0150), were more disobedient (U=217.0; p=.0123), more anxious (U=132.5; p=.0002), had more problems with relationships with children (U=190.0; p=.0021) and were more dependent (U=185.0; p=.0033). The acyanotic children were more disobedient than both the BMT group (U=313.0; p=.0268) and the healthy group (U=390.5; p=.0067). (See Appendix C: Table 14).

Looking at the children in each of the groups in terms of the numbers of areas in which they were felt to have problems, there were fewer cyanotic children with no problems and a higher proportion of cyanotic children with problems in more than 3 areas than in the other groups (Table 7.10).

The prevalence of behaviour problems was 21%, 17% and 4% for the cardiac, BMT and healthy groups respectively. The cardiac group had a significantly higher prevalence of behaviour problems than the healthy group (chi-squared = 4.46206; p=.0347). Comparison of the cyanotic and acyanotic subgroups with the reference groups indicated that the cyanotic group had a higher prevalence of behaviour problems than the healthy group (chi-squared = 10.07423; p=.0015). There were no significant differences between the BMT group and the total cardiac group or either of the cardiac subgroups.

For those children on whom Rutter A and parental interview data were available, there was no association for any of the groups or for the cyanotic and acyanotic subgroups between the presence or absence of behaviour problems on the 2 measures.

POST-OPERATIVE BEHAVIOUR: NUMBERS OF AREAS IN WHICH CHILDREN WERE RATED AS HAVING PROBLEMS					
	CYANOTIC (n=14)	ACYANOTIC (n=25)	TOTAL CARDIAC (n=39)	BMT (n=32)	HEALTHY (n=41)
No problems	2 (14%)	10 (40%)	12 (31%)	12 (38%)	24 (59%)
Problems in 1 area	3 (21%)	5 (20%)	8 (21%)	11 (34%)	10 (24%)
Problems in 2 areas	0 (0%)	6 (24%)	6 (15%)	3 (9%)	5 (12%)
Problems in 3 areas	2 (14%)	2 (8%)	4 (10%)	4 (13%)	1 (2%)
Problems in > 3 areas	7 (50%)	2 (8%)	9 (23%)	2 (6%)	1 (2%)

Table 7.10

7.6.3. Changes Over Time: Cardiac Group

There were no changes on any of the parental interview items for the 11 cyanotic patients on whom data were collected before and after surgery. For the 25 acyanotic patients parents felt that there were significantly more problems with disobedience (Z= -2.0226; p=.0431) after surgery compared with pre-operatively.

7.6.4. Changes Over Time: Comparison with the Reference Groups

Parental interview data were collected on 36 cardiac patients, 26 BMT patients and 36 healthy children on both test occasions. Children in the cardiac group were rated as being more disobedient (Z= -2.0226; p=.0431) and more active (Z= -2.3664; p=.0180) after

surgery compared with beforehand. Parents of BMT patients felt that their children had significantly fewer problems in new situations (Z= -1.9604; p=.0500) post-operatively. In the healthy group children were rated as having fewer problems in new situations (Z=-2.3953; p=.0166) and in relationships with children (Z=-2.2014; p=.0277) at the second test occasion.

To examine the possible influence of age on behaviour, the sample was split into two age groups according to the overall median age. For the younger age group there were no significant changes over time for the total cardiac, BMT, healthy or cyanotic groups. The acyanotic group were felt to be less anxious after surgery (Z=-2.1325; p=.0330). In the older age group there were no significant changes over time for the cyanotic and acyanotic subgroups. Both the BMT and healthy groups had fewer problems in new situations at the second test occasion (Z=-2.0226; p=.0431; Z=-2.5205; p=.0117 for the BMT and healthy groups respectively). The BMT group were also felt to be less dependent (Z=-1.9604; p=.0500). The healthy group had fewer problems in relationships with children at the second test occasion (Z=-2.2014; p=.0277). The only significant change in the cardiac group was that they were more active after surgery (Z=-2.0226; p=.0431).

7.7. POST-OPERATIVE SELF-PERCEPTION

7.7.1. Cardiac Group

Twenty-nine of the 30 children (98%) aged between 5 and 17 years completed the self-perception test at follow-up, 7 of whom had cyanotic lesions and 22 had acyanotic defects. The only significant difference between the 2 subgroups was on the weak-strong construct, where the cyanotic patients rated themselves as stronger than the acyanotic group did

(U=39.0; p=.0365) (See Appendix C: Table 15). When the cardiac sample was divided into two according to the median age (7.75 years), cyanotic children in the younger age group rated themselves as more lonely than those in the acyanotic group (U=5.5; p=.0295). In the older age group acyanotic children liked themselves less (U=7.5; p=.0441) and rated themselves as more angry (U=8.0; p=.0442) than the cyanotic children.

There were no significant differences between the cyanotic and acyanotic groups on their ratings of their "ideal self" on any of the 8 constructs (See Appendix C: Table 16).

Finally a score was computed for each construct which reflected the difference between "self" and "ideal self" ratings, but again there were no significant differences between the 2 subgroups (See Appendix C: Table 17).

7.7.2. Comparison with the Reference Groups

Twenty-four of the 30 eligible BMT patients and all 34 of the eligible children in the healthy group completed the self-perception test. The cardiac children rated themselves as more frightened (U=251.5; p=.0297), more angry (U=264.5; p=.0540) and more ill (U=259.0; p=.0543) than the BMT group. There were no significant differences between the cardiac and healthy groups (See Appendix C: Table 18). The BMT group rated themselves as more miserable (U=272.0; p=.0073) and more angry (U=229.5; p=.0013) compared with the healthy group. The total self perception score in the BMT group was lower (more negative) than that of the healthy group (U=237.0; p=.0207).

When the 3 groups were divided into two according to the median age of the total sample (9.5 years), the cardiac children in the younger age group did not differ significantly from those in either of the 2 reference groups. In the older age group, the cardiac children rated themselves as more frightened (U=43.0; p=.0297) and less good (U=39.5; p=.0299) than those in the BMT group. There were no significant differences between the cardiac and healthy groups.

The only significant difference on ratings of "ideal self" between the 3 groups was that the cardiac group rated their ideal self as more angry than the BMT group did (U=265.0; p=.0376) (See Appendix C: Table 19).

The only significant difference in the computed score, reflecting the difference between self and ideal-self, was between the cardiac and healthy groups on the angry-calm construct (U=338.5; p=.0394) (See Appendix C: Table 20).

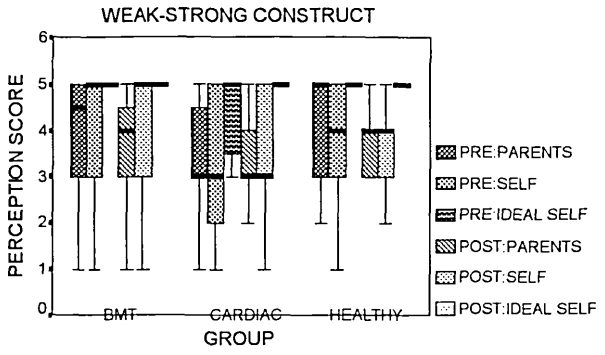
7.7.3. Changes Over Time: Cardiac Group

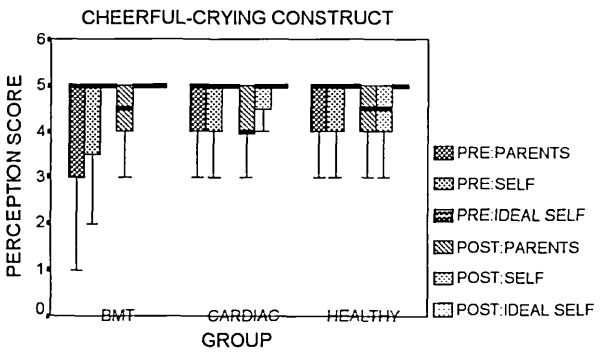
For the 7 cyanotic patients who completed the self-perception task before and after surgery, the only parameter on which the change in score approached significance was on the ill-well construct (Z= -1.8869; p=.0592). There were no significant changes over time on any of the ideal self scores. Eighteen patients with acyanotic lesions completed the task on both occasions. They perceived themselves to be significantly less frightened (Z= -1.9876; p=.0469) and less ill (Z= -2.0449; p=.0409) post-operatively and their total score was higher (more positive) at follow-up (Z=-2.3669; p=.0179). There were no significant changes over time on any of the ideal self scores (figures 2-9).

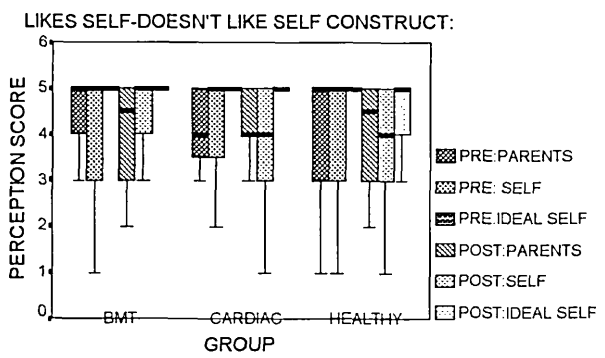
Figures 2-9 are box and whisker plots and represent the pre- and post-operative perceptions on the eight constructs. The bars represent centiles (25th and 75th), the lines represent outliers and the horizontal solid bar is the median. In each instance the sequence is pre-operative perception (parents, child:self and child:ideal self) and post-operative perception (parents, child:self and child:ideal self).

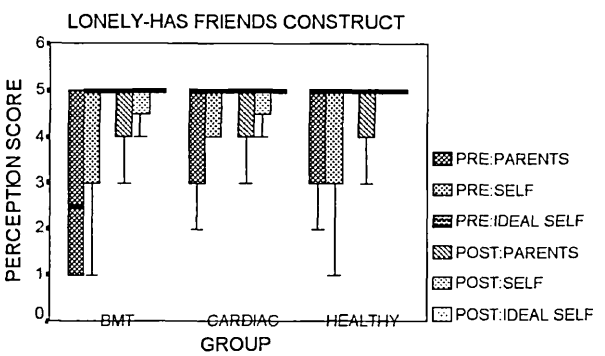
7.7.4. Changes Over Time: Comparison with the Reference Groups

Twenty-five cardiac patients, 23 BMT patients and 30 healthy children completed the self-perception test pre- and post-operatively. The cardiac patients rated themselves as significantly less ill (Z= -2.7219; p=.0065) and also rated their ideal self as significantly less ill (Z= -1.9917; p=.0464) after surgery compared with beforehand. Their total self perception score was higher at follow-up (Z =-2.0286; p =.0425). The BMT patients perceived themselves to be significantly more cheerful (Z= -2.0732; p=.0382), less frightened (Z= -2.3412; p=.0192), less angry (Z= -2.2014; p=.0277) and less ill (Z= -3.0594; p=.0022) after treatment and their total score was higher at follow-up (Z=-2.4990; p=.0125). Children in the healthy group rated themselves as significantly more angry (Z= -2.2363; p=.0253) at follow-up. There were no significant changes over time in ideal self-perception scores in either the BMT or healthy groups.

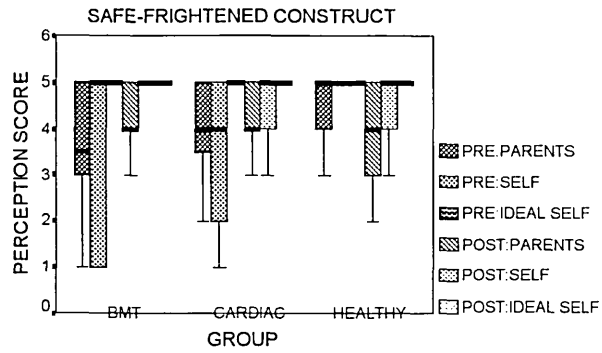

WEAK-STRONG CONSTRUCT
PERCEPTION SCORE
0
1
2
3
4
5
6
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF

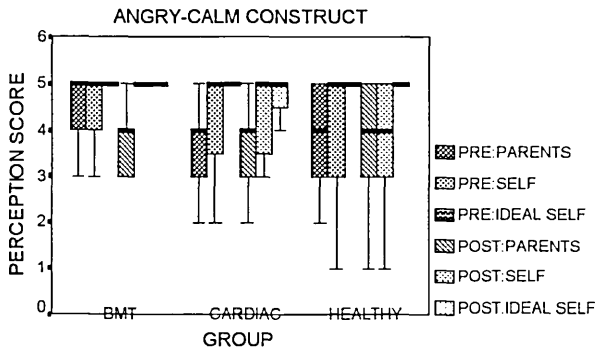

CHEERFUL-CRYING CONSTRUCT
PERCEPTION SCORE
0
1
2
3
4
5
6
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF

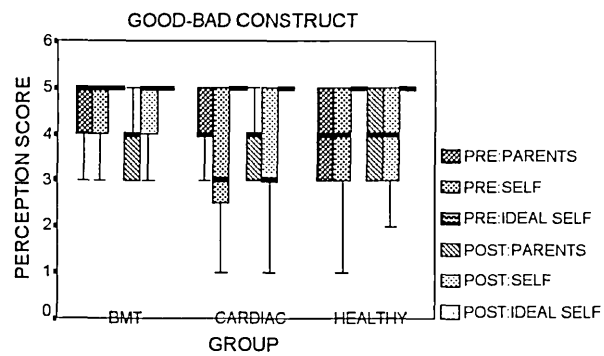

LIKES SELF-DOESN'T LIKE SELF CONSTRUCT:
PERCEPTION SCORE
0
1
2
3
4
5
6
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE: SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF

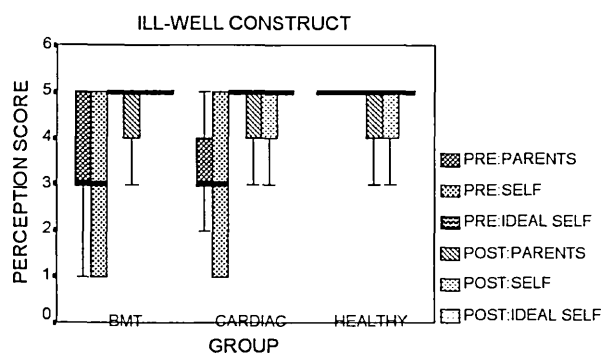

LONELY-HAS FRIENDS CONSTRUCT
PERCEPTION SCORE
0
1
2
3
4
5
6
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF

Figs. 2-5


SAFE-FRIGHTENED CONSTRUCT
PERCEPTION SCORE
6
5
4
3
2
1
0
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF


ANGRY-CALM CONSTRUCT
PERCEPTION SCORE
6
5
4
3
2
1
0
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST.IDEAL SELF


GOOD-BAD CONSTRUCT
PERCEPTION SCORE
6
5
4
3
2
1
0
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF


ILL-WELL CONSTRUCT
PERCEPTION SCORE
6
5
4
3
2
1
0
BMT
CARDIAC
HEALTHY
GROUP
PRE:PARENTS
PRE:SELF
PRE:IDEAL SELF
POST:PARENTS
POST:SELF
POST:IDEAL SELF

Figs. 6-9

7.7.5. Self-Perception and Behaviour Problems

There was no correlation between total pre-operative self-perception scores and post-operative Rutter A scores in any of the 3 groups. In the cardiac group there was a significant correlation between total post-operative self-perception scores and the presence or absence of behaviour problems on the Rutter A ($r = .5883$; $p=.000$; $n=28$). The presence of behaviour problems was associated with a more negative self-perception. There was no relationship between post-operative self-perception and Rutter A scores for either of the 2 reference groups. In terms of the neurotic and antisocial subscales of the Rutter A, post-operative self perception scores were significantly correlated with the neurotic subscale score in the cardiac group ($r=-.4817$; $p=.005$) but not in the 2 reference groups. There was no correlation between antisocial subscale scores and post-operative self perception scores in any of the 3 groups.

7.8. POST-OPERATIVE PARENTAL PERCEPTION

7.8.1. Cardiac Group

Data on 39 of the 40 eligible children (98%) were collected, where parents were asked to rate their children on the same 8 constructs as the children had rated themselves. In addition, a rating of personality was also given. There were no significant differences in parental ratings on any of the constructs between the cyanotic ($n=14$) and acyanotic ($n=25$) groups (See Appendix C: Table 21). When the sample was divided into two according to the median age (6.85 years), parents of cyanotic children in the younger age group rated them as more ill ($U=48.0$; $p=.0488$) than parents in the acyanotic group rated their children. There were no significant differences in parental ratings of older children in the two subgroups.

Comparison of parents' and children's ratings indicated a greater degree of agreement than pre-operatively. In the cyanotic group there were significant correlations on the doesn't like self-likes self, lonely-has friends, frightened-safe and angry-calm constructs and on the total score. In the acyanotic group correlations were significant on the crying-cheerful, doesn't like self-likes self and lonely-has friends constructs and on the total score (Table 7.11).

7.8.2. Comparison with the Reference Groups

Comparison of the cardiac, BMT (n=29) and healthy (n=41) groups indicated no significant differences between the cardiac and reference groups on any measures of parental perception (See Appendix C: Table 22), although parents in the BMT group rated their children as more angry (U=440.0; p=.0482) than parents of children in the healthy group. When the sample was divided into younger and older children (median age: 7.75 years), parents of the younger cardiac children rated them as more ill (U=77.0; p=.0105) compared with parental ratings in the BMT group. There were no significant differences between parental ratings of the cardiac and healthy children in the younger age group. In the older age group there were no differences between parental ratings in the cardiac and BMT groups but parents of the cardiac children did rate their children as weaker (U=101.0; p=.0316) compared with parental ratings of the healthy children.

POST-OPERATIVE DATA: CHILD AND PARENTAL PERCEPTION: CORRELATION: TOTAL SAMPLE		
	CYANOTIC (n=7)	ACYANOTIC (n=21)
WEAK-STRONG	r= .3654 p= .210	r= .3448 p= .063
CRYING-CHEERFUL	r= .4524 p> .05	r= .7671 p= .000
DOESN'T LIKE SELF-LIKES SELF	r= .7943 p= .016	r= .5267 p= .007
LONELY-HAS FRIENDS	r= .8627 p= .006	r= .4623 p= .017
FRIGHTENED-SAFE	r= .9270 p= .001	r= .2401 p= .147
ANGRY-CALM	r= .7508 p= .026	r= .2947 p= .097
BAD-GOOD	r= .1442 p= .379	r= -.1080 p= .321
ILL-WELL	r= .0685 p= .442	r= .0649 p= .390
TOTAL SCORE	r= .8105 p= .014	r= .3673 p= .051

Table 7.11

Comparison of parents' and children's ratings indicated some agreement. In the cardiac group as a whole there were significant correlations on the crying-cheerful, doesn't like self-likes self, lonely-has friends, frightened-safe and angry-calm constructs and on the total score. In the BMT group there were significant correlations on the weak-strong, lonely-has friends and frightened-safe constructs and on the total score and in the healthy group on the weak-strong, doesn't like self-likes self and angry-calm constructs and on the total score (Table 7.12).

POST-OPERATIVE DATA: CHILD AND PARENTAL PERCEPTION: CORRELATION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS			
	CARDIAC (n=28)	BMT (n=23)	HEALTHY (n=34)
WEAK-STRONG	r= .0884 p= .327	r= .7310 p= .000	r= .3306 p= .028
CRYING-CHEERFUL	r= .5421 p= .001	r=-.0453 p= .419	r= .1888 p= .142
DOESN'T LIKE SELF-LIKES SELF	r= .6008 p= .000	r= .0621 p= .389	r= .3936 p= .011
LONELY-HAS FRIENDS	r= .6174 p= .000	r= .5166 p= .006	r= .1002 p= .286
FRIGHTENED-SAFE	r= .4765 p= .005	r= .4786 p= .010	r= .2557 p= .072
ANGRY-CALM	r= .4776 p= .005	r=-.2259 p= .150	r= .2989 p= .043
BAD-GOOD	r=-.0305 p= .439	r=-.1724 p= .216	r= .0448 p= .401
ILL-WELL	r= .0831 p= .337	r= .1804 p= .211	r=-.0246 p= .445
TOTAL SCORE	r= .6137 p= .000	r= .7318 p= .000	r= .3465 p= .022

Table 7.12

7.8.3. Changes Over Time: Cardiac Group

There were no significant changes on any of the constructs in parental ratings of the 11 cyanotic patients on whom data were collected pre- and post-operatively. Twenty-three children with acyanotic lesions were rated by their parents both pre- and post-operatively. The only significant change was on the ill-well construct, on which parents felt that their children were significantly less ill (Z= -2.1339; p=.0329) at the follow-up assessment.

7.8.4. Changes Over Time: Comparison with the Reference Groups

Parents of the 34 cardiac patients rated their children as less good (Z= -1.9882; p=.0468) and less ill (Z= -2.6157; p=.0089) post-operatively. For the 25 BMT patients on whom data were collected both pre- and post-operatively, parents rated their children as being significantly less lonely (Z= -2.7872; p=.0053), less frightened (Z= -2.2363; p=.0253), less good (Z= -2.1583; p=.0309) and less ill (Z= -2.7605; p=.0058) at follow-up. There were no significant changes over time in parental ratings of the 36 healthy children.

7.9. DISCUSSION OF THE RESULTS

7.9.1. Developmental Function: 0 - 4.5 Years

The findings of poorer performance on all of the developmental parameters in the cyanotic group contradict those of other researchers, who have found significant improvements in cognitive function for cyanotic patients who have had corrective surgery (Linde et al, 1970), although this latter study focused on older children. There are few prospective studies evaluating the impact of cardiac surgery on development in children under 3 years of age, but retrospective studies comparing children with cyanotic and acyanotic lesions who underwent corrective surgery in infancy have also found significant differences between the two groups, with children with cyanotic lesions performing less well (Newburger et al, 1984; Hesz & Clark, 1988). Retarded developmental outcome following surgery in infancy has also been linked to a number of family variables, such as socioeconomic status (O'Dougherty et al, 1983). Maternal overprotectiveness in particular is also relevant for the cyanotic group, with mothers of 3 of the 6 cyanotic patients in the younger age group specifically mentioning that their children were very dependent and clingy and that they didn't mix with other children. From the interviews it was apparent that these mothers were themselves anxious and

unwilling to allow their children to be more active and engage in more exploratory behaviour. In contrast, the more marked improvement noted by parents in physical condition in the acyanotic patients may have resulted in decreased parental overprotectiveness and a greater willingness to allow their child to develop appropriately. A further factor for explaining the differences between the 2 subgroups is that 6 children (86%) with cyanotic lesions had been rehospitalised since surgery, compared with none in the acyanotic group. Hospitalisation, particularly between the ages of 6 months and 4 years, is acknowledged to be potentially harmful emotionally (Prugh et al, 1953; Vernon et al, 1966) and is also likely to compromise cognitive development. Furthermore, a number of the children had to undergo further procedures under general anaesthesia. Increasing numbers of procedures under general anaesthesia and periods of hospitalisation have been associated with adverse effects on the cognitive development of pre-school aged children (Ludman et al, 1993). Although the cyanotic group did not show a loss of skills over time, they did not achieve the rate of development expected in one year.

The significantly lower scores obtained by the cardiac group on 3 of the subscales and on the overall IQ compared with the healthy group are accounted for by the significant differences between the healthy and cyanotic groups. There were no differences between the healthy and acyanotic groups. These results are similar to those for older children of Kramer et al (1989), who found that children with congenital heart disease and no physical handicap did not differ from healthy children, whereas those with congenital heart disease and a degree of physical impairment had lower IQ scores than healthy children. As expected, the healthy group did not show any significant changes in performance over time, although on both test occasions they achieved higher scores than would be expected from the standardised norms for the test

(Griffiths, 1954; 1970). The most likely explanation for this is that the standardised norms for the tests are no longer appropriate. A study of 447 infants tested from 1978-1982 using the same scales found a significantly higher DQ than that originally suggested for the scales (Hanson, Aldridge Smith & Hume, 1985).

Pre-operatively, the presence of a heart condition, with surgery pending, rather than the type of lesion, differentiated the cardiac and healthy groups. Post-operatively, there are clear differences according to the nature of the underlying disorder, with children with corrected acyanotic lesions indistinguishable in terms of developmental function from the healthy group. A variety of factors have been suggested to account for such findings - such as physical condition, hospitalisation and maternal attitudes (Linde et al, 1970; O'Dougherty et al, 1983; Kramer et al, 1989). The results of this research support such a multifactorial aetiology.

Although the BMT group had experienced significant amounts of hospitalisation and traumatic treatment regimens, there did not appear to be any deleterious effects on developmental functioning. The acyanotic, BMT and healthy groups were largely comparable. A number of patients in the BMT group had acquired disorders which had been successfully treated. One explanation for the differences with the cyanotic group is that the condition of the cyanotic group is congenital but, unlike the acyanotic group, their medical progress after surgery had been less straightforward and in 1 case (14%) surgery had been palliative. Thus after treatment, in contrast to the pre-operative findings, the nature of the underlying illness is a relevant factor in determining the effects on development. Grouping all cardiac patients together post-operatively is misleading - the evidence is that the presence of a cyanotic lesion potentially can damage the developing brain, whereas for those with

acyanotic lesions successfully repaired in infancy, the outlook is more promising. Despite the different nature of the treatment protocols for cardiac surgery and BMT patients, the lack of significant differences between the acyanotic and BMT groups suggests that the actual treatment is of less significance in determining developmental outcome than the nature of the underlying disorder. For other groups of chronically sick children with onset of their disease in the first year of life, development has remained delayed one year after corrective surgery and it is suggested that for such children "normalisation" of development does not occur rapidly (Stewart, Uauy, Waller, Kennard, Benser & Andrews, 1989).

7.9.2. Cognitive Ability: 4.5 - 17 Years

Within the older age group, the cyanotic group were performing less well than the acyanotic group on all of the cognitive parameters. The cyanotic group did not show any significant changes in performance over time, although scores on all of the parameters were lower at follow-up. In the acyanotic group the results were more mixed. There was a significant improvement in the recall of digits score but a significant deterioration in the matrices score. The improvement in recall of digits (a short-term memory test) could indicate improved concentration post-operatively. With surgery pending, the aspect of testing most likely to have been influenced by the situation pre-operatively was short-term memory. The significant improvement after surgery supports this view.

The finding of an overall deterioration in performance in the older cyanotic group is similar to that for the younger age group. In general, the degree of change in the acyanotic group was quite small, as expected. The acyanotic group were performing closer to their true potential initially and were therefore not expected to show a lot of change after surgery.

Once again the findings of the cyanotic group contradict those of other researchers who found improvement in cognitive function after surgery (Linde et al, 1970). However, the poor academic functioning of the cyanotic patients supports the finding that, even in children with surgically corrected cyanotic lesions, performance on academic functioning is significantly worse than that of other groups of children (Wright & Nolan, 1994).

Within our study there were only a small number of cyanotic patients, which might have influenced the results. Furthermore, 4 cyanotic patients (36%) had been readmitted to hospital compared with 3 (13%) in the acyanotic group. Although there was no significant difference in the time after surgery before restarting school, the cyanotic patients did miss more school, which would also be expected to affect their performance. A lack of schooling would be expected to influence the incidence of reading problems, where comparisons are made with age-related norms, and is likely to be a contributory factor to the significantly higher prevalence of reading problems in the cyanotic group. Other factors include parental attitude - one mother whose child had had successful corrective surgery for a complex cyanotic lesion commented that she was "very careful" with her daughter, admitting that after having a sick child for 9 years it was very difficult to treat her normally and "let her get on".

As found pre-operatively, at follow-up the acyanotic group were largely indistinguishable in terms of cognitive function from the healthy group. Medical intervention does not appear to have had a major impact on performance. In contrast, the cyanotic group did differ from the healthy group on a number of parameters, although the fact that differences in IQ were present before and after treatment again suggests that the intervention was of less significance than the underlying lesion. Post-operatively, the significant differences between the cyanotic

and healthy groups on reading IQ and prevalence of reading problems are likely to be attributable to the schooling missed by the cyanotic patients.

As pre-operatively, the BMT and cyanotic groups demonstrated similar differences with the healthy group, predominantly in areas related to academic functioning. Scores for all of the groups were within the normal range, although, as mentioned previously, this needs to be interpreted with caution. The lack of change in the BMT group again suggests that it is the effects of the underlying disorder, rather than the treatment of BMT, which is the significant factor for cognitive function. These findings also support the view that chronic illness and subsequent treatment interfere with schooling and affect academic performance (Pless & Pinkerton, 1975; Eiser, 1980(a); Mearig, 1985; Peckham, 1988).

