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Declining mortality from congenital heart disease in Malta

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ABSTRACT: Congenital heart disease (CHD) is the most common congenital anomaly occurring in approximately 10/1000 live births. Analysis of official Maltese Health Division statistics listing CHD as the primary cause of death on death-certificates has shown a significant fall in mortality from CHD in Malta from 1952 to 1993 (r= -0.84, p<0.0001). This decline has persisted despite the steady incidence of CHD and has not yet plateaued. The decreasing mortality from CHD may be attributed to advances in paediatric cardiology and paediatric cardiac surgery over the past four decades.

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Introduction

Congenital heart disease (CHD) is the most frequent of all congenital anomalies[1] and therefore constitutes an important public health issue. In the last four decades, tremendous progress has been achieved in the diagnosis and treatment of CHD world-wide. Similar developments in paediatric cardiology services have become available in Malta and these improvements should be reflected by a declining specific mortality rate from CHD. Crude cause-specific mortality statistics for the Maltese Islands have been available since the turn of the twentieth century. However, these are absolute figures which do not reflect mortality rates and give no indication of changing trends. The accuracy of this data is dependent on the doctor certifying the true cause of death. This study analysed cause-specific mortality rates from CHD in the Maltese Islands for the years 1912-93, and investigated the changing trends in CHD mortality for the Maltese population.

Material and Methods

Population statistics and mortality data were collected from official annual government publications[2,3,4,5,6] for the period 1912-93.

Population data

For the purposes of this study, demographic data, particularly the age-specific population figures, were based on the official censuses conducted every decade with interruption during the period 1941-51 (due to the Second World War) and during the period 1971-81. For the latter period, the mean population of the two censuses conducted in 1967 and 1985 was taken as representative of the population at this time. The ten-year mean age-specific mortality rate from CHD was calculated.

Deaths from CHD

Mortality data was subdivided according to patient age. The publications for the periods 1897-1911 and 1940-50 did not list CHD as a specific disorder, but included CHD under the general heading of 'deaths from congenital disorders'. Mortality data from CHD for these years was therefore not available. Separate neonatal mortality data was only available after 1951. Data regarding cause-specific stillbirth mortality is not documented in official national statistics and was therefore not available.

Mortality rates

The ten-year mean early childhood (<5 years) mortality rates were based on the total live births reported during the year of the censuses. The specific annual mortality rate from CHD was calculated using the estimated end of year population.

The published statistics do not break down mortality from CHD by different lesions, therefore the specific mortality rates for individual lesions could not be calculated.

Mortality rates were calculated per 100,000 population. These were further analysed according to the age at death and these results were calculated per 1000 live births.

Statistical analysis

Pearson correlation was used to calculate the association between specific mortality from CHD and year. A p value of <0.05 was taken to represent a significant correlation.

Results

Annual trends in specific mortality rates

The annual specific mortality rates for CHD show an

overall decrease in mortality from CHD (Figure 1). However, during the 81 year study period, there were marked fluctuations in rates:

1912-52

The specific mortality rate from CHD rose from 2.34 per 100,000 population in 1912-20 to 4.43 in 1931-40. Rates could not be calculated for the period 1941-51 due to the absence of a population census during World War II.

1952-93

The rate rose to 7.65 per 100,000 population in the period 1951-60, and thereafter gradually fell to 2.38 per 100,000 population in 1989-90 (Table 1). The trend in decreasing specific mortality rate from CHD for the period 1952-93 was significant ($r=-0.84$, $p<0.0001$).

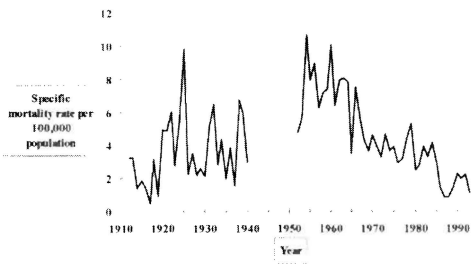

Fig. 1 - Specific mortality rate from congenital heart disease in the Maltese Islands

Annual specific mortality rates by age

The specific mortality rates were further analysed by age at death (Table 2). There was a declining trend in mortality from 1950-91 in all ages except in the early neonatal period (<1 week of age). The majority of deaths from CHD occurred in the first year of life. The specific mortality rate from CHD in the first year averaged 1.18 per 1000 livebirths in 1981-90, while the mortality in children age 1-5 years averaged 1.30 per 1000 livebirths for the same decade. This higher mortality after the first year of life can be noted throughout all the decades studied, both pre- and post-1940s (Table 2).

Discussion

Routine mortality statistics have a useful, albeit limited role in evaluating the health status of a community. These statistics are dependent on the correct diagnosis being recorded by the doctor certifying the cause of death. Bearing these limitations in mind, trends in cause-specific mortality rates can provide a means of audit of health services, and can be used to evaluate the impact of innovative medical interventions.

Any comparison of the data on incidence and mortality from CHD in Malta with that of other countries must be viewed with caution. The definition of CHD encompasses a heterogeneous group of lesions which results in variations between different national registries. The lowest reported incidence of CHD is from West Flanders with 1.79 cases per 1000 births and the highest is Glasgow with 11.4 cases per 1000 births1. The incidence for Malta for the period 1983-85 was estimated at 3.47 per 1,000 live births but follow-up was only up to the age of 1 month[7]. The latter is almost certainly an underestimate due to underascertainment of cases as the incidence of CHD is 8-9/1000 live births[8]. The childhood specific mortality from CHD for the corresponding period was 1.3 per 1,000 live births. Therefore, it appears that approximately 14% of children born with CHD die from this disorder.

