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Cardiology in the Young (2017), **27**, 2025–2028
doi:10.1017/S1047951117002220

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Original Article

The 2017 Seventh World Congress of Pediatric Cardiology & Cardiac Surgery: week in review – adults with CHD*

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Abstract For the first time in 2017 adults with CHD was included in the World Congress of Pediatric Cardiology & Cardiac Surgery. With growing numbers and more complex patients reaching adulthood, there is a growing need for more attention to this subspecialty. Although survival is excellent and now over 90% of patients reach adulthood, many have residual problems and complications. Heart failure and arrhythmias are most commonly encountered. Life-expectancy is nearly normal for mild lesions, but remains reduced in moderate-complex lesions and re-interventions are often needed. As most patients want to live a normal life, sports participation and pregnancy become very important issues. Finally, although innovative treatments are being developed, including for end-stage heart failure, we have to refine strategies for optimal care, including during the end-stage lives of our patients. This article provides an overview of a selection of topics in the field of adults with CHD presented during the 2017 Seventh World Congress of Pediatric Cardiology & Cardiac Surgery (WCPCCS) in Barcelona.

Keywords: Adult CHD; pregnancy; heart failure; arrhythmias; outcome

Received: 21 November 2017; Accepted: 5 December 2017

Organisation of care

The organisation of care for adults with CHD patients differs in different parts of the world. In the United States of America and Canada, a major problem is to keep patients in follow-up, as many live far from specialist medical providers. In the United States of America financial issues also play a role, and because patients are financially responsible for their medical care, many cannot afford interventions or even advanced imaging investigations, such as cardiac magnetic resonance or CT. In many countries in Europe most patients are under regular follow-up and financial limitations do not play a major role. The impact of socio-economic factors is not well studied within the field of CHD. In the United States of America, an adults with CHD subspecialty examination for cardiologists has been introduced. This is an important step in the quality of care provided to adults with CHD patients. There is an ongoing discussion concerning the role of paediatric versus adult cardiologists in the care of adult patients. Clearly, paediatric cardiologists are best trained in the embryology and anatomic aspects of our profession, whereas adult cardiologists are typically trained in treatment of heart failure and arrhythmias, and can treat these specific problems and comorbidities when patients reach an advanced age. Historically, paediatric cardiologists organised the care not only for children but also for young adults. However, there is a clear trend that cardiologists take over the responsibility for adult patients. The most important factor is clearly the competence and individual skills of the doctor taking care of the patients. In European countries, for example in the Netherlands, the care for children and adults with CHD patients is structured by the government. Only a few centres have permission to perform interventions (surgical or percutaneous) in these patients,

*Presented at the 2017 Seventh World Congress of Pediatric Cardiology & Cardiac Surgery (WCPCCS 2017), Barcelona, Spain, July 16–21, 2017. Presented Friday, July 21, 2017.

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https://doi.org/10.1017/S1047951117002220 Published online by Cambridge University Press

and are appointed by the Ministry of Health. Thus, all interventions are performed in one of these "Congenital Expert Centers". Patients can be regularly followed in other hospitals, but it must be an affiliated hospital, working in close cooperation (shared care) with one of the recognised expert centres and with clear regulations. In these affiliated hospitals cardiologists also have to be trained in CHD. In the remaining (not-affiliated) hospitals patients can be seen and treated in the context of a mild lesion or in case of emergency (arrhythmia). Although not all patients are followed regularly, the majority have a follow-up plan well in place. In many Asian countries also the care for adults with CHD patients is well in place, for instance, in Taiwan a large country-wide programme is running.

Survival

Following the introduction of cardiopulmonary bypass surgery, nowadays 80% of our patients reach 40 years of age. For children born in the current era, these numbers are probably even better. This has resulted in a change in the proportion of paediatric and adult patients. In 1965 about 30% of congenital patients were adults, in 1985 this increased to 50%, and in 2005 already 60% of all congenital patients were adults.[1] The most important causes of death at adult age are heart failure – increasingly important, sudden cardiac death, and perioperative mortality. The improved survival rates result in an increase in very old congenital patients.[2] Not only patients with simple lesions reach older age, but also patients with moderate and even complex lesions have a better chance of reaching 60 years and older.