7.9.3. Behaviour at Home: 3 - 5 Years

Within the younger age group there were no significant differences between the cyanotic and acyanotic subgroups or between the cardiac, BMT and healthy groups in the prevalence of problem behaviour. There were also no significant changes over time or significant associations between pre- and post-operative scores in any of the groups. The small numbers again make interpretation difficult. Neither of the two illness groups had a higher prevalence of problem behaviour than the healthy group, and post-operatively the pattern of areas of difficulty mentioned by parents in all three groups was similar. It is, however, not possible to say that there has been a change in problems because of the small numbers seen on both occasions, before and after treatment. The other important factor in interpreting the results is normal developmental changes over the course of a year and the fact that different behaviours predominate at different ages (Richman, 1988). Although pre-school aged

children with chronic illness who have undergone hospitalisation and stressful invasive procedures are recognised to be at risk of behaviour problems (Prugh et al, 1953; Goslin, 1978; Richman, 1988), the post-operative results from this study are based on too small a sample size to support or refute this view.

7.9.4. Behaviour at Home: 5 - 17 Years

For the older age group there were no significant differences between the cyanotic and acyanotic groups in the prevalence of post-operative problem behaviours, but within the acyanotic group those who were symptomatic had a significantly higher prevalence of problem behaviour at home than those who were asymptomatic. Possible explanations for this are that those children who were symptomatic prior to surgery were finding it difficult to adjust to being "normal", whereas for those who were asymptomatic and well beforehand, there was little change. Mothers of the previously symptomatic patients may also be unable to treat their children normally, leading to adjustment difficulties in the child.

There were no significant changes over time in the number of children who were reported to have behaviour problems. However, in the cyanotic group the presence of behaviour problems before surgery was a significant predictor of behaviour problems after surgery, whereas in the acyanotic group it was not. These results do not support the suggestion that there are improvements in adjustment of children with congenital heart disease following surgery (Landtman et al, 1959; Landtman, 1973), particularly for cyanotic patients (Linde et al, 1970). A possible explantation for the differences between our results and those of previous researchers is the difference in assessment tools. In the Landtman studies (1959; 1973) the measures were qualitative. In the Linde et al study (1970) "adjustment" was

measured on global rating scales and specific behaviours were not assessed. For the cyanotic group in our study in particular, behaviour problems were stable over time, suggesting that it is the effects of the cardiac condition which is of specific relevance and that the intervention of cardiac surgery has little impact on the prevalence of problem behaviours, at least in the short term. Central nervous system impairment, which is more prevalent in cyanotic patients (Silbert et al, 1969) was not looked at specifically in this study, but it has been suggested that it is linked to poor overall psychological functioning (DeMaso et al, 1990), and this may be a further contributory factor in the findings of this study. Significantly more adjustment difficulties have been reported in children with corrected cyanotic lesions compared with controls (Wright & Nolan, 1994), which supports the results of this study.

On individual items of the Rutter A scale, parents of cyanotic patients felt that post-operatively there were more difficulties in terms of irritability, misery, anxiety, sleeping problems and disobedience in particular (see Appendix D: Table 27). This suggests an increase in depressive symptomatology, and one explanation for this is that the surgery had not afforded the children the benefits they had expected, resulting in disillusionment and depression. A greater number of the cyanotic group had experienced medical problems and further hospitalisation since their surgery, which they had probably not anticipated, and this might be expected to result in increased feelings of depression and anxiety.

Although not significant, the prevalence of problem behaviour at home was higher in the cardiac group than in the healthy group. As pre-operatively, of those children with behaviour problems in the healthy group, a higher percentage had antisocial patterns of behaviour than in the cardiac group. Low rates of antisocial patterns of behaviour have been previously

reported for children with surgically corrected congenital heart disease (Stucki, Stocker, Hammerli, Rufenacht, Stucki, Weber & Schupbach, 1992). Overall, parents of the healthy group rated their children as having more problems with stealing, fighting, disobedience and lying. The finding of a higher prevalence of problem behaviour in cardiac children after surgery compared with healthy children supports other findings in the literature (Utens et al, 1993). In contrast to the cardiac group, behaviour problems at the first test occasion were not significantly associated with behaviour problems at follow-up in the healthy group. This was due to a reduction in the prevalence of behaviour problems in the healthy group, with no child developing new behaviour problems over time.

Whilst the cardiac and BMT groups did not differ significantly in the prevalence of problem behaviour at home, both had a higher rate of problems than the healthy group, with the differences reaching significance for the BMT and healthy groups. Rates for the cyanotic and BMT groups were comparable and are likely to be a reflection of the more serious initial illness, treatment and ongoing difficulties in these two groups compared with the acyanotic group. These results correspond with other findings in the literature that children who have undergone stressful hospital treatments for chronic conditions are more likely to show a deviant behaviour pattern compared with their healthy contemporaries (Stuber et al, 1991). There was an increase in the number of BMT children with behaviour problems, suggesting that the treatment of BMT had had an impact on adjustment. This is in contrast to the findings for the cardiac - and in particular, cyanotic - group, for whom the lesion, rather than the surgery, seemed to be of greater significance.

7.9.5. Behaviour at School: 5 - 17 Years

The cyanotic and acyanotic groups did not differ in the prevalence of problem behaviour at school. For the cardiac group as a whole, pre-operative adjustment was a significant predictor of post-operative adjustment. Whilst this remained true for the acyanotic group, it was not the case for the cyanotic patients. This suggests that for the less severely impaired children, behaviour problems at school were stable over time and the intervention of cardiac surgery had little impact. Within this acyanotic group the less apparent nature of the symptoms and effects on physical appearance means that teachers are less likely to have been influenced in their perception of the child's behaviour by his/her condition. Following the surgery, the differences in the child's appearance are likely to have been negligible. Although in most cases a different teacher completed the questionnaire at the two assessments (due to a natural change of class), the impact of the child's condition is likely to have been similar on both occasions. In contrast, the surgery had a more dramatic impact on both symptoms and appearance for a number of the cyanotic children, resulting in a differing impact on the teachers. It is therefore difficult to assess whether the lack of association between pre- and post-operative scores in the cyanotic group is due to changes related to the surgery or whether it is reflecting the fact that two different teachers completed the measure.

As pre-operatively, there were no significant differences post-operatively in the prevalence of problem behaviour between the cardiac and healthy groups, which corroborates other findings of teacher-rated adjustment of children with surgically repaired lesions (Wright & Nolan, 1984). However, the prevalence of problem behaviour was lower in the healthy group (7%), at a level comparable to that for the normal population (Rutter et al, 1970). There was no significant association between pre- and post-operative Rutter B scores in the

healthy group, mainly due to the reduction in the number of children with behaviour problems at the second test occasion. The two most likely explanations for the change in prevalence of problem behaviour are that firstly different teachers completed the questionnaires on each occasion and secondly that there is some effect of an increase in age. The lack of a reduction in the number of cardiac children with behaviour problems, in the light of the healthy group results, therefore needs to be interpreted with caution. The initial assumption that there have been no adverse effects of surgery on adjustment at school - due to the lack of an increase in adjustment difficulties - is not necessarily correct if a reduction is expected for the healthy population. However, the cardiac group were younger, so if age is a factor in the prevalence of problem behaviour for the normal population then this also needs consideration. What is clear is that successful, corrective cardiac surgery has not led to a reduction in teacher-rated adjustment difficulties at school.

The prevalence of adjustment difficulties at school was similar in the cardiac and BMT groups. Both of the illness groups had a higher prevalence (non-significant) of adjustment difficulties than the healthy group, which is consistent with other findings for chronically ill children (Pless & Pinkerton, 1975; Eiser, 1985; Noll, Bukowski, Rogosch, LeRoy & Kulkarni, 1990; Wray & Yacoub, 1991). Teachers' additional comments on the questionnaires indicated that, for both illness groups, poor concentration and a lack of motivation were particular problems.

The lack of significant differences between the two illness groups and the healthy group is likely to be due to the relatively small sample sizes, in view of the demonstrated trends. Larger samples would also enable a more detailed analysis of the cyanotic and acyanotic

groups and more clarification of the potential impact of corrective surgery on adjustment at school.

7.9.6. Parental Interview

Following surgery, parents rated the cyanotic children as having significantly more temper tantrums and being significantly more anxious and less active than the acyanotic children. Within the cyanotic group a greater number had been rehospitalised since discharge and in some cases further surgery was indicated, which was a rather different picture to the acyanotic group, who had generally had an excellent medical outcome. The differing medical courses is likely not only to contribute to direct differences between the children themselves, but also to parents' perceptions of their children. Further analysis of the parental interview data indicated that the number of areas in which parents felt that their children had problems was higher in the cyanotic group and a smaller percentage of cyanotic children were felt to have no problems compared with the acyanotic group. This suggests that, from the parents' perspective, cyanotic lesions and subsequent surgery do have an effect, with a greater number of facets of behaviour being affected than for children with acyanotic lesions.

Whilst there were no significant differences over time in parental ratings of the cyanotic group, parents of the acyanotic children felt that they were significantly more disobedient after surgery. From the interviews it was apparent that this was not related so much to the surgery as to the improvement in their physical condition and the fact that the children wanted to be more active. Parents sometimes found this difficult to deal with and anxiety provoking and in some instances tried to be over-protective and restrictive, resulting in rebellion and more disobedient behaviour from the child. A further explanation is that these

children were behaving normally, but parents saw it as disobedience because they were not used to their child behaving in this manner. Overall, the cardiac group were rated as being more disobedient (accounted for by the acyanotic group) and more active after surgery. The increase in activity levels is expected given that in the majority of cases surgery had been corrective.

A significantly higher prevalence of behaviour problems was reported on the parental interview for the cyanotic children, but there was no association between the presence of behaviour problems on the Rutter A scales and parental interview for either the cyanotic or acyanotic groups. This again suggests that the Rutter A scale was not detecting areas of concern to parents.

Cardiac children were rated as having significantly more problems than healthy children in the areas of disobedience, anxiety, dependency and peer relationships, and this was largely attributable to the cyanotic group, who furthermore had more temper tantrums and difficulties with new situations than the healthy group. The only significant difference between the aycanotic and healthy group was that the acyanotic group were rated as being more disobedient, an explanation for which was given earlier. The lack of difference between the acyanotic and healthy group supports the view that the acyanotic group were essentially "normal" after surgery and also, encouragingly, that this was endorsed by parental perceptions. In contrast, the cyanotic patients were rated as having more problems in six of the eight areas of behaviour. The explanation for this probably centres on a combination of genuine differences in behaviour - due to the disease and parenting practices - and parental perceptions of the child. For those children who had had a good outcome from surgery,

increased activity and a wish for independence may well have been dealt with by over-anxious and restrictive responses, resulting in increased aggressive behaviours. In those children who had had a less than optimal outcome, passive, anxious and over-dependent behaviour continued or even increased. Hospitalisation and further treatment, together with further prolonged absence from school, are likely to have contributed to feelings of helplessness and social isolation. The greater difficulty in new situations is also likely to have been attributable to the younger age of the cyanotic group compared with the healthy group and an unwillingness on the part of the parents to encourage - or even let - their child into new situations. The higher prevalence of behaviour problems in the cardiac group compared with the healthy group is again largely attributable to the cyanotic children and is further evidence of the difficulties experienced by these children and families, even after successful surgery. The marked differences between the cyanotic and healthy groups and the similarities between the acyanotic and healthy groups post-operatively, in contrast to the pre-operative findings, suggest that the nature of the lesion, rather than the presence of the lesion, is of greater significance in determining adjustment following surgery.

In terms of changes over time, the healthy group showed a reduction in the prevalence of problems with new situations and peer relationships. Both of these are likely to be influenced by age, so this finding is expected. When the samples were split into two age bands the improvements were seen in the older, but not the younger, children. In contrast, such differences did not occur in the cardiac group. This is probably due to the younger age of the cardiac children, particularly as changes were not found for the younger children in the healthy group.

Overall, the cardiac group had significantly more problems in the areas of new situations, disobedience and relationships with adults compared with the BMT group, largely accounted for by the cyanotic patients. The cyanotic group were also more dependent and less active. Compared with the healthy group, the BMT group were more anxious and had more difficulties in peer relationships. Their greater anxiety is likely to be due to their illness, treatment and uncertain future. The disruption to schooling and normal social interactions, together with effects of the treatment on their physical appearance, are probable causes for their disrupted peer relationships. Older, but not younger, children in the BMT group had fewer difficulties in new situations and were less dependent after treatment compared with beforehand, and these changes are likely to be age related. The younger age of the cyanotic patients is also a contributory factor to many of the differences with the BMT group. The fact that many of these cyanotic children were still physically small is also likely to influence the way in which they were treated, and to contribute to parental over-protective behaviour. The BMT group generally showed more similarities with the acyanotic group, despite the more traumatic and intrusive nature of the BMT treatment. This suggests that parental behaviours and perceptions are particularly important for parents of cyanotic children and that factors such as the congenital nature of the lesion and early age at diagnosis continue to have an impact, even after corrective surgery.

7.9.7. Self-Perception

Post-operatively, there were few differences between the ways in which the cyanotic and acyanotic groups perceived themselves, although older acyanotic children rated themselves as more angry and liked themselves less than the cyanotic group. This may be a reaction to their surgery. The acyanotic group had been less incapacitated initially and in many cases had

been asymptomatic but they still had to undergo the trauma of major surgery, which had also left them with permanent physical disfigurement. For children who were leading relatively normal lives, the experience of hospitalisation and intrusive, disfiguring surgery might be expected to result in anger and a less favourable body image. In contrast, the cyanotic children were probably more tolerant of their need for surgery because they were more symptomatic pre-operatively and were also more used to dealing with the effects of congenital heart disease on their physical appearance. Following surgery, the cardiac group rated themselves and their ideal self as less ill. This was largely attributable to changes in the acyanotic group and suggests that, as the threats of impending hospitalisation and treatment have been removed, they can now allow themselves to express a more positive view of their ideal health status. The finding that the acyanotic group rated themselves as less frightened post-operatively, whilst expected because the treatment is behind them, also suggests that their anxiety about the surgery beforehand may have resulted in their using denial about their ideal health status. The use of denial as an adaptive coping mechanism has been previously reported for ill children (Koocher & O'Malley, 1981; Allen & Zigler, 1986; Engstrom, 1992.)

There were no significant differences between the cardiac and healthy groups on any measures of self-perception. Pre-operatively, the cardiac children had perceived themselves as weaker, more frightened and more ill than the healthy group. The lack of difference at follow-up is likely to be largely due to the positive changes in the acyanotic group and reinforces other results of this study that, following surgery, the acyanotic group is largely indistinguishable from the healthy group.

Following surgery, the cardiac children rated themselves as more frightened, angry and ill than the BMT children. As mentioned earlier, it was the acyanotic, rather than the cyanotic, children who were more angry and it would seem that the less severely ill children had reacted with a greater degree of anger to their treatment. The perception of the cardiac group that they were more ill than the BMT group may be due to the fact that some of the children still had to undergo further treatment and were still physically affected by their condition. Even after surgery, mothers of cardiac children have been found to underestimate their child's physical capabilities (Casey et al, 1994) and to be overprotective (Garson et al, 1974; Garson et al, 1978), which would also be expected to influence the child's self perception. The finding that both illness groups perceived themselves to be less ill after treatment is not only reassuring from the point of view of the children's psychological functioning, but also reinforces the use of such an instrument for the assessment of such children.

7.9.8. Parental Perception

There were no significant differences in parental perceptions of cyanotic and acyanotic children after surgery, supporting the view that actual disease severity is not the most important factor in determining how mothers perceive their children. In contrast to pre-operative perceptions, there was a higher degree of concordance between children's and parents' ratings, particularly on the constructs dealing with the child's psychological state. There was no agreement on the ill-well construct. This contradicts findings for children with juvenile rheumatoid arthritis, where children and parents showed agreement on ratings of the physical aspects of the disease, but differed in their reports of the psychological impact (Billings, Moos, Miller & Gottlieb, 1987). Overall, parents of cardiac children rated them

as less ill and less good post-operatively compared with before surgery. Parents of the cyanotic group did not perceive any changes in their children over time, but the acyanotic group were rated as being less ill at follow-up. The cyanotic group on whom data were available pre- and post-operatively was small, and although a number of these children had had successful cardiac surgery, for some, surgery had been palliative and further operations would still be necessary, therefore explaining why there was no change in parents' perceptions of health status. In contrast, the generally successful corrective surgery in the acyanotic group would be expected to result in parents rating their children as less ill. The finding that the children were perceived to be less good after surgery reinforces parents reporting increased disobedience (section 7.9.6.), discussed earlier.

There were no significant differences between the total cardiac and healthy groups on post-operative measures of parental perception. Pre-operatively, the two groups differed - appropriately - on the ill-well construct. The lack of difference on this construct post-operatively is expected in view of the change in parental perceptions of health status in the cardiac group. It is likely to be largely attributable to the more positive perceptions of parents of the acyanotic children. As expected, there were no changes over time on any measures of parental perception of the healthy group.

The lack of difference between the cardiac and BMT groups at follow-up appears to be mainly due to the more positive perception of parents of the BMT children after treatment compared with beforehand. The BMT children were rated as being angrier than the healthy children, which reinforces the finding that a higher proportion of BMT children respond with aggression in a stressful situation (see section 5.3.3.2.). Post-operative follow-up after BMT

involves repeated hospital visits, blood tests etc., so a greater expression of anger in BMT children compared with healthy children is not unexpected.

Overall, the results indicate that, in assessing the psychological impact of the intervention of cardiac surgery for children with congenital heart disease, it is essential to look at the group in terms of their initial diagnosis. In contrast to the findings of previous researchers, children in this study - particularly those with cyanotic lesions - did not show significant improvements in performance on psychological parameters after surgery. Before surgery there were few differences between those children with cyanotic and acyanotic lesions. Post-operatively, there were a number of significant differences between the two subgroups, with those with acyanotic lesions being indistinguishable on a number of measures from a group of healthy children. The results of the post-operative comparison with another group of chronically ill children undergoing intrusive treatment indicated that there was greater variation within the cardiac group than there was between the two illness (cardiac and BMT) groups.

CHAPTER 8

DATA ABOUT THE PARENTS

8.1. INTRODUCTION

The results obtained on the parents will be presented in turn on each area of functioning tested - psychological distress, marital relationship, perceived locus of control and coping. Pre-operative results, post-operative results and changes over time will be presented for each area. Results on the parents of the cardiac group will be given first and then comparisons made with the parents of the children in the two reference groups. The degree of satisfaction with medical and non-medical aspects of care and parental concerns for the future will be presented for parents of the cardiac and BMT children. Finally, the results will be briefly discussed in terms of the existing literature on parents of children with chronic illness and congenital heart disease in particular.

8.2. PSYCHOLOGICAL DISTRESS

8.2.1. Pre-operative General Health Questionnaire Results

8.2.1.1. Cardiac Group

Fifty-four of the 74 mothers (73%) and 48 of the 71 fathers (68%) completed the GHQ. Thirty-five mothers (65%) and 23 fathers (48%) obtained scores of 5 or more, indicating significant levels of psychological distress (Table 8.1). There were no significant differences in the prevalence of psychological distress between parents of cyanotic and acyanotic children.

PRE- AND POST-OPERATIVE LEVELS OF PSYCHOLOGICAL DISTRESS AND CHANGES OVER TIME

	CYANOTIC		ACYANOTIC		CARDIAC		BMT		HEALTHY		COMMENTS
	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	
PRE-OPERATIVE SCORE ≥ 5	(n=19) << 14 (74%)	(n=14) ++ 4 (29%)	(n=35) ^^ 21 (60%)	(n=34) >> 19 (56%)	(n=54) + 35 (63%)	(n=48) ^ 23 (48%)	(n=64) < 50 (78%)	(n=47) > ++ 31 (67%)	(n=66) + ^^ << 19 (29%)	(n=56) ^> >> 7 (13%)	+ X^2 = 14.15366 p = .0002 < X^2 = 29.80665 p = .0000 ^ X^2 = 14.11680 p = .0002 > X^2 = 29.11075 p = .0000 ++X^2 = 4.73105 p = .0296 <<X^2 = 10.70152 p = .0011 ^^X^2 = 8.05596 p = .0045 >>X^2 = 17.32704 p = .000
POST-OPERATIVE SCORE ≥5	(n=18) 7 (39%)	(n=13) 2 (15%)	(n=35) 6 (17%)	(n=33) 6 (18%)	(n=53) 13 (25%)	(n=46) 8 (17%)	(n=32) 7 (22%)	(n=21) 5 (24%)	(n=46) 12 (26%)	(n=38) 5 (13%)	
CHANGES OVER TIME: CHI-SQ = p = WILCOXON: Z = p =	(n=16) Fisher's Exact: .23077 -1.4676 .1422	(n=12) Fisher's Exact: .09091 -1.3416 .1797	(n=31) 2.39569 .1217 -3.2958 .0010	(n=30) .0000 1.0000 -2.6371 .0084	(n=47) 4.05321 .0441 -3.5279 .0004	(n=42) 1.06158 .3029 -2.8961 .0038	(n=32) .23510 .6278 -4.0145 .0001	(n=20) Fisher's Exact: .22136 -1.8904 .0587	(n=41) .0000 1.0000 - .4708 .6378	(n=34) 8.25575 .0041 - .5345 .5930	see explanations in sections 4.6.2. and 8.2.3.

Table 8.1

8.2.1.2. Comparison with the Reference Groups

Sixty-four of the 74 mothers (86%) and 47 of the 68 fathers (69%) in the BMT group and 66 of the 75 mothers (88%) and 56 of the 64 fathers (88%) in the healthy group completed the GHQ. Comparison of the cyanotic and acyanotic subgroups and reference groups indicated that fathers in the BMT group had significantly higher rates of psychological distress than fathers in the cyanotic group. Mothers in both the cyanotic and acyanotic subgroups had higher rates of distress than mothers in the healthy group and fathers in the acyanotic subgroup had higher rates of distress than fathers in the healthy group. Comparison of the total cardiac and reference groups indicated that there were no significant differences between the cardiac and BMT groups, but both mothers and fathers in the cardiac group had significantly higher rates of psychological distress than mothers or fathers in the healthy group (Table 8.1).

8.2.1.3. Parents Not Followed-Up

Six of the 54 mothers (11%) and 2 of the 48 fathers (4%) completing the GHQ pre-operatively were not followed-up. Five of the 6 mothers and both fathers were lost to follow-up. Comparison of the follow-up and non-followed up samples indicated that there were no significant differences in the rates of psychological distress for either mothers or fathers.

8.2.2. Post-operative General Health Questionnaire Results

8.2.2.1. Cardiac Group

Completed GHQ questionnaires were returned by 53 of the 54 mothers (98%) and 46 of the 53 fathers (87%). Thirteen mothers (25%) and 8 fathers (17%) obtained scores indicative

of significant levels of psychological distress (Table 8.1). There were no significant differences in the prevalence of psychological distress between parents in the cyanotic and acyanotic subgroups.

8.2.2.2. Comparison with the Reference Groups

Thirty-two of the 42 mothers (76%) and 21 of the 38 fathers (55%) in the BMT group and 46 of the 49 mothers (94%) and 38 of the 41 fathers (93%) in the healthy group completed the GHQ at follow-up. There were no significant differences in the rates of psychological distress between the cyanotic and acyanotic subgroups and the 2 reference groups or between the total cardiac, BMT and healthy groups (Table 8.1).

8.2.3. Changes Over Time

Changes over time were assessed in two ways (see page 172). Firstly, the association between pre- and post-operative psychological distress was assessed with the chi-squared test; secondly, a Wilcoxon test was performed to measure the direction of change between the pre- and post-operative ratings. Completed GHQ questionnaires were received on both test occasions on 47 mothers and 42 fathers in the cardiac group, 32 mothers and 20 fathers in the BMT group and 41 mothers and 34 fathers in the healthy group. There was a significant decrease over time in the prevalence of levels of psychological distress in mothers in the acyanotic subgroup and total cardiac and BMT groups and fathers in the acyanotic subgroup and total cardiac group, with the reduction in psychological distress levels almost reaching significance for fathers in the BMT group. There were no significant changes over time in the healthy group. For mothers in the total cardiac group there was also a significant association between the presence of significant levels of psychological distress pre-

operatively and 12 months later, but this did not apply to mothers in the BMT or healthy groups. For the fathers, there was no association between pre- and post-operative levels in either of the cardiac subgroups or in the total cardiac or BMT groups, but the association was significant for fathers in the healthy group (Table 8.1).

8.2.4. Relationship Between Post-Operative Maternal Distress Levels and Adjustment of the Patients

In the cardiac group there was a significant association between the presence of psychological distress in mothers post-operatively and the presence of a significant degree of problem behaviour in their children (chi-squared=6.000; p=.0143). The association was not significant in either of the 2 reference groups.

8.3. MARITAL RELATIONSHIP

8.3.1. Pre-operative Dyadic Adjustment Scale Results

8.3.1.1. Cardiac Group

Fifty-two of the 70 mothers (74%) and 47 of the 70 fathers (67%) eligible for the measure completed the marital relationship questionnaire. The mean total scores and scores on the item describing overall marital satisfaction (item 9) are given in Table 8.2. There were no significant differences between parents of the cyanotic and acyanotic children.

8.3.1.2. Comparison with the Reference Groups

Fifty-four of the 67 mothers (81%) and 48 of the 67 fathers (72%) in the BMT group and 57 of the 64 mothers (89%) and 56 of the 64 fathers (88%) in the healthy group who were eligible for the measure completed the marital relationship questionnaire. There were no

PRE- AND POST-OPERATIVE SCORES ON THE MARITAL RELATIONSHIP QUESTIONNAIRE AND CHANGES OVER TIME

	CYANOTIC		ACYANOTIC		CARDIAC		BMT		HEALTHY		COMMENTS
	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	
Pre-operative total score	(n=16)	(n=14)	(n=36)	(n=33)	(n=52)	(n=47)	(n=54)	(n=48)	(n=57)	(n=56)	
mean:	38.563	39.429	39.333	38.364	39.096	38.681	37.852	39.333	37.649	38.393	
S.D.:	7.694	5.155	6.676	5.556	6.937	5.451	7.272	6.285	6.342	5.179	
Pre-operative score on item 9	(n=16)	(n=14)	(n=35)	(n=33)	(n=51)	(n=47)	(n=53)	(n=45)	(n=56)	(n=56)	
mean:	3.625	3.786	3.486	3.485	3.529	3.575	3.359	3.667	3.732	3.857	
S.D.:	1.147	.893	1.173	1.176	1.155	1.098	1.242	1.398	1.198	1.052	
Post-operative total score	(n=15)	(n=13)	(n=34)	(n=33)	(n=49)	(n=46)	(n=26)	(n=19)	(n=38)	(n=37)	
mean:	41.333	41.385	39.971	39.727	40.388**	40.196	36.269**	37.263	38.290	38.162	** p<.05
S.D.:	3.309	4.538	5.351	5.456	4.825	5.218	7.661	6.332	5.594	5.585	
Post-operative score: item 9	(n=15)	(n=12)	(n=34)	(n=31)	(n=49)	(n=43)	(n=26)	(n=19)	(n=37)	(n=36)	
mean:	4.333	4.167	3.941	3.807	4.061**	3.907	3.269**	3.421	3.568	3.417	** p<.05
S.D.:	.900	1.030	1.205	1.078	1.126	1.065	1.218	.961	1.501	1.228	
Changes over time: total score	(n=14)	(n=12)	(n=31)	(n=29)	(n=45)	(n=41)	(n=25)	(n=19)a	(n=34)	(n=33)a	
t=	.98	2.21	.09	1.68	.57	2.56	- 1.76	- .90	- .22	- 1.51	a p=.049
p=	.347	.049	.932	.104	.575	.015	.091	.378	.825	.141	
Correlation between pre- r=	.6379	.7957	.7277	.7675	.7076	.7754	.7652	.4643	.6347	.8687	
and post-operative scores p=	.007	.001	.000	.000	.000	.000	.000	.023	.000	.000	
Changes over time: item 9	(n=14)	(n=11)	(n=30)	(n=28)c	(n=44)	(n=39)	(n=25)b	(n=17)	(n=32)b	(n=33)c	
t=	2.46	1.94	2.09	2.07	2.95	2.58	- 1.41	- .46	- 1.38	- 2.35	b p=.036
p=	.029	.082	.045	.048	.005	.014	.170	.651	.177	.025	c p=.005
Correlation between pre- r=	.6371	.9018	.5546	.6657	.5906	.7270	.3001	.5519	.5633	.6370	
and post-operative scores p=	.004	.000	.001	.000	.000	.000	.073	.011	.000	.000	

Table 8.2

significant differences between the cyanotic and acyanotic subgroups and the reference groups or between the total cardiac, BMT and healthy groups on the mean total scores or on the mean score for the marital satisfaction item (Table 8.2).

