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Review article

Health in adults with congenital heart disease

Judith A.A.E. Cuypers [a,*], Elisabeth M.W.J. Utens [b], Jolien W. Roos-Hesselink [a]

[a] Department of Cardiology, Erasmus MC, Rotterdam, The Netherlands
[b] Department of Child and Adolescent Psychiatry/Psychology, Erasmus MC—Sophia Children's Hospital, Rotterdam, The Netherlands

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ABSTRACT

Since the introduction of cardiac surgery, the prospects for children born with a cardiac defect have improved spectacularly. Many reach adulthood and the population of adults with congenital heart disease is increasing and ageing. However, repair of congenital heart disease does not mean cure. Many adults with congenital heart disease encounter late complications. Late morbidity can be related to the congenital heart defect itself, but may also be the consequence of the surgical or medical treatment or longstanding alterations in hemodynamics, neurodevelopment and psychosocial development. This narrative review describes the cardiac and non-cardiac long-term morbidity in the adult population with congenital heart disease.

Contents

1. Introduction

Congenital heart defects are the most common congenital defects, accounting for one third of all major congenital anomalies. The current incidence is about 8 per 1000 live births [1]. About 50% of these children will have a mild defect which requires no immediate treatment. Some defects, for example small ventricular septal defects, may even close spontaneously in childhood. The

* Corresponding author at: Department of Cardiology, Erasmus Medical Center, P.O. Box 2040, 3015 CA Rotterdam, The Netherlands.
E-mail address: j.cuypers@erasmusmc.nl (J.A.A.E. Cuypers).

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other half will need treatment early in life. Before cardiac surgery became possible, many of these children died in childhood and only 15% of all children born with a heart defect reached adult age. Especially children born with more complex, cyanotic heart disease, had little chance to survive their first year. Nowadays, due to the great successes of cardiac surgery and perioperative care, over 90% of children born with a cardiac defect will reach adulthood [2,3].

2. Changing epidemiology of congenital heart disease

Since the introduction of cardiac surgery in the mid-1950s, there have been tremendous improvements in diagnosis, surgical treatment and perioperative care. This has resulted in a large increase in the prevalence of adult congenital heart disease [4]. Since long, adults with congenital heart disease have indeed outnumbered the children with congenital heart disease [3]. On top of that, the amount of adult patients with complex congenital heart disease is increasing. Only the number of adult patients with unrepaired cyanotic congenital heart disease and those with Eisenmenger physiology is decreasing due to the better recognition and repair of congenital heart defects [5]. The median age of adult patients with congenital heart disease is gradually increasing too. More and more patients reach middle age and even patients above 60 are not a rarity any more. Their prevalence has been reported to have increased sixfold since 2000 [6]. All these changes will have a substantial impact on healthcare resources. As in all tertiary centers for congenital heart disease, we have seen a clear increase in outpatient clinic visits, which is represented in Fig. 1. Also, the older patients with congenital heart disease will need more hospitalizations and interventions than younger patients [6].

3. Contemporary survival of patients with congenital heart disease

Historically, survival was assumed to be decreased in all forms of congenital heart disease, whether simple or complex [7]. Perioperative mortality, which was substantial in the earliest era of congenital cardiac surgery, will have contributed to this idea. Nowadays, life expectancy has improved substantially and the age at death is increasing [8]. Long-term survival prospects in patients with mild, isolated congenital heart defects are excellent and not different from the general population. In patients with complex defects, survival prospects are significantly worse [9]. This was confirmed by the results of a longitudinal cohort study in our center: survival up to 40 years after ASD repair was similar to the general population (91%) [10], and only mildly decreased after VSD repair (86%) [11], but after repair of Fallot or Mustard operation for transposition of the great arteries, it was clearly decreased (72% resp. 68%) [12,13].

4. Causes of death in adult congenital heart disease

Mortality in congenital heart disease is mainly cardiovascular, more specifically due to heart failure and sudden cardiac death [14]. Life expectancy decreases with disease severity. With increasing life expectancy of congenital heart disease patients, non-cardiac causes of death will of course become more prevalent.

5. Residual cardiac problems after repair of congenital heart disease

Due to the greatly improved survival, focus has shifted from mere quantity of life to quality of life. Although in the earliest eras of surgical repair many patients were considered “cured”, we have since learned that this is not the case. Cardiac function often is not completely normal and many patients may have late complications [10–13].

5.1. Reoperations

Reoperations may be necessary for residual defects, that were not or could not be repaired at the time of the initial surgery. New abnormalities may also develop as a result of the surgical techniques used. These sequellae may need re-intervention many years after the initial operation. For example, severe pulmonary valve regurgitation is frequently encountered late after repair of tetralogy of Fallot, because large pericardial patches were originally used to widen the obstructed right ventricular outflow tract and pulmonary valve. The resulting pulmonary regurgitation was thought to be harmless, but proved not to be benign at all [15]. Reoperation for severe pulmonary regurgitation is now the most frequent re-intervention in adults after Fallot repair. In our study, re-intervention rates varied from 2% after ASD repair to 45% after Fallot repair and the Mustard operation [10–13].

5.2. Arrhythmias

Arrhythmias do occur in patients with complex as well as simple congenital heart disease. The incidence is increasing with follow-up duration. Arrhythmias can be related to surgical scars, anatomical abnormalities, or chamber dilatation due to hemodynamic overload. Atrial arrhythmias are usually benign, but can be ill tolerated or even lethal in patients with complex lesions such as single ventricle anatomy (Fontan circulation) or a systemic right ventricle, as is the case in patients after an atrial switch operation for transposition of the great arteries [16]. Ventricular arrhythmias are less often encountered but are potentially life-threatening. In our study, ventricular tachycardia had occurred in 6% of the patients after Fallot repair and Mustard operation after 40 years of follow-up. Indications for ICD implantation are less well established in this patient group. Bradycardia and AV-blocks do also occur, and may require pacemaker implantation. In our data, the cumulative incidence of pacemaker implantation after 40 years varied from 6% in the patients after ASD or VSD repair, to 33% after the Mustard operation [10–13].

5.3. Heart failure

Heart failure is infrequently seen in childhood, but may develop in adult life. Ventricular function may be damaged due to perioperative factors, or by longstanding hypoxemia in cyanotic defects before repair took place. After repair, ventricular volume or pressure overload due to residual abnormalities may compromise ventricular function in the long run. Especially in patients after atrial switch operation for transposition of the great arteries, who consequently have their right ventricle serving as the systemic ventricle, ventricular function is known to deteriorate over time [17,18]. In our study, only 2% of all patients had a normal systemic right ventricular function 35 years after the initial surgery [13]. Nowadays, atrial switch operations for TGA have been replaced by the arterial switch operation. However, many patients who underwent an atrial switch operation are still alive today. Although the outcome after the arterial switch is better, a subgroup of these patients will develop ventricular dysfunction [19,20].

5.4. Sudden cardiac death (SCD)

Sudden cardiac death (SCD) occurs more often in congenital heart disease patients than in the general population. Its estimated overall annual incidence is 0.09% [21]. Patients with repaired