Essentials of paediatric cardiac surgery

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Abstract

The history of cardiac surgery began with procedures to treat congenital heart disease, from the Blalock—Taussig shunt in the 1940s to the first use of extracorporeal circulation in the 1950s. The whole speciality of cardiothoracic surgery has grown from the work of these early pioneers and the wider applications of cardiopulmonary bypass have led to surgery for coronary artery disease and degenerative valvular disease in adults. Surgery for congenital heart disease now accounts for 10—15% of all cardiac surgery and continues to be at the forefront of the speciality in terms of innovation and evolving approaches. The speciality has advanced in parallel with advances in neonatal medicine and paediatric intensive care that have enabled an increasingly more complex range of conditions to become treatable. Outcomes have steadily improved, although this remains one of the most high-risk areas of modern surgery. Major congenital heart disease is a very serious condition and only 15% of children survived infancy prior to the surgical era. Now over 85% of children survive into adulthood and, for the first time, there are now more adults alive with congenital heart disease than there are children. Caring for these patients has led to the development of 'adult congenital heart disease' as an emerging specialist area. Surgery has become increasingly specialized and technically demanding with a trend to repair lesions at an earlier age and to aim for complete anatomical repair in infancy rather than a series of staged procedures. Over two-thirds of all procedures are now carried out before the age of 1 year.

Keywords Cardiac surgery; cardiothoracic surgery; congenital heart disease; neonatal surgery

Introduction

The heart is the most common major organ system to be affected by congenital abnormalities with an incidence of 0.8 per 100 live births. Although common, the majority of conditions are minor and do not require surgical intervention.

Congenital heart disease can be broadly classified into defects in septation of the circulation (ventricular septal defects, atrial septal defects, truncus arteriosus, etc.) and under-development/atresia of the structures and chambers of the heart (e.g. pulmonary atresia, hypoplasia of the aortic arch). Often multiple lesions occur together.

Another classification is to divide lesions into cyanotic and acyanotic conditions. Cyanotic conditions imply that there is an obligatory right to left shunt within the circulation One of the most challenging groups to manage are circulations in which one of the ventricles is underdeveloped such that it cannot support the circulation unaided — these are referred to as 'functionally univentricular' circulations and, although they account for only 5% of all congenital heart disease, they provide a considerable amount of the surgical workload. The most common conditions are listed in Table 1.[1]

Surgery aims to restore normal anatomy wherever possible. However, where the anatomy is not correctable or too complex to consider single-stage correction, it may be necessary to perform procedures that achieve a safe, balanced circulation without necessarily achieving normal physiology. These include systemic pulmonary artery shunts to augment pulmonary blood flow or pulmonary artery banding to limit pulmonary blood flow in cases of over-circulation. These are referred to as 'palliative' procedures because they are not corrective; however, the term should be used with caution in view of the more common usage of the term in the setting of terminal disease. Many of these procedures are temporizing measures until definitive repair is performed at a later age.

Most procedures are performed using cardiopulmonary bypass ('open heart surgery') via a midline sternotomy, although 10—15% do not require bypass and may be performed via sternotomy or thoracotomy ('closed procedures'). These include simple coarctation repairs, arterial shunts and pacemaker procedures. Major tracheal abnormalities can also be corrected on cardiopulmonary bypass since as there is no need to ventilate the patient while the tracheal reconstruction is carried out.

Over 3500 open heart procedures are performed in children every year in the UK.[2] One third of surgical procedures are now performed in neonates (i.e. under 30 days of age) with a further one third performed in infancy (i.e. under 1 year of age). In complex lesions the surgeon needs to balance the risk of a long bypass procedure with alternative strategies that palliate the circulation and allow for completion of the repair at an older age. However, there is an increasing trend towards planning for complete anatomical correction at a younger age and bypass technology has advanced such that cardiopulmonary bypass in children under 2.5 kg is now regarded as routine.