8.3.1.3. Parents Not Followed-Up

Four of the 52 mothers (8%) and 2 of the 47 fathers (4%) who completed the measure pre-operatively were not followed-up, all of whom were lost to follow-up. Whilst those fathers not followed up did not have significantly different scores from the follow-up sample, the mean total score for the mothers who were not followed up was significantly lower than that of the mothers who were followed-up (mean total score for follow-up sample: 39.833, S.D. 5.740; mean total score for mothers not followed up: 30.250; S.D. 13.75; t=2.83; p=.007).

8.3.2. Post-operative Dyadic Adjustment Scale Results

8.3.2.1. Cardiac Group

Forty-nine of the 53 mothers (92%) and 46 of the 53 fathers (87%) completed the marital relationship questionnaire 12 months after treatment. The mean total scores and mean score for item 9 are given in Table 8.2. There were no significant differences between parents in the 2 cardiac subgroups.

8.3.2.2. Comparison with the Reference Groups

Twenty-six of the 37 mothers (70%) and 19 of the 37 fathers (51%) in the BMT group and 38 of the 41 mothers (93%) and 37 of the 41 fathers (90%) in the healthy group eligible for the measure completed the questionnaire at the second test occasion. There were no significant differences between parents in either of the cardiac subgroups and the reference

groups. Mothers in the total cardiac group obtained a significantly higher mean total score and mean score on the item dealing with overall marital satisfaction than mothers in the BMT group, indicating a higher degree of marital satisfaction for mothers in the cardiac group. There were no significant differences between the fathers' scores in the total cardiac, BMT and healthy groups (Table 8.2).

8.3.3. Changes Over Time

Forty-five mothers and 41 fathers in the cardiac group, 25 mothers and 19 fathers in the BMT group and 34 mothers and 33 fathers in the healthy group completed the measure at both test occasions. There was a significant increase in the mean score of the item dealing with overall marital satisfaction for mothers in both cardiac subgroups and fathers in the acyanotic subgroup. Fathers in the cyanotic subgroup also had a significantly higher mean total score at the second test occasion. In the total cardiac group there was a significant increase in the mean score of the item dealing with overall marital satisfaction for both mothers and fathers and fathers also had a significantly higher mean total score at the second test occasion. There were no significant changes over time in the BMT group, but in the healthy group there was a significant decrease in fathers' mean item score for marital satisfaction (Table 8.2). There was a significant correlation between pre- and post-operative mean total scores for mothers and fathers in both cardiac subgroups and in the total cardiac, BMT and healthy groups, indicating the stability of these scores over time. There was a significant group x time interaction on fathers' mean total score ($p=.046$) (Fig. 10) and on fathers' (Fig. 11) and mothers' (Fig. 12) mean score on the overall marital satisfaction item ($p=.015$ for both mothers and fathers). Relative to the healthy group, there was a significant

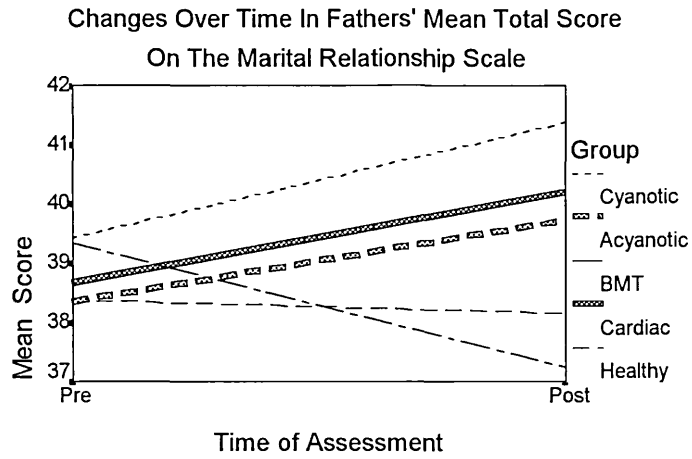


Fig. 10

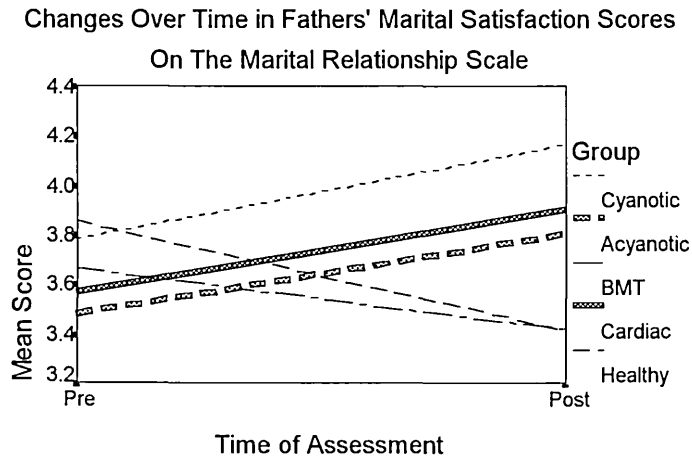


Fig. 11

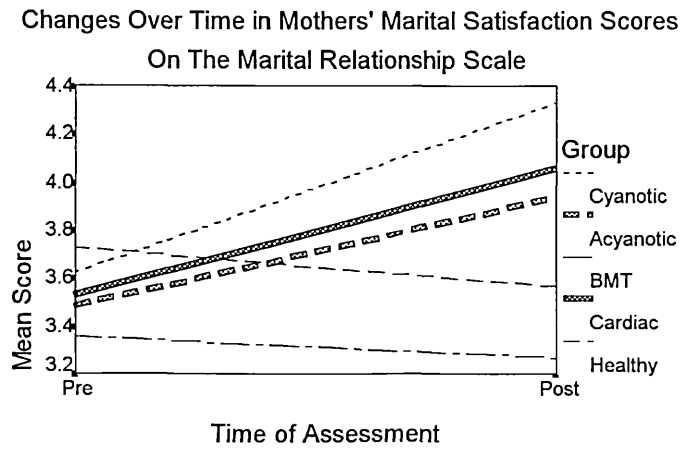


Fig. 12

change in fathers' mean total score and mothers' mean score on the overall marital satisfaction item in the BMT group. Relative to the healthy group there was also a significant change in fathers' mean score on the overall marital satisfaction item in the acyanotic group.

8.4. PERCEIVED LOCUS OF CONTROL

8.4.1. Pre-operative Locus of Control Scale Results

8.4.1.1. Cardiac Group

Fifty-four of the 74 mothers (73%) and 46 of the 71 fathers (65%) completed the locus of control questionnaire pre-operatively. Mean total scores are given in Table 8.3. There were no significant differences between parents of cyanotic and acyanotic children.

8.4.1.2. Comparison with the Reference Groups

Fifty-three of the 74 mothers (72%) and 42 of the 68 fathers (62%) in the BMT group and 64 of the 75 mothers (85%) and 56 of the 64 fathers (88%) in the healthy group completed the locus of control questionnaire. Mothers in the BMT group obtained significantly higher scores than mothers in the acyanotic subgroup, total cardiac group and healthy group, indicating that BMT mothers had a more external locus of control than mothers in the other groups (Table 8.3).

8.4.1.3. Parents Not Followed-Up

Six of the 54 mothers (11%) and 2 of the 46 fathers (4%) who were seen pre-operatively were not followed-up. Five of the 6 mothers and both fathers were lost to follow-up. There were no significant differences in the mean total scores between those parents who were followed up and those who were not.

PRE- AND POST-OPERATIVE SCORES ON THE LOCUS OF CONTROL QUESTIONNAIRE AND CHANGES OVER TIME

	CYANOTIC		ACYANOTIC		CARDIAC		BMT		HEALTHY		COMMENTS
	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	mothers	fathers	
Pre-operative total score	(n=18)	(n=13)	(n=36)	(n=33)	(n=54)	(n=46)	(n=53)	(n=42)	(n=64)	(n=56)	~ p<.001
			~				~		~		
mean:	12.944	13.539	11.361	12.273	11.889~	12.630	14.434~	13.405	12.125	12.179	
S.D.:	2.960	2.757	3.035	3.891	3.076	3.623	3.285	4.548	2.984	3.465	
Post-operative total score	(n=18)	(n=13)	(n=35)	(n=33)	(n=53)	(n=46)	(n=31)	(n=21)	(n=45)	(n=37)	* p<.01
			*				*				
mean:	12.556	12.923	11.143	11.667	11.623*	12.022	13.677*	12.381	12.778	10.703	
S.D.:	3.166	3.593	2.892	3.129	3.033	3.276	2.713	2.991	2.883	3.222	
Changes over time	(n=15)	(n=12)	(n=32)	(n=29)	(n=47)	(n=41)	(n=28)	(n=21)	(n=39)	(n=33)	
t=	.65	1.13	.18	.48	.50	.96	1.01	1.77	- 2.24	.33	
p=	.525	.283	.860	.638	.617	.342	.319	.091	.031	.741	
r=	.5602	.7825	.5089	.7227	.5455	.7237	.5864	.8387	.4231	.5916	
p=	.015	.001	.001	.000	.000	.000	.001	.000	.004	.000	

Table 8.3

8.4.2. Post-operative Locus of Control Scale Results

8.4.2.1. Cardiac Group

Fifty-three of the 54 mothers (98%) and 46 of the 53 fathers (87%) completed the locus of control questionnaire post-operatively. Mean total scores are given in Table 8.3. There were no significant differences between parents in the cyanotic and acyanotic subgroups.

8.4.2.2. Comparison with the Reference Groups

Thirty-one of the 42 mothers (74%) and 21 of the 38 fathers (55%) in the BMT group and 45 of the 49 mothers (92%) and 37 of the 41 fathers (90%) in the healthy group completed the measure at follow-up. Mothers in the BMT group had a significantly higher score than mothers in the acyanotic subgroup. The only significant difference between the total cardiac, BMT and healthy groups was that the mothers in the BMT group had significantly higher scores than the mothers in the cardiac group (Table 8.3).

8.4.3. Changes Over Time

Forty-seven mothers and 41 fathers in the cardiac group, 28 mothers and 21 fathers in the BMT group and 39 mothers and 33 fathers in the healthy group completed the locus of control questionnaire at both test occasions. There were no significant changes over time in either of the cardiac subgroups or in the total cardiac or BMT group, but mothers in the healthy group had a significantly higher score at follow-up compared with the first assessment. There was a significant correlation between pre- and post operative scores for mothers and fathers in both cardiac subgroups and in the total cardiac, BMT and healthy groups (Table 8.3).

8.5. COPING

8.5.1. Pre-operative Utrecht Coping List Results

8.5.1.1. Cardiac Group

Fifty-five of the 74 mothers (74%) and 48 of the 71 fathers (68%) completed the coping scale. There were no significant differences between the mothers of the cyanotic (n=19) and acyanotic (n=36) children or between the fathers in the cyanotic (n=14) and acyanotic (n=34) groups in the use of any particular coping strategy (Tables 8.4 and 8.5). Of the different coping mechanisms, mothers and fathers in both subgroups used comforting cognitions and actively solving most frequently. Mothers in the cyanotic group used expression of emotions and reacting with depression least frequently, whilst mothers in the acyanotic subgroup used avoidance and reacting with depression least frequently. Fathers in both subgroups used palliative and depressive mechanisms of coping least frequently.

8.5.1.2. Comparison with the Reference Groups

Forty-nine of the 74 mothers (66%) and 40 of the 68 fathers (59%) in the BMT group and 65 of the 75 mothers (87%) and 55 of the 64 fathers (86%) in the healthy group completed the coping scale. There were no significant differences between the total cardiac and reference groups or between the cyanotic and acyanotic subgroups and reference groups in the use of any particular coping strategy. Fathers in the BMT group used palliation and depression significantly more than fathers in the healthy group. Use of comforting cognitions was the coping strategy adopted most frequently by mothers in the total cardiac and BMT groups and reacting with depression was the least frequently used coping strategy used by all groups of mothers. Fathers in the total cardiac, BMT and

MOTHERS' PRE- AND POST-OPERATIVE SCORES ON THE COPING SUBSCALES AND CHANGES OVER TIME: MEANS AND STANDARD DEVIATIONS

COPING SUBSCALE	CYANOTIC (n=16)			ACYANOTIC (n=32)			TOTAL CARDIAC (n=48)			BMT (n=26)			HEALTHY (n=40)			COMMENTS
	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	
Actively Solving	2.414 .458	2.438 .371	.0234 .4088	2.536 .601	2.532 .580	-.0040 .4202	2.495 .555	2.500 .516	.0053 .4121	2.587 .543	2.418 .606	-.1683 .4582	2.480> .484	2.311> .410	-.1689 .3336	> t = 3.08 p = .004
Palliation	2.154 .402	2.202 .475	.0481 .2955	2.117 .440	2.096 .462	-.0208 .3995	2.128 .425	2.128 .462	.0000 .3690	2.250< .382	2.085< .301	-.1650^ .3657	2.072 .341	2.078 .330	.0063^ .3100	^ t = -2.1546 p = .034 < t = 2.26 p = .033
Avoidance	2.156 .330	2.125 .339	-.0347 .1609	2.023 .368	2.025 .404	-.0036 .3907	2.073 .358	2.059 .383	-.0142 .3290	2.178 .373	2.076 .331	-.1022 .4832	2.132 .446	2.041 .362	-.0906 .2921	
Social Support	2.411 .487	2.367 .451	-.0444 .3644	2.344 .549	2.350 .568	.0054 .5435	2.366 .525	2.355 .528	-.0109 .4886	2.308 .537	2.192 .577	-.1154 .6106	2.376 .515	2.350 .462	-.0256 .4284	
Depression	1.589 .383	1.670 .416	.0804 .2808	1.505 .352	1.478 .376	-.0268 .3045	1.533 .361	1.542 .396	.0089 .2982	1.566 .407	1.514 .332	-.0514 .4142	1.568 .470	1.511 .374	-.0571 .4140	
Expression of Emotions	2.021 .463	2.000 .455	-.0208 .4939	2.172 .682	2.247 .564	.0753 .6538	2.121 .616	2.163 .538	.0426 .6004	2.115 .489	2.141 .559	.0256 .6319	2.359> .435	2.171> .366	-.1880 .3884	> t = 3.02 p = .004
Comforting Cognitions	2.622 .517	2.711 .677	.0889 .4792	2.602 .712	2.441 .623	-.1613 .6317	2.609 .649	2.529 .646	-.0797 .5926	2.693 .673	2.487 .655	-.1154 .6250	2.433 .646	2.258 .531	-.1750 .7510	

Table 8.4

FATHERS' PRE- AND POST-OPERATIVE SCORES ON THE COPING SUBSCALES AND CHANGES OVER TIME: MEANS AND STANDARD DEVIATIONS

COPING SUBSCALE	CYANOTIC (n=12)			ACYANOTIC (n=30)			TOTAL CARDIAC (n=42)			BMT (n=18)			HEALTHY (n=34)			COMMENTS
	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	Pre	Post	Change	
Actively Solving	2.573 .496	2.563 .339	-.0104 .4246	2.591 .498	2.522 .498	-.0690 .3513	2.585 .491	2.534 .453	-.0518 .3697	2.792 .541	2.590 .388	-.2014 .4312	2.543 .510	2.438 .483	-.1055 .3465	
Palliation	1.896 .416	1.885 .289	-.0104 .3633	1.971 .371	2.000 .395	.0292 .3695	1.949 .381	1.967 .368	.0179 .3637	2.199 .464	2.184* .422	-.0147 .3152	1.852 .385	1.822* .430	-.0303 .2865	* p< .05
Avoidance	2.046 .309	2.000 .367	-.0463 .2657	2.011 .387	2.026 .307	.0148 .2986	2.021 .363	2.019 .321	-.0026 .2877	2.080 .446	2.043 .411	-.0370 .5562	1.893 .381	1.953 .388	.0609 .2965	
Social Support	2.139^ .382	1.861^ .460	-.2778 .4342	2.133 .468	2.083 .536	-.0500 .3666	2.135 .441	2.020 .520	-.1151 .3956	2.167 .607	2.147 .580	-.0196 .5800	2.121 .553	2.086 .468	-.0354 .4285	^ t= 2.22 p= .049
Depression	1.571 .468	1.512 .598	-.0595 .3074	1.571 .352	1.514 .361	-.0571 .2992	1.571 .383	1.514 .434	-.0578 .2978	1.810 .407	1.611 .469	-.1984 .5590	1.476 .444	1.455 .395	-.0216 .3116	
Expression of Emotions	2.083 .588	2.194 .437	.1111 .5918	2.244 .612	2.289 .469	.0444 .4931	2.198 .603	2.262 .457	-.0635 .5166	1.870 .617	2.037 .604	.1667 .4884	1.990 .502	2.000 .569	.0090 .5888	
Comforting Cognitions	2.528 .810	2.306 .822	-.2222 .6409	2.244 .600	2.256 .565	.0111 .5968	2.325 .669	2.270 .639	-.0556 .6112	2.537 .606	2.685* .718	.1481 .7773	2.192 .589	2.081* .589	-.1111 .5046	* p< .05

Table 8.5

healthy groups used actively solving most frequently and reacted with depression least frequently.

8.5.1.3. Parents Not Followed-Up

Six of the 55 mothers (11%) and 2 of the 48 fathers (4%) completing the coping scale pre-operatively were not followed-up. Five of the 6 mothers and both fathers were lost to follow-up. Comparison of the follow-up and non-followed up samples indicated that there were no significant differences in the use of any particular coping strategy for either mothers or fathers.

8.5.2. Post-operative Utrecht Coping List Results

8.5.2.1. Cardiac Group

Completed coping questionnaires were returned by 53 of the 54 mothers (98%) and 46 of the 53 fathers (87%). There were no significant differences in the use of any particular coping strategy between parents in the cyanotic and acyanotic subgroups (Tables 8.4 and 8.5). Use of comforting cognitions and actively solving were the most frequently used coping mechanisms by mothers and fathers in both the cyanotic and acyanotic subgroups and reacting with depression was the least frequently used strategy by all groups of parents.

8.5.2.2. Comparison with the Reference Groups

Thirty-two of the 42 mothers (76%) and 20 of the 38 fathers (53%) in the BMT group and 45 of the 49 mothers (92%) and 38 of the 41 fathers (93%) in the healthy group completed the coping scale at follow-up. The only significant difference between parents in any of the

groups was that fathers in the BMT group used palliation and comforting cognitions significantly more than fathers in the healthy group.

8.5.3. Changes Over Time

Forty-eight mothers and 42 fathers in the cardiac group, 26 mothers and 18 fathers in the BMT group and 40 mothers and 34 fathers in the healthy group completed the measure at both test occasions. There were no significant changes over time for mothers in the cyanotic and acyanotic subgroups or total cardiac group, but mothers in the BMT group used palliation significantly less after treatment than beforehand (Table 8.4). Mothers in the healthy group used actively solving and expression of emotions significantly less at follow-up compared with the first assessment. The only significant change for any group of fathers was in the cyanotic group, with a decrease in the use of social support at follow-up compared with before surgery (Table 8.5). Repeated measures analysis of variance indicated that there were no significant group x time interactions on any of the coping strategies for mothers or fathers apart from the result that, relative to the healthy group, there was a change in the use of palliation by mothers in the BMT group.

8.6. PERCEPTION OF DISEASE SEVERITY

8.6.1. Relationship Between Mothers' and Clinicians' Perception of Disease Severity

There was no significant relationship in the cardiac group between mothers' and clinicians' perception of disease severity (chi-squared = 4.1620; p=.1248).

8.6.2. Mothers' Distress Levels and Their Perception of Disease Severity

Within the cardiac group there was no significant difference in mothers' perception of disease severity between those mothers who obtained scores indicative of psychological distress and those who did not have scores indicative of psychological distress, irrespective of clinicians' ratings of disease severity (chi-squared = 8.9074; p=.0635).

8.7. DEGREE OF EXPRESSED SATISFACTION WITH MEDICAL AND NON-MEDICAL ASPECTS OF CARE, AND CONCERNS FOR THE FUTURE

At follow-up, parents of the cardiac and BMT patients were asked whether or not they were satisfied with various aspects of the service offered at the time of their child's treatment. They were also asked whether or not they had concerns about their child's health in terms of the future. The results are presented in Table 8.6.

PARENTAL SATISFACTION WITH MEDICAL AND NON-MEDICAL ASPECTS OF CARE AND CONCERNS FOR THE FUTURE

		CYANOTIC	ACYANOTIC	CARDIAC	BMT	COMMENTS
Communication:	Satisfied	10 (56%)	14 (40%)	24 (45%)***	29 (74%)***	***:X^2= 6.63274
	Dissatisfied	8 (44%)	21 (60%)	29 (55%)	10 (26%)	p = .0100
Accommodation:	Satisfied	9 (50%)	20 (57%)	29 (55%)	26 (67%)	
	Dissatisfied	9 (50%)	15 (43%)	24 (45%)	13 (33%)	
Facilities:	Satisfied	11 (61%)	19 (54%)	30 (57%)	23 (59%)	
	Dissatisfied	7 (39%)	16 (46%)	23 (43%)	16 (41%)	
Support:	Satisfied	8 (44%)	23 (66%)	31 (58%)***	36 (92%)***	***:X^2=11.33076
	Dissatisfied	10 (56%)	12 (34%)	22 (42%)	3 (8%)	p = .0008
Medical Management:	Satisfied	13 (72%)	25 (71%)	38 (72%)	33 (85%)	
	Dissatisfied	5 (28%)	10 (29%)	15 (28%)	6 (15%)	
Future Concerns:	Yes	15 (83%)***	16 (46%)***	31 (58%)	25 (64%)	***:X^2= 5.46582
	No	3 (17%)	19 (54%)	22 (42%)	14 (36%)	p = .0194

Table 8.6

A significantly higher proportion of parents of cyanotic children expressed concerns about the future compared with parents of acyanotic children. Parents of cardiac children reported a higher degree of dissatisfaction with communication with staff and support received than parents of BMT children.

8.8. DISCUSSION OF THE RESULTS

8.8.1. Levels of Psychological Distress

The lack of difference in the prevalence of pre-operative psychological distress between parents in the cyanotic and acyanotic subgroups supports previous findings for mothers of cardiac children that the severity of their child's heart condition has little bearing on anxiety levels (Linde et al, 1966; Kitchen, 1978). Comparison of the cardiac, BMT and healthy groups yielded no significant differences between parents in the cardiac and BMT groups but, as expected, significantly higher rates of psychological distress in parents of the ill children compared with parents of the healthy children. These results are in agreement with findings of mental health difficulties for mothers of other groups of chronically ill children (Wallander et al, 1989; Hughes & Lieberman, 1990; Thompson, Gustafson, Hamlett & Spock, 1992; Thompson, Gil, Burbach, Keith & Kinney, 1993; Bradford, 1994). The reported rate of maternal distress in the healthy group (29%) is similar to that reported for random community samples (Goldberg, 1978).

At follow-up there was a reduction in the prevalence of psychological distress for parents in all of the illness groups, although this was not significant for mothers or fathers in the cyanotic group. As expected, there were no significant changes over time in the healthy group. Following cardiac surgery parental anxiety levels have been found to reduce

(Landtman et al, 1960; Linde et al, 1970). In our study the reduction in psychological distress was less marked in parents of cyanotic children than in parents of acyanotic children, which is likely to be partly attributable to the ongoing medical problems experienced by some of the cyanotic children.

For mothers in the cardiac group overall, pre-operative GHQ scores were predictive of post-operative GHQ scores, suggesting that factors other than the child's medical condition and treatment are important in determining psychological distress. Other studies have found that illness variables account for a relatively small percentage of the variation in maternal psychological distress (Thompson et al, 1992) and a number of non-disease related factors which influence maternal adjustment have been identified (Varni & Wallander, 1988; Wallander et al, 1989), such as maternal coping and temperament, as well as factors such as perception of disease severity. Research on parental adjustment to congenital heart disease and subsequent cardiac surgery has not focused on the role of parental variables but clearly this is an area for future investigation.

8.8.2. Marital Relationship

Research on the effects of congenital heart disease on parents' marital relationship has tended to focus on the prevalence of divorce in such families (Finley et al, 1979; Silbert et al, 1982). Divorce rates were not found to be higher than for the normal population and were not related to the severity of the child's cardiac lesion (Finley et al, 1979). Within our study, 93% of the cardiac children came from two-parent homes, compared with 85% of the healthy children, which supports previous findings. The lack of difference between parents of

cyanotic and acyanotic children supports the view that disease severity does not have a bearing on parental reporting about their relationship.

Pre-operative comparison of the cardiac, BMT and healthy groups yielded no differences in scores on the marital questionnaire, in contrast to other findings of significantly higher marital distress in parents of ill children compared with those of healthy children (Silbert et al, 1982; Barbarin et al, 1985; Dahlquist et al, 1993). Other research has shown that coping with disease can have a positive effect on parents' relationships (Pless & Satterwhite, 1975(a); Vance et al, 1980) but in view of the lack of data collected before the child became ill, such conclusions cannot be applied to this study.

Following treatment, there were no significant differences between parents in any of the illness groups and parents in the healthy group, but mothers in the cardiac group had significantly higher scores on both the total scale and marital satisfaction item than mothers in the BMT group. In the cardiac group both mothers and fathers reported a significant increase in degree of marital happiness after treatment, suggesting that the intervention of largely corrective surgery had had some positive effects. However, the finding for all groups that pre-operative scores were significantly correlated with post-operative scores also indicates the stability of marital relationships over time, irrespective of the child's health status or treatment interventions.

8.8.3. Locus of Control

The focus of previous work on parents' locus of control has been on the relationship between parental locus of control, in terms of parenting beliefs, and adjustment of the child (Campis

et al, 1986; DeMaso et al, 1991). Work with both adult and paediatric patients has suggested better adjustment in those with an internal, rather than an external, locus of control (Alogna, 1980; Moffatt & Pless, 1983) and there is also evidence for chronic illness in children being associated with a more external locus of control in the children (Kellerman et al, 1980; Perrin & Shapiro, 1985; Greenberg, Kazak & Meadows, 1989). However, there is little information about parents' own locus of control beliefs. In the current study there were no significant differences in locus of control scores between parents of cyanotic and acyanotic children, again suggesting that disease severity is not important in determining locus of control beliefs. However, mothers of BMT patients had a more external locus of control than mothers in the cardiac group both before and after treatment. The significant correlation between locus of control scores at the two test occasions for mothers and fathers in all groups indicates the stability of locus of control beliefs over time. Interestingly, mothers in the healthy group had a more external locus of control at follow-up compared with the first assessment, whereas there were no significant changes over time in the illness groups. Perhaps the strategies adopted by parents of ill children in dealing with the illness and treatment enable them to deal with life stresses without events altering their perceptions and beliefs, whereas mothers of healthy children are more vulnerable to the influence and impact of everyday life events.

8.8.4. Coping

There is little information about coping strategies utilised by parents of cardiac children, but in studies of other groups of chronically ill children parental coping strategies have been found to influence adjustment in the children (Sanger et al, 1991; Sloper, Larcombe & Charlton, 1994) and to vary according to the specific disease involved (Eiser & Havermans, 1992). Coping mechanisms have also been linked to parental adjustment, with poor

adjustment being associated with more use of palliative coping methods (Thompson et al, 1992; Thompson, Gil, Gustafson, George, Keith, Spock & Kinney, 1994).

