

Simple congenital heart disease: a complex challenge for public health

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Congenital heart disease (CHD) represents a broad spectrum of conditions, from simple defects with an excellent prognosis, to the complex and severe, which require multiple procedures and have uncertain long-term outcomes. As outcomes in cardiac surgery have improved, research has tended to focus on more complex cardiac lesions, rather than mild defects where results were already excellent (1). Simple defects have been defined as ventricular septal defect (VSD), atrial septal defect (ASD), pulmonary stenosis (PS), patent ductus arteriosus (PDA), aortic stenosis (AS) and aortic coarctation in older children (2). In the case of many simple forms of CHD, survival has been said to be normal (3) and guidelines do not even recommend cardiology follow-up (4,5). Have these recommendations been made on sound evidence? Or has it simply become a truism that these patients have the same life expectancy as the general population?

Several reports have demonstrated that patients with simple CHD have equivalent survival to the general population (6,7). This is a view echoed in guidelines and major cardiology and cardiac surgery textbooks (3,8,9). However, although these studies have excellent longitudinal follow-up and include large cohorts of patients, the number of patients with each individual diagnosis is small (6,7). As a result, they are prone to having large confidence intervals (7), and may simply not show a difference in survival compared to the general population due to the small number of patients. More recently, large studies have begun to cast doubt on the assertion that patients with mild CHD will have a normal life expectancy (1,10,11). A large systematic review by Verheugt and colleagues (1)

found that survival was decreased, in all forms of CHD. They specifically noted that patients with mild CHD were underrepresented in the literature, and had poorer survival than the general population (1).

In this context, we read with interest a recent paper by Videbæk and colleagues (12), which provides long-term, population-based outcomes of patients with simple CHD. Simple CHDs were defined as isolated and uncomplicated ASD, VSD, PDA and mild PS. Children with comorbidities, such as pulmonary artery hypertension, were excluded. This study is unique as it is a population-wide cross-section of patients that were all assessed by a single cardiologist, between 1963 and 1973. Furthermore, follow-up is complete thanks to Denmark's national registry of patients. The study included 1,241 patients with 58,422 patient years of follow-up.

At a median age of 47 years, they demonstrated that the risk of death in those with simple CHD was doubled the risk of the general population. Furthermore, the risk of sudden unexpected death was increased approximately 4-fold, and the risk of cardiac death was increased 6-fold. Furthermore, there was nearly a 6-fold increase in rates of morbidity, and in particular increased rates of cardiac surgery, heart failure, endocarditis, pulmonary hypertension, ventricular tachycardia and stroke. While these figures seem somewhat alarming, it must be remembered that they represent quite small absolute risks: the overall mortality rate in the simple CHD cohort was 2.2 per 1,000 patient years. Interestingly, the risk of mortality was remarkably consistent among the groups of patients, with each of the individual cardiac diagnoses (ASD, VSD, PS and PDA) having an individual

mortality rate in the range of 2.1–2.2 per 1,000 patient years.

There are several limitations of the study, which must be borne in mind when interpreting these data. Firstly, the patients included were diagnosed over 40 years ago, and patterns of treatment and diagnosis have changed over time. However, this will invariably be the case in studies with such a long follow-up. Furthermore, the patients were all diagnosed by a single cardiologist, which ensures consistency of diagnosis, but raises questions of generalizability, especially since these patients were recruited in a time when echocardiography was not yet available.

Nevertheless, this is an important study as it challenges the view that patients with simple CHD are cured. This information contributes to a growing body of evidence that defects perceived as simple and cured, may not be as benign as surgeons and cardiologists believe. In our own experience, children with partial atrioventricular septal defect, often grouped with mild CHD, have excellent survival, but a reoperation rate of approximately 25% at 20 years follow-up (13,14). Furthermore, only 43% of adult patients were under the care of a cardiologist (13).

This information is of particular importance due to the shifting demographic of patients with CHD. More than half of patients with CHD are now adults (15) and it is estimated that the total population of adults with CHD in the United States is in the range of 1.3–1.4 million patients (1,16). Of these patients, it is estimated that 750,000 have simple congenital defects (16). Previously this group had been considered cured, and would not have been required to attend specialist long-term follow-up. Clearly, the findings of Videbæk (12) are potentially very important at a population level. If all of these patients were to require specialist follow-up it would have substantial implications for the cost of health care, as well as creating additional workforce strain on the relatively small community of adult congenital cardiologists. In order to carefully inform recommendations regarding follow-up there would ideally be a way to stratify patients with simple CHD, so that follow-up could be focused on those at greater risk. For example, Kuijpers *et al.* (10) demonstrated that males, but not females, had decreased survival following closure of simple ASD. Furthermore it is known that older age at the time of ASD closure is an important risk factor for death (6). More work is required in this area to delineate, which patients are at higher risk and hence require closer follow-up.

Furthermore, these findings have important implications at the level of the individual patient-clinician interaction. It is no longer appropriate for clinicians to tell patients

that they are cured of their disease by surgery in the case of simple CHD. While there is good reason to be optimistic about their long-term outcomes, a word of caution regarding the slightly higher risk of arrhythmias and death, as well as the need for reoperation is appropriate. This allows patients to be more vigilant about their health, with the potential that they may seek review earlier in the course of any late complications, hopefully mitigating some of the increased risk.

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Footnote

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