

Changing trends in surgery for ventricular septal defect in Malta

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ABSTRACT: Congenital heart defects (CHD) comprise the commonest group of malformations and ventricular septal defect (VSD) is the most prevalent type of CHD most frequently requiring corrective surgery. There has been an increase in the number of Maltese patients operated for this condition over 1930-1994 which has now stabilised. Peri-operative mortality has declined, paralleled by a significant decline in age at surgery. The birth prevalence of VSD requiring surgery is 0.83/1000 live births with a surgical rate of 0.88 operations/1000 live births including primary operations and reoperations. Approximately 122,000 children are born with this condition annually world-wide at an estimated potential surgical cost of £2,280,000,000.

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Introduction

Congenital heart disease (CHD) is a label for a heterogeneous set of lesions which comprise the commonest group of congenital malformations, and ventricular septal defect (VSD) is the commonest form of CHD¹. Malta is an ideal setting for studies dealing with historical trends in treatment due to the relatively closed and captive nature of the population.

This paper identifies all Maltese patients diagnosed as having haemodynamically significant VSD requiring intervention born up to the end of 1994 and analyses trends in age at surgery, method of surgical closure, operative centre, perioperative mortality, reoperations for VSD and overall trends in number of operations for VSD.

Methods

Definitions

CHD was defined as a structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance¹. Cases with multiple CHD diagnoses had their lesions classified hierarchically. The most haemodynamically important lesion was considered the primary diagnosis throughout.

VSD was defined as a defect in the interventricular septum. Patients with a primary diagnosis of VSD were graded by severity into two groups. Mild VSDs were those which did not require surgical intervention. Severe VSDs were those which required intervention and are the subject of this study. Perimembranous defects were those located in the upper, perimembranous portion of the interventricular septum, related to the tricuspid valve on the right side of the heart and to the aortic valve on the left side of the heart. Muscular defects were those located more inferiorly in the interventricular septum, bounded entirely by the muscular septum throughout

their circumference.

Diagnosis of VSD was only accepted for inclusion into this study if made by echocardiography (available in Malta since 1988), cardiac catheterisation, surgery or post-mortem. Recent studies dealing with the epidemiology of CHD have only included patients diagnosed as having CHD by one year of age². This condition was not applied due to the historical nature of the study.

Patients

The catchment area for this study was the Maltese archipelago. St. Luke's Hospital is the only regional hospital in Malta. It caters for the investigation of all patients suspected as having CHD, and their follow-up. Termination of pregnancy is illegal, hence, there is no organised antenatal screening programme for CHD.

Data sources

Patients with severe VSD were identified from copies of all paediatric echocardiogram reports, registers of patients referred for investigation or treatment abroad, local elective catheter lists, visiting consultant cardiologist clinic lists, visiting consultant cardiac surgical lists and post-mortem reports. Newly diagnosed cases of severe VSD born between October and December 1994 were entered prospectively.

Statistics

Official Maltese publications were used to obtain total monthly livebirths from 1990 to 1994³.

Pearson correlation was used to analyse correlation of events with time.

A p value of 0.05 was taken to represent statistical significance.

Software

Patients with CHD were entered onto a dedicated dBASE database (MAPCAD: Maltese Paediatric Cardiology Database). Diagnoses and interventions were coded with a Version 2 Read Codes Browser⁴. A series of dBASE procedures were used to extract data from MAPCAD. The data was analysed with Excel 5.0 and SPSS.

Results 1930-1994

There were 85 cases of severe VSD in this period.

Severe VSD-unoperated

10 patients were not operated. The reasons for non-operation were late diagnosis with irreversible pulmonary hypertension (n=5, year of birth: 1930-1986), Down's syndrome (n=3, year of birth: 1967-1973) and multiple malformations (n=2, year of birth: 1979-1991).

Reoperation for VSD

Ten of the 75 operated VSDs were reoperated (13%). Reasons for reoperation were redo operations (n=4, year of 1st operation: 1976-1993), debanding of previous pulmonary artery band with closure of multiple VSDs (n=3, year of 1st operation: 1966-1993), re-exploration in the early postoperative period for tamponade (n=1, year of operation: 1987), pacemaker insertion for iatrogenic heart block (n=1, year of operation: 1968), and aortic valve replacement for associated progressive aortic valvar stenosis (n=1).

5 yearly operation totals and peri-operative mortality

The 5 yearly total number of operated VSDs has progressively increased from 1 case in 1955-1959 to 28 cases in 1990-1994 (Figure 1). The yearly mean number of operations for VSD has levelled out at 5.6 cases per

annum in 1990-1994. Five-yearly percentage peri-operative mortality decreased steadily from 25% in 1975-1979 to 0% in 1990-1994. Further analysis of data showed a progressive decline in mean age at surgery in the eras 1965-69, 1970-74, 1975-89 and 1990-94.

Age at surgery

There was a significant trend towards earlier age at surgery over the period under study ($p < 0.0001$) (Figure 2). This trend was also present but not statistically significant since introduction of echocardiography (1988-1994, n=25, $r = -0.36$, $p = 0.07$).