Aortic pathology

Many CHDs are associated with aortic dilatation or aortic pathology. This is well recognised, for example, in Marfan syndrome and in the context of a bicuspid aortic valve, but can also be found in patients with tetralogy of Fallot, single ventricle with pulmonary stenosis, arterial trunk, transposition of the great arteries, or hypoplastic left heart syndrome. Histological abnormalities of the aortic wall, the so called "cystic medial necrosis", has been reported in some of these congenital defects and included elastic fibre fragmentation.[3,4] Progressive aortic-root dilatation has been described, not only in aortic syndromes, such as Marfan, but also in patients with repaired tetralogy of Fallot.[5] Predicting factors for aortic dilatation were longer shunt-to-repair time, male gender, pulmonary atresia, and right aortic arch. However, aortic dissection is rarely found in Fallot patients and aortic dilatation was the indication for reoperation in only 2% of Fallot patients.[6] Controversies exist concerning preventive medical treatment for aortic dilatation. For example, the debate on the effect of β-blocker on aortic dilatation in Marfan patients ranges from "On the basis of meta-analysis, there is no evidence that β-blockade therapy has clinical benefit in Marfan"[7] to "Despite insufficient evidence of its benefit, the prescription of β-blockade has become a standard practice in the prevention of aortic dilatation and dissection in Marfan",[8] whereas others state that "β-blockade appears to limit aortic dilatation in children with Marfan. This treatment should be recommended as soon as the diagnosis is made".[9] It seems that the debate is still ongoing. In addition, other medications have been studied and show conflicting results. Among children and young adults with Marfan syndrome who were randomly assigned to losartan or atenolol, no significant difference in the rate of aortic-root dilatation over a 3-year period was found.[10]

Arrhythmias

One of the most common occurring complications is arrhythmia. Up to 20% of adult congenital patients suffer from supraventricular tachyarrhythmias. In addition, they are at increased risk for bradyarrhythmias, ventricular tachyarrhythmias, and sudden cardiac death. Atrial tachyarrhythmias may be caused by different mechanisms, including focal and re-entrant mechanisms. Catheter ablation is generally the preferred therapy for supraventricular arrhythmias when medication is not effective. The procedural success rate of catheter ablation is around 80%, and although many patients have recurrent supraventricular tachyarrhythmias, in the long term 70% of patients is in sinus rhythm.[11] The role of anticoagulation – especially non-vitamin K antagonist oral anticoagulants – is unclear in CHD and more research has to be performed in this field. Appropriate algorithms for risk assessment and treatment decisions still need to be established. Ventricular tachycardia ablation is promising and often feasible, but the prognostic value is unclear.

Resynchronisation therapy is also a promising but underdeveloped field. In a small recent study, acute right ventricular resynchronisation improved haemodynamics in children with repaired tetralogy of Fallot.[12] This was done by atrial-triggered right-ventricular free-wall pacing in complete fusion with spontaneous ventricular activation to achieve maximal QRS-duration shortening and resulted in higher pulse pressures and lower central venous pressures. This technique can be useful especially in acute postoperative heart failure management.

Ventricular function and heart failure

Heart failure is an important cause for morbidity and mortality in adults with CHD. In 20–40% of these

https://doi.org/10.1017/S1047951117002220 Published online by Cambridge University Press

