# A Service for Grown up Patients with Congenital Heart Disease (GUCH) in Malta

**Prof Victor GRECH, Dr Oscar AQUILINA, Prof Jane SOMERVILLE**

## Introduction

Untreated congenital heart disease takes a heavy toll for those born with such anomalies, with approximately 65% dying in infancy and 15-20% reaching adolescence and adulthood.[1] Malta has had the same incidence of congenital heart disease in live births as other European countries, i.e. 0.8/1000 live births.[2] It has been fortunate to be able to give children optimal therapy by the ability to send infants and children to London for treatment including open heart surgery over the last 40 years. The majority have returned improved or cured, with diminishing mortality rates (figure 1) and increasing survival rates in the region of 95%.[3] Many have passed adolescence becoming parents and even grandparents, and are or have been in full employment, contributing to the community.

Potentially, once such patients leave childhood, their contact with paediatricians is lost and they often may have no regular medical supervision. Frequently, this is important, particularly for those with complex problems as well as those who have had valves replaced.

The importance of this new and ever expanding cohort of grown-up congenital heart (GUCH) patients is known but establishing optimal medical services for GUCH patients over the last 25 years in the UK has been difficult with the UK's established example followed with reluctance. In 1988, the European Society of Cardiology formed a special working group (number 22) to lead activities in Europe and assist policy making with guidelines.[4] Canada has formed a good network and has produced exemplary units lead by Toronto and the USA, despite two Bethesda reports lags behind.

**Key Words**

Heart defects, congenital

Grown-up congenital heart disease (GUCH)

Cardiology Service, Hospital

Cardiac Care Facilities/manpower/ *organization & administration

Mater Dei Hospital has established a clinic for adult congenital heart patients and those who need to change from paediatrics to adult cardiology care. This clinic was established by two of us (VG and OA) and has recently been joined by Professor Jane Somerville who pioneered the speciality and was responsible for establishing the services in UK and in Europe, as well as other parts of the world.

The outpatients clinic is held monthly, and is open to all patients with congenital heart disease aged 14 years and over, males and females, and also patients with Marfan, Turner, Noonan and other syndromes wherein heart and blood vessel pathologies are implicated.

The clinic is growing and there are now several hundred patients on the clinic database which has been maintained since 1994.[5] The intention of this article is to draw attention to this new service and encourage referral of all patients with congenital heart disease for assessment of their condition and any medical needs, such that Maltese GUCH patients can receive appropriate and optimal followup if required. The likely requirements of this service is reviewed by surgical survival rates for congenital heart disease for Maltese patients.

## Methods

The Maltese Paediatric Cardiology Database was queried to obtain all patients operated for congenital heart disease. Cardiac conditions were subdivided into two:

Severe CHD included those lesions with all valves and chambers present. These lesions can usually be completely repaired with a biventricular circulation i.e. with one ventricle supporting the systemic circulation and another ventricle supporting the pulmonary circulation.

Complex CHD included those lesions with valve and/or chamber atresia and/or hypoplasia. Generally, complex lesions can only achieve extended palliation via extensive surgery with a univentricular circulation i.e. with one ventricle supporting the systemic circulation

and no ventricular support to the pulmonary circulation. Flow across the lungs is achieved by passive flow from the venae cavae.

### Patients

All patients diagnosed as having CHD, who were born up to 2004 inclusive, and operated for CHD up to 2004 inclusive were included in this study.

### Population

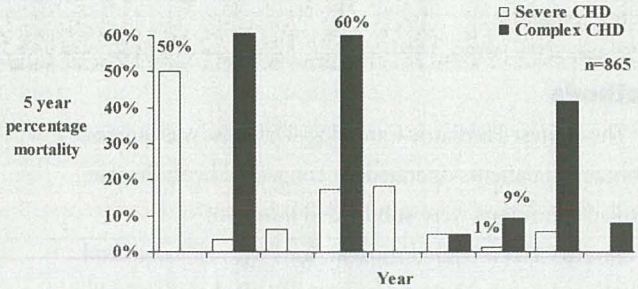
The catchment area for this study were the Maltese Islands. Mater Dei Hospital is the only regional hospital in Malta, and caters for the investigation of all patients suspected of having CHD, and their follow up.

## Results

865 operations were documented for severe and complex lesions. The first operation for CHD on a Maltese patient was performed locally by a Maltese team and this was ligation of a patent ductus arteriosus in 1947. There was a five year hiatus after which patients began to be referred to The Hospital for Sick Children, London (GOSH) and to Guy's Hospital, London. Cases began to be referred to St. Mary's Hospital, London from 1966 up to the early 1980s. Referrals then began to be sent to Hammersmith and reverted primarily back to GOSH in 1988.

The percentage 5-year perioperative mortality decreased throughout the period under study overall, and for both severe and complex CHD.