Method of surgery and centre

The method of VSD closure has changed from predominantly direct suture up to the late 1970s to closure almost exclusively by synthetic patch thereafter. Maltese patients were operated mainly at St. Mary's Hospital (Paddington, London) up to the early-1980s, when the most commonly utilised centre became the Hospital for Sick Children (Great Ormond Street), London.

1990-1994

The annual VSD figures showed progressively fewer VSDs prior to 1990, while the numbers of defects stabilised over 1990-1994. This period was therefore chosen for epidemiological calculations as ascertainment since 1990 appeared to be complete. All severe defects were diagnosed before one year of age allowing standard calculations to be made regarding epidemiological aspects of VSD.

The population of Malta is approximately 360,000 and there were 26,117 livebirths in 1990-1994.

This study showed 22 live births with severe VSD in the same period with a birth prevalence of 0.83/1000 live births. Twelve were peri-membranous defects, seven were muscular defects and three had mixed defects. Three had multiple VSDs (0.11/1000 live-births). Only one of these required 2-stage repair with pulmonary

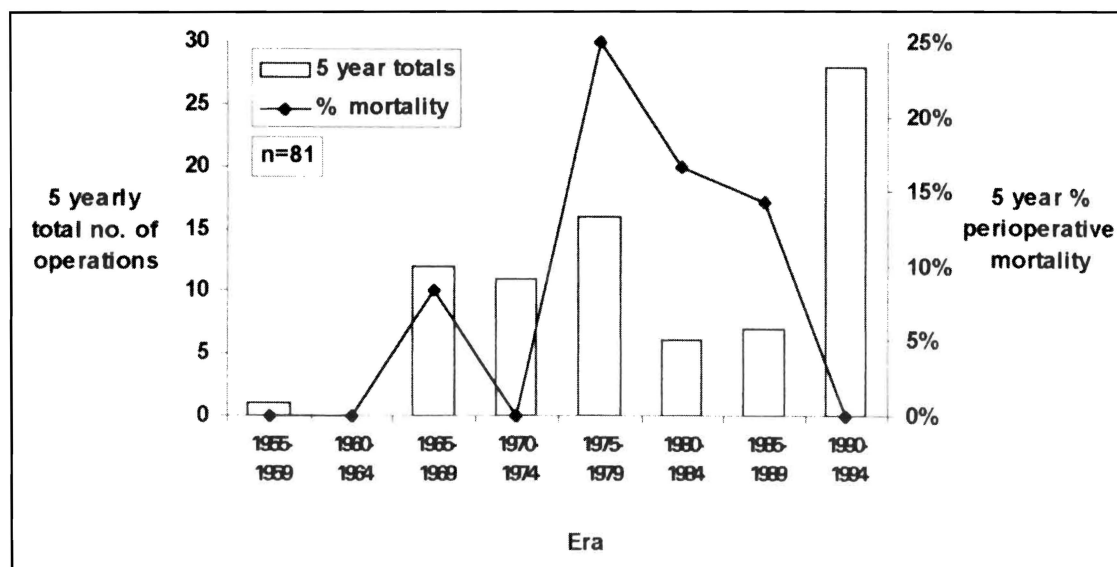


Figure 1: 5 year percentage peri-operative mortality for patients operated for VSD. (4 operations performed in 1995 not shown).

artery banding prior to total definitive repair. The overall operation rate was 0.88 operations/1000 live-births (including one patient operated twice).

Down's syndrome accounted for 32% of severe VSD (n=7) and one other patient had Holt-Oram syndrome. The latter had multiple VSDs.

Discussion

Without intervention, 90-95% of patients with CHD will die before adolescence^{5,6}.

VSD is the commonest form of CHD requiring surgical intervention and can therefore be used as an indicator of surgical paediatric cardiology services.

The developments in local paediatric cardiology services have run in parallel with services in other countries. Since diagnostic and interventional decisions were undertaken by visiting consultant cardiologists and cardiac surgeons, trends found locally can be extrapolated to larger countries with similar paediatric cardiology services.

1930-1994

There has been a progressive increase in the number of operated cases of VSD in the period reviewed which has only levelled off since 1990. A striking decline in peri-operative mortality for VSD was paralleled by a trend towards earlier age at surgery. Factors contributing to this include improved surgical expertise, experience and equipment, and increased local medical awareness of CHD and the possibility of successful treatment^{7,8}.

The decrease in operations after 1975 with subsequent pick-up of number of operations was initially thought to be due to the reorganisation of the Maltese medical services in the decade following 1977. However, further analysis of data showed a progressive decline in mean age at surgery in the eras 1965-69, 1970-74, 1975-89 and 1990-94. Although diagnosis may have been delayed in the period 1975-89 with subsequent late pick-up in 1990-94, this would tend to be obscured by a trend towards earlier age at diagnosis (and hence surgery) due to

improvement in medical and technological trends in the last period.

Other trends in surgical management of VSD include a move towards synthetic patch closure of defects rather than direct suture since the late 1970s to avoid heart block. The only case of surgically induced heart block in this series occurred in 1968.