Although the coping questionnaire used in this study was a general questionnaire about coping, it was felt that the child's illness was the most salient factor in parents' lives and that the coping strategies adopted by parents would apply to their coping with the illness too. As with other measures of parental functioning, the coping results of the current study indicated no significant differences either within the cardiac group or between the cardiac and BMT group in the use of any particular coping strategy either before or after treatment. Before treatment, parents in the cardiac, BMT and healthy groups utilised the same coping mechanisms with greatest and least frequency, suggesting that neither the presence nor nature of the chronic illness was influential in determining the type and frequency of strategies used. In general, there were few changes in any of the groups over time, suggesting that coping strategies are idiosyncratic to individuals. The only significant change over time in the cardiac group was that fathers of the cyanotic patients used social support less at follow-up compared with beforehand. One reason for this may be the change in situation, with social support being more readily available prior to surgery. Fathers may also have felt that it was more acceptable to look for support from others when surgery was pending, rather than at one year after treatment when the surgery was behind them. The only change in the BMT group was the reduction in the use of palliative coping mechanisms by the mothers. Palliative strategies involve seeking temporary distraction and reducing arousal levels. At the time of the first assessment a number of the BMT patients had been in hospital for a while and mothers of these children in particular looked for distraction in activities such as knitting and

embroidery. In contrast, at the follow-up mothers were more likely to be active in domestic chores and organising family life and were therefore less likely to actively seek distraction.

In summary, there were few differences in measures of parental functioning between the different illness groups. Furthermore, there were few changes over time, suggesting that the treatment interventions were of less significance than either factors attributable to the presence of chronic illness or individual parental characteristics. The relationship between parental functioning and functioning of the child, and the role of specific factors in determining how parents and children cope, are areas requiring further investigation.

CHAPTER 9

DATA ABOUT THE SIBLINGS

The results obtained on the siblings will be presented in turn of each area of functioning tested - behaviour at home, behaviour at school, parental interview and occurrence of school problems. Adjustment after treatment, rated by parents, will also be presented for the 2 illness groups. Pre-operative results, post-operative results and changes over time will be presented on each area of functioning. In each area results of the cardiac siblings will be presented first and then comparisons will be made with the siblings in the 2 reference groups. Parental ratings of behaviour problems at home in the siblings will also be compared with their ratings of behaviour problems in the patients. Finally, the results will be briefly discussed in terms of the existing literature on siblings of children with chronic illness and congenital heart disease in particular.

FUNCTIONING IN THE SIBLINGS

A description of pre-operative age distribution, sex and birth order of siblings in each of the 3 groups is given in Table 5.7, page 191. Sixty cardiac siblings, 70 BMT siblings and 68 siblings in the healthy group were followed up post-operatively.

9.1. UNDER 3'S

No validated measures of behaviour were used for siblings under 3 years of age (pre-operatively, n=12, n=27 and n=20 for the cardiac, BMT and healthy groups respectively). At one year after treatment the number of siblings under 3 years of age was 4, 6 and 10 in the cardiac, BMT and healthy groups respectively.

9.2. BEHAVIOUR AT HOME: 3 - 5 YEARS

9.2.1. Pre-operative Richman BCL Results

9.2.1.1. Cardiac Group

Twelve siblings were in the age range of 3 - 5 years. Completed Richman BCL questionnaires were received on 6 (50%), none of whom obtained a score indicative of behaviour problems at home.

9.2.1.2. Comparison with the Reference Groups

Twelve BMT siblings and 15 siblings in the healthy group were eligible for completion of the Richman BCL and questionnaires were received on 4 children in the BMT group and 11 in the healthy group. There were no significant differences in the prevalence of problem behaviour between the 3 groups (0%, 25% and 9% in the cardiac, BMT and healthy groups respectively).

9.2.1.3. Children not Followed-up

Of the 12 cardiac siblings assessed pre-operatively, 6 were not followed up because the patient died during or soon after surgery (n=3) or the family were lost to follow-up. Completed questionnaires were received on 2 of the 3 siblings lost to follow-up but on none of those where the patient died.

9.2.2. Post-operative Richman BCL Results

9.2.2.1. Cardiac Group

Five siblings in the cardiac group were eligible for completion of the Richman BCL and questionnaires were received back on 4 (80%), none of whom obtained a score above the cut-off point.

9.2.2.2. Comparison with the Reference Groups

Completed Richman BCL's were received on 8 (67%) of the 12 eligible BMT siblings and on 7 (88%) of the 8 eligible siblings in the healthy group. There were no significant differences in the prevalence of problem behaviour between the 3 groups (0%, 13% and 25% for the cardiac, BMT and healthy groups respectively).

9.2.3. Changes Over Time

Completed Richman BCL's were received at both test occasions on 2 cardiac siblings, 1 BMT sibling and 4 siblings in the healthy group. None of these children had scores above the cut-off point at either test occasion.

9.3. BEHAVIOUR AT HOME: 5 - 17 YEARS

9.3.1. Pre-operative Rutter A Results

9.3.1.1. Cardiac Group

Sixty-two cardiac siblings were between 5 and 18 years of age. Completed Rutter A questionnaires were received on 45 (73%). Eight (18%) obtained a score of 13 or more, indicative of a significant degree of problem behaviour at home. Three (37.5%) of the children had neurotic problems, 4 (50%) had an antisocial pattern of behaviour and 1 sibling

had behaviour problems of an undifferentiated nature (Table 9.1). Fifteen children had siblings with cyanotic lesions and 30 had siblings with acyanotic lesions. There were no differences in the prevalence of sibling behaviour problems in the 2 subgroups.

9.3.1.2. Comparison with the Reference Groups

Completed Rutter A questionnaires were received on 56 of 69 BMT siblings (81%) and 58 of 70 siblings in the healthy group (84%). There were no significant differences in the prevalence of problem behaviours at home in the three groups (18%, 14% and 16% for the cardiac, BMT and healthy groups respectively). There was a lower prevalence of antisocial type behaviour problems in the reference groups compared with the cardiac group, but the differences were not significant (Table 9.1).

9.3.1.3. Comparison of Sibling and Patient Rutter A Results

Total scores and the prevalence of behaviour problems were compared in the patients and their siblings. In the acyanotic subgroup, total cardiac, BMT and healthy groups there was a significant correlation between sibling and patient total scores (Table 9.2). There was no significant association between the presence or absence of behaviour problems in the patient and siblings in the cyanotic and acyanotic subgroups or in the total cardiac or BMT group, but the association was significant in the healthy group.

SIBLING RUTTER A RESULTS					
	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVE: SCORE ≥ 13	(n=15) 2 (13%)	(n=30) 6 (20%)	(n=45) 8 (18%)	(n=56) 8 (14%)	(n=58) 9 (16%)
COMMENTS	neurotic = 1 antisocial = 1 undiff. = 0	neurotic = 2 antisocial = 3 undiff. = 1	neurotic = 3 antisocial = 4 undiff. = 1	neurotic = 4 antisocial = 2 undiff. = 2	neurotic = 5 antisocial = 2 undiff. = 2
POST-OPERATIVE: SCORE ≥13	(n=16) 3 (19%)	(n=34) 3 (9%)	(n=50) 6 (12%)	(n=42) 5 (12%)	(n=48) 9 (19%)
COMMENTS	neurotic = 1 antisocial = 1 undiff. = 1	neurotic = 2 antisocial = 0 undiff. = 1	neurotic = 3 antisocial = 1 undiff. = 2	neurotic = 2 antisocial = 2 undiff. = 1	neurotic = 4 antisocial = 4 undiff. = 1
CHANGE OVER TIME IN SCORE	(n=15)	(n=26)	(n=41)	(n=36)	(n=35)
Z =	0.000	.9129	.7388	1.6036	0.00
p =	1.000	.3613	.4631	.1088	1.00
chi-sq =	Fisher Exact:	.15388	4.43771	13.30603	6.63762
p =	.25714	.6949	.0352	.0003	.0100

Table 9.1

PARENTAL ASSESSMENT OF THE PREVALENCE OF BEHAVIOUR PROBLEMS AT HOME: COMPARISONS BETWEEN PATIENTS AND SIBLINGS					
	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVE RUTTER A TOTAL SCORES r = p =	(n=10) .0949 .397	(n=18) .6075 .004	(n=28) .5615 .002	(n=41) .4513 .002	(n=49) .4432 .001
PRE-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS ON THE RUTTER A chi-sq = p =	(n=10) Fisher Exact .50000	(n=18) Fisher Exact: .40523	(n=28) 1.51706 .2181	(n=41) 1.35068 .2452	(n=49) 5.77786 .0162
PRE-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS ON THE INTERVIEW chi-sq = p =	(n=14) Fisher Exact: .60440	(n=37) 2.05599 .1516	(n=51) 1.20811 .2717	(n=63) .01250 .9110	(n=74) 1.59715 .2063
POST-OPERATIVE RUTTER A TOTAL SCORES r = p =	(n=11) .4891 .063	(n=26) .1442 .241	(n=37) .2061 .110	(n=32) .5671 .000	(n=40) .2598 .053
POST-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS ON THE RUTTER A chi-sq = p =	(n=11) Fisher Exact .26190	(n=26) .0000 1.0000	(n=37) .39572 .5293	(n=32) 2.43256 .1188	(n=40) .13775 .7105
POST-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS ON THE INTERVIEW chi-sq = p =	(n=17) Fisher Exact: .01471	(n=42) .08514 .7705	(n=59) 1.75624 .1851	(n=60) 7.58113 .0059	(n=69) .0000 1.0000

Table 9.2

9.3.1.4. Children not Followed-up

Of the 62 cardiac siblings seen pre-operatively, 15 (24%) were not followed up. Seven were lost to follow-up and in the other 8 instances the patient died. Questionnaires were not received on any of the siblings in families where the patient died but were received on 2 of the 7 siblings in families lost to follow-up, one of whom had behaviour problems at home of an antisocial nature.

9.3.2. Post-operative Rutter A Results

9.3.2.1. Cardiac Group

Completed Rutter A questionnaires were received on 50 of the 51 eligible children, 6 (12%) of whom obtained a score indicative of a significant degree of problem behaviour at home. Classification of the nature of the behaviour problems indicated that 3 (50%) of the children had behaviour problems of a neurotic nature, 1 (17%) had an antisocial pattern of behaviour and 2 (33%) siblings had behaviour problems of an undifferentiated nature (Table 9.1). Sixteen of the children had siblings with cyanotic lesions and 34 had siblings with acyanotic lesions. There were no differences in the prevalence of sibling behaviour problems in the 2 subgroups.

9.3.2.2. Comparison with the Reference Groups

Fifty-two BMT siblings and 50 siblings in the healthy group were eligible for completion of the Rutter A and questionnaires were received back on 42 (81%) BMT siblings and 48 (96%) siblings in the healthy group. Five (12%) BMT siblings and 9 (19%) healthy group siblings obtained scores above the cut-off point, but there was no significant difference in the prevalence of problem behaviour at home between the three groups. There was a higher

prevalence of antisocial type behaviour problems in the reference groups compared with the cardiac group, but the differences were not significant (Table 9.1).

9.3.2.3. Comparison of Sibling and Patient Post-operative Rutter A Results

The correlation of patient and sibling total scores was not significant in either the cyanotic or acyanotic subgroups or in the total cardiac group, but it was significant in the BMT group and almost reached significance in the healthy group (Table 9.2). There was no significant association in the cyanotic and acyanotic subgroups or in the total cardiac, BMT and healthy groups between the presence or absence of behaviour problems in the patient and their sibling(s).

9.3.3. Changes Over Time

There were no significant changes over time on the Rutter A scale in any of the groups (Table 9.1). There was a significant association between the presence or absence of behaviour problems before treatment and 12 months later in each of the 3 groups, indicating the stability of behaviour problems over time, but this association was not significant in the cyanotic and acyanotic subgroups.

9.4. BEHAVIOUR AT SCHOOL

9.4.1. Pre-operative Rutter B Results

9.4.1.1. Cardiac Group

School attendance and type of education are given in Table 5.8, page 192. Fifty-nine cardiac siblings were eligible for completion of a Rutter B scale and questionnaires were sent to the teachers of 48, all of which were returned. Questionnaires were not sent in 3 instances where

families refused permission and in 8 cases the patient died during surgery so the teachers were not approached. Of the 48 children on whom data were available, 8 (17%) obtained a score of 9 or more, indicative of a significant degree of problem behaviour at school. Five (62.5%) had a neurotic pattern of behaviour and 3 (37.5%) an antisocial behaviour pattern (Table 9.3). Sixteen of the children had siblings with cyanotic lesions and 32 had siblings with acyanotic lesions. There were no differences in the prevalence of sibling behaviour problems in the 2 subgroups.

9.4.1.2. Comparison with the Reference Groups

Rutter B questionnaires were sent to 65 of the 66 teachers of the eligible BMT siblings (in the remaining case parents refused permission for the teachers to be approached) and to the teachers of all 68 eligible siblings in the healthy group. Completed forms were returned on 59 (91%) BMT siblings and 67 (99%) siblings in the healthy group. Comparison of the three groups indicated no significant differences in the prevalence of problem behaviour at school. The siblings in the healthy group had a similar distribution of behavioural type to the cardiac siblings but in the BMT group no child had an antisocial pattern of behaviour (Table 9.3).

9.4.1.3. Children not Followed-up

Of the 48 cardiac siblings assessed pre-operatively, 6 were not seen post-operatively, all of whom were lost to follow up. Two of these children had a significant degree of problem behaviour at school and in both cases this was of a neurotic nature.

SIBLING RUTTER B RESULTS					
	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVE: SCORE ≥ 9	(n=16) 2 (13%)	(n=32) 6 (19%)	(n=48) 8 (17%)	(n=59) 5 (8%)	(n=67) 12 (18%)
COMMENTS	neurotic = 1 antisocial = 1 undiff = 0	neurotic = 4 antisocial = 2 undiff = 0	neurotic = 5 antisocial = 3 undiff. = 0	neurotic = 4 antisocial = 0 undiff. = 1	neurotic = 6 antisocial = 5 undiff. = 1
POST-OPERATIVE: SCORE ≥ 9	(n=14) 3 (21%)	(n=29) 3 (10%)	(n=43) 6 (14%)	(n=43) 5 (12%)	(n=43) 7 (16%)
COMMENTS	neurotic = 2 antisocial = 1 undiff = 0	neurotic = 2 antisocial = 1 undiff = 0	neurotic = 4 antisocial = 2 undiff. = 0	neurotic = 4 antisocial = 1 undiff. = 0	neurotic = 2 antisocial = 4 undiff. = 1
CHANGE OVER TIME IN SCORE	(n=14)	(n=24)	(n=38)	(n=38)	(n=37)
Z =	.5345	1.3416	.4045	.4045	.7338
p =	.5930	.1797	.6858	.6858	.4631
chi-sq =	Fisher Exact:	5.34545	5.06787	2.31827	3.64081
p =	.39560	.0208	.0244	.1279	.0564

Table 9.3

9.4.2. Post-operative Rutter B Results

9.4.2.1. Cardiac Group

Fifty-six siblings were 3 years or older and eligible for education. Two teenagers had left school but the remaining 54 were all receiving some form of education, one of whom was at a special school. Of the 49 children eligible for completion of a Rutter B scale, questionnaires were sent to the teachers of 46 and were received back on 43 (94%). Six of the 43 (14%) obtained a score of 9 or more, indicative of a significant degree of problem behaviour at school. Four (67%) had a neurotic pattern of behaviour and 2 (33%) had an antisocial pattern of behaviour (Table 9.3). Fourteen of the children had siblings with cyanotic lesions and 29 with acyanotic lesions. There were no differences in the prevalence of sibling behaviour problems in the 2 groups.

9.4.2.2. Comparison with the Reference Groups

Sixty-four BMT siblings and 58 siblings of healthy children were eligible for education. Five BMT siblings between the ages of 3 and 5 years were not attending nursery/playgroup and a further 4 had left school. In the healthy group 4 teenagers had left school. With the exception of 2 siblings in the healthy group at special school, all of the remaining siblings in both reference groups were at normal school. Rutter B questionnaires were sent to the teachers of all eligible siblings - 48 in the BMT group and 46 in the healthy group - and were received back on 43 in each group. Comparison of the cardiac and reference groups indicated no significant differences in the prevalence of problem behaviour at school. In the healthy group there was a higher percentage of children with antisocial behaviour problems and fewer children with a neurotic pattern of behaviour compared with the cardiac and BMT groups (Table 9.3).

9.4.3. Changes Over Time

There were no significant changes over time on the Rutter B scale in any of the 3 groups (Table 9.3). In the acyanotic subgroup and total cardiac group there was a significant association between the presence or absence of behaviour problems pre- and post-operatively, but there was no significant association in the cyanotic subgroup or BMT group. In the healthy group the association between the presence or absence of behaviour problems at each test occasion almost reached significance (Table 9.3).

9.5. PARENTAL INTERVIEW

For 10 cardiac siblings the interview data were not collected preoperatively because the patient died during surgery, before an interview with the parents had taken place.

9.5.1. Pre-operative Parental Interview Results

9.5.1.1. Cardiac Group

Parents of 16 (21%) of the 76 siblings on whom data were collected felt that their child had behaviour problems at home. Twenty-onr of the children had siblings with cyanotic lesions and 55 with acyanotic lesions. There were no significant differences in the prevalence of sibling behaviour problems at home between the 2 subgroups. For those children on whom Rutter A data were available, there was a significant association between the presence or absence of problem behaviour on the Rutter A and the presence or absence of behaviour problems at home according to the interview for the acyanotic subgroup and the total cardiac group, but not for the cyanotic subgroup. (Table 9.4).

9.5.1.2. Comparison with the Reference Groups

Parents of 44 (42%) of the 106 BMT siblings and 14 (13%) of the 104 healthy group siblings on whom data were collected felt that their child had behaviour problems at home. There was a significant association between Rutter A scores and the presence of behaviour problems at home in the healthy group but not in the BMT group (Table 9.4). The prevalence of behaviour problems at home from the interview data was significantly higher in the BMT group than in the cardiac group (chi-squared = 8.08440; p=.0045) and the healthy group (chi-squared = 20.30220; p=.0000) but there were no significant differences between the cardiac and healthy groups.

9.5.1.3. Comparison of Pre-operative Parental Interview Sibling and Patient Results

There was no association in any of the groups between the presence or absence of behaviour problems on the parental interview for the patient and their sibling(s) (Table 9.2).

9.5.1.4. Children not Followed-up

Seventeen of the 76 cardiac siblings on whom parental interview data were collected pre-operatively were not seen post-operatively, 15 of whom were lost to follow-up. Parents of 3 of these siblings felt that their child had behaviour problems at home.

9.5.2. Post-operative Parental Interview Results

9.5.2.1. Cardiac Group

Parents of 13 (22%) of the 59 siblings on whom data were collected felt that their child had behaviour problems at home. Seventeen of the children had siblings with cyanotic lesions and

PREVALENCE OF SIBLING BEHAVIOUR PROBLEMS ACCORDING TO THE PARENTAL INTERVIEW AND ASSOCIATION BETWEEN THE PRESENCE OR ABSENCE OF BEHAVIOUR PROBLEMS ON THE RUTTER A SCALES AND ON THE PARENTAL INTERVIEW

	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS	(n=21) 5 (23%)	(n=55) 11 (20%)	(n=76) 16 (21%)	(n=106) 44 (42%)	(n=104) 14 (13%)
ASSOCIATION BETWEEN PRE-OPERATIVE INTERVIEW AND RUTTER A SCORES	(n=15) Fisher Exact: p = .09524	(n=30) X^2 = 13.58135 p = .0002	(n=45) X^2 = 17.812 p = .0000	(n=57) X^2 = 2.34148 p = .1260	(n=48) X^2 = 18.82353 p = .0000
POST-OPERATIVE PREVALENCE OF BEHAVIOUR PROBLEMS	(n=17) 7 (41%)	(n=42) 6 (14%)	(n=59) 13 (22%)	(n=61) 8 (13%)	(n=68) 15 (22%)
ASSOCIATION BETWEEN POST-OPERATIVE INTERVIEW AND RUTTER A SCORES	(n=16) Fisher Exact: p = .06250	(n=33) X^2 = .0000 p = 1.0000	(n=49) X^2 = 1.09088 p = .2963	(n=42) X^2 = .0000 p = 1.0000	(n=49) X^2 = 19.93011 p = .0000
CHANGES OVER TIME IN THE PREVALENCE OF BEHAVIOUR PROBLEMS	(n=17) Z = - .9129 p = .3613 Fisher Exact: p = .05995	(n=42) Z = - .5606 p = .5751 X^2 = .16085 p = .6884	(n=59) Z = 0.00 p = 1.00 X^2 = 3.98943 p = .0458	(n=60) Z = 2.7373 p = .0062 X^2 = .11457 p = .7350	(n=69) Z = - .8022 p = .4236 X^2 = 14.18586 p = .002

Table 9.4

42 had siblings with acyanotic lesions. The difference between the 2 subgroups in the prevalence of sibling behaviour problems almost reached significance (chi-squared = 3.64888; p=.0561). For those children on whom Rutter A data were available, there was no significant association between the presence of behaviour problems according to the interview and on the Rutter A in the cyanotic or acyanotic subgroups or total cardiac group (Table 9.4).

9.5.2.2. Comparison with the Reference Groups

Parents of 8 (13%) of the 61 BMT siblings and 15 (22%) of the 68 healthy group siblings on whom data were collected felt that their child had behaviour problems at home. There was no significant association between Rutter A scores and the presence of behaviour problems at home from interview data in the BMT group but the association was significant in the healthy group (Table 9.4). The prevalence of behaviour problems at home from interview data was not significantly different in the three groups.

9.5.2.3. Comparison of Post-operative Parental Interview Sibling and Patient Results

There was no significant association in the acyanotic subgroup or total cardiac or healthy groups between the presence or absence of behaviour problems on the parental interview for the patient and their sibling(s), but the association was significant in the cyanotic subgroup and in the BMT group (Table 9.2).

9.5.3. Changes Over Time

There were no significant changes over time in the presence or absence of behaviour problems in the cyanotic and acyanotic subgroups or in the cardiac and healthy groups, but

there was a significant decrease in the presence of behaviour problems in the BMT group (Table 9.4). In the cardiac and healthy groups, but not in the BMT group, there was a significant association between the presence of behaviour problems at the two test occasions. The association was not significant in either the cyanotic or acyanotic subgroups.

9.6. SCHOOL PROBLEMS

9.6.1. Pre-operative Results

9.6.1.1. Cardiac Group

Parents of 52 of the 59 siblings felt that their child had no problems at school. Two were reported to have academic problems, 3 adjustment problems and a further 2 to have both academic and adjustment problems (Table 9.5). There were no significant differences in the prevalence of school problems between the cyanotic and acyanotic subgroups. In the acyanotic subgroup and total cardiac group there was a significant association between the presence of problem behaviour at school measured on the Rutter B and parental assessment of academic problems, but not between Rutter B scores and adjustment problems (Table 9.6).

9.6.1.2. Comparison with the Reference Groups

Parental ratings of school problems for the three groups are given in Table 9.5. There was no significant association between Rutter B scores and the presence of academic or adjustment problems in the BMT group. In the healthy group there was a significant association between Rutter B scores and adjustment problems (Table 9.6). There were no significant differences between the 3 groups in parental assessment of the occurrence of school problems.

SIBLING SCHOOL PROBLEMS					
	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVELY	(n=18)	(n=41)	(n=59)	(n=71)	(n=81)
NONE :	15 (83%)	37 (90%)	52 (88%)	62 (87%)	72 (89%)
ACADEMIC :	1 (6%)	1 (2%)	2 (3%)	4 (6%)	2 (2%)
ADJUSTMENT :	1 (6%)	2 (5%)	3 (5%)	2 (3%)	5 (6%)
ACADEMIC & ADJUSTMENT :	1 (6%)	1 (2%)	2 (3%)	3 (4%)	2 (2%)
POST-OPERATIVELY	(n=17)	(n=37)	(n=54)	(n=51)	(n=55)
NONE :	13 (76%)	26 (70%)	39 (72%)	44 (86%)	44 (80%)
ACADEMIC :	3 (18%)	1 (3%)	4 (7%)	4 (8%)	1 (2%)
ADJUSTMENT :	1 (6%)	6 (16%)	7 (13%)	2 (4%)	6 (11%)
ACADEMIC & ADJUSTMENT :	0 (0%)	4 (11%)	4 (7%)	1 (2%)	4 (7%)
CHANGES OVER TIME	(n=16)	(n=33)	(n=49)	(n=44)	(n=50)
ACADEMIC : Z =	-1.3416	-1.6036	- 2.0226	0.00	- 1.3416
p =	.1797	.1058	.0431	1.00	.1797
chi-sq =	Fisher Exact:	5.93183	10.50305	15.18688	19.07013
p =	.18750	.0149	.0012	.0001	.0000
ADJUSTMENT : Z =	.0000	-2.2014	- 1.8904	0.00	- 1.2136
p =	1.0000	.0277	.0587	1.00	.2249
chi-sq =	Fisher Exact:	5.22882	4.75080	.0000	12.05357
p =	.93750	.0222	.0293	1.000	.0005

Table 9.5

9.6.1.3. Children not Followed-up

Eight cardiac siblings on whom these data were collected pre-operatively were not seen post-operatively - all being lost to follow-up - and none of them were rated as having problems at school.

9.6.2. Post-operative Results

9.6.2.1. Cardiac Group

Parents of 39 of the 54 siblings felt that their child had no problems at school. Four were reported to have academic problems, 7 to have adjustment problems and a further 4 to have both academic and adjustment problems (Table 9.5). There was no association between the presence of problems on the Rutter B and parent-rated academic or adjustment problems in either of the 2 subgroups or total cardiac group (Table 9.6).

9.6.2.2. Comparison with the Reference Groups

Parental ratings of school problems for the three groups are given in Table 9.5. There was no significant association between Rutter B scores and the presence of academic or adjustment problems in either of the reference groups (Table 9.6). There were no significant differences between the 3 groups in parental assessment of the prevalence of school problems.

9.6.3. Changes Over Time

In the total cardiac group parents reported an increase in the prevalence of both academic and adjustment problems at school and in the acyanotic subgroup there was a reported increase in the prevalence of adjustment problems (Table 9.5). There were no significant

ASSOCIATION BETWEEN THE PRESENCE OF BEHAVIOUR PROBLEMS ON THE RUTTER B SCALE AND THE PRESENCE OF ACADEMIC OR ADJUSTMENT PROBLEMS AT SCHOOL, ACCORDING TO THE PARENTAL INTERVIEW					
	CYANOTIC	ACYANOTIC	CARDIAC	BMT	HEALTHY
PRE-OPERATIVE: RUTTER B AND ACADEMIC PROBLEMS	(n=16)	(n=32)	(n=48)	(n=56)	(n=67)
CHI-SQUARED:	Fisher Exact:	4.43077	10.24000	.11740	1.37781
p:	.12500	.0353	.0014	.7319	.2405
PRE-OPERATIVE: RUTTER B AND ADJUSTMENT PROBLEMS	(n=16)	(n=32)	(n=48)	(n=56)	(n=67)
CHI-SQUARED:	Fisher Exact:	.0000	.00000	.00000	8.43748
p:	.87500	1.0000	1.0000	1.0000	.0037
POST-OPERATIVE: RUTTER B AND ACADEMIC PROBLEMS	(n=14)	(n=29)	(n=43)	(n=42)	(n=42)
CHI-SQUARED:	Fisher Exact:	.0000	.18833	.01963	.00000
p:	.54670	1.0000	.6643	.8886	1.0000
POST-OPERATIVE: RUTTER B AND ADJUSTMENT PROBLEMS	(n=14)	(n=29)	(n=43)	(n=42)	(n=42)
CHI-SQUARED:	Fisher Exact:	.56234	.01189	.0000	.35000
p:	.78571	.4533	.9132	1.000	.5541

Table 9.6

changes over time in the occurrence of school problems in either of the reference groups or in the cyanotic subgroup.

In the acyanotic, but not cyanotic, subgroup and in the total cardiac, BMT and healthy groups there was a significant association between the presence of academic problems pre- and post-operatively. In the acyanotic subgroup and total cardiac and healthy groups, but not in the cyanotic or BMT groups, there was also a significant association between the presence of adjustment problems at the two test occasions (Table 9.5).

9.7. ADJUSTMENT FOLLOWING TREATMENT

9.7.1. Cardiac Group

At the 12 month follow-up parents of 24 (40%) of the 60 siblings retrospectively felt that the child had had adjustment difficulties following the patient's return from hospital. Frequently mentioned behaviours included withdrawal, anxieties, attention seeking and jealousy. In the vast majority of cases these adjustment problems had been of a relatively short duration (approximately one month) and were not ongoing at the post- operative follow-up. This is confirmed by the lack of association between early post-operative adjustment problems and behaviour problems on the Rutter A at 12 months (chi-squared = .00205; p=.9639). There were no differences in the prevalence of adjustment difficulties between the 17 siblings of children with cyanotic lesions and the 42 siblings of children with acyanotic lesions.

