Congenital heart disease in the teenage patient

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Abstract

Due to increasing incidence and improved survival, greater numbers of children with congenital heart disease (CHD) are surviving to their teenage years and beyond. This patient group have complex medical needs, which interact with the emotional and developmental challenges of adolescence. Teenagers with CHD are at increased risk of both neurodevelopmental and emotional disorders, and it is important that all healthcare professionals who deal with the group are aware of these issues. Quality of life is also significantly impaired in this group, and strategies should be considered to maximise this and other holistic outcomes. As teenage patients become adults, their care is transitioned to adult services, a potentially stressful time for these patients. It is important that this process is well understood, and managed in a streamlined and organised manner to minimise numbers lost to follow-up. It is at this stage that patient educational information, probably previously given to parents or carers, is reiterated to the patient themselves. This should include advice around the patient's specific lesion, as well as general advice about exercise tolerance, endocarditis risk, and pregnancy.

Keywords adolescent; cardiology; congenital; heart defects; paediatrics; quality of life; transition to adult care

Introduction

Over recent decades there has been a dramatic change in the congenital heart disease (CHD) patient population. Paediatricians and general practitioners are now far more likely to deal with older patients, including teenagers, who have survived severe CHD. Such patients form a particularly challenging and complex group. On top of a potentially life-threatening illness with complicated and debilitating interventions and medications, the patient is entering an emotionally and physically challenging life phase. Adolescents are attempting to grapple with the challenges of independent life, whilst negotiating transitions in healthcare settings and providers. It is essential that the care provider interacts with the patient in the context of these profound changes.

A wide spectrum of congenital heart lesions exists in the adolescent CHD population, ranging from the benign lesion with no effect on longevity to life-limiting conditions that are often highly symptomatic. In addition to problems that affect CHD patients of all ages, such as heart failure, pulmonary hypertension, arrhythmias, and infective endocarditis, teenagers have special problems and considerations which will be presented in this review.

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Epidemiology

The current incidence of congenital heart disease (CHD) is estimated to be between 4 and 10 per 1000 live births. For reasons that are not fully understood, this incidence rate seems to be increasing over time, probably due largely to improved case ascertainment. However, it may also be possible that changing risk factor exposure patterns may be causing an increase in the true incidence.

Mortality from CHD remains a significant cause of death in the developed world, accounting for between 30 and 50% of all deaths due to congenital anomalies. However, the mortality rates for specific defects have undergone a dramatic transformation since the early development of the speciality of paediatric cardiology in the 1930s. From a starting point of almost 100% mortality during infancy or childhood for the most severe defects, currently over 90% of children with cardiac lesions survive until adulthood.

The result of this increase in birth incidence, coupled with a drastically reduced mortality rate for most lesions, is an increase in the numbers of teenage and adult patients with congenital heart disease. In fact, these changes have been so dramatic that worldwide there are now more adults living with CHD than children. The exact prevalence of CHD in the adult population is not known, but is estimated to be around 3000 per million people.

This change in the demographic makeup of the CHD population is of relevance to paediatricians, as well as adult physicians. Before reaching adulthood, these patients must necessarily negotiate their adolescent years. During this often difficult time, most patients will initially remain under the care of paediatric cardiologists and general paediatricians, before undergoing the process of transition to adult care services. The large increase in adult CHD patients has thus been preceded by a similarly large increase in the numbers of teenagers living with CHD, hence the focus of this review.

In earlier eras those patients surviving to adolescence were usually those who had been 'cured', either surgically or by spontaneous resolution of their lesion. These patients require a relatively minimal level of medical input. However, in the modern age a large proportion of the increase in this CHD adolescent population is made up of survivors of multiple palliative surgical procedures, including single-ventricle circulations. This is a unique patient group with a variety of specific and complex medical needs, and is very intensive in terms of demands placed on the medical services.

Psychosocial, neurodevelopmental, and emotional issues

Adolescent patients with severe CHD will have almost certainly undergone multiple surgical procedures, often utilising cardiopulmonary bypass and deep hypothermic circulatory arrest. In addition to this, if not diagnosed antenatally, they may have initially presented in the neonatal period with profound cyanosis,

acidosis, and hypoxic-ischaemic brain injury. In univentricular patients, even those who avoided this initial presenting insult will have had several years of chronic hypoxia before the completion of their Fontan circulation.

The results of these factors on the developing brain are not completely known. Both children and adults with CHD have been shown to have lower IQ than unaffected controls. In addition to this, survivors of complex CHD have been shown to exhibit a characteristic pattern of neurodevelopmental impairment, including mild cognitive impairment, but also abnormal development of language and communication skills, reduced attentive ability, and dysfunctional executive processing. A relationship between severity of CHD and adverse neurological outcome has been demonstrated, with the biggest problems being seen in the most complex lesions, presumably for the reasons outlined above.