Pre-1941

The ten-year average mortality rates indicate an overall increase in deaths from CHD from 1912 to 1941. The mortality rates are less than those recorded after 1951. Indeed, the specific mortality in 1912 was 2.34 per 100,000 population as compared with 2.38 per 100,000 population in 1993. This almost certainly reflects diagnostic inaccuracies, with less cases being identified as dying from CHD before 1941. The problems preclude accurate interpretation and comparison of the figures obtained between the pre-1941 and post-1951 periods.

Post-1951

Between the years 1941-51, no calculations were possible due to the lack of a census during this decade.

Following 1951, the specific mortality rates for CHD in Malta declined significantly from 7.65 per 100,000 population in 1951 to 2.38 in 1993. This declining trend does not yet show any signs of reaching a plateau.

On analysis of these trends by age at death, the early neonatal mortality appears to have been little affected (Table 2). However, the death rate from CHD in the first two years of life has decreased considerably.

Changes in trend in specific mortality rate in late childhood (5-14 years) are difficult to assess due to the small numbers involved which cause wide fluctuations. However, there does appear to be a fall in the specific mortality rates in the 5-24 year age groups in the last

Table 1 - Age-specific mortality rates for CHD:
Decennial average per 100,000 age-specific population (census year)

YEAR	<5 yrs	5-14 yrs	15-24 yrs	25-44 yrs	>45 yrs	TOTAL
1911-20	17.85	0.49	0.27	0.21	0	2.34
1921-30	39.18	0.68	0	0	0	4.36
1931-40	37.28	1.01	0	0	0	4.43
1941-50	-	-	-	-	-	-
1951-60	56.23	1.10	1.63	1.20	0.14	7.65
1961-70	58.92	1.93	1.88	0.54	0.36	5.95
1971-80	36.20	1.63	2.43	0.44	0.11	3.57
1981-90	31.62	0.18	0.20	0.74	0.10	2.38

[data for 1911 and 1941-51 unavailable: means for 1911-20 and 1951-60 based on nine year averages]

['-' denotes data unavailable]

Table 2 - CHILDHOOD SPECIFIC MORTALITY RATES FOR CHD: Decennial average per 1000 livebirths (census year)

YEAR	<1 week	1-4 weeks	4 wks-1 yr	<1 year totals	1-2 years	2-3 years	3-4 years	4-5 years	<5 year totals
1911-20	-	-	-	0.55	0.05	0.03	0	0	0.63
1921-30	-	-	-	0.95	0.10	0.04	0.03	0.05	1.17
1931-40	-	-	-	1.05	0.17	0.04	0.01	0.04	1.31
1941-50	-	-	-	-	-	-	-	-	-
1951-60	0.42	0.35	1.23	2.00	0.24	0.09	0.09	0.06	2.48
1961-70	0.85	0.58	1.17	2.59	0.25	0.10	0.04	0.02	3.00
1971-80	0.49	0.21	0.66	1.36	0.11	0.04	0	0.04	1.55
1981-90	0.72	0.20	0.26	1.18	0.02	0.06	0.02	0.02	1.30

[data for 1911 and 1941-51 unavailable: means for 1911-20 and 1951-60 based on nine year averages]
['-' denotes data unavailable]

decade (Table 1), although this change is not yet significant.

In this study, the decline in mortality rates may have been caused by changes in the paediatric cardiology services in the country in the last four decades. World-wide, surgery in the management of CHD has improved considerably since 1939[9] when the first patent ductus arteriosus was successfully ligated[10]. This was followed by the creation of a subclavian artery to right pulmonary artery shunt for palliation of Fallot's Tetralogy[11] (1945), hypothermic open heart surgery for closure of secundum atrial septal defect[12] (1953), and total cardiopulmonary bypass for closure of a secundum ASD (1953)[13]. In Malta, the first ligation of a patent ductus arteriosus was performed by Prof. P.P. Debono in 1947, while the first "blue baby" was sent to the UK for operation in 1950[14]. Since 1989, a comprehensive programme of cardiac surgery for children was established whereby a visiting specialist team from Great Ormond Street Hospital for Children (London) performs a surgical list on an alternate year basis[15]. The establishment of a fully operational local adult cardiothoracic service allows for greater flexibility in the surgical service provided and may, in time, result in a greater number of cases of CHD being operated locally.

The advance in surgical management of CHD was paralleled by improvements in the medical care of these patients including the availability of drugs used to control heart failure, arrhythmias and other problems associated with CHD. In addition, since 1966, the local medical services were augmented with regular visits by consultant cardiologists from the United Kingdom[15]. Currently, consultant paediatric cardiologists from two UK tertiary referral centres visit four times a year and perform outpatient clinics, diagnostic and interventional catheter studies, including electrophysiological mapping and ablation. Having created the links with referral centres abroad, patients requiring further investigations and surgery are transferred to the U.K. Furthermore, several local paediatricians have completed specialist training and, of these, three are fully trained in paediatric echocardiography[16].

Congenital heart disease is one area where improvement in health services, with the inevitable associated financial costs, has had a significant impact on outcome in terms of mortality. Additional improvements in the care of children with CHD will further diminish the mortality of this common group of congenital disorders.

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