9.7.2. Comparison with the BMT Group

In the BMT group, as for the total cardiac group, parents of 22 (34%) of the 65 siblings on whom data were collected felt that there had been adjustment problems on the patient's return home. The types of behaviours mentioned were similar to those in the cardiac group and descriptive reports indicated that in some cases there were ongoing difficulties, although the majority had resolved by the 12 month follow-up. There was no significant association between early post-operative adjustment problems and a significant degree of problem behaviour on the Rutter A at 12 months (chi-squared = .70946; p=.3996).

9.8. DISCUSSION OF THE RESULTS

The literature describing the impact of chronic illness on siblings is inconsistent in its findings. Even within the very small number of studies of siblings of children with congenital heart disease, there are discrepancies and contradictions.

9.8.1. Behaviour at Home

The results of this study generally support the view that the presence of a chronically sick child does not invariably have a negative impact on the healthy siblings (Gayton et al, 1977; Ferrari, 1984; Fielding et al, 1985; Gallo et al, 1992), although the small sample sizes may be a reason for the lack of significant differences between the groups. On the structured measures of behaviour at home, the cardiac, BMT and healthy groups did not differ significantly in the prevalence of problem behaviour at the first test occasion. However, all three groups had higher rates of problem behaviour than reported for the normal population (Rutter et al, 1970). Although not significantly different, siblings of cardiac patients were reported to have a higher prevalence of antisocial type behaviour problems. Hostility,

jealousy and resentment have been previously reported for siblings of children with congenital heart disease (Maxwell & Gane, 1962; Boon, 1972), which corresponds with some parental reporting of sibling behaviour in this study: "He has been jealous of E because of all of the attention E received and he can be quite aggressive towards E" (15 year old sibling of 11 year old patient). Other parents reported that the healthy siblings were kinder and more understanding towards their ill sibling: "His eldest brother feels sorry for him and has been a bit nicer to him" (18 year old sibling of 11 year old patient). This has also been previously reported for siblings of chronically ill children (Iles, 1979; Taylor, 1980) and for siblings of cardiac patients in particular (Faux, 1991).

Within all three groups (although not in the cyanotic subgroup) there was a significant association between patient and sibling Rutter A total scores. This suggests that maternal factors may be important in the reporting of behaviour problems (Weissman et al, 1980; Ghodsian, Zajicek & Wolkind, 1984) and that family factors have an important influence on the adjustment of the sibling (Cox, 1988; Dumas et al, 1989; Gath, 1989).

After treatment the prevalence of problem behaviour at home did not differ between the three groups and there were no significant changes over time. However, within each group, but not within the cyanotic and acyanotic subgroups, there was a significant association between the presence of behaviour problems at the two test occasions. Whilst this clearly indicates the stability of behaviour problems over time for all three groups of siblings, it also highlights the similarity of the total cardiac/BMT and healthy groups and further supports the view that intrusive treatment regimens do not necessarily have a negative impact on sibling behaviour. Although on standardised measures the siblings of the ill children appeared relatively well

adjusted, there was variation in the scores. Descriptive data indicated that some siblings were particularly distressed, so caution is needed in basing all conclusions on the questionnaire data alone (Stewart et al, 1992). The lack of association between the presence of behaviour problems at the two test occasions in the cyanotic and acyanotic subgroups indicates the importance of assessing within group variability as well as looking at broad diagnostic categories. The smaller numbers of siblings within the cardiac subgroups is likely to be a contributory factor to the findings.

9.8.2. Behaviour at School

In terms of behaviour at school, the lack of difference between the three groups in the prevalence of problem behaviour on both test occasions again refutes the view of significant adverse psychological effects of chronic illness on healthy siblings. Behaviour problems were stable over time in the acyanotic subgroup, total cardiac and healthy groups, but not in the cyanotic subgroup or BMT group. The lack of change over time in the total cardiac group again suggests that the intervention of cardiac surgery does not have a significant impact on sibling behaviour at school. However, the fact that behaviour problems were stable over time in the acyanotic but not cyanotic or BMT groups suggests that siblings of less seriously ill children respond in a similar way to siblings of healthy children, but that children whose sibling is more seriously and overtly ill respond in a different way.

9.8.3. Parental Interview

On the parental interview, parents of BMT siblings reported a significantly higher prevalence of problem behaviour pre-operatively than parents of siblings in the cardiac or healthy groups. This may have been due to the fact that a number of the BMT siblings were marrow

donors and so were also having to face the prospect of hospitalisation themselves, together with their feelings about being donors. In the acyanotic subgroup and total cardiac and healthy groups the significant association between the presence of behaviour problems on the Rutter A and parental interview indicates that the Rutter A was a sufficiently sensitive measure to detect those children whom parents felt had behaviour problems, whereas in the cyanotic subgroup and BMT group this was not the case. The similar patterns of behaviour in the acyanotic and healthy groups suggests that the siblings of acyanotic patients had developed adaptive coping mechanisms for dealing with their sick siblings' illness, although again this may also be a reflection of the less serious nature of their siblings' illness. Despite successful surgery, behaviour problems in the siblings in the total cardiac group were stable over time, indicating that other factors were more important than the intervention of surgery. Continued maternal over-protection of the patient, mothers' mental state and coping mechanisms and siblings' own continued anxieties and resentment are all likely to contribute. A number of parents reported that, whilst the siblings still had behaviour problems, they were less severe than pre-operatively: "She has been very affected by L's condition. She felt pushed into the background and became withdrawn, unsettled and introverted. Now she is more settled but she still doesn't eat properly and she still has problems" (11 year old sister of 5 year old patient). It may be that one year is too short a time for the siblings of these patients to adjust fully to the successful treatment of their brother or sister, particularly if they have lived with congenital heart disease for a long time - or even for their whole life - and "normality" is still a difficult concept to grasp. In contrast, siblings of BMT children may have experienced a significantly shorter time of illness in the family and although they exhibit more behaviour problems initially - often in response to the diagnosis and immediate implementation of treatment- successful BMT leads to a fairly rapid resolution of a number

of those behaviours. Similar numbers of cardiac and BMT siblings were reported to have adjustment difficulties when the patient went home from hospital, although these more acute behavioural responses tended to be of relatively short duration and would seem to be more a reaction to the specific treatment and hospitalisation rather than to the underlying disease.

9.8.4. School Problems

Parents of siblings of other groups of chronically ill children have reported more schooling difficulties than parents of healthy children (Reynolds et al, 1988), but this was not true at the first test occasion in this study. Parental ratings of academic difficulties, but not adjustment, in siblings of acyanotic patients were associated with teachers' reports of behaviour problems. It might be expected that adjustment, rather than academic, difficulties and behaviour problems would be significantly correlated - and, indeed, this was the case for the healthy group. The acyanotic group finding suggests that parents were interpreting academic "failure" as a learning difficulty, rather than being due to behavioural problems - perhaps because this was a more acceptable explanation. Whilst the cardiac, BMT and healthy groups did not differ at follow-up in the prevalence of academic and adjustment problems, parents of siblings of cardiac patients reported an increase in both academic and adjustment problems, with 28% of children having difficulties in at least one area, which is similar to the rates reported by Reynolds et al (1988). In all three groups the significant association between academic problems at the two test occasions indicates that the two illness groups were responding in a similar way to the healthy group. The lack of association in the cyanotic subgroup is likely to be due, at least in part, to the small sample size. However, the significant increase in academic problems in the cardiac group suggests that the experience of their siblings' cardiac surgery and resulting disruption did have some impact

on academic functioning. In the acyanotic subgroup and total cardiac and healthy groups adjustment problems were also stable over time. Successful cardiac surgery does not result in early improvements by siblings at school - rather, the increase in problems suggests that the intervention of cardiac surgery can have a negative impact on siblings in the school environment, at least from the parents' perspective.

CHAPTER 10

CRITIQUE OF THE STUDY AND IMPLICATIONS OF THE RESULTS

10.1 DISCUSSION OF THE METHODOLOGY OF THE STUDY

10.1.1. Review of Aims and Design of the Study

The aim of the study was to evaluate the impact of congenital heart disease and subsequent cardiac surgery on the psychosocial functioning of the child and family. Seventy-five children with congenital heart disease and their families were assessed pre-operatively and 54 were followed-up one year after surgery. For comparison, a group of children with another type of life-threatening illness who were undergoing an intrusive, potentially fatal treatment regimen were seen, together with a group of healthy children with no medical problems. The cross-sectional and longitudinal components of this study distinguish it from previously published work with children with congenital heart disease undergoing cardiac surgery. Many of the existing studies are retrospective and lack reference groups, particularly within the younger age range. Another methodological innovation in the present study is its multifactorial framework, in which all family members are included.

Within the field of chronic illness in children the focus of understanding and research is now moving from a deficit centred approach to one which looks at the adaptive responses of children and families. One criticism of this study is that it is directed more towards assessing maladjustment, rather than adjustment and coping. However, so little work has been done on children with congenital heart disease (or those undergoing BMT) - particularly in comparison with the vast literature on children with cancer, diabetes, asthma etc. - that one of the initial aims of this study was to explore factors likely to be involved in congenital heart disease and cardiac surgery. Furthermore, based on previous findings, a specific set of

hypotheses were proposed and were addressed within a multifactorial framework, with the aim of clarifying some of the earlier work.

10.1.2. Methodology

10.1.2.1. Longitudinal Assessment

10.1.2.1.1. Selection of Cardiac Cases

The sample size (n=75) was large relative to many of the previously published studies and the sequential selection of cases ruled out possible selection bias and provided a sample representative of the population of children with congenital heart disease. However, selecting cases in this way meant that there was a large age range, resulting in quite small sample sizes within narrowly defined age bands. The group was heterogeneous with respect to diagnosis, type of surgery, bypass duration and other clinical variables, but the sample size was too small to control for all of these. The group was also not homogeneous with respect to the measured background characteristics. However, one of the objectives of the study was to examine the impact of congenital heart disease and cardiac surgery within a multifactorial framework. Selecting a totally homogeneous sample was not practical; furthermore, such a sample would be of limited value in generalising to the population of children with congenital heart disease.

There were only three children with mental handicap. To exclude them would have resulted in a biased sample unrepresentative of the overall population, but to include them in the analysis of cognitive function and behaviour would also have distorted the results. Unfortunately, the small number of such children means that no conclusions can be drawn

about the effects of congenital heart disease and subsequent cardiac surgery on this specific group.

Children were selected from three cardiac centres in London. The centres differed in terms of medical protocols and treatment regimens, facilities for children and families and approaches to visiting, accommodation for family members etc. However, the inclusion of children in a multicentre study reduces the risk of introducing some clinic biases. Variation in outcome within a single disease category has been at least partly attributed to specific differences in individual clinic populations (Lavigne & Faier-Routman, 1992) so the inclusion of children from several centres should result in findings more representative of the overall population of children with congenital heart disease.

10.1.2.1.2. Timing of Assessments

Pre-operatively, patients were seen in a hospital setting 1-2 days before surgery. Ideally, children would have been seen at home before they were admitted, but this was not possible for practical reasons. A number of children lived a considerable distance from the hospital; furthermore, families were often given very little warning of the exact date for admission. Assessing the children at a time when surgery was imminent may have influenced certain aspects - such as short term memory and measures of self perception. However, the environment and timing of assessment were the same for all cardiac patients. The pre-operative assessment focused retrospectively on functioning prior to the hospital admission. Parents were also distressed at this time, which may have influenced their recall and reporting of their own and their children's functioning.

10.1.2.1.3. Patients Lost to Follow-up

Ten children were lost to follow-up. In general, these patients did not differ on pre-operative measures from those who survived surgery, so it could be assumed that results of post-operative functioning were representative of the total sample who survived surgery. A number of those not followed up lived a considerable distance from the treating hospital and did not turn up at their local out-patient appointment with the treating consultant. This may have implications for their future compliance with treatment and perhaps is indicative of some of the problems faced by families treated far from home.

10.1.2.2. Cross-sectional Assessment

10.1.2.2.1. Selection of Comparison Groups

There are a number of difficulties in selecting appropriate comparison groups. In order to specifically assess the impact of cardiac surgery, the most appropriate control group would have been a group of children matched on background and medical variables who did not undergo cardiac surgery. As there is no viable alternative to surgical intervention, such a control group cannot be gathered. A group of children who met the criteria of having a chronic illness and undergoing a potentially life-saving treatment (BMT) were available for selection as a reference group. Within a non-categorical approach to illness, which suggests that there are more similarities than differences between different illness groups, such a group of children are an acceptable reference group.

10.1.2.2.2. Actual Comparability of CHD and BMT Groups

The two illness groups did differ on a number of medical parameters. One of the most important differences was that the congenital heart disease group all had a congenital

disorder, usually diagnosed at birth, whereas half of the BMT group had acquired disorders with a recent onset and diagnosis. Such differences have been identified previously as important influences on outcome. The two groups had differing disease courses, with those with acquired disorders tending to have a more rapid deterioration. In terms of intervention, the cardiac group underwent more invasive treatment with respect to having major surgery, but their length of hospital stay tended to be shorter and their post-operative recovery quicker than in the BMT group. Although the cardiac group all spent a short time in the intensive care unit, they did not have to endure the rigours and restrictions of isolation and reverse barrier nursing experienced by the BMT group. There were deaths in both groups, but those in the cardiac group happened during surgery or in the perioperative period. In contrast, there were a number of late deaths in the BMT group. However, both groups of children had to undergo stressful, intrusive and painful interventions. They both experienced hospitalisation and disruption to family and school life and the threat of dying was present for them all. For both the children and their families in the two groups, many of the stressors of chronic illness and subsequent treatment were similar.

10.1.2.2.3. Matching of the Three Groups

The only matching criterion on which the three groups differed was in the age of the patient, with the cardiac group being significantly younger than the reference groups. The diagnosis of congenital heart disease in infancy means that many children are - and have to be - treated at a young age, particularly as this has been considered to be beneficial for subsequent development and cognitive functioning in children with cyanotic lesions. In contrast, the acquired nature of many of the underlying disorders in the BMT group meant that age at diagnosis was very variable, with some diagnosed as teenagers. For those children in the

BMT group with genetic disorders, the mean age at onset of treatment was similar to that of the cardiac group. Although the genetic subgroup would have been a better comparison group in terms of age at diagnosis and treatment, this would have introduced a selection bias into the BMT group and, from a practical viewpoint, it would have taken significantly longer to gather a sufficient number of cases.

The groups did differ in age (cardiac: mean 5.2 years; S.D. 3.8 years; BMT: mean 6.6 years; S.D. 4.2 years; healthy: mean 6.7 years; S.D. 4.2 years) and although the differences were not large, they were statistically significant ($p<.05$). However, clinically the difference in mean ages of the total groups is of little relevance with respect to psychological outcome. It is of far greater clinical significance to look at the patients within smaller age bands. When the groups were split into more meaningful age bands - according to the basis on which the tests were conducted - the differences in age were no longer significant.

One criticism of the selection process is that the children in the cardiac group were not individually matched with the reference patients. Whilst practically it was not possible to individually match the cardiac and BMT groups, it would have been possible to match the cardiac and healthy groups. However, the original design of the study meant that the BMT and healthy groups were individually matched. One repercussion of this matching procedure was that within the healthy group in general only those children matched with a BMT survivor were followed up.

Of the available children and families, a comparable proportion were seen from each group, although a higher percentage declined to participate in the healthy group. With the

consecutive selection of cases in the two illness groups it was important that of those available, comparable proportions participated and that the reasons for non-participation were similar in each group, so that sampling bias was minimised. The two groups were very similar in the distribution of non-participating cases.

There were differences between the three study groups on a number of demographic and family factors, including residency, mean parental ages, unemployment and family size. Whilst the majority of the children in the healthy group came from the London area, a high proportion of the BMT group were resident more than 30 miles from London, although frequently in other cities. The distance of the family home from the hospital has implications for additional disruption to family life. The higher rate of unemployment seen in the two illness groups has been previously reported for parents of other chronically sick children (Burton, 1975). In some cases parents had given up their job to look after their child or be with them for their treatment. The differing employment rates may also be due to the relative prosperity and higher employment found in London compared with other areas of the U.K.

Cardiac children were more frequently only children than either the BMT or healthy children and family size was significantly smaller in the cardiac group. Other researchers have found that the birth of a child with congenital heart disease can result in the postponement of further pregnancies (Boon, 1972). The significant difference in family size is likely to be due to the congenital nature of the lesion, impact of a diagnosis in infancy and caretaking demands.

The three groups were comparable on many other background variables - such as social class, parents' marital state and siblings' mean age, sex and ordinal position distribution (despite the

fact that the siblings were not individually matched). A higher percentage of siblings in the BMT group had chronic illnesses than in the references groups. This was due to the fact that a number of the siblings had the same genetic disease as their brother or sister undergoing BMT.

Practically, it is impossible to match different groups of children on all background variables. Initial matching criteria of sex of the patient and social class did not differentiate the three groups, and the difference in mean age of the total groups was considered to be unimportant clinically. The comparability of the three groups on demographic and family variables was therefore considered to be acceptable.

10.1.2.2.4. Setting

Whilst both illness groups were seen in hospital, the healthy group were seen at home. Ideally the cardiac and BMT groups should also have been seen at home, but this was not practical (for the reasons noted above). It was also considered inappropriate to expect the healthy children and families to travel to a hospital to be seen. Being seen in a home environment might be expected to be less stressful, so this may have influenced the comparison of the illness and healthy groups.

In order to control the testing environment, both of the illness groups were seen for follow-up in a hospital setting. This was invariably during an outpatient visit, and whilst they may have had some anxieties about returning to the hospital, they did know that they would go home after the appointment. It is therefore possible that, although the initial and follow-up

assessments were both in a hospital environment, there were subtle differences due to the inpatient/outpatient distinction.

A number of the cardiac group were seen for follow-up at their local referring hospital, whereas the BMT group were all seen at the treating hospital. Again, although this was unavoidable for practical reasons, there may have been subtle differences between the two groups.

10.1.2.3. Measures Used

The Ruth Griffiths Scales (Griffiths, 1954; 1970) have been extensively used in British studies and were standardised on a British population. As with many developmental scales, there have been criticisms about the correlation between results obtained on the Griffiths scales and results obtained on other measures of cognitive function at an older age (Hindley & Owen, 1978). One of the main reasons for this discrepancy is that infant tests concentrate on assessing motor skills. The Ruth Griffiths scales have been in use for some decades, and more recently developed tests that concentrate on the infant's ability to deal with information are better predictors of later IQ (Murphy, 1987).

The British Ability Scales (Elliott, 1983) are currently one of the most popular scales used in Britain. Their suitability for use over a large age range and the fact that they were standardised on a British population were particular factors which resulted in their selection for this study. A number of the scales of the BAS have been criticised for containing a high level of test and item bias, which it is felt undermines the validity of the BAS for girls in particular (Margrain, 1985). However, most of the scales which have been reported to have

a gender-specific orientation are not used in the short form IQ assessment, which was the method adopted for this study.

Recently, there has been considerable criticism of IQ tests and what they actually measure (Flynn, 1987; Murphy, 1987). What appears clear is that standardised norms for developmental and cognitive tests are increasing (Cockburn & Ounsted, 1983; Hanson et al, 1985; Fulton et al, 1987; Fuggle et al, 1991) and that over time individual IQ scores change (Murphy, 1987; Hindley & Owen, 1978). Despite such criticisms, there are as yet no better tests of general cognitive function. Current tests therefore continue to have a role in areas such as chronic illness and whether or not children are performing within the normal range (Murphy, 1987).

The Richman (Richman & Graham, 1971; Richman, 1979) and Rutter (Rutter et al, 1970) scales were selected because of their standardisation on British samples and their wide use with groups of chronically ill children in British studies. However, there are a number of potential criticisms of the scales. Firstly, the inclusion of a number of somatic items, particularly on the Rutter A scale, which in those with chronic illness may reflect actual illness symptoms rather than behaviour problems. In studies using the Child Behaviour Checklist (CBCL) (Achenbach & Edelbrock, 1983) with groups of chronically ill children, elevated scores on the somatic scale have been reported (Wallander, Varni, Babani, Tweddle Banis & Thompson Wilcox, 1988; MacLean, Perrin, Gortmaker & Pierre, 1992). Confounding of illness symptoms and psychiatric symptomatology is unavoidable with behaviour rating scales (such as the CBCL and Rutter scales) that do not take into account the context (chronic physical illness) of the reported problems (MacLean et al, 1992).

Secondly, it has been suggested that such measures have limited sensitivity to identify mild adjustment problems of the type most often encountered in children with chronic physical disease (Perrin, Stein & Drotar, 1991). Thirdly, such scales may not reflect parental concern. In a study using the Richman BCL Stallard (1993) found that the number of parents reporting concern about one or more aspects of their child's behaviour was significantly higher than the number of children identified as having behaviour problems according to the scoring criteria. In some areas, such as bed-wetting, behaviours which were considered to be "marked problems" according to the scoring criteria were not considered to be problems by parents. Results from our study confirmed this view, indicating that children who obtained scores indicative of behaviour problems on the Rutter A were not necessarily rated as having behaviour problems on the parental interview, suggesting that the Rutter A was not measuring perceived areas of concern for parents. Finally, some parents and teachers have been reported as finding the focus of the Rutter items "disconcerting" due to the emphasis on undesirable traits (Goodman, 1994).

The measure of self-perception used in this study was devised specifically for this research. There has been considerable criticism of the fact that many studies of chronic illness do not include any measures completed by the children themselves but rely exclusively on parental or teacher or physician reports. Maternal perceptions of child behaviour are probably a function of both child and maternal characteristics (Wallander et al, 1988) and it has been suggested that parents under stress may tend to overestimate inappropriate child behaviour (Brody & Forehand, 1986), which may be particularly relevant for the parents of chronically sick children. The tool used in this study used constructs related to the self perception of chronically ill children (Burns & Zweig, 1980) and enabled an evaluation not only of how the

children perceived themselves, but also how they would like to be. This latter aspect is particularly important in evaluating quality of life for these children. More traditional measures of self perception enable comparisons to be made between different groups of children - including healthy children - but they do not take into account how the ratings relate to the way the child would like to be. Although real and ideal self images have been measured previously (Allen & Zigler, 1986), such studies are rare. The measure used in this study also had the advantage of allowing parents to rate their children on the same constructs, thus enabling direct comparison of child and parent responses. Use of cartoon drawings to illustrate the poles of the constructs enabled the measure to be used with relatively young children, so that a wide age range could be covered.

Areas of parental functioning were assessed with four measures. The GHQ (Goldberg, 1972; 1978) and DAS (Spanier, 1976) have been widely used in studies of chronic illness. The coping instrument (UCL: Schreurs, 1987) has not been extensively used in either British or American studies, although it does have acceptable validity and reliability. The locus of control measure was created for this study, but all items included were from other validated measures. All of the six mean item scores were significantly correlated with the total score, indicating that the same concept was being measured throughout.

10.1.3.4. Compliance with Research and Completion of Measures

Overall, acceptable rates of compliance with the research were obtained and completion rates of the questionnaires were satisfactory. The study was viewed positively by both families and staff. Indeed, many families welcomed participating and seemed to benefit from the opportunity to discuss their feelings and relay the details of their child's illness and its impact

on the family. A number of parents expressed a desire to be involved in longer term follow-up and were anxious to be part of a research programme that could help others in the future. After the conclusion of data collection, some of the families maintained contact by letter and telephone, seemingly reluctant to sever the links. Staff in all centres were cooperative and helpful in facilitating the smooth running of the data collection. In many instances they too welcomed the involvement of the research worker, particularly if they had specific concerns about individual children and families. In some of the centres the need for support of the staff was highlighted by the regular contact with the research workers - particularly in those units where there was, as yet, no psychological support mechanism.

10.1.3.5. Further Limitations in Study Design

As is the case with many other studies of the effects of chronic illness and subsequent hospitalisation, invariably it was the mothers who were seen for research purposes. Fathers were frequently not seen. The reasons for this were mainly of a practical nature - fathers often stayed at home to look after other children, tending to visit in the evenings or at weekends. However, fathers did fill in a series of questionnaires, so there was some direct information provided. Siblings were also not usually seen, so it was necessary to rely exclusively on parent/teacher reports. This is also a frequently reported shortcoming, but for practical reasons it was unavoidable.

Due to the lack of any suitable measures of behaviour for children under 3 years, a large percentage of the cardiac group in particular were not assessed with regard to behaviour. A further concern was the difficulty in obtaining an objective, global rating of the child's physical condition, completed by physicians. A number of studies of cardiac patients have

used the New York Health Association (NYHA) classification, but this was not considered to be sensitive enough, with virtually all patients being classed in NYHA category 1 or 2. The doctors were therefore asked to make a rating of the urgency of treatment pre-operatively, but no objective ratings were collected at follow-up. In retrospect, a linear scale of illness severity should have been designed for this study, which could have been used before and after treatment for both illness groups. Furthermore, parents could have also rated their child's health status on the same scale, enabling direct comparison of parents' and physicians' perceptions of disease severity.

The study design enabled us to explore factors likely to be involved in congenital heart disease and cardiac surgery within a multifactorial framework. Some of the earlier research has been clarified and areas for future investigation can be proposed, based on the findings of this study.

10.2. DISCUSSION OF THE RESULTS OF THE STUDY

The results have already been discussed within the individual chapters. The focus of the first part of this section is the discussion of the specific hypotheses. The study will then be discussed within a more general framework.

10.2.1. Hypotheses

Table 10.1 summarises the hypotheses, together with the results.

SUMMARY OF HYPOTHESES AND RESULTS

NUMBER	HYPOTHESIS	RESULTS
1	Children with cyanotic lesions will perform at a lower level of intellectual functioning pre-operatively compared with children with acyanotic defects.	Developmental Testing: 0 - 3.5 Years No significant differences between cyanotic and acyanotic children. Cognitive Testing: 3.6 - 17 Years Significant differences between cyanotic and acyanotic groups on matrices and speed of information processing subtests, overall IQ, arithmetic, reading and spelling. HYPOTHESIS SUPPORTED FOR OLDER, BUT NOT YOUNGER, AGE GROUP.
2	Corrective surgery will result in a greater increase in cognitive function for children with cyanotic defects compared with those with acyanotic lesions.	Patients with cyanotic lesions did not show a greater increase in developmental or cognitive function than those with acyanotic defects. HYPOTHESIS NOT SUPPORTED

3	Age at time of repair will be positively correlated with a greater degree of post-operative cognitive impairment in children with cyanotic defects but for children with acyanotic lesions there will be no association between age at repair and degree of cognitive impairment.	Developmental Testing: 0-4.5 Years Cyanotic Group: significant positive correlation between age and performance on eye-hand coordination and performance subtests. No correlation between age and performance in acyanotic group. Cognitive Testing: 4.6-17 Years Age was negatively correlated with performance on the arithmetic attainment for both cyanotic and acyanotic groups. HYPOTHESIS NOT SUPPORTED
4	There will be a higher prevalence of behaviour problems both before and at 12 months after surgery among those patients with a cyanotic lesion compared with those with an acyanotic lesion, and in the group with acyanotic defects the prevalence of behaviour problems will be higher in those with symptomatic, compared with asymptomatic, lesions.	There was a higher prevalence of behaviour problems in the cyanotic children compared with the acyanotic children, but the differences did not reach statistical significance. Within the acyanotic group there was a significantly higher prevalence of behaviour problems post-operatively in the symptomatic group. HYPOTHESIS NOT SUPPORTED FOR THE DIFFERENCES BETWEEN CYANOTIC AND ACYANOTIC PATIENTS. HYPOTHESIS SUPPORTED FOR POST-OPERATIVE DIFFERENCES BETWEEN SYMPTOMATIC AND ASYMPTOMATIC GROUPS

5	Positive self perception, both before and at 12 months after treatment, will be correlated with Rutter A scores at 12 months.	Pre-operative self perception was not correlated with post-operative Rutter A scores in any of the 3 groups. Post-operative self perception was positively correlated with post-operative Rutter A scores in the cardiac group, but not in the other 2 groups. HYPOTHESIS SUPPORTED FOR POST-OPERATIVE FINDINGS IN THE CARDIAC GROUP, BUT NOT SUPPORTED FOR THE 2 REFERENCE GROUPS
6	For the older age group (5-17 years) those who score as poorly adjusted at 12 months on the Rutter A or B scales will have a longer mean time after surgery before returning to school compared with those whose adjustment is within normal limits.	There was no significant correlation between time before returning to school and Rutter A or Rutter B scores in either the cardiac or BMT groups. HYPOTHESIS NOT SUPPORTED
7	Time in hospital since discharge will be correlated with Rutter A and B scores at 12 months.	Time in hospital since discharge was correlated with behaviour problems at home in the cardiac group. HYPOTHESIS SUPPORTED
8	Pre-operative adjustment of the child will be significantly associated with post-operative adjustment.	There was a significant association between pre- and post-operative adjustment at home and at school in the cardiac group but not in either of the reference groups. HYPOTHESIS SUPPORTED FOR CARDIAC GROUP BUT NOT FOR 2 REFERENCE GROUPS

9	There will be no relationship between mothers' and clinicians' perception of disease severity.	There was no significant association between mothers' and clinicians' perception of disease severity. HYPOTHESIS SUPPORTED
10	Maternal levels of psychological distress at 12 months will be negatively correlated with adjustment in the child at 12 months.	The presence of maternal psychological distress was significantly correlated with behaviour problems at home in the cardiac group but not in either of the 2 reference groups. HYPOTHESIS SUPPORTED FOR CARDIAC GROUP BUT NOT FOR REFERENCE GROUPS.
11	Pre-operatively, those mothers showing psychological distress will perceive their child's disease as more severe compared with those mothers showing no psychological distress, regardless of clinicians' assessment of actual urgency of treatment.	Regardless of clinicians' assessment of actual urgency of treatment, mothers showing psychological distress pre-operatively did not perceive their child's disease as more severe compared with those mothers showing no psychological distress. HYPOTHESIS NOT SUPPORTED
12	Pre-operative parental distress will be significantly associated with post-operative distress.	There was a significant association between pre- and post-operative levels of psychological distress for mothers of the cardiac group and fathers of the healthy group, but not for mothers in either of the 2 reference groups or for fathers in either of the 2 illness groups. HYPOTHESIS SUPPORTED FOR MOTHERS OF THE CARDIAC GROUP AND FATHERS OF THE HEALTHY GROUP

Table 10.1

A further hypothesis, not included in the table, was that psychosocial functioning of the child, siblings and parents will be more compromised in the cardiac group compared with the healthy group, but the cardiac and BMT groups will not differ significantly from each other. This has been discussed throughout in the context of the specific results and other hypotheses, where appropriate.

