



CME ARTICLE

Long-term cardio-respiratory consequences of heart disease in childhood

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EDUCATIONAL AIMS

- To appreciate that many children with surgically corrected structural heart disease have residual cardio-pulmonary limitations into adulthood.
- To develop a familiarity with the range of respiratory limitations in relation to the cardiac condition that may be anticipated in children with significant structural heart disease.
- To appreciate that formal exercise testing is an important consideration in children with congenital or acquired heart disease in adolescence.
- To understand that children with cardiomyopathies and/or arrhythmias may present with a mixture of non-specific respiratory and cardiac symptoms prior to the correct diagnosis being confirmed.

KEYWORDS

congenital heart disease;
spirometry;
exercise capacity;
arrhythmias

Summary The dynamic interaction between the heart and lungs leads to a degree of respiratory co-morbidity including both restrictive and obstructive airway abnormalities, which may be overlooked in children with congenital and acquired heart disease. The improving imaging techniques of the heart, both foetal and post-natal coupled with minimally invasive techniques for device implantation and better operative techniques for complex congenital heart disease have resulted in more children with longitudinally documented structural heart disease surviving into their adult years. Children presenting with cardiomyopathy or arrhythmias, as well as those with repaired cardiac disease, can be offered advice with regard to formal exercise testing and participation in sports, which may be particularly helpful in the adolescent years. Furthermore, through the interest of some adult cardiologists in paediatric heart disease over the past 20 years, facilities for the smooth transition of care to adult services are improving.

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Structural heart disease is being diagnosed with increasing frequency before birth during the second and third trimesters. A significant proportion will still present in the neonatal period and subtler lesions, such as atrial septal defects (ASDs), may not present until later childhood or

adulthood. This is primarily a result of better foetal imaging and echocardiographic technology that has become more widely available as well as better recognition of the underlying lesions by the echocardiographic technicians.¹ Coupled with better non-invasive techniques for device implantation [e.g. atrial septal defects (ASDs) and persistent patent ductus arteriosus (PDAs)] and operative techniques for complex congenital heart disease (e.g. early arterial switches for transposition of the great arteries), more children with heart disease are surviving well into their adult years.

REPORTING OF OPERATIVE MORTALITY FOR COMPLEX CONGENITAL HEART DISEASE

Generally speaking, many larger cardiac surgical centres from all over the world will report excellent results. Single-centre results, such as mortality rates of 1.8% from a single centre ($n = 171$)² for arterial switch operations in neonates may differ from larger multicentre databases from North America at 2.0% ($n = 199$)³ and Europe at 11.2% ($n = 169$)⁴ for the same procedure. Similarly, mortality for total anomalous pulmonary connection results for a single centre at 5.3% ($n = 38$)⁵ differ from a multicentre database in North America ($n = 122$) at 9.0% and Europe ($n = 17$) at 41.2%.⁴ Single-centre results may well appear considerably better than multicentre databases because of the talents of particular surgeons, an inherent reporting bias favouring positive outcomes, larger patient numbers and particular expertise for specific operations built on many years of cumulative experience.⁶ As suggested by Tweddle and Spray,⁶ data from the Society of Thoracic Surgeons Congenital Heart Surgery Database for the time period 1998–2001, reported a neonatal cardiac surgical hospital mortality of 11.2% in over 2800 cases.³ From the same period, the Paediatric European Cardio-Thoracic Surgical Registry reported a mortality approaching 18% in 574 neonates undergoing cardiac surgery.⁴ However, many parents of infants with significant structural heart disease may not consider much beyond the immediate surgical mortality, and only later come to appreciate that many children will have significant cardio-respiratory limitations as well as neuro-cognitive abnormalities into their adult years.

LARGE LEFT TO RIGHT SHUNTS: ASD, VSD AND PDA

Considered as a group, these conditions are the most common structural lesions presenting for assessment and management in young children. When they occur in isolation, they are usually readily diagnosed with echocardiography and managed with observation, device implantation or surgery. They have an excellent prognosis with the precautionary strategy of subacute bacterial endocarditis prophylaxis being

an important clinical consideration for patients with a ventricular septal defect (VSD) or PDA. However, the more subtle effects on cardiac function, pulmonary function and exercise performance are not widely appreciated.

ASDs

A common clinical observation is that asymptomatic children with a large atrial shunt report improved exercise tolerance after closure of the ASD.⁷ Prior to closure of an ASD between the preschool and teenage years, there will typically be a volume load on the right heart and pulmonary circulation with right ventricular dilatation and an absence of pulmonary hypertension. The timing of intervention (device placement or surgery) is designed to precede the development of significant pulmonary hypertension, right heart failure and atrial arrhythmias and thereby normalise life expectancy.⁷ Not all is straightforward, as one, albeit small, study has demonstrated that even well children who undergo closure of a secundum ASD will have abnormalities persisting after surgery.⁸

Prior to closure there are only minor changes in spirometry and haemodynamics compared with healthy children at comparable exercise levels. Interestingly, in both children⁷ and adults,⁹ prior to ASD closure there is a large reduction in maximum oxygen uptake under exercise conditions, which does not normalise within 4 months of ASD closure. However, in the adult study, when subjects were followed up for 10 years after ASD closure, their maximum oxygen uptake had normalised.⁹ A potential contributory mechanism for the delay in improvement may involve reduced expiratory flows in the small airways (mean expiratory flow at 25% forced expiration) seen as a result of elevated pulmonary blood flow that corrects within months in children but may take longer to improve in adults who have endured higher pulmonary blood flow for a considerably longer period.⁷

One study has attempted to stratify the risk for patients with an ASD having abnormal lung function in relation to their pulmonary artery pressure (PAP). Schofield and colleagues¹⁰ demonstrated that in 44 patients with ASD, those whose pulmonary artery pressure was >33 mmHg had significantly decreased vital capacity and forced expiratory volume at 1 second (FEV1), which did not recover during the post-operative period.

VSDs

The size of the VSD, thus the degree of severity of the left to right shunt, and the influence of the timing of surgical procedures (primary closure versus pulmonary artery banding and subsequent VSD closure) may result in different pulmonary function outcomes.¹¹ In children, pulmonary function tests conducted prior to VSD closure commonly reported reduced or normal dynamic compliance with a normal functional residual capacity.^{11–13}

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