This is of particular relevance to adolescent CHD patients, as it is at this stage that these developmental problems may begin to manifest themselves. As the patient starts to establish an independent personality and life, separate from their parents, they may find that these complications, although subtle, become a real impediment to their goals.

In addition to direct effects, these neurodevelopmental problems may also have secondary repercussions on the emotional and behavioural state of the teenage patient. Adolescence is a time of profound hormonal and neurological changes, and is a period when behavioural problems are relatively frequent in the general population. Younger children with CHD are known to have an increased incidence of withdrawn and aggressive behaviour patterns. These patterns will worsen in some patients during adolescence, and this may prove challenging to healthcare providers, particularly as the patient begins to attain some degree of independence over their health and healthcare decisions.

As well as behavioural challenges, chronic medical conditions are associated with an increased prevalence of emotional disorders. It has been estimated that up to a third of adult patients with CHD have a psychiatric disorder, compared to around a fifth of the general adult population. Adolescent survivors of severe CHD have often suffered from repeated hospitalisation, resulting in social isolation and interruption to schooling. There may also be constant reminders of their medical problems, such as ongoing cyanosis, scarring, and poor exercise tolerance. Such lesions are often diagnosed very early in infancy, with a resulting disruption of normal parental interaction with the child during a crucial period in the psychological development of the child.

The most common psychological problems seen in teenagers with CHD are depression and anxiety. Feelings of low self-esteem and inferiority are relatively common, in part related to reduced levels of physical functioning and altered body image due to scarring or dysmorphism. Anxiety often focuses around fears regarding specific cardiac mortality, and worries about future treatments and operations. Interestingly, studies have shown that despite the dramatic reductions in mortality due to serious CHD, no such improvements have been seen in the psychological health of these patients, highlighting this as an area of potential improvement in adolescent care.

Several official bodies including the American College of Cardiology and the American Heart Association recommend that all CHD patients, including teenagers, should be formally assessed for cognitive, mood and behavioural disorders, both on an individual and family level. Although effective treatments are available for mental illnesses such as depression and anxiety, very little research has been performed comparing different treatment modalities in the context of CHD. One recent Cochrane review of depression therapies in CHD found no randomised controlled trials that met the inclusion criteria. However, it remains clear that this is an important issue in the care of adolescents with CHD, and should not be ignored.

Quality of life

As the survival of patients with CHD has improved in recent years, research attention has shifted away from simple measures of mortality and morbidity, towards more holistic measures of outcome such as health-related quality of life (QoL). This is defined as "the impact of a specific illness on a patient's ability to function in various life contexts, and draw personal satisfaction from physical, psychological, and social functioning perspectives". There is a large body of data examining the impact of CHD on QoL in children, adolescents, and adults, but the results are conflicting. This is most likely the result of the use of varying QoL instruments, heterogeneous patient groups, and a wide variety in methodological quality.

The most rigorous studies, involving the use of unaffected control groups, have shown a significantly impaired QoL in patients with CHD compared to their healthy peers. Unsurprisingly, the magnitude of this effect has been shown to be related to the seriousness of the initial diagnosis (although this relationship has not been reproduced in some studies). Children with mild CHD, not requiring any intervention through to adulthood have been shown to have good QoL, comparable with healthy peers. However, in contrast, those with single-ventricle circulations (and those requiring repeated interventions) have been shown to have significantly impaired QoL. Attempts have been made to quantify the degree to which CHD negatively impacts on QoL in comparison with other chronic diseases. Studies have found that the degree to which patients are affected with severe CHDs is similar in magnitude to end-stage renal failure and cystic fibrosis.

There are interesting data implying that paediatric patients with healthy siblings seem to report lower quality of life than those without, suggesting that patients may compare their abilities unfavourably with their 'normal' sibling. This effect may worsen during adolescence, as patients become more aware of the impact of CHD on their life in comparison with their healthy peers. In addition, acceptance by peers and participation in group activities reaches maximal importance during the teenage years. This is demonstrated in several studies that show a gradual worsening of QoL as the patient transitions from the childhood to teenage years. Also, a divergence is seen between patient-assessed and parent-assessed QoL scores during this time, perhaps revealing an attempt by the adolescent to establish independence, and rebel against parentally-set limits.

However, some studies have a more positive view of the teenage years. It has been described that some patients report an improvement in their quality of life during this period as they gradually gain independence and control over their health. Despite this, adolescents with CHD are shown to be less likely to