The hypotheses were postulated on the basis of previous research, both with children with chronic illness in general and specifically with congenital heart disease. The hypotheses concerned with developmental and cognitive functioning in the cardiac group were not supported. Pre-operatively there were no differences in developmental functioning in the younger age group between those children with cyanotic and acyanotic lesions. There is little published information on the development of younger children with congenital heart disease, but the hypothesis was predominantly based on results of research with older children. The lack of difference at this early stage of development indicates that the expected deleterious effects associated with cyanosis are not yet evident. However, within the older age group there were significant differences between the cyanotic and acyanotic groups on a number of cognitive parameters, with the cyanotic patients performing at a significantly lower level. The difference in findings for the two age groups suggests that there may be a critical age for correction of cyanotic lesions, after which there are negative effects associated with the presence of cyanosis for subsequent development and cognition. As well as direct effects of hypoxia and other cardiac-related factors, it is also likely that more prolonged cyanosis influences parenting attitudes, treatment of the child by teachers and peers, self-esteem etc., all of which could influence cognitive and academic performance. Greater increases after surgery in cognitive performance scores for children with cyanotic, compared with acyanotic

lesions were not found (hypothesis 2). Furthermore, there was no association between age at time of repair and subsequent cognitive functioning in cyanotic patients (hypothesis 3). Several factors may have contributed to these findings. A number of cyanotic patients still had residual cardiac problems and the fact that their lesions were not corrected would mean that improvements in performance after surgery would not necessarily be expected. The number of cyanotic patients was small and the group was not homogeneous with respect to diagnosis. Studies which have linked age at time of repair with subsequent cognitive functioning have been based on children with a specific condition (transposition of the great arteries), so it is possible that the correction of the defect is not the only relevant factor. Within the younger group the age range at the time of correction of the defects was smaller than that of Newburger et al (1984), so it is possible that the differences in age in our study were not great enough to expect any association between age at repair and subsequent cognitive function. Within the older age group there were only 11 patients with cyanotic lesions who were followed up, with an age range at surgery of 3.6 years to 10.4 years, and other factors such as hospitalisation, social class and parental attitudes are also likely to have been influential, possibly rendering insignificant the association between age at repair and post-operative performance.

Both before and after surgery, the prevalence of behaviour problems was higher, but not significantly so, in the cyanotic, compared with the acyanotic, group (hypothesis 4). The lack of significant difference between the cyanotic and acyanotic groups is likely to be attributable, at least in part, to the relatively small sample sizes, particularly in the cyanotic group. These results could therefore be accounted for by a type II statistical error.

The finding of a significant correlation between post-operative self perception and adjustment (measured by the Rutter A scales) in the cardiac but not in the reference groups (hypothesis 5) may be due to the different nature of the behaviour problems of the three groups. Within the cardiac group, of those children with behaviour problems, four (80%) had problems of a neurotic nature; none had an antisocial pattern of behaviour. In contrast, in the two reference groups, 50% of the children with problem behaviour had antisocial patterns of behaviour. There may be a relationship between neurotic behaviours and self-perception, but not between antisocial behaviours and self-perception.

There was no significant correlation between post-operative adjustment and time before returning to school in either of the two illness groups. In general, the cardiac children returned to school within a couple of months of surgery and there was relatively little variation in the length of absence. This is likely to be due to the fact that follow-up policies in cardiac centres were similar, with the usual recommendation being that, for children who were physically well, return to school was allowed once the sternum had healed (usually about 6 weeks after surgery), with an initial limitation on contact sports. Most of the literature on the effects of chronic illness on schooling focuses on children with cancer. It remains unclear how "corrective" treatment for chronic conditions, such as cardiac surgery, affects return to school and the reintegration of the child back into the school environment.

It was suggested that further hospitalisation after initial surgery would be correlated with behaviour problems (hypothesis 7). This hypothesis was supported for the cardiac group. In general, the children in the cardiac group and their parents had expected that the surgery would resolve their heart problems and that further hospitalisation would not be necessary.

The finding that there was an association between subsequent hospitalisation and adjustment problems is likely to be attributable not only to the child, but also to the parents. Those parents whose children had developed subsequent medical problems reported feeling anxious and also expressed specific concerns about the future and their child's health. It is well documented that maternal mental health influences not only their reporting of child behaviour problems (Fergusson, Horwood, Gretton & Shannon, 1985) but also the actual occurrence of adjustment problems in the children (Rutter & Quinton, 1984; Zuckerman & Beardslee, 1987; Mulhern, Fairclough, Smith & Douglas, 1992; Thompson, Gustafson, George & Spock, 1994). It is therefore not possible to assess the degree to which the need for further treatment is related directly to adjustment difficulties in the child.

Pre-operative adjustment was significantly associated with post-operative adjustment in the cardiac group and specifically for the cyanotic patients. This suggests that the intervention of cardiac surgery has less impact on post-operative adjustment than the presence of a "heart problem". These children had lived with their condition since birth and the cyanotic patients in particular had experienced significant effects on their appearance, peer relationships, school attendance etc. Furthermore, their families had also had to cope with the condition for some time. Although a number of the children were significantly improved after surgery, behaviour problems were stable over time. Corrective surgery had not resulted in significant improvements in behaviour, although it may be that positive effects of treatment on behaviour were not yet evident 12 months after surgery and that longer term follow-up would indicate improvements. One contributing factor to this was the mothers' unchanged perception of their children. However, behaviour problems at school, assessed by teachers, were also

stable over time, suggesting that there were characteristics of the children themselves which determined their adjustment.

In the BMT group there was an increase after treatment in the number of children with behaviour problems at home, suggesting that the treatment itself had had a detrimental effect on some of the children. Again, maternal anxiety and perception of the sick child are likely to have contributed to the reporting of behaviour problems.

For the cardiac group, it was suggested that there would be no relationship between mothers' and clinicians' perception of disease severity (hypothesis 9). Urgency of surgery was used as a measure of clinicians' perception of disease severity, which was judged by clinicians as being an accurate indicator of the child's current health status. Furthermore, doctors reported that the degree of urgency of surgery was communicated to parents as a rating of how sick the child was. The results are consistent with other research on maternal perception of disease severity (Bergmann & Stamm, 1967; Offord et al, 1972; Casey et al, 1994) although another recent study found that mothers did have accurate perceptions of disease severity (DeMaso et al, 1991). However, assessments of maternal perception of disease severity in the current study were carried out just 1-2 days prior to surgery, which is likely to have influenced mothers' ratings of their child's condition.

There was support for the hypothesis that post-operative maternal levels of psychological distress would be correlated with behaviour problems at home in the cardiac group (hypothesis 10), although the hypothesis was not supported for either of the reference groups. Poor adjustment in cardiac (Linde et al, 1966; Kong et al, 1986) and other groups

of chronically ill (Ludman, Spitz & Kiely, 1994; Sloper et al, 1994) children has previously been related to maternal distress and anxiety, although again the role of maternal mental health in the reporting of behaviour problems is not clear. The finding that there was no correlation between maternal GHQ scores and children's Rutter A scores in the healthy group suggests that the role of maternal mental health in the occurrence of behaviour problems is not a simple cause and effect. In the cardiac group it is probable that both maternal distress and adjustment in the child were in part individually linked to aspects of the disease and/or treatment. The lack of a relationship between maternal and child adjustment in the BMT group suggests that specific disease and/or treatment factors had less of a direct impact on mother and child than in the cardiac group.

Parents' perceptions of disease severity have been found to be influential in determining their own adjustment (Linde et al, 1970; Parcel et al, 1979; Jessop, Riessman & Stein, 1988) but in the current study mothers showing psychological distress did not perceive their child's disease to be more severe than mothers who did not show psychological distress (hypothesis 11). Other researchers have found that the relationship between perception of disease severity and adjustment is not always direct and that in some instances parents who perceived their child's illness as less severe than it actually was had more problems (Frydman, 1980). The lack of a relationship between these two variables indicates the importance of assessing the impact of chronic illness within a multifactorial framework, rather than looking at individual factors in isolation within a simple cause and effect relationship.

There was support for the hypothesis that pre-operative levels of maternal distress would be significantly associated with post-operative distress (hypothesis 12) in the cardiac group, but

not in either of the two reference groups. The lack of an association in the healthy group suggests that distress levels are not stable over time and are likely to be influenced by a variety of changing family and sociodemographic factors. In the cardiac group, but not in the BMT group, the stability of distress levels over time is likely to be attributable, at least in part, to some aspects of the disease, such as the congenital nature of the lesion, and to the emotional significance of the involvement of the heart. There was a significant reduction over time in the prevalence of psychological distress for mothers and fathers in both of the illness groups, but not in the healthy group, suggesting that successful treatment had had a positive impact on parental levels of distress.

10.2.2. Discussion of the Results within a Theoretical Framework

There is relatively little published literature about the psychosocial impact of congenital heart disease and subsequent cardiac surgery for the child and his/her family. The essentially exploratory nature of this study indicates that, in common with other chronic illnesses, children with congenital heart disease and their families are at increased risk of adjustment difficulties. Cardiac surgery is a potential stressor, but it does not have a uniform impact. Rather, its effects are mediated by a myriad of demographic, social, environmental and medical factors.

Within the literature on chronic illness there are two main theoretical perspectives. The disease specific approach, which until recently was the dominant method for identifying children with chronic conditions, considers individual conditions as discrete entities, with each disease differing from every other. Treatment of and adjustment to each disease is dependent on its specific characteristics, with research focusing on the identification of these.

From a conceptual point of view it seems unlikely that each of the thousands of chronic conditions has distinct effects on children and families (Perrin, Newacheck, Pless, Drotar, Gortmaker, Leventhal, Perrin, Stein, Walker & Weitzman, 1993).

Empirical studies have demonstrated the existence of commonalities in the experience of having a chronic condition, aside from characteristics specific to particular conditions (Nolan & Pless, 1986; Pless & Nolan, 1991). This alternative perspective is the "non-categorical" approach, which focuses on factors shared by many conditions, children and families and groups children with diverse conditions across, rather than within, diagnostic categories (Pless & Pinkerton, 1975; Stein & Jessop, 1982(a)). Applying such an approach to research and health policies requires an "effective operational definition of a chronic condition" (Perrin et al, 1993). In contrast to the disease specific approach, in which definitions focus on the presence of a particular condition, definitions based on the non-categorical approach generally focus on the impact of the condition on the child and may therefore reflect aspects such as the child's functional status or ongoing requirements for medication etc., rather than the mere presence of a condition. Use of the consequences of disorders allows consistency across diagnoses, describes the impact of morbidity and is adaptable to meet specific purposes (Stein, Bauman, Westbrook, Coupey & Ireys, 1993)

Pre-operatively, on many of the psychological parameters evaluated, the cardiac and BMT groups did not differ significantly, supporting a non-categorical approach to illness. In this study, the cardiac and BMT groups were not comparable on a number of disease parameters, such as the illness severity, congenital or acquired nature of the disease, time since diagnosis, the extent of the illness-imposed restrictions etc. - yet essentially the two groups

demonstrated very similar psychosocial characteristics. However, the two groups did share the fact that they both consisted of children with a chronic illness who were facing the prospect of imminent, intrusive, potentially life-threatening treatment and hospitalisation.

Within the cardiac group itself, although heterogeneous with respect to illness severity and the presence of cyanosis or other symptoms, the cyanotic and acyanotic groups differed on very few parameters except for some measures of cognitive testing within the older age group. In general, the pre-operative results of the cardiac group support the view that it is the presence, rather than the nature, of the underlying disorder which is of significance.

Pre-operative comparisons of both illness groups and the healthy group indicated that both the cardiac and BMT groups experienced more problems on a number of parameters than the healthy group. In some areas, such as the developmental testing and parental interview, both the cardiac and BMT groups demonstrated similar differences when compared with the healthy group.

Post-operatively, the cardiac and BMT groups again demonstrated a number of similarities. However, within the cardiac group there were a number of significant differences between the cyanotic and acyanotic patients, particularly on the developmental and cognitive parameters. In contrast to the pre-operative findings, the nature of the initial diagnosis was a relevant factor within the cardiac group in determining psychosocial functioning after treatment. The acyanotic group showed more similarities with the healthy group than with the cyanotic group. The BMT group showed similarities with the acyanotic group on developmental and cognitive parameters, whereas the cyanotic group demonstrated a number

of significant differences with both the BMT and acyanotic groups. Some of these differences in the pattern of pre- and post-operative comparisons are likely to be attributable to the fact that some of the acyanotic and BMT patients no longer had (or were considered by parents/themselves to have) a "chronic illness". The problem of classifying children who have conditions with long intervals of quiescence that may or may not represent a "cure", such as children with repaired congenital heart disease, has been identified (Stein et al, 1993). In the cyanotic group, although many had had corrective surgery, several still had ongoing problems and for others, who were physically well, there were still some effects on their physical appearance - such as small stature. Stein and colleagues (1993) have proposed a framework for identifying children with ongoing health problems. Within this framework a number of children in the current study with corrected acyanotic defects would not be considered to have a chronic condition due to the fact that they were not limited in function, activities or social role compared with healthy peers, were not dependent on medication and did not now need ongoing medical care. In contrast, children with corrected cyanotic lesions still had limitations in areas of cognition and emotional development, many were still requiring medication and for some the physical effects and limitations of the lesions were still evident. Classification of children according to the consequences of their current health status, in accordance with a non-categorical approach rather than by diagnosis of congenital heart disease, permits a more meaningful interpretation of our results.

These results indicate that within a single disease category there may be more variation in terms of the psychological impact than there is between different disease categories and that the effects can change with time/treatment. Much of the published research on chronic illness has looked at the impact of a specific disease, such as cystic fibrosis or diabetes, without

considering the variability within the sample and the experience of individual children. The results of this study suggest that neither a disease-specific nor a non-categorical approach is totally adequate - rather, the focus should combine aspects of both approaches. There are undoubtedly some factors which are specific to the study of congenital heart disease and cardiac surgery - e.g. the involvement of the heart and the emotional significance of that, the presence of cyanosis and the resulting physical, physiological and psychological implications, and the need for open heart surgery, with possible repercussions of the bypass procedure etc. Other aspects of congenital heart disease are shared with a few other diseases, such as the effects on physical appearance and the need for surgery, whilst there are yet other variables common to the majority of children with chronic illness - e.g. need for checkups, hospital admissions, coping with the reactions of others etc.

The study results indicate that there are a number of risk factors associated with congenital heart disease - such as the congenital nature of the illness - and with cyanotic heart disease in particular, that need further clarification. A number of the hypotheses were not supported and some of the results were not in agreement with those of previous researchers. Whilst small sample sizes is likely to be a contributory factor, it is also probable that some of the earlier work which focused on one or two variables was too simplistic in its approach.

10.3. IMPLICATIONS OF THE STUDY RESULTS AND FUTURE DIRECTIONS

As a chronic condition, congenital heart disease creates many of the same generic stresses for the child and their family as do other chronic illnesses. However, there are also some unique stressors with which the child and family have to cope, such as heart failure. Congenital heart disease is a heterogeneous group of conditions with a wide variation in

physical effects in affected children. However, there are indications that there are also some factors, such as cyanosis, which if present are specific stressors.

The original model chosen for this study contained aspects of a disease specific approach (in terms of some of the specific medical and surgical variables related to congenital heart disease and cardiac surgery in particular) as well as some variables based on the general literature, favouring a non-categorical approach (see pages 82, 367). The sample size in this study precluded evaluating the specific impact of some potentially important variables, but the use of the model as a basis for assessing the impact of congenital heart disease and cardiac surgery on the child and family enabled some preliminary evaluation of the role of some of these variables. The study demonstrated that the use of such a model is justified and valid, in that it was possible to test the proposed objectives.

The results of this research corroborate those of much of the literature on the effects of chronic illness in general. Although children with congenital heart disease and their families are at increased risk of adjustment difficulties compared with the healthy population, more importantly, the majority of children and families appear to cope with congenital heart disease and subsequent cardiac surgery.

10.3.1. Implications for Further Research

Further research with a larger sample is now needed in order to assess how psychosocial morbidity is related to any treatment, environmental or family characteristics. Furthermore, those variables that predict adjustment and adaptive coping responses to congenital heart disease and cardiac surgery need to be identified. Although congenital heart disease is one

of the most commonly occurring chronic health conditions in children, it has been the focus of relatively little research. The current study, by identifying that there are psychosocial implications of congenital heart disease and cardiac surgery for the child and family, has conclusively shown that further research with this particular population is warranted.

It is clear that a multifactorial approach to the study of coping with congenital heart disease and cardiac surgery is required. A child's chronic illness does not affect the child alone (Kazak, 1989). Instead, all family members are affected and the changes imposed on individuals also affect the rest of the family. For example, the presence of a child with congenital heart disease not only has a direct effect on parents and other siblings, but the impact on the parents, such as having to devote more time to the sick child, also affects siblings. In the same way that disease processes and medical interventions are not static, so functioning of all family members fluctuates over time, so there is also a dynamic component involved in the psychosocial functioning of the child and family.

Within the current study, the effects of chronic disease and the intervention of stressful, potentially fatal treatment regimens on psychosocial functioning were evaluated and assessments made at two points in time. Although cardiac surgery did not appear to have a significant impact on psychosocial functioning in the short term, the longer term implications were not examined and need further investigation. Other researchers have found that children with corrected cyanotic lesions continue to be an "at risk" group for adjustment difficulties (Wright & Nolan, 1994), but even in children with corrected acyanotic lesions there are suggestions that there may be repercussions in later life (Brandhagen et al, 1991).

10.3.2. Research Within A Developmental Framework

The literature on the effects of chronic illness clearly indicates the importance of assessing the adjustment and coping of children within a developmental framework (Hagen, Anderson & Barclay, 1986; Eiser, 1990(b)). Children's understanding of the causes and implications of illness and compliance with treatment vary with age. Evaluation of the impact of illness on children must therefore be carried out from a developmentally sensitive perspective. One criticism of much of the developmentally based work is that cognitive and emotional responses to illness are assumed to follow a sequence of stages, with little consideration being given to the effects of social and cultural factors, or the previous experience of illness (Eiser, 1989). It is also important to recognise that basic physiological processes change with age and that biological changes can influence the child's psychological response to illness (Coupey & Cohen, 1984).

Although the age range of this study, in common with many others, was quite extensive, children were assessed within more clinically relevant, narrower age bands. Further research would benefit from assessing larger numbers of children within narrower age ranges, so that greater emphasis could be given to the normal developmental changes of childhood and the ways in which illness at a particular age may affect them.

As well as considering the adjustment of the child within a developmental framework, the developmental stage of the family also needs to be taken into account. Factors which are relevant for families of babies are different to those pertinent for the family of the adolescent. The child's developmental stage influences the response of others towards the child (Hagen

et al, 1986) and the emotional and caretaking demands placed on families are also dependent on the age of the child and the developmental stage of the family.

10.3.3. Methods of Assessment

10.3.3.1. Longitudinal and Cross-Sectional Measurement

Although the impact of congenital heart disease on the child and family was assessed pre-operatively, there is clearly a need to evaluate the effects from the time of diagnosis. Improvements in coping over time have been documented in other chronic illnesses (Kupst, Schulman, Maurer, Honig, Morgan & Fochtman, 1984; Kupst & Schulman, 1988). However, within the limited research on the effects of congenital heart disease, there are no long-term follow-up studies which assess family adaptation from the time of diagnosis. Within our study a number of children had palliative, rather than corrective, surgery and it would seem to be important to evaluate the long term implications of such differences.

The use of a group of children undergoing BMT for comparison highlighted the similarities in a number of psychological parameters between different illness groups. Such cross-sectional studies provide valuable information about the impact of chronic illness and stressful hospital treatment but future research would benefit from comparison of children with congenital heart disease and their families with other groups of children with congenital problems undergoing major surgical procedures, such as those with biliary atresia. This would enable, within a non-categorical framework, further clarification of the role of factors such as the presence of a congenital lesion and diagnosis in infancy.

10.3.3.2. Sources of Information

Research on the impact of chronic illness should involve all family members. Too often, information is provided solely by mothers, with fathers and siblings rarely having any direct involvement. A number of studies have found discrepancies between mothers' and fathers' reporting of problems (Earls, 1980; Eiser et al, 1992) and relying on one parent only for information is likely to yield only a partial picture (Eiser et al, 1992). Although there are many practical difficulties associated with active participation by other family members, efforts should still be made to do this. Within our study, fathers were asked to complete a series of questionnaires on themselves only, although few studies have included any measures of paternal functioning, and all of the information obtained on the siblings was provided by parents and teachers. Future research needs to focus on ways of promoting the direct involvement of each family member, particularly as fathers and siblings of children with congenital heart disease have been rarely included in studies.

Much of the work evaluating the impact of disease on the paediatric patient has relied on mothers', physicians' and teachers' reports, although the use of different informants can result in conflicting results (Perrin, Ramsey & Sandler, 1987; Noll, LeRoy, Bukowski, Rogosch & Kulkarni, 1991). However, it is essential that the children should contribute information themselves. Children do not necessarily share their parents' perspective on their illness (Levenson, Copeland, Morrow, Pfefferbaum & Silberberg, 1983; Eiser, 1993) and relying solely on parental reports can result in a distorted, biased view of the impact on the child. Our study found a lack of agreement between parents' and children's reporting on measures of the child's self-perception, supporting previous findings on the lack of inter-informant

agreement (Weissman et al, 1980; Nelms, 1986; Ennett et al, 1991; Engstrom, 1992; Wachtel et al, 1995).

The perceptions of the physicians also need to be evaluated, in order to obtain some objective ratings of disease severity. Assessments by physicians on a longitudinal basis would also enable actual changes in disease state to be correlated with changes in psychosocial functioning of the child and family. Although it has been suggested that actual disease severity has less bearing on adaptation than perception of disease severity, few studies have actually looked at the progression of disease state over time and how changes influence the coping of children and their families.

10.3.4. Congenital Heart Disease and Family Functioning

Family functioning in families of children with congenital heart disease has not been looked at previously, although in other groups of chronically ill children family functioning has been found to be a better predictor of child adjustment than the presence of the illness (e.g. Lewis and Khaw, 1982). In particular, high levels of family conflict (Wertlieb, Hauser & Jacobson, 1986) and low levels of family cohesion (Lavigne, Nolan & McLone, 1988) are associated with behaviour symptomatology. When a family has limited resources available to it, that is associated with greater parental depression (Timko, Stovel & Moos, 1992). Furthermore, family functioning has been found to have indirect effects on the physical health of chronically ill children (Patterson, McCubbin & Warwick, 1990). Family strain is also a strong predictor of teacher-rated adjustment of children with surgically palliated complex congenital heart disease (Casey et al (in press)). Such evidence indicates the need to assess the role of family functioning in the adjustment of children with congenital heart disease and to evaluate the

family in terms of relationships, cohesion and other interactions between family members, rather than just looking at individuals in isolation. The importance of conducting family interviews to assess family coping, rather than assessing individuals' perceptions of the family, has also been highlighted (Hauser, 1990).

10.3.5. Identification of Adaptive Responses to Congenital Heart Disease

Further research needs to move away from a deficit-centred approach towards identifying how the majority of children and families cope with congenital heart disease. Few of the assessment tools used to evaluate the impact of chronic illness have norms established for patterns within the chronically ill population (Masters, Cerreto & Mendlowitz, 1983) - instead, many have been designed for psychiatric populations. Appropriate selection of measures is therefore imperative so that responses which are psychologically "healthy" within the sphere of chronic illness are not deemed to be pathological due to comparisons with inappropriate standards. In an age of financial constraints and stretched resources, it is necessary to be able to identify those children and families who would maximally benefit from therapeutic interventions and to target these interventions appropriately. Increasing our understanding of adaptive coping responses and the factors that determine them will also enable professionals to understand the mechanisms of maladaptive coping responses.

A final requirement of any further research devised to plan and implement therapeutic interventions is the systematic evaluation of the efficacy and effectiveness of such interventions.

10.3.6. Clinical Implications of the Findings

10.3.6.1. Intervention

The diagnosis of congenital heart disease can be devastating for parents. The finding that families of affected children have more difficulties and experience more stress than those with other congenital problems (Emery, 1989) suggests that early therapeutic intervention should be specifically targeted towards families of newly diagnosed children. Early psychosocial intervention has been found to be effective for mothers of other groups of chronically ill children (Kupst, Schulman, Maurer, Morgan, Honig & Fochtman, 1983). Within this study, the finding of a relationship between parental mental state and adjustment of the child further supports early intervention with the parents, in order to try and alleviate some of their difficulties and prevent subsequent adjustment difficulties for the children themselves. The previously documented relationship between maternal anxiety, guilt and overprotective behaviour, and adjustment and anxiety in the child (Linde et al, 1971; Offord et al, 1972; Kong et al, 1986) also suggests that offering appropriate interventions to mothers, and in particular to those identified as being "at risk" of feeling extreme anxiety and/or guilt, in an effort to help them modify their responses, may have a direct, positive effect on the child.

The provision of routine, non-crisis oriented psychological help for chronically ill children and families is still rare (Eiser, 1993). Significant barriers to referral are contributed by families, medical staff and mental health providers (Sabbeth & Stein, 1990). However, intervention at an early stage can reduce later dysfunction, and thus the later burden on child, family and professionals. Furthermore, the provision of routine psychological support, rather than only at a time of crisis, will reduce the likelihood of adding to the child's and family's burden of being "different" and of feeling that they are being identified as failing to cope. If

families feel that such help is a concomitant of chronic illness and it is offered at the outset, they will hopefully be more able to accept and benefit from it.

A further, frequently overlooked, issue is the importance of recognising that, on top of disease related factors, the child and family also have normal, everyday problems with which to cope (McCubbin, Nevin, Cauble, Larsen, Comeau & Patterson, 1982). In the context of the disease itself, such day to day difficulties, such as problems with peer relationships, may be considered trivial by others, but they are as significant for the sick child as for the healthy child. Family conflict and marital problems are also part of daily life for healthy families, but too often families of sick children and professionals attribute any problem to the illness. Such an approach not only adds to the burden of the illness felt by the child, but can also preclude recognising and resolving difficulties in an appropriate manner. The results of this and other studies support the view that these are normal families dealing with abnormal circumstances.