patients heart failure is the cause of death.[1] Several causes contribute to the high risk of heart failure. Volume load can be caused by shunts or valve regurgitation, pressure overload is encountered in aortic or pulmonary stenosis, and these stenoses can be at valve level, but also at other levels, for instance, in aortic coarctation or peripheral pulmonary stenosis. Arrhythmias, such as atrial fibrillation, occur more often and contribute to deterioration of the haemodynamic situation. Finally, coronary artery disease, cardiomyopathy, and pericardial disease are seen more often in adults with CHD patients compared with the general population. Some factors result from the embryonically abnormal anatomic substrate, but cyanosis, myocardial dysfunction, and operative scars play an important role in the development of heart failure. For risk stratification, Nt-proBNP has recently been shown to be of value in adult CHD patients. Nt-proBNP is not only associated with ventricular function and exercise capacity, recently it was proven to be predictive for mortality and heart failure. In patients with a low Nt-proBNP no events occurred.[13,14] Treatment of heart failure in adult CHD patients is extrapolated from other acquired causes of heart failure, but is not evidence-based, simply because no good data exist. Of course, where possible, treatment of the underlying disease must be undertaken. However, all trials of medical therapies have not shown clear benefit in the adult CHD population. On theoretical grounds Angiotensin-converting enzyme-inhibitors and β-blockers can be used, extrapolating the benefits found in patients with acquired heart disease. However, the few studies performed in adult CHD patients have failed to show clear benefit.[15,16] As these studies were typically small with a relatively short duration of follow-up, we cannot conclude that these therapies have no effect, but clearly larger trials are needed, and at this moment there is no evidence for positive effects. In patients with a systemic right ventricle, the use of Angiotensin II receptor blockers showed no significant effect on the defined endpoint. However, in a sub-analysis of patients who were symptomatic, a positive effect was found.

Mechanical circulatory support may be an option in the (near-)future. However, a recent study of biventricular or total artificial heart support in patients with tetralogy of Fallot showed a high incidence of complications and also here the risk-benefit ratio is less favourable compared with other patient groups.[17]

With increasing numbers, more adults with CHD patients need hospital admission; moreover, the in-hospital mortality due to adults with CHD shows a clear rise.[18] End-of-life care including bereavement care needs attention and this crucial part of our profession becomes an important part of our clinical work (Figure 1). Patients often have unrealistic expectations for options like assist devices or heart transplantation and physicians are often too late in initiating communications on end-of-life.[19] The attitude of care providers is crucial to timely initiate important conversations, no longer on what can be done, but now focusing on how the patient wants to die. These conversations are very difficult but at the same time very important and often very rewarding.

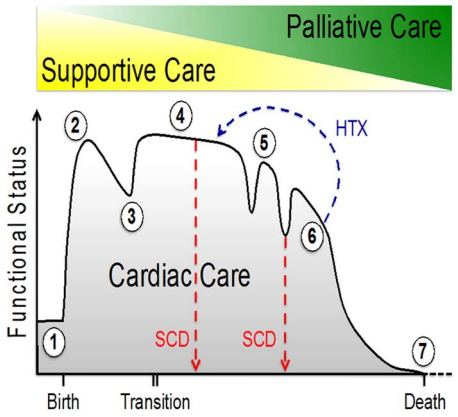

Figure 1.
Lifelong care for patients with congenital heart disease. (1) Parental prenatal support. (2) Initial surgical repair/palliation. (3) Re-interventions during childhood or adolescence. (4) Plateau of variable lengths in adulthood. (5) Variable adverse cardiac events and functional decline with variable slope, intermittent exacerbations that respond to rescue efforts, and/or adult re-interventions or procedures. (6) Refractory symptoms and limited function. (7) End-of-life care including bereavement care. HTX = heart transplantation; SCD = sudden cardiac death.

Finally, a fantastic presentation was provided by Dr Carole Warnes from the Mayo Clinic with the title "The courage to be imperfect". In this presentation she expressed her profound respect for pioneering cardiothoracic surgeons; courageous having to devise new surgical techniques and overcoming many challenges, such that now we have such excellent surgical outcomes in our patients. Cardiologists also have to be courageous; they have to recognise their lack of knowledge and have the courage to ask for help and refer when necessary. Finally, our patients, who learn so much resilience, compassion, and empathy, surely have the most courage of all, and continue to teach us so much about what is important and the value of life and our lives (Fig 1).

Acknowledgements

None.

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

https://doi.org/10.1017/S1047951117002220 Published online by Cambridge University Press

Conflicts of Interest

None.

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