The operative centre was predominantly St. Mary's Hospital (London) up to the early-1980s. This reflects the change in visiting cardiologists performing paediatric clinics in Malta. Prior to 1985, children were seen by Dr. Besterman, an adult cardiologist based at St. Mary's Hospital. Children who required cardiac surgery were therefore transferred to that centre. In 1985, Dr. Hallidie-Smith, a paediatric cardiologist, began to see all children with CHD. This consultant was initially based at the Hammersmith Hospital (London), and later moved to Great Ormond Street. Naturally children were operated in these centres. Few children have been operated in Malta as the natural history of these lesions is that of heart failure in infancy, hence surgery is not elective. For this reason, cardiac surgery for VSD in infants has only been carried out in UK tertiary centres as a semi-urgent procedure.

1990-1994

Global cost

The current, open market cost for cardiopulmonary bypass surgery in the UK (GOSH and Harley Street Clinic, London) is £15,000 for procedures where admission to intensive care does not exceed 7 days and hospital stay overall does not exceed 14 days (currency in sterling), and VSD falls within this category.

The birth prevalence of severe VSD was 0.83/1000 live-births with an operation rate of 0.88 operations/1000 live-births.

The birth prevalence of VSD is the same in different parts of the world^{1,6}. The current annual live births world-wide total around 144,000,000⁹. This works out to an annual number of live births with VSD of around 119,000 and an annual number of operations of 127,000

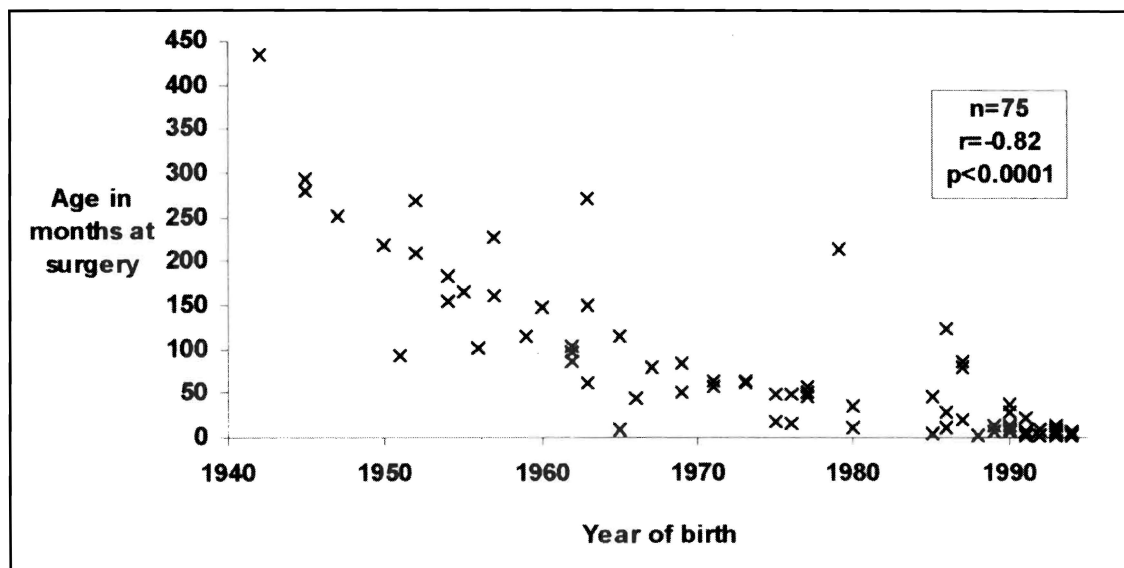


Figure 2: Age in months at first operation for VSD.

world-wide.

Given the above data, the potential world cost of treating this condition in terms of surgery alone is £1,900,800,000. This does not include the cost of preoperative medical follow-up including investigation and possible treatment of heart failure, parental loss of work to attend hospital visits and antibiotic prophylaxis.

However, in many countries, paediatric cardiology facilities are unavailable or incomplete. This results in a large number of these children dying of heart failure in infancy, or from irreversible pulmonary hypertension (Eisenmenger syndrome) in their second to third decade of life, or from bacterial endocarditis.

Local cost

The national budget recurrent expenditure for 1997 is estimated at LM493,000,000. The capital and recurrent expenditure for the Health budget for 1997 (including recurrent expenditure for the care of the elderly) is estimated at LM57,000,000. Surgical treatment for VSD would account for 0.02% of the total budget and 0.2% of the Health budget. In real terms, surgery for VSD costs Malta far less due to the reciprocal Health agreement between Malta and the UK which allows 180 new Maltese patients per annum to be treated in the UK as NHS patients at no extra cost to Malta. Since surgery for CHD is one of the most expensive interventions undertaken on patients referred for treatment abroad, CHD patients are invariably included in this category.

Conclusion

VSD which requires surgery is diagnosed early on in life by non-invasive echocardiography and carries a very low perioperative mortality. Malformation statistics from small countries with high ascertainment of lesions can give insights in past trends of intervention world-wide and can estimate costs of treatment on a national or global basis.

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