If, as the results of our study indicate, the presence of pre-operative adjustment problems is predictive of later post-operative problems, particularly for patients with cyanotic lesions, there are clear implications for intervention at an early stage. Although the indications are that cardiac surgery has little impact on psychosocial functioning, resolution of pre-operative adjustment difficulties may enable children and families to maximise the benefits offered by corrective surgery.

10.3.6.2. Communication

As previously stated, it is imperative that a developmental framework is adopted in which to work with chronically ill children. This is also relevant in terms of the provision of

information and support. The findings in this study of a high degree of parental dissatisfaction with communication with staff and provision of counselling/support concur with earlier findings (e.g. Apley et al, 1967; Silverman, 1983) and highlight an area of need. Written information was rarely offered at any of the centres participating in our study and parents typically felt that information they did receive was not tailored to their requirements or emotional state. Not all of the children received preparation prior to surgery and in some instances little or no information had been given to them about any aspects of their treatment. It is well documented that information and preparation need to be tailored to the developmental level of the child, but this often did not happen.

A further aspect of communication concerns that between professionals. Medical and nursing staff need to become more aware of the specific concerns of children and families and the factors that are relevant to and for them. Whilst there is a change in the approach to the care of chronically ill children and families, with a multidisciplinary team approach now being more widely adopted, there is still scope for improved communication within such teams. The ward culture can have a major impact on children and families and influence their coping, although research has seldom included the psychological functioning of the professional group as a protective or risk factor (Mercer, 1994). Treatment of children in specialist centres involving collaborative care has been found to result in improvement in disease status (Stiller & Draper, 1989; Bloomfield & Farquhar, 1990) and such findings have implications for the child with congenital heart disease.

10.3.6.3. Community Liaison

In general there is a different perception of chronic illness in the community and in hospitals. Hospitals deal with diseases and give a disease-specific view, whilst communities focus on generic "chronic illness". Within a non-categorical framework many factors are relevant for the general population of chronically ill children, such as restrictions imposed by the disease and the need for hospital appointments. Whilst it is important to have general systems of care for such children and to be able to provide appropriate, community based services, there is also a need to recognise some of the unique aspects of congenital heart disease. The importance of recognising the salient characteristics of individual illnesses and their effects on children and families, rather than studying children as a composite "chronically ill" category has been previously acknowledged (Perrin et al, 1987; Lewis, Haiken & Hoyt, 1994; Mullins, Chaney, Hartman, Olson, Youll, Reyes & Blackett, 1995). The emotional significance attached to the heart and the popularly held belief that a heart problem will result in a heart attack and possible sudden death can result in some very specific difficulties for children with congenital heart disease. Fear and anxiety on the part of teachers can result in further isolation of such children and rejection by peers. For children with other chronic illnesses it has been suggested that schools may be overprotective or unwilling to take responsibility for the child (Lansdown & Goldman, 1988) and this is also likely to be relevant for children with congenital heart disease. A reluctance to encourage the children in case it precipitates an "attack" and a tendency to treat them as younger because of their small, frail stature, can add to the academic difficulties they already have. Although much of the research on the psychological effects of congenital heart disease has focused on IQ and academic performance, there seems to be little evidence of any progress in terms of recognition of such problems in school and the provision of appropriate help for these

children. The results of the current study corroborate previous findings of academic difficulties at school, with obvious implications for practical management. Schools clearly need to be more aware of the particular problems that may be associated with congenital heart disease and there needs to be greater involvement of, and support offered to, staff dealing with such children. Although information leaflets have been specifically prepared for teachers of children with congenital heart disease (Information for Teachers About Children with Heart Conditions - published by Heart Care), it seems that few teachers are routinely supplied with them if they have an affected child in their class. Obviously there is still a lot of scope for improving and increasing communication between health professionals and the community and for integrating the range of services available for chronically ill children - and in particular, for children with congenital heart disease - and their families.

APPENDICES

APPENDIX A

LOCUS OF CONTROL SCALE

1. Becoming a success is a matter of:

Hard Work	_ _ _ _	Luck

2. When things start to go wrong, they are often:

Things you do yourself	_ _ _ _	Things which are happening to you

3. I have the feeling that I have:

Very little influence	_ _ _ _	Very much influence

over the things which are happening to me.

4. The one who gets to be boss often depends on :

Who was lucky to be there in the first place	_ _ _ _	Who was the most capable person

5. As far as world affairs are concerned, we:

Are the victims of forces that we can neither understand nor control	_ _ _ _	Can control world events by taking an active part in political and social affairs

6. Generally, I feel that I have:

Very much control	_ _ _ _	Very little control

over the direction my life is taking.

UTRECHT COPING LIST

Each question is rated in terms of seldom/never, sometimes, often or very often.

Dealing with problems and unpleasant events:

Explanation

People in general have different ways of reacting when they are confronted with problems or unpleasant events.

One's reaction is often dependent on the nature and the severity of the problem, or on the moment at which the event occurs.

On the next pages you will find a number of descriptions which indicate different ways in which one can think or react when facing a problem.

Please indicate on each sentence how often you would find yourself reacting in this way. Do this by putting, for each sentence, a cross in one box.

Items

1. Telling oneself that things could be worse.
2. Trying to relax.
3. Isolating oneself totally from other people.
4. Showing one's annoyance.
5. Taking a gloomy view of the situation.
6. Becoming involved in other activities to keep your mind off the problem.
7. Showing one's anger with those responsible for the problem.
8. Giving-in in order to avoid difficult situations.
9. Resigning oneself to the situation.
10. Sharing one's worries with someone.
11. Making a direct intervention when problems occur.
12. Telling oneself that things will turn out right after all.

13. Considering a problem as a challenge.

14. Trying to dispel one's worries temporarily by taking a break.

15. Waiting to see what will happen.

16. Trying to reduce your tension by increased smoking, eating, drinking, physical exercise etc.

17. Looking for distraction.

18. Finding out all about the problem.

19. Trying to avoid difficult situations as much as possible.

20. Remaining optimistic about the future.

21. Trying to keep cool in difficult situations.

22. Considering different solutions to the problem.

23. Using a direct approach in order to solve the problem.

24. Worrying about the past.

25. Seeking cheerful company when one is worried or upset.

26. Trying to get away from the situation.

27. Working off one's tension.

28. Waiting for better times to come.

29. Asking someone to help.

30. Taking relaxing medication when one feels tense or upset.

31. Taking refuge in fantasies.

32. Making several alternative plans for handling a problem.

33. Being totally preoccupied with the problem.

34. Directing one's thoughts towards other matters.

35. Trying to make oneself feel better one way or the other.

36. Telling oneself that other people also have their problems from time to time.

37. Realising every cloud has a silver lining.

38. Showing one's feelings.

39. Seeking sympathy and comfort from somebody.

40. Letting the problem get on top of oneself.

41. Trying to see the humourous aspects of the problem.

42. Showing that there are things which are bothering you.

43. Discussing the problem with friends or family.

44. Letting things sort themselves out.

45. Not worrying, things usually work out fine.

46. Feeling unable to do anything.

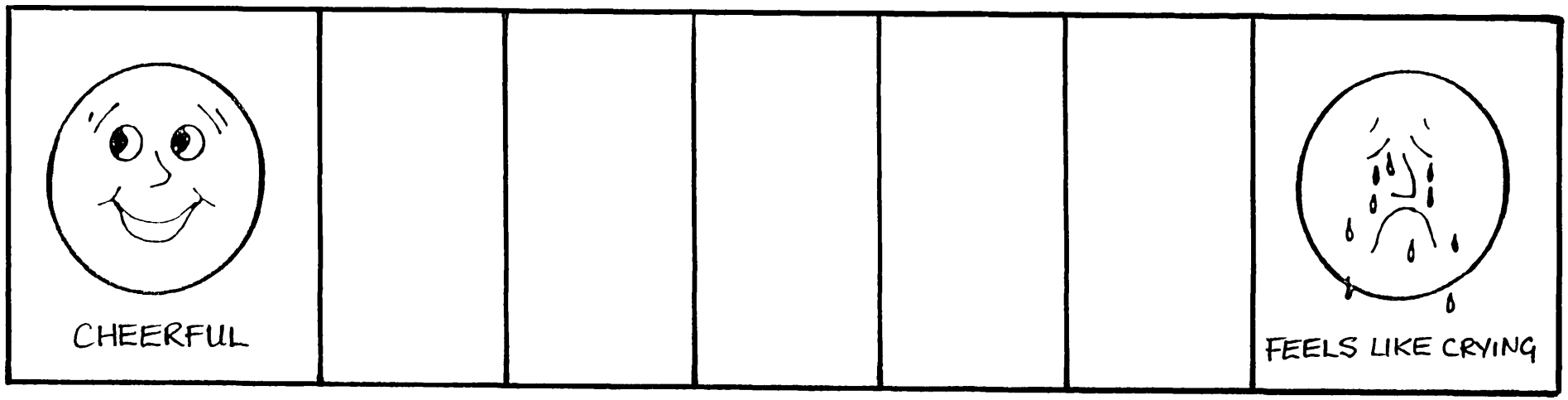
47. Taking courage into yourself when confronted with difficulties.

COPING SUBSCALES

SUBSCALE	ITEMS
Actively solving/approaching problem	11, 13, 18, 21, 22, 23, 32, 41
Palliative responses - arousal reduction, dealing with emotions, finding a temporary escape.	2, 6, 14, 16, 17, 34, 35, 47
Avoidance/waiting	8, 9, 12, 15, 19, 26, 40, 44, 45
Looking for social support	10, 29, 38, 39, 42, 43
Collapsing/depression; depressive reaction	3, 5, 24, 30, 31, 33, 46
Expression of emotions and anger	4, 7, 27
Use of comforting cognitions	1, 36, 37

Three items not part of any factor: 20, 25, 28

Fig. 13 An Example of One of the Constructs Used to Measure Self Perception



APPENDIX B

TABLE 1

PRE-OPERATIVE AGES OF CHILDREN IN EACH SUBGROUP PERFORMING EACH OF THE SUBTESTS AND SCHOOL ATTAINMENTS

	CYANOTIC	ACYANOTIC	t	p
RECALL OF DIGITS AGE: S.D. (YRS):	7.56 3.58 (n=16)	7.47 2.98 (n=29)	- .09	.926
VISUAL RECOGNITION AGE: S.D. (YRS):	3.94 .54 (n= 5)	4.26 .59 (n= 5)	- .89	.398
NAMING VOCABULARY AGE: S.D. (YRS):	5.01 1.28 (n=10)	5.69 1.08 (n=21)	-1.55	.132
VERBAL REASONING AGE: S.D. (YRS):	3.94 .54 (n= 5)	4.26 .59 (n= 5)	- .89	.398
MATRICES AGE: S.D. (YRS):	9.00 3.04 (n=12)	7.66 2.61 (n=24)	1.37	.179
SIMILARITIES AGE: S.D. (YRS):	9.00 3.04 (n=12)	7.66 2.61 (n=24)	1.37	.179
SPEED OF INFORMATION PROCESSING AGE: S.D. (YRS):	11.09 2.08 (n= 7)	11.17 2.55 (n= 9)	- .07	.947
BAS IQ AGE: S.D. (YRS):	7.51 3.47 (n=17)	7.33 3.02 (n=30)	.18	.854
ARITHMETIC AGE: S.D. (YRS):	7.76 3.42 (n=16)	7.08 2.71 (n=29)	.74	.463
READING AGE: S.D. (YRS):	9.00 3.04 (n=12)	7.66 2.61 (n=24)	1.37	.179
SPELLING AGE: S.D. (YRS):	9.00 3.04 (n=12)	7.66 2.61 (n=24)	1.37	.179

TABLE 2

25, 50 and 75 centiles

PRE-OPERATIVE PARENTAL INTERVIEW ITEMS: TOTAL SAMPLE				
	CYANOTIC (n=12)	ACYANOTIC (n=32)	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 1.50 2.00	1.00 2.00 2.00	179.0	.7055
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	184.0	.7229
DISOBEDIENCE	1.00 1.00 3.00	1.00 1.00 1.00	152.0	.1147
ANXIETIES AND WORRIES	1.00 2.00 2.75	1.00 1.50 2.00	148.0	.2046
RELATIONSHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 1.00	182.5	.7219
RELATIONSHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	183.0	.6907
DEPENDENCY	1.00 2.00 3.00	1.00 1.00 1.75	122.5	.0291
ACTIVITY	1.00 1.00 3.00	1.00 1.00 1.00	129.0	.0133

TABLE 3

25, 50 and 75 centiles.

PRE-OPERATIVE PARENTAL INTERVIEW ITEMS: CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 44)	BMT (n= 54)	H (n= 54)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 2.00 2.00	1.00 2.00 2.00	1.00 2.00 2.00	939.0	.3093	1072.5	.5627
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	1048.0	.4074	1098.0	.5398
DIS-OBEDIENCE	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	1006.0	.1324	1024.0	.1235
ANXIETIES AND WORRIES	1.00 2.00 2.00	1.00 1.00 2.00	1.00 1.00 1.00	962.0	.1832	725.0	.0002
RELATION-SHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 2.00	1.00 1.00 1.00	1036.0	.5181	1034.5	.2019
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	1034.0	.3496	1051.0	.1760
DEPENDENCY	1.00 1.00 2.00	1.00 1.00 2.00	1.00 1.00 1.00	944.5	.3813	817.0	.0004
ACTIVITY	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	977.0	.1970	974.0	.0183

TABLE 4

25, 50 and 75 centiles.

PRE-OPERATIVE PARENTAL INTERVIEW ITEMS: CYANOTIC, BMT AND HEALTHY GROUPS							
	CY (n=12)	BMT (n= 54)	H (n= 54)	MANN-WHITNEY ANOVA: CYANOTIC VS BMT		MANN-WHITNEY ANOVA: CYANOTIC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 1.50 2.00	1.00 2.00 2.00	1.00 2.00 2.00	273.0	.7570	309.0	.9547
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	296.0	.8040	290.0	.4840
DIS-OBEDIENCE	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	228.0	.0179	232.0	.0163
ANXIETIES AND WORRIES	1.00 2.00 2.75	1.00 1.00 2.00	1.00 1.00 1.00	216.0	.0779	159.5	.0004
RELATION-SHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 2.00	1.00 1.00 1.00	271.0	.4984	294.0	.5895
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	272.0	.3304	276.5	.1893
DEPENDENCY	1.00 2.00 3.00	1.00 1.00 2.00	1.00 1.00 1.00	180.0	.0216	145.5	.0000
ACTIVITY	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	191.5	.0039	190.0	.0001

TABLE 5

25, 50 and 75 centiles.

PRE-OPERATIVE PARENTAL INTERVIEW ITEMS: ACYANOTIC, BMT AND HEALTHY GROUPS							
	ACY (n= 32)	BMT (n= 54)	H (n= 54)	MANN-WHITNEY ANOVA: ACYANOTIC VS BMT		MANN-WHITNEY ANOVA: ACYANOTIC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 2.00 2.00	1.00 2.00 2.00	1.00 2.00 2.00	666.0	.2631	763.5	.4862
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	752.0	.3711	808.0	.6799
DIS-OBEDIENCE	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	778.0	.4868	792.0	.4687
ANXIETIES AND WORRIES	1.00 1.50 2.00	1.00 1.00 2.00	1.00 1.00 1.00	746.0	.4592	565.5	.0017
RELATION-SHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 2.00	1.00 1.00 1.00	765.0	.6624	740.5	.1798
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	762.0	.4824	774.5	.2688
DEPENDENCY	1.00 1.00 1.75	1.00 1.00 2.00	1.00 1.00 1.00	739.5	.8717	671.5	.0116
ACTIVITY	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	782.5	.9777	784.0	.2809

TABLE 6

25, 50 and 75 centiles.

PRE-OPERATIVE SELF PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n=11)	ACYANOTIC (n=20)	U	p
WEAK-STRONG	1.00 1.00 5.00	1.25 3.00 4.00	102.0	.7320
CRYING-CHEERFUL	1.00 5.00 5.00	3.50 5.00 5.00	90.0	.3001
DOESN'T LIKE SELF-LIKES SELF	1.00 5.00 5.00	4.00 5.00 5.00	104.0	.7769
LONELY-HAS FRIENDS	3.00 5.00 5.00	4.00 5.00 5.00	109.0	.9588
FRIGHTENED-SAFE	1.00 2.00 5.00	2.25 5.00 5.00	81.0	.1937
ANGRY-CALM	5.00 5.00 5.00	3.25 5.00 5.00	95.0	.4200
BAD-GOOD	1.00 5.00 5.00	3.00 4.50 5.00	103.5	.7702
ILL-WELL	1.00 1.00 5.00	1.25 3.50 5.00	79.0	.1783
TOTAL SCORE	22.00 28.00 32.00	27.25 32.00 35.00	78.0	.1855

TABLE 7

25, 50 and 75 centiles.

PRE-OPERATIVE "IDEAL" SELF PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n=11)	ACYANOTIC (n=20)	U	p
WEAK-STRONG	5.00 5.00 5.00	3.25 5.00 5.00	93.5	.3913
CRYING-CHEERFUL	5.00 5.00 5.00	5.00 5.00 5.00	98.5	.4574
DOESN'T LIKE SELF-LIKES SELF	5.00 5.00 5.00	3.25 5.00 5.00	86.5	.1843
LONELY-HAS FRIENDS	5.00 5.00 5.00	5.00 5.00 5.00	103.5	.6450
FRIGHTENED-SAFE	5.00 5.00 5.00	5.00 5.00 5.00	103.5	.6450
ANGRY-CALM	5.00 5.00 5.00	5.00 5.00 5.00	88.0	.1189
BAD-GOOD	3.00 5.00 5.00	5.00 5.00 5.00	92.0	.3311
ILL-WELL	3.00 5.00 5.00	5.00 5.00 5.00	96.0	.4011
TOTAL SCORE	36.00 38.00 40.00	34.50 38.00 40.00	97.0	.5787

TABLE 8

25, 50 and 75 centiles.

PRE-OPERATIVE DATA: DIFFERENCE BETWEEN CHILD'S SELF AND IDEAL SELF PERCEPTION SCORES: TOTAL SAMPLE

	CYANOTIC (n=11)	ACYANOTIC (n=20)	U	p
WEAK-STRONG	-4.00 -2.00 .00	-3.75 -1.50 - .25	102.5	.7513
CRYING-CHEERFUL	-2.00 .00 .00	-1.50 .00 .00	78.5	.1440
DOESN'T LIKE SELF-LIKES SELF	-4.00 .00 .00	-1.00 .00 .00	101.5	.7081
LONELY-HAS FRIENDS	.00 .00 .00	-1.00 .00 .00	105.5	.8277
FRIGHTENED-SAFE	-4.00 -2.00 .00	-2.75 .00 .00	81.0	.2065
ANGRY-CALM	.00 .00 .00	-1.50 .00 .00	102.5	.7169
BAD-GOOD	-2.00 .00 .00	-2.00 .00 .00	101.5	.7081
ILL-WELL	-4.00 -2.00 .00	-3.75 - .50 .00	88.0	.3461
TOTAL SCORE	-14.00 -10.00 - 5.00	-11.75 - 5.00 .00	76.5	.1660

TABLE 9

25, 50 and 75 centiles

PRE-OPERATIVE "IDEAL" SELF PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 31)	BMT (n= 41)	H (n= 44)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	P	U	P
WEAK-STRONG	4.00 5.00 5.00	4.50 5.00 5.00	5.00 5.00 5.00	545.5	.6533	610.5	.2922
CRYING-CHEERFUL	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	584.0	.9245	650.0	.7698
DOESN'T LIKE SELF-LIKES SELF	5.00 5.00 5.00	4.00 5.00 5.00	5.00 5.00 5.00	535.5	.5527	639.5	.6730
LONELY-HAS FRIENDS	5.00 5.00 5.00	4.75 5.00 5.00	5.00 5.00 5.00	521.0	.2287	655.0	.8320
FRIGHT-ENED-SAFE	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	586.0	.9507	597.5	.0823
ANGRY-CALM	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	522.5	.3440	627.0	.4999
BAD-GOOD	3.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	537.5	.9317	606.5	.3606
ILL-WELL	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	554.5	.9491	653.0	.8233
TOTAL SCORE	26.00 31.00 35.00	25.25 31.00 36.00	32.00 34.50 38.75	523.0	.6592	427.5	.0060

TABLE 10

25, 50 and 75 centiles.

PRE-OPERATIVE PARENTAL PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n=12)	ACYANOTIC (n=30)	U	p
WEAK-STRONG	3.00 3.00 4.75	3.00 4.00 5.00	128.5	.1329
CRYING-CHEERFUL	4.25 5.00 5.00	4.00 5.00 5.00	165.0	.6085
DOESN'T LIKE SELF-LIKES SELF	3.00 4.00 5.00	4.00 5.00 5.00	144.5	.2777
LONELY-HAS FRIENDS	2.25 4.00 5.00	4.00 5.00 5.00	119.0	.0609
FRIGHTENED-SAFE	3.00 4.00 5.00	4.00 5.00 5.00	142.0	.2473
ANGRY-CALM	3.00 4.00 4.00	3.00 3.00 4.00	145.5	.2961
BAD-GOOD	3.25 4.00 4.75	3.00 4.00 5.00	180.0	1.0000
ILL-WELL	3.00 3.00 3.75	3.00 4.00 5.00	116.5	.0646
TOTAL SCORE	28.00 29.50 35.00	30.00 33.00 36.00	123.0	.1104
PERSONALITY	3.00 3.50 4.00	3.00 3.00 4.00	184.0	.9538

APPENDIX C

TABLE 11

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL INTERVIEW ITEMS: TOTAL SAMPLE				
	CYANOTIC (n=14)	ACYANOTIC (n=25)	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 2.00 2.00	1.00 1.00 2.00	137.5	.2091
TEMPER TANTRUMS	1.00 1.00 3.00	1.00 1.00 1.00	132.0	.0298
DISOBEDIENCE	1.00 1.00 3.00	1.00 1.00 3.00	174.0	.9700
ANXIETIES AND WORRIES	1.00 2.00 3.00	1.00 1.00 1.50	97.5	.0197
RELATIONSHIPS WITH CHILDREN	1.00 1.00 2.00	1.00 1.00 1.00	125.5	.0726
RELATIONSHIPS WITH ADULTS	1.00 1.00 1.25	1.00 1.00 1.00	150.5	.2163
DEPENDENCY	1.00 1.00 3.00	1.00 1.00 1.00	124.0	.0505
ACTIVITY	1.00 1.00 1.25	1.00 1.00 1.00	137.5	.0175

TABLE 12

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL INTERVIEW ITEMS: CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 39)	BMT (n= 32)	H (n= 41)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 1.00 2.00	1.00 1.00 1.00	1.00 1.00 1.50	411.5	.0192	653.0	.0885
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	602.5	.6501	736.0	.2117
DIS-OBEDIENCE	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	487.0	.0181	607.5	.0039
ANXIETIES AND WORRIES	1.00 1.00 2.00	1.00 1.00 2.00	1.00 1.00 1.00	566.0	.5719	577.0	.0068
RELATION-SHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 2.00	1.00 1.00 1.00	533.0	.3352	673.5	.0357
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	544.0	.0371	735.0	.2049
DEPENDENCY	1.00 1.00 2.00	1.00 1.00 1.00	1.00 1.00 1.00	543.0	.1765	663.5	.0477
ACTIVITY	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	594.5	.3936	793.5	.8993

TABLE 13

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL INTERVIEW ITEMS: CYANOTIC, BMT AND HEALTHY GROUPS							
	CY (n= 14)	BMT (n= 32)	H (n= 41)	MANN-WHITNEY ANOVA: CYANOTIC VS BMT		MANN WHITNEY ANOVA: CYANOTIC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 2.00 2.00	1.00 1.00 1.00	1.00 1.00 1.50	119.5	.0068	196.0	.0324
TEMPER TANTRUMS	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	181.0	.0990	219.0	.0150
DIS-OBEDIENCE	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	174.0	.0408	217.0	.0123
ANXIETIES AND WORRIES	1.00 2.00 3.00	1.00 1.00 2.00	1.00 1.00 1.00	167.0	.2617	132.5	.0002
RELATION-SHIPS WITH CHILDREN	1.00 1.00 2.00	1.00 1.00 2.00	1.00 1.00 1.00	189.5	.6927	190.0	.0021
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.25	1.00 1.00 1.00	1.00 1.00 1.00	176.0	.0074	238.5	.0601
DEPENDENCY	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	155.0	.0218	185.0	.0033
ACTIVITY	1.00 1.00 1.25	1.00 1.00 1.00	1.00 1.00 1.00	182.0	.0402	243.5	.1202

TABLE 14

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL INTERVIEW ITEMS: ACYANOTIC, BMT AND HEALTHY GROUPS							
	ACY (n= 25)	BMT (n= 32)	H (n= 41)	MANN-WHITNEY ANOVA: ACYANOTIC VS BMT		MANN WHITNEY ANOVA: ACYANOTIC VS HEALTHY	
				U	p	U	p
BEHAVIOUR IN NEW SITUATIONS	1.00 1.00 2.00	1.00 1.00 1.00	1.00 1.00 1.50	292.0	.1082	457.0	.3533
TEMPER TANTRUMS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	378.5	.4346	508.0	.8691
DIS-OBEDIENCE	1.00 1.00 3.00	1.00 1.00 1.00	1.00 1.00 1.00	313.0	.0268	390.5	.0067
ANXIETIES AND WORRIES	1.00 1.00 1.50	1.00 1.00 2.00	1.00 1.00 1.00	317.0	.1143	444.5	.1492
RELATION-SHIPS WITH CHILDREN	1.00 1.00 1.00	1.00 1.00 2.00	1.00 1.00 1.00	319.5	.0927	483.5	.2882
RELATION-SHIPS WITH ADULTS	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	368.0	.1064	496.5	.6089
DEPENDENCY	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	388.0	.7491	478.5	.4275
ACTIVITY	1.00 1.00 1.00	1.00 1.00 1.00	1.00 1.00 1.00	387.5	.3768	475.0	.1695

TABLE 15

25, 50 and 75 centiles

POST-OPERATIVE SELF PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n= 7)	ACYANOTIC (n=22)	U	p
WEAK-STRONG	5.00 5.00 5.00	3.00 3.00 5.00	39.0	.0365
CRYING-CHEERFUL	5.00 5.00 5.00	4.00 5.00 5.00	56.0	.1297
DOESN'T LIKE SELF-LIKES SELF	1.00 5.00 5.00	3.00 4.50 5.00	75.5	.9338
LONELY-HAS FRIENDS	1.00 5.00 5.00	4.75 5.00 5.00	67.5	.5180
FRIGHTENED-SAFE	1.00 4.00 5.00	4.00 5.00 5.00	52.0	.1511
ANGRY-CALM	1.00 5.00 5.00	3.75 5.00 5.00	75.5	.9299
BAD-GOOD	1.00 5.00 5.00	3.00 4.00 5.00	73.0	.8257
ILL-WELL	4.00 5.00 5.00	3.75 5.00 5.00	75.0	.9009
TOTAL SCORE	20.00 33.00 40.00	29.50 35.00 38.00	72.5	.8169

TABLE 16

25, 50 and 75 centiles

POST-OPERATIVE "IDEAL" SELF PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n= 6)	ACYANOTIC (n=22)	U	p
WEAK-STRONG	5.00 5.00 5.00	4.75 5.00 5.00	51.0	.2058
CRYING-CHEERFUL	5.00 5.00 5.00	5.00 5.00 5.00	57.0	.3479
DOESN'T LIKE SELF-LIKES SELF	5.00 5.00 5.00	5.00 5.00 5.00	57.0	.3481
LONELY-HAS FRIENDS	5.00 5.00 5.00	5.00 5.00 5.00	54.0	.2695
FRIGHTENED-SAFE	4.75 5.00 5.00	5.00 5.00 5.00	61.5	.6388
ANGRY-CALM	3.25 5.00 5.00	4.75 5.00 5.00	57.0	.5056
BAD-GOOD	3.25 5.00 5.00	5.00 5.00 5.00	55.5	.4122
ILL-WELL	5.00 5.00 5.00	5.00 5.00 5.00	63.0	.6015
TOTAL SCORE	35.75 39.00 40.00	37.00 40.00 40.00	57.5	.5974

