Adult congenital heart disease

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Abstract

Continued advances in the understanding and management of congenital heart disease (CHD) mean that over 90% of children born with CHD now survive to adulthood. This in turn results in greater numbers of adult patients presenting for medical and surgical care at non-specialist centres. A simple classification of adult congenital heart disease (ACHD) according to complexity can help clinicians to understand the implications of the specific cardiac anomaly encountered. Issues relating to the conduct of anaesthesia in ACHD patient include careful attention to euvolaemia, the preservation of sinus rhythm and cardiac output, and in complex patients manipulating the balance between systemic and pulmonary blood flows. Additionally, effective antibiotic prophylaxis and the prevention of either excessive bleeding or thromboembolism are vitally important. It should not be forgotten that although many patients with simple or repaired cardiac lesions may be very well managed in a non-specialist unit, those with Eisenmenger's syndrome or severe pulmonary hypertension have an extremely high risk of death in the perioperative period, and in all but life-threatening situations should always be managed within specialist centres.

Keywords adult congenital heart disease; antibiotic prophylaxis; bleeding; classification according to complexity; Eisenmenger's syndrome; pulmonary hypertension; thromboprophylaxis

Royal College of Anaesthetists CPD Matrix: 2A03

Learning objectives

After reading this article, you should be able to:

- understand a complexity-based classification of adult congenital heart disease (ACHD)
- list three important physiological implications of long-standing congenital heart disease
- identify those patients who should be referred to a specialist unit for care
- recognize the risks associated with pregnancy in patients with different complexity ACHD

Adult congenital heart disease (ACHD), also known as grown-up congenital heart disease (GUCH) refers to patients who have survived into adulthood despite being born with structural abnormalities of the heart or great vessels.[1] Prior to the advent of corrective surgery or interventional cardiology, as many as 80% of these patients died in childhood, whereas the latest reports suggest that over 90% of children born with CHD now survive to adulthood.

Population estimates suggest there are approximately 1.2 million ACHD patients in Europe and a further 2 million in the USA. In the UK there are more than 250,000 CHD patients over the age of 16 years; exceeding the total number of children with the disease. Approximately 90% of these patients are diagnosed in infancy or childhood but 10% are not diagnosed until adulthood (e.g. secundum atrial septal defect, Ebstein's anomaly, coarctation of the aorta, and congenitally corrected transposition).

Approximately 40% of ACHD patients have simple lesions, or have undergone curative treatment[2] requiring little or no ongoing specialist cardiac care. A further 35–40% have moderate complexity lesions and require access to specialist input, and the remaining 20–25% have complex lesions that require lifelong specialist supervision and support.

Classification

In childhood it is common to classify CHD by anatomical analysis, or the presence or absence of cyanosis. Patients with ACHD present with the lesion distribution shown in Table 1, and are better classified according to simple, moderate or severe complexity (Table 2). This allows stratification of risk and the level of specialist care commonly required, as well as meaningful analysis of morbidity and mortality data.

Approximately 10% of patients present with ACHD in adult life, mostly with simple disease. However there have been occasional case reports of late-presenting moderate or severe complexity disease.

Even with specialist care, patients with ACHD have a higher burden of morbidity and a shorter life expectancy than those with structurally normal hearts. The majority of these deaths (77%) relate to increased cardiovascular risk; 45% due to heart failure, and a further 19% from sudden cardiac death.[3]

There is considerable variation in the occurrence of different lesions in ACHD and in their chronic sequelae; and it is beyond the scope of this review to describe them all, however some key conditions are discussed in more detail below (Table 2).

Simple lesions

Simple ACHD may be divided into small unrepaired septal defects without significant shunt, larger previously repaired septal defects, previously closed patent ductus arteriosus, and isolated valvular disease.

Of these simple lesions, ventricular septal defect (VSD) is the most common in childhood, however as 40% close spontaneously by the age of 2 and 90% by the age of 10 they are less common in adults. Atrial septal defects (ASD) are less common at birth but are less likely to close during childhood and therefore present more frequently in adults; these are usually repaired surgically or with percutaneous device implantation.

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Distribution of diagnoses of patients with adult congenital heart disease in the Dutch national registry (CONCOR programme)[3]

Diagnosis	Percentage %
Atrial septal defect	17
Ventricular septal defect	14
Tetralogy of Fallot	11
Coarctation	10
Aortic stenosis	8
Pulmonary stenosis	7
Bicuspid aortic valve	6
Marfan's syndrome	5
Transposition of the great arteries	5
Pulmonary atresia	2
AVSD	2
Ebstein's anomaly	2
Other diagnosis	11

Table 1

Isolated valvular heart disease usually has a slow rate of progression and the risk of cardiac death is low if the patient is asymptomatic.[4] However progressive ventricular dysfunction can occur without symptoms and these patients should remain under long-term cardiological review even if surgery is not contemplated.