A great deal of corrective surgery requires some sort of reconstruction or enlargement of underdeveloped or absent structures in the heart, thus one of the great challenges in congenital heart surgery is the need to allow for growth and to use autologous tissues wherever possible. Inevitably, with many conditions children will outgrow artificial valves and conduits leading to a series of re-operations during childhood and the need for life-long specialist follow-up. Current research is focused on using stem-cell technology to grow patches and valves from autologous tissue or to use biological 'scaffolds' that can ultimately be seeded and repopulated by native cells.[3]

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Common congenital heart conditions and incidence

Lesion	Abbreviation	Average incidence (% of patients with cong heart disease)	Range (%)
Ventricular septal defect	VSD	30.1	24—34.6
Persistence of arterial duct	PDA	9	5.5—12.6
Atrial septal defect	ASD	7.5	4.3—11.2
Atrioventricular septal defect	AVSD	3.9	2.4—7.4
Pulmonary stenosis	PS	7.2	2.7—10.8
Aortic stenosis	AS	5.2	3.7—8.4
Coarctation of aorta	CoA	6.2	3.4—9.8
Transposition of great arteries	TGA	4.8	2.6—7.8
Tetralogy of Fallot	Fallot's	5.6	3.7—8.6
Common arterial trunk	Truncus	1.2	0—2.5
Tricuspid atresia	TA	2.3	0—4.5
Hypoplastic left heart	HLHS	2	0.6—3.4
Double inlet ventricle	DILV/DIRV	0.75	0—1.7
Double outlet right ventricle	DORV	0.2	0—1
Totally anomalous pulmonary venous drainage	TAPVD	1.1	0—2.8
Miscellaneous		13.7	2.8—21.7

Adapted from Anderson RH et al., 1989.

Table 1

Examples of commonly repaired lesions

These two examples show what is now routinely achieved in paediatric heart surgery and the importance of life-long follow-up:

1) **Tetralogy of Fallot:** This is the most common cyanotic heart condition and accounts for 5—10% of all congenital heart disease. The condition is characterized by a large ventricular septal defect together with multi-level obstruction to the outflow tract of the right ventricle. Untreated, children become increasingly cyanosed as the muscular obstruction to the outflow tract worsens. The anatomy is shown in Figure 1a.

   Historically, the condition was originally palliated with a systemic pulmonary artery shunt (the Blalock—Taussig shunt) to provide additional pulmonary blood flow until the child was considered old enough for definite repair. The shunt procedure is now much less commonly performed, and usually reserved for small neonates where complete repair may be difficult. Nowadays, surgical repair is generally performed at 4—5 months of age when the child is around 6—7 kg in weight. The operation is performed via a median sternotomy on cardiopulmonary bypass, cannulating both the superior and inferior vena cava so that the surgeon can work through the right atrium. The VSD is closed using a prosthetic patch, working through the tricuspid valve via the right atrium (see Figure 2a). The right outflow tract is enlarged by dividing and resecting muscle bundles but usually also requires the placement of a patch across small pulmonary artery to establish an adequate sized outflow tract to the right ventricle (Figures 1b and 2).

   The procedure restores normal anatomy and physiology and is associated with a 98—99% survival in the modern era.[4] However, children need regular follow-up as they tend to develop increasing pulmonary regurgitation across the reconstructed outflow tract with consequent dilatation of the right heart. This is well tolerated and produces minimal symptoms, but most patients come to need a pulmonary valve replacement in early adulthood to restore pulmonary competence. This is a good example of the need for specialist follow-up and life-long surveillance in congenital heart disease.

2) **Transposition of the great arteries:** In this condition the aorta arises from the right ventricle and the pulmonary artery from the left ventricle, creating two parallel circulations. Babies are usually profoundly cyanosed and only able to survive due to a degree of mixing of the circulation that occurs through a patent foramen ovale and a patent ductus arteriosus (see Figure 3). Without intervention the condition is lethal. Babies are now frequently diagnosed ante-natally so that delivery and immediate post-natal management can be planned. The ductus arteriosus is kept open with an infusion of prostaglandin and the foramen ovale can be enlarged by passing an angioplasty balloon across it via the femoral vein (balloon atrial spetostomy). These procedures achieve optimal mixing of the circulation and stabilization of the circulation.

   Surgical repair is then planned within the first two weeks of life. It is important to intervene this early because the left ventricle is supporting the pulmonary circulation (working against a low resistance) and will rapidly lose muscle mass and 'involutes'. If surgery is delayed too long then the left ventricle will have become too weak to support the systemic circulation. Surgery achieves anatomical correction by switching the great arteries back into their correct arrangement. This is performed on cardiopulmonary bypass during which the great arteries are transected, mobilized and re-anastomosed. However, the most complex part of the surgery involves the coronary arteries, which arise from the native aorta (arising from the right ventricle): these have to be separately excised from the aorta,