TABLE 17

25, 50 and 75 centiles

POST-OPERATIVE DATA: DIFFERENCE BETWEEN CHILD'S SELF AND IDEAL SELF PERCEPTION SCORES: TOTAL SAMPLE				
	CYANOTIC (n=6)	ACYANOTIC (n=22)	U	p
WEAK-STRONG	-1.00 .00 .00	-2.00 -1.00 .00	38.5	.1031
CRYING-CHEERFUL	.00 .00 .00	- .25 .00 .00	57.0	.5072
DOESN'T LIKE SELF-LIKES SELF	-1.75 .00 .00	-1.00 .00 .00	62.5	.8297
LONELY-HAS FRIENDS	-1.00 .00 .00	.00 .00 .00	60.0	.6580
FRIGHTENED-SAFE	-4.00 .00 .00	-1.00 .00 .00	59.0	.6543
ANGRY-CALM	-1.00 .00 1.75	-1.00 .00 .00	46.5	.2324
BAD-GOOD	-2.50 .00 .25	-2.00 - .50 .00	59.5	.6972
ILL-WELL	- .25 .00 .00	-1.25 .00 .00	56.5	.4839
TOTAL SCORE	-1.25 2.50 11.50	.00 3.50 8.25	63.5	.8879

TABLE 18

25, 50 and 75 centiles

POST-OPERATIVE SELF PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 29)	BMT (n= 24)	H (n= 34)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	3.00 3.00 5.00	3.00 5.00 5.00	3.00 4.00 5.00	312.0	.6692	489.5	.9595
CRYING-CHEERFUL	5.00 5.00 5.00	5.00 5.00 5.00	4.00 5.00 5.00	309.0	.2627	379.5	.0595
DOESN'T LIKE SELF-LIKES SELF	3.00 5.00 5.00	4.00 5.00 5.00	3.00 4.50 5.00	293.0	.2636	469.0	.7197
LONELY-HAS FRIENDS	4.50 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	339.5	.8356	474.0	.7183
FRIGHT-ENED-SAFE	4.00 5.00 5.00	5.00 5.00 5.00	4.00 5.00 5.00	251.5	.0297	422.0	.2468
ANGRY-CALM	3.50 5.00 5.00	5.00 5.00 5.00	3.00 4.00 5.00	264.5	.0540	394.5	.1418
BAD-GOOD	3.00 4.00 5.00	3.25 5.00 5.00	3.00 4.00 5.00	283.5	.1984	482.5	.8761
ILL-WELL	4.00 5.00 5.00	5.00 5.00 5.00	4.00 5.00 5.00	259.0	.0543	465.5	.6337
TOTAL SCORE	29.00 34.00 39.00	34.00 38.00 40.00	30.75 33.50 37.00	246.0	.1596	458.0	.6278

TABLE 19

25, 50 and 75 centiles

POST-OPERATIVE "IDEAL" SELF PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 28)	BMT (n= 24)	H (n= 34)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	299.5	.5382	474.5	.9745
CRYING-CHEERFUL	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	316.0	.5358	442.5	.4389
DOESN'T LIKE SELF-LIKES SELF	5.00 5.00 5.00	5.00 5.00 5.00	4.00 5.00 5.00	332.0	.8947	405.0	.1447
LONELY-HAS FRIENDS	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	330.0	.8527	423.5	.1158
FRIGHT-ENED-SAFE	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	315.0	.4041	452.0	.4719
ANGRY-CALM	4.25 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	265.0	.0376	400.0	.0926
BAD-GOOD	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	292.5	.2032	438.5	.4250
ILL-WELL	5.00 5.00 5.00	5.00 5.00 5.00	5.00 5.00 5.00	319.5	.8881	436.5	.2365
TOTAL SCORE	37.00 40.00 40.00	38.00 40.00 40.00	37.75 40.00 40.00	287.5	.6543	468.0	.9013

TABLE 20

25, 50 and 75 centiles

POST-OPERATIVE DATA: DIFFERENCE BETWEEN CHILD'S SELF AND IDEAL SELF PERCEPTION SCORES: COMPARISON BETWEEN CARDIAC, BMT AND HEALTHY GROUPS

	C (n= 28)	BMT (n= 24)	H (n= 34)	MANN WHITNEY ANOVA: CARDIAC VS BMT		MANN WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	-2.00 -1.00 .00	-2.00 .00 .00	-2.00 -1.00 .00	263.0	.2337	474.5	.9823
CRYING-CHEERFUL	.00 .00 .00	.00 .00 .00	-1.00 .00 .00	284.0	.2091	397.5	.1973
DOESN'T LIKE SELF-LIKES SELF	-1.00 .00 .00	- .75 .00 .00	-1.00 .00 .00	287.5	.3144	451.0	.6914
LONELY-HAS FRIENDS	.00 .00 .00	.00 .00 .00	.00 .00 .00	323.0	.7464	457.0	.7132
FRIGHT-ENED-SAFE	-1.00 .00 .00	.00 .00 .00	-1.00 .00 .00	264.5	.1004	432.5	.4631
ANGRY-CALM	-1.00 .00 .00	.00 .00 .00	-2.00 -1.00 .00	310.5	.5739	338.5	.0394
BAD-GOOD	-2.00 .00 .00	-1.75 .00 .00	-2.00 -1.00 .00	301.5	.4912	459.0	.7986
ILL-WELL	- .75 .00 .00	.00 .00 .00	.00 .00 .00	259.0	.0851	435.0	.4504
TOTAL SCORE	.00 3.50 8.75	.00 1.50 6.00	2.00 4.50 8.00	248.0	.2360	434.0	.5507

TABLE 21

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL PERCEPTION: TOTAL SAMPLE				
	CYANOTIC (n=14)	ACYANOTIC (n=25)	U	p
WEAK-STRONG	2.75 3.00 4.00	3.00 4.00 5.00	117.0	.0770
CRYING-CHEERFUL	3.00 4.00 5.00	4.00 5.00 5.00	138.5	.2401
DOESN'T LIKE SELF-LIKES SELF	4.00 5.00 5.00	4.00 4.00 5.00	157.0	.8522
LONELY-HAS FRIENDS	3.00 4.50 5.00	4.00 5.00 5.00	126.5	.1018
FRIGHTENED-SAFE	3.00 4.50 5.00	4.00 5.00 5.00	153.0	.4795
ANGRY-CALM	3.00 3.50 5.00	3.00 3.00 4.00	162.0	.6860
BAD-GOOD	3.75 4.00 5.00	3.00 4.00 5.00	157.0	.5791
ILL-WELL	3.00 4.00 5.00	4.00 5.00 5.00	120.5	.0754
TOTAL SCORE	27.50 29.00 37.50	30.50 33.00 36.00	136.5	.4219
PERSONALITY	2.00 3.00 4.00	3.00 3.00 4.00	153.0	.4958

TABLE 22

25, 50 and 75 centiles

POST-OPERATIVE PARENTAL PERCEPTION: COMPARISON OF CARDIAC, BMT AND HEALTHY GROUPS							
	C (n= 39)	BMT (n= 29)	H (n= 41)	MANN-WHITNEY ANOVA: CARDIAC VS BMT		MANN-WHITNEY ANOVA: CARDIAC VS HEALTHY	
				U	p	U	p
WEAK-STRONG	3.00 4.00 5.00	3.00 4.00 5.00	3.50 4.00 5.00	531.5	.6594	640.0	.1056
CRYING-CHEERFUL	4.00 5.00 5.00	4.00 4.00 5.00	4.00 4.00 5.00	528.5	.6168	758.0	.6634
DOESN'T LIKE SELF-LIKES SELF	4.00 4.50 5.00	3.00 5.00 5.00	4.00 5.00 5.00	542.5	.9060	720.5	.5222
LONELY-HAS FRIENDS	4.00 5.00 5.00	4.00 5.00 5.00	4.00 5.00 5.00	534.5	.6610	779.5	.8231
FRIGHT-ENED-SAFE	4.00 5.00 5.00	4.00 4.00 5.00	3.50 4.00 5.00	554.5	.8818	762.5	.6997
ANGRY-CALM	3.00 3.00 4.00	3.00 4.00 4.00	3.00 4.00 5.00	564.0	.9842	614.0	.0592
BAD-GOOD	3.00 4.00 5.00	3.00 4.00 5.00	3.00 4.00 4.00	546.5	.8040	753.0	.6344
ILL-WELL	4.00 5.00 5.00	4.50 5.00 5.00	4.00 5.00 5.00	442.5	.0720	705.5	.2982
TOTAL SCORE	29.00 33.00 37.00	29.50 34.00 36.00	31.00 34.00 37.00	549.5	.9848	707.5	.4817
PERSON-ALITY	3.00 3.00 4.00	3.00 4.00 5.00	3.00 4.00 4.00	440.0	.1055	646.5	.1205

APPENDIX D

TABLE 23

ITEMS OF THE RICHMAN SCALE: PRE-OPERATIVE FREQUENCIES					
	CY (n=2)	ACY (n=6)	CARDIAC (n=8)	BMT (n=5)	HEALTHY (n=8)
Poor appetite	1 (50%)	5 (83%)	6 (75%)	3 (60%)	2 (25%)
Faddy with eating	0 (0%)	6 (100%)	6 (75%)	4 (80%)	6 (75%)
Wets the bed at night	0 (0%)	3 (50%)	3 (38%)	1 (20%)	1 (13%)
Wets during the day	0 (0%)	0 (0%)	0 (0%)	1 (20%)	0 (0%)
Soils pants	0 (0%)	1 (17%)	1 (13%)	2 (40%)	0 (0%)
Difficulties with settling at bedtime	2 (100%)	4 (67%)	6 (75%)	1 (40%)	2 (25%)
Difficulties with waking at night	2 (100%)	4 (67%)	6 (75%)	4 (80%)	3 (38%)
Sleeps with parent because upset or won't sleep alone	1 (50%)	4 (67%)	5 (63%)	4 (80%)	2 (25%)
Not active enough/too active	0 (0%)	0 (0%)	0 (0%)	1 (20%)	1 (13%)
Difficulties with concentration during play	1 (50%)	2 (33%)	3 (38%)	0 (0%)	2 (25%)
Clingy	0 (0%)	3 (50%)	3 (38%)	2 (40%)	2 (25%)
Demands a lot of attention	1 (50%)	2 (33%)	3 (38%)	3 (60%)	3 (38%)
Difficult to manage or control	1 (50%)	2 (33%)	3 (38%)	4 (80%)	1 (13%)
Problems with temper tantrums	1 (50%)	5 (83%)	6 (75%)	4 (80%)	4 (50%)
Miserable/irritable	1 (50%)	1 (17%)	2 (25%)	2 (40%)	0 (0%)
Worried	2 (100%)	0 (0%)	2 (25%)	2 (40%)	3 (38%)
Fearful	2 (100%)	1 (17%)	3 (38%)	0 (0%)	1 (13%)
Difficulties with brothers or sisters	0 (0%)	1 (17%)	1 (13%)	0 (0%)	3 (38%)
Difficulties playing with other children	0 (0%)	1 (17%)	1 (13%)	0 (0%)	0 (0%)

TABLE 24

ITEMS OF THE RUTTER A SCALE: PRE-OPERATIVE FREQUENCIES					
	CY (n=8)	ACY (n=21)	CARDIAC (n=29)	BMT (n=40)	HEALTHY (n=42)
Complained of headaches	3 (38%)	11 (52%)	14 (48%)	19 (48%)	28 (67%)
Stomach ache or vomiting	5 (63%)	9 (43%)	14 (48%)	22 (55%)	32 (76%)
Asthma or attacks of wheezing	0 (0%)	2 (10%)	2 (7%)	6 (15%)	2 (5%)
Wet the bed or pants	0 (0%)	5 (24%)	5 (17%)	6 (15%)	8 (19%)
Soiled or lost control of bowels	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (2%)
Temper tantrums (complete loss of temper, with shouting, angry movements, etc.)	2 (25%)	11 (52%)	13 (45%)	16 (40%)	18 (43%)
Tears on arrival at school or refused to go into school building	1 (13%)	4 (19%)	5 (17%)	5 (13%)	5 (12%)
Truanted from school	0 (0%)	0 (0%)	0 (0%)	0 (0%)	4 (10%)
Stammer or stutter	0 (0%)	1 (5%)	1 (3%)	1 (3%)	2 (5%)
Any difficulty with speech, apart from stammering or stuttering	0 (0%)	2 (10%)	2 (7%)	1 (3%)	2 (5%)
Did he/she ever steal things?	0 (0%)	0 (0%)	0 (0%)	0 (0%)	7 (17%)
Was there any eating difficulty?	3 (38%)	8 (38%)	11 (38%)	20 (50%)	10 (24%)
Was there any sleeping difficulty?	0 (0%)	7 (33%)	7 (24%)	11 (28%)	12 (29%)
Very restless, had difficulty staying seated for long	3 (38%)	7 (33%)	10 (35%)	16 (40%)	17 (41%)
Squirmy, fidgety child	2 (25%)	7 (33%)	9 (31%)	6 (15%)	12 (29%)
Often destroyed own or others' property	0 (0%)	1 (5%)	1 (3%)	2 (5%)	5 (12%)
Frequently fought or extremely quarrelsome with other children	0 (0%)	3 (14%)	3 (10%)	5 (13%)	14 (33%)

ITEMS OF THE RUTTER A SCALE: PRE-OPERATIVE FREQUENCIES (contd.)					
	CY (n=8)	ACY (n=21)	CARDIAC (n=29)	BMT (n=40)	HEALTHY (n=42)
Not much liked by other children	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (2%)
Often worried, worried about many things	3 (38%)	10 (48%)	13 (45%)	20 (50%)	21 (50%)
Tended to be on own, rather solitary	3 (38%)	3 (14%)	6 (21%)	18 (45%)	8 (19%)
Irritable, quick to fly off the handle	1 (13%)	11 (52%)	12 (41%)	13 (33%)	13 (31%)
Often appeared miserable, tearful or distressed	0 (0%)	6 (29%)	6 (21%)	8 (20%)	7 (17%)
Had twitches, mannerisms, tics of face or body	0 (0%)	2 (10%)	2 (7%)	1 (3%)	1 (2%)
Frequently sucked thumb or fingers	1 (13%)	4 (19%)	5 (17%)	15 (38%)	9 (21%)
Frequently bit nails or fingers	2 (25%)	5 (24%)	7 (24%)	10 (25%)	9 (21%)
Often disobedient	1 (13%)	8 (38%)	9 (31%)	11 (28%)	20 (48%)
Could not settle to anything for more than a few moments	2 (25%)	6 (29%)	8 (28%)	10 (25%)	8 (19%)
Tended to be fearful or afraid of new things or situations	5 (63%)	5 (24%)	10 (35%)	15 (38%)	13 (31%)
Fussy or over-particular child	1 (13%)	7 (33%)	8 (28%)	13 (33%)	12 (29%)
Often told lies	0 (0%)	3 (14%)	3 (10%)	4 (10%)	12 (29%)
Bullied other children	0 (0%)	1 (5%)	1 (3%)	3 (8%)	2 (5%)

TABLE 25

ITEMS OF THE RUTTER B SCALE: PRE-OPERATIVE FREQUENCIES					
	CY (n=9)	ACY (n=24)	CARDIAC (n=33)	BMT (n=30)	HEALTHY (n=44)
Very restless, had difficulty staying seated for long	3 (33%)	4 (17%)	7 (21%)	7 (23%)	10 (23%)
Truanted from school	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (2%)
Squirmy, fidgety child	3 (33%)	6 (25%)	9 (27%)	6 (20%)	7 (16%)
Often destroyed or damaged own or others' property	0 (0%)	1 (4%)	1 (3%)	1 (3%)	0 (0%)
Frequently fought or extremely quarrelsome with other children	0 (0%)	3 (13%)	3 (9%)	3 (10%)	5 (11%)
Not much liked by other children	1 (11%)	3 (13%)	4 (12%)	4 (13%)	3 (7%)
Often worried, worried about many things	4 (44%)	12 (50%)	16 (49%)	14 (47%)	14 (32%)
Tended to be on own, rather solitary	3 (33%)	5 (21%)	8 (24%)	7 (23%)	9 (21%)
Irritable, touchy, quick to fly off the handle	0 (0%)	0 (0%)	0 (0%)	4 (13%)	7 (16%)
Often appeared miserable, tearful or distressed	2 (22%)	5 (21%)	7 (21%)	8 (27%)	6 (14%)
Had twitches, mannerisms, ties of face or body	2 (22%)	2 (8%)	4 (12%)	3 (10%)	1 (2%)
Frequently sucked thumb or fingers	0 (0%)	1 (4%)	1 (3%)	3 (10%)	3 (7%)
Frequently bit nails or fingers	1 (11%)	1 (4%)	2 (6%)	5 (17%)	3 (7%)
Tended to be absent from school for trivial reasons	1 (11%)	3 (13%)	4 (12%)	2 (7%)	9 (21%)
Often disobedient	2 (22%)	3 (13%)	5 (15%)	6 (20%)	11 (25%)
Could not settle	3 (33%)	6 (25%)	9 (27%)	9 (30%)	10 (23%)

ITEMS OF THE RUTTER B SCALE: PRE-OPERATIVE FREQUENCIES (contd.)					
	CY (n=9)	ACY (n=24)	CARDIAC (n=33)	BMT (n=30)	HEALTHY (n=44)
Tended to be fearful or afraid of new things or situations	2 (22%)	9 (38%)	11 (33%)	9 (30%)	8 (18%)
Fussy or over-particular child	4 (44%)	1 (4%)	5 (15%)	10 (33%)	12 (27%)
Often told lies	0 (0%)	1 (4%)	1 (3%)	2 (7%)	3 (7%)
Has stolen things on one or more occasions in the past 12 months	1 (11%)	0 (0%)	1 (3%)	0 (0%)	3 (7%)
Unresponsive, inert or apathetic	3 (33%)	5 (21%)	8 (24%)	9 (30%)	7 (16%)
Often complained of aches or pains	4 (44%)	2 (8%)	6 (18%)	8 (27%)	4 (9%)
Has had tears on arrival at school or has refused to come into the building in the past 12 months	1 (11%)	2 (8%)	3 (9%)	4 (13%)	1 (2%)
Has a stutter or stammer	0 (0%)	2 (8%)	2 (6%)	1 (3%)	2 (5%)
Resentful or aggressive when corrected	0 (0%)	2 (8%)	2 (6%)	5 (17%)	6 (14%)
Bullied other children	0 (0%)	0 (0%)	0 (0%)	0 (0%)	4 (9%)

TABLE 26

ITEMS OF THE RICHMAN SCALE: POST-OPERATIVE FREQUENCIES					
	CY (n=5)	ACY (n=3)	CARDIAC (n=8)	BMT (n=8)	HEALTHY (n=7)
Poor appetite	2 (40%)	2 (67%)	4 (50%)	1 (13%)	4 (57%)
Faddy with eating	3 (60%)	3 (100%)	6 (75%)	5 (63%)	6 (86%)
Wets the bed at night	1 (20%)	1 (33%)	2 (25%)	3 (38%)	1 (14%)
Wets during the day	2 (40%)	0 (0%)	2 (25%)	1 (13%)	0 (0%)
Soils pants	0 (0%)	0 (0%)	0 (0%)	1 (13%)	0 (0%)
Difficulties with settling at bedtime	2 (40%)	3 (100%)	5 (63%)	3 (38%)	3 (43%)
Difficulties with waking at night	2 (40%)	3 (100%)	5 (63%)	7 (88%)	6 (86%)
Sleeps with parent because upset or won't sleep alone	2 (40%)	2 (67%)	4 (50%)	6 (75%)	3 (43%)
Not active enough/too active	2 (40%)	0 (0%)	2 (25%)	1 (13%)	1 (14%)
Difficulties with concentration during play	1 (20%)	0 (0%)	1 (13%)	2 (25%)	3 (43%)
Clingy	1 (20%)	0 (0%)	1 (13%)	2 (25%)	2 (29%)
Demands a lot of attention	2 (40%)	0 (0%)	2 (25%)	2 (25%)	2 (29%)
Difficult to manage or control	1 (20%)	2 (67%)	3 (38%)	6 (75%)	4 (57%)
Problems with temper tantrums	3 (60%)	2 (67%)	5 (63%)	6 (75%)	5 (71%)
Miserable/irritable	1 (20%)	0 (0%)	1 (13%)	1 (13%)	1 (14%)
Worried	4 (80%)	1 (33%)	5 (63%)	4 (50%)	1 (14%)
Fearful	3 (60%)	1 (33%)	4 (50%)	1 (13%)	0 (0%)
Difficulties with brothers or sisters	0 (0%)	2 (67%)	2 (25%)	2 (25%)	0 (0%)
Difficulties playing with other children	0 (0%)	1 (33%)	1 (13%)	0 (0%)	1 (14%)

TABLE 27

ITEMS OF THE RUTTER A SCALE: POST-OPERATIVE FREQUENCIES					
	CY (n=8)	ACY (n=22)	CARDIAC (n=30)	BMT (n=24)	HEALTHY (n=31)
Complained of headaches	4 (50%)	13 (59%)	17 (57%)	12 (50%)	20 (65%)
Stomach ache or vomiting	3 (38%)	7 (32%)	10 (33%)	13 (54%)	16 (52%)
Asthma or attacks of wheezing	1 (13%)	3 (14%)	4 (13%)	3 (13%)	5 (16%)
Wet the bed or pants	1 (13%)	5 (23%)	6 (20%)	2 (8%)	6 (19%)
Soiled or lost control of bowels	0 (0%)	1 (5%)	1 (3%)	2 (8%)	2 (6%)
Temper tantrums (complete loss of temper, with shouting, angry movements, etc.)	4 (50%)	9 (41%)	13 (43%)	9 (38%)	13 (42%)
Tears on arrival at school or refused to go into school building	3 (38%)	2 (9%)	5 (17%)	3 (13%)	1 (3%)
Truanted from school	0 (0%)	0 (0%)	0 (0%)	1 (4%)	2 (6%)
Stammer or stutter	0 (0%)	1 (5%)	1 (3%)	1 (4%)	1 (3%)
Any difficulty with speech, apart from stammering or stuttering	2 (25%)	0 (0%)	2 (7%)	1 (4%)	3 (10%)
Did he/she ever steal things?	0 (0%)	0 (0%)	0 (0%)	0 (0%)	3 (10%)
Was there any eating difficulty?	5 (63%)	5 (23%)	10 (33%)	13 (54%)	8 (26%)
Was there any sleeping difficulty?	3 (38%)	7 (32%)	10 (33%)	7 (29%)	8 (26%)
Very restless, had difficulty staying seated for long	2 (25%)	9 (41%)	11 (37%)	7 (29%)	7 (23%)
Squirmy, fidgety child	2 (25%)	6 (27%)	8 (27%)	4 (17%)	4 (13%)
Often destroyed own or others' property	1 (13%)	2 (9%)	3 (10%)	0 (0%)	2 (6%)
Frequently fought or extremely quarrelsome with other children	2 (25%)	0 (0%)	2 (7%)	6 (25%)	8 (26%)

ITEMS OF THE RUTTER A SCALE: POST-OPERATIVE FREQUENCIES (contd.)					
	CY (n=8)	ACY (n=22)	CARDIAC (n=30)	BMT (n=24)	HEALTHY (n=31)
Not much liked by other children	1 (13%)	2 (9%)	3 (10%)	0 (0%)	1 (3%)
Often worried, worried about many things	4 (50%)	11 (50%)	15 (50%)	10 (42%)	13 (42%)
Tended to be on own, rather solitary	3 (38%)	2 (9%)	5 (17%)	6 (25%)	9 (29%)
Irritable, quick to fly off the handle	2 (25%)	7 (32%)	10 (33%)	9 (38%)	8 (26%)
Often appeared miserable, tearful or distressed	3 (38%)	3 (14%)	6 (20%)	7 (29%)	3 (10%)
Had twitches, mannerisms, tics of face or body	0 (0%)	1 (5%)	1 (3%)	0 (0%)	0 (0%)
Frequently sucked thumb or fingers	1 (13%)	3 (14%)	4 (13%)	7 (29%)	5 (16%)
Frequently bit nails or fingers	1 (13%)	4 (18%)	5 (17%)	4 (17%)	7 (23%)
Often disobedient	4 (50%)	7 (32%)	11 (37%)	8 (33%)	18 (58%)
Could not settle to anything for more than a few moments	2 (25%)	5 (23%)	7 (23%)	9 (38%)	4 (13%)
Tended to be fearful or afraid of new things or situations	5 (63%)	8 (36%)	13 (43%)	7 (29%)	6 (19%)
Fussy or over-particular child	2 (25%)	4 (18%)	6 (20%)	12 (50%)	3 (10%)
Often told lies	1 (13%)	4 (18%)	5 (17%)	1 (4%)	7 (23%)
Bullied other children	2 (25%)	0 (0%)	2 (7%)	1 (4%)	1 (3%)

TABLE 28

ITEMS OF THE RUTTER B SCALE: POST-OPERATIVE FREQUENCIES					
	CY (n=8)	ACY (n=21)	CARDIAC (n=29)	BMT (n=22)	HEALTHY (n=31)
Very restless, had difficulty staying seated for long	2 (25%)	7 (33%)	9 (31%)	5 (23%)	8 (26%)
Truanted from school	0 (0%)	1 (5%)	1 (3%)	0 (0%)	0 (0%)
Squirmy, fidgety child	2 (25%)	3 (14%)	5 (17%)	5 (23%)	6 (19%)
Often destroyed or damaged own or others' property	0 (0%)	0 (0%)	0 (0%)	1 (5%)	2 (6%)
Frequently fought or extremely quarrelsome with other children	2 (25%)	1 (5%)	3 (10%)	3 (14%)	3 (10%)
Not much liked by other children	0 (0%)	2 (10%)	2 (7%)	2 (9%)	1 (3%)
Often worried, worried about many things	3 (38%)	10 (48%)	13 (45%)	10 (45%)	5 (16%)
Tended to be on own, rather solitary	1 (13%)	7 (33%)	8 (28%)	7 (32%)	2 (6%)
Irritable, touchy, quick to fly off the handle	1 (13%)	1 (5%)	2 (7%)	5 (23%)	5 (16%)
Often appeared miserable, tearful or distressed	2 (25%)	3 (14%)	5 (17%)	2 (9%)	1 (3%)
Had twitches, mannerisms, tics of face or body	2 (25%)	2 (10%)	4 (14%)	1 (5%)	0 (0%)
Frequently sucked thumb or fingers	0 (0%)	1 (5%)	1 (3%)	1 (5%)	3 (10%)
Frequently bit nails or fingers	2 (25%)	1 (5%)	3 (10%)	2 (9%)	5 (16%)
Tended to be absent from school for trivial reasons	1 (13%)	0 (0%)	1 (3%)	0 (0%)	1 (3%)
Often disobedient	1 (13%)	2 (10%)	3 (10%)	4 (18%)	10 (32%)
Could not settle	2 (25%)	5 (24%)	7 (24%)	5 (23%)	4 (13%)
Tended to be fearful or afraid of new things or situations	3 (38%)	6 (29%)	9 (31%)	3 (14%)	1 (3%)

ITEMS OF THE RUTTER B SCALE: POST-OPERATIVE FREQUENCIES (contd.)					
	CY (n=8)	ACY (n=21)	CARDIAC (n=29)	BMT (n=22)	HEALTHY (n=31)
Fussy or over-particular child	0 (0%)	1 (5%)	1 (3%)	1 (5%)	2 (6%)
Often told lies	0 (0%)	0 (0%)	0 (0%)	1 (5%)	4 (13%)
Has stolen things on one or more occasions in the past 12 months	1 (13%)	0 (0%)	1 (3%)	1 (5%)	0 (0%)
Unresponsive, inert or apathetic	2 (25%)	4 (19%)	6 (21%)	2 (9%)	2 (6%)
Often complained of aches or pains	2 (25%)	2 (10%)	4 (14%)	4 (18%)	2 (6%)
Has had tears on arrival at school or has refused to come into the building in the past 12 months	1 (13%)	2 (10%)	3 (10%)	1 (5%)	0 (0%)
Has a stutter or stammer	0 (0%)	1 (5%)	1 (3%)	0 (0%)	0 (0%)
Resentful or aggressive when corrected	1 (13%)	2 (10%)	3 (10%)	4 (18%)	9 (29%)
Bullied other children	0 (0%)	1 (5%)	1 (3%)	1 (5%)	4 (13%)

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