Patients with simple lesions may otherwise require little ongoing specialist support. At least one review in adulthood is recommended, however, as they remain at increased risk of arrhythmias and ventricular dysfunction.

Moderate complexity lesions

The commonest moderate complexity lesion seen in adults is tetralogy of Fallot (ToF). This is characterized by right ventricular outflow tract (RVOT) obstruction, VSD, right ventricular hypertrophy and an aorta overriding the ventricular septum. Repair of ToF is generally carried out within the first year of life and usually consists of VSD repair and patch enlargement of the RVOT. If the patch traverses the pulmonary valve, the valve will be regurgitant resulting in right ventricular overload and progressive dysfunction with increased risk of arrhythmias and sudden death. Valve-sparing surgery is therefore preferred wherever possible. The 40-year survival of ToF patients is more than 90% but arrhythmia and sudden death are important late sequelae; additionally these patients are relatively symptom free in early adulthood and may be lost to follow-up despite possibly needing further intervention. Adult pulmonary valve replacement in ToF patients has been shown to reduce right ventricular size, improve long-term function, and reduce the risk of ventricular tachycardia (VT). This may however be high-risk surgery and transcatheter valve replacement has been more recently introduced with encouraging results.

Coarctation of the aorta, if severe, presents in the neonatal period. Milder forms can present in adult life, where the pathognomonic triad of hypertension, reduced femoral pulses and notching of the ribs on chest X-ray may be seen. Severe neonatal coarctation is life threatening, but even the milder forms repaired in adulthood are associated with significant morbidity including re-coarctation and persistent hypertension. A common association (85%) is a bicuspid aortic valve which may ultimately become regurgitant or stenotic. For these reasons such patients should have regular follow-up by a cardiologist. If re-coarctation, significant hypertension, and valvar disease have been excluded, these patients may be treated as functionally normal for the purposes of anaesthesia.

Ebstein's anomaly is a congenital displacement of the valve leaflets of the tricuspid valve into the right ventricle. The leaflets are variably restricted resulting in mild to severe tricuspid regurgitation, 'atrialization' of the morphologic right ventricle, and ultimately right heart failure. Ebstein's anomaly is frequently associated with ASD, patent foramen ovale (PFO) and Wolff–Parkinson–White syndrome, as well as non-cardiac abnormalities that may also require surgical intervention. Thus although Ebstein's anomaly accounts for only 1–2% of CHD it is seen disproportionately commonly in adult patients presenting for other procedures. Surgical repair or replacement of the tricuspid valve may be carried out at any age, however there remains significant susceptibility to perioperative arrhythmias and sudden cardiac death.

The last frequently occurring group of moderate complexity diseases is right- and left-ventricular outflow tract (RVOT and LVOT) obstruction. In each case these can be classified as valvar, subvalvar or supravalvar. Regardless of the anatomy the functional outcome includes ventricular hypertrophy and, eventually, failure; presenting with peripheral or pulmonary oedema. Additionally, long-standing LVOT may present with left ventricular subendocardial ischaemia secondary to hypertrophy, and RVOT may present with arrhythmias in the context of mild to moderate desaturation.

With all moderate complexity ACHD, it is wise to discuss patients presenting for non-cardiac surgery with their specialist centre to gain information regarding their previous history, current functional status, and future management plan, prior to proceeding with anaesthesia.

Severe complexity lesions

Complex ACHD can be divided into three main groups: patients who have undergone palliative surgery, patients with rare or complex anomalies not amenable to correction, and patients with an uncorrected but functionally balanced circulation. All ACHD patients with cyanotic disease will fall into one of these categories.

The commonest palliative procedures currently employed are the Fontan circulation, total cavopulmonary connection (TCPC), and systemic to pulmonary shunts. The Fontan and TCPC result in a univentricular circulation, with the superior and inferior venae cavae attached to the pulmonary artery either via the right atrium, or a conduit, respectively. These procedures are used in the treatment of tricuspid or pulmonary atresia, hypoplastic left heart syndrome, and unbalanced large atrioventricular septal defects and have significant limitations, particularly if the systemic ventricle is morphologically right sided.

The Fontan and TCPC result in an entirely passive pulmonary circulation, therefore forward flow and an adequate cardiac