*Figure 1*




*5 yearly percentage early (1 month) percentage mortality for surgery for congenital heart disease in Malta. Complex lesions: univentricular hearts (e.g. tricuspid and pulmonary atresia). Severe lesions: biventricular hearts (e.g. ventricular septal defect closures, tetralogy of Fallot and coarctation repairs and arterial switch procedures for transposition of the great arteries). Extended from Grech V and Elliott MJ 1998.3xx*

## Discussion

### Characteristics of the population

This is a young population with cardiac problems and has replaced rheumatic heart disease that was so prevalent 30-50 years ago. The needs of GUCH patients are different from the commoner type of adult cardiac patient with hypertension and coronary artery disease. They are younger, have many medical problems in systems other than the cardiovascular, as well as more social adaptive problems which require advice and support. These patients include the more complex conditions i.e. tetralogy, coarctation, atrioventricular septal defect, transposition of the great arteries, total anomalous pulmonary venous drainage, truncus arteriosus and univentricular hearts and does not include relatively simple and straightforward lesions such as pulmonary stenosis and ventricular septal defect.

A liaison nurse specialist is vital in this service so that patients can make direct contact for medical advice and appropriate referral. General services, all available at Mater Dei Hospital, are required by 20-25%, particularly obstetric services. The supervision of pregnancy and delivery and assessment of offspring and siblings requires the support of both cardiologist and obstetrician experienced in the care of those at cardiac risk in pregnancy.[6]

The GUCH patient population is slowly ageing. In the longer established units, 30% are over 40 years old and have lesions with present the health service with the usual spectrum of comorbidities. The main problems affecting GUCH patients are:

### Cardiac problems

1. Arrhythmias (commonest)
2. Increasing cyanosis
3. Heart failure
4. Endocarditis
5. Emboli
6. Deteriorating symptomatic state
7. Need for reinvestigation

Facilities needed for the followup of GUCH patients include:

1. Usual cardiac tests e.g. ECG, CXR, exercise testing
2. Echocardiography by expert staff including transoesophageal echocardiography. Equipment must be modern with facilities to measure flow, volume and Doppler for both flow and tissues.
3. Electrophysiology
4. Pacing
5. Cardiac catheterisation

6. Sophisticated imaging techniques such as magnetic resonance imaging requires someone familiar with congenital heart disease and appropriate software.

Malta has most of these, but the services must be brought together for the sake of our GUCH population who must, in turn, be encouraged to attend specialist GUCH clinics. It is certain from the number of survivors from our database review that a significant number of our GUCH population is lost to followup. A recent local attempt to study all patients beyond infancy who had had tetralogy of Fallot repair only succeeded in tracing 57 patients out of a total of 100, with 43 therefore lost to followup.[6] This is in keeping with studies in larger countries which have clearly demonstrated that these patients exist and attend non-specialist clinics despite some of them having very complex heart disease.[7]

The plight and problems of GUCH patients have been recently highlighted locally in a conference at the Mediterranean Conference Centre in December 2008 (Mediterranean GUCH course), wherein an authoritative faculty discussed various aspects of this select population (www.maltime.com under past events).

A simple calculation based on published epidemiological data for the Maltese Islands, and assuming 95% survival shows that our pool of GUCH patients will incrementally increase by 12 patients per year, every year.

With the clinic now established, it is important to consolidate the service and make it efficient, embracing all the Maltese GUCHs. Patients with GUCH should therefore be referred to the GUCH clinic at Mater Dei Hospital for specialist assessment and followup.

## References


1. MacMahon B, McKeown T, Record RG. The incidence and life expectation of children with heart disease. Br Heart J 1953;15:121.
2. Grech V. Spectrum of congenital heart disease in Malta: an excess of lesions causing right ventricular outflow tract obstruction in a population based study. Eur Heart J 1998;19:521-525.
3. Grech V, Elliott MJE. The evolution of surgical trends in congenital heart disease: a population based study. Int J Cardiol 1998;66:285-292.
4. Report of the British Cardiac Society Working Party. Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart. 2002;88 Suppl 1:1-14.
5. Grech V, Pace J. Automation of follow-up and data analysis of paediatric heart disease in Malta. Int J Cardiol. 1999;68:145-9.
6. Galea N, Aquilina O, Grech V. Risk factors for QRS prolongation after repaired tetralogy of Fallot. Hellenic J Cardiol. 2006;47:66-71.
7. Bowker TJ, Hunter AS, Williams GJ. Does "grown-up" congenital heart disease in district hospitals differ from that in regional cardiac centres? A pilot for a national survey. Heart 1998;79(suppl I):26.



Dr Victor GRECH

*Paediatric Department, Mater Dei Hospital, Malta*

Email: victor.e.grech@gov.mt