Guide for Advanced Nursing Care of the Adult with Congenital Heart Disease

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Foreword

Do we need a textbook on the nursing care of adult congenital heart disease (ACHD) patients? Surely those of us who look after this complex family of patients are aware of how important collaboration and a multidisciplinary approach are; therefore the answer is yes, we need it.

In this volume Serena Flocco and co-editors coordinated a great panel of authors from many centers that encourage all of us to think more deeply about how important nursing approach is to these patients.

Epidemiology, anatomy, and pathophysiology are treated exhaustively and give a complete overview of the problem.

The first step in taking care of ACHD patients is to help them and their families to skip from childhood to adulthood; therefore the excellent chapter dedicated to transition is a milestone for nursing training because the nurses should lead this process.

The care of our ACHD patients is a lifelong story; therefore Part III of the textbook is very important indeed because the authors focus on curing these patients in different settings like the outpatient clinic, the hospital ward, the cath lab, and the critical area.

The fourth part is dedicated to a complex subject of heart failure, outlining a hot topic like cardiac transplantation.

ACHD patients need also to be helped during their daily life steps: planning or trying to avoid pregnancy, when they want to exercise, sport, etc. These points are treated in very comprehensive Parts V and VI.

Not surprisingly, there is a part completely dedicated to patients' education with particular attention to the hot topic of their health engagement; this part is fundamental because counseling may improve their quality of life.

The textbook concludes with a chapter devoted to nursing care in the final stage of their life which is a very delicate issue; a combined approach with both physician and nurse is the best option for developing an advanced care planning program dedicated to it.

The truly final part of this amazing editorial job is the one that remembers all of us of the important role of all the nurses in taking care of our CHD patients during the SARS-COV 2 pandemic, and the implication for them.

Overall, this excellent source of information and discussion will stimulate all the readers interested in the cure of ACHD patients to think beyond the frequent limits

of our management suggesting a different and more scientific manner to approach them. This textbook is a valuable help approaching the complex challenge that these patients represent for all of us.

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Massimo Chessa

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Part I

Epidemiology, Anatomy and Pathophysiology

Incidence of Congenital Heart Disease and Relevance in Adulthood

1

Basma Abdelkader Hammad, Alexandra Arvanitaki, and Michael A. Gatzoulis

1.1 Epidemiology of Congenital Heart Disease

Congenital heart disease is an array of anatomical defects that range from simple defects, which close spontaneously during early years of life, to complex cyanotic defects that necessitate early surgical repair. Incidence is approximately 7 confirmed cases per 1000 or 1 in every 145 babies born [1]. This figure obviously varies according to the population studied but is an approximation for many Western countries [2]. Table 1.1 illustrates the different congenital anatomical defects and corresponding prevalence.

Around two-thirds of all congenital heart disease is diagnosed in infancy, 30% in children, and 10% in adults (those over 16 years of age). Nowadays, there are more adults with congenital lesions that need surveillance and re-intervention in their adult life, and this implicates the delivery of care in adult congenital heart disease service [3].

The delivery of service to congenital heart disease expands from pediatric age till transition into adult care service. Most adults with congenital heart lesions are usually seen by the pediatric cardiologist. These patients have been the beneficiaries of advances in pediatric cardiology and cardiac surgery services exemplified by the fact that 96% of children with congenital cardiac lesions who survive infancy will live to at least 15 years of age [4, 5].

Approximately 17% of congenital cardiac conditions occur in association with a recognized syndrome that "causes" the defect [3]. However, the genetic

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Table 1.1 Incidence of congenital heart disease

Lesion	%
Ventricular septal defect (VSD)	28.8
Atrial septal defect (ASD)	18
Patent ductus arteriosus (PDA)	9.6
Atrioventricular septal defect (AVSD)	5
Pulmonary stenosis (PS)	5.5
Tetralogy of Fallot (TOF)	3.7
Aortic coarctation	3.7
Aortic valve stenosis (AS), bicuspid AV	2.4
Complete transposition of the great arteries (d-TGA)	3.4
Single ventricle	<1

contribution to congenital lesions is much greater. Over the last decade, numerous genetic loci and chromosomal abnormalities have been identified for a whole range of conditions. One only needs to look at the recurrence rate for mothers with congenital heart disease to realize that familial and genetic factors contribute to many of the most common lesions.

There is, however, a great geographic variation regarding the incidence of CHD, which remains high in developing countries, located in Africa and Asia (up to 2.5%), and lower (about 1.2%) in most developed countries. This difference is mainly attributed to the establishment of fetal screening programs and termination of pregnancy in middle- and higher-income countries. The etiology of CHD is multifactorial, due to both genetic predisposition and environmental influences throughout fetal and early life [6]. Although Turner syndrome, trisomy 21, a 22q11 deletion, and other syndromes are seen in 20% of CHD cases, the majority of CHD is non-syndromic. In terms of environmental factors, maternal exposure to toxins and chemicals such as pesticides, herbicides, and lithium, smoking, alcohol, obesity, diabetes mellitus, malnutrition, rubella, and low socioeconomic status have been associated with increased risk of CHD.

There is a great genotype–phenotype heterogeneity with a high causative variance relative to low frequency of specific CHD phenotypes [7, 8], which renders disease classification difficult. CHD defects are classified as simple, moderate, or severe, in terms of complexity [9]. The most common CHD subtypes worldwide are the simple defects, atrial septal defect (ASD) and ventricular septal defect (VSD), which account for about one-third of CHD. On the other hand, the incidence of newborns with single ventricle physiology and other severe CHD types has declined over the past 30 years among all regions, likely associated with prenatal screening [10].

Over the last decades, major breakthroughs in pediatric cardiology, cardiac surgery, and interventional cardiology have dramatically improved the course of CHD, with a substantial decline in early mortality globally and especially in high-income countries [11]. In the 1950s, survival of children born with CHD was limited to 15%. Nowadays, more than 90% of these children survive well into adulthood. Therefore, there has been a shift in the disease landscape from infancy and

childhood toward adulthood, with adults accounting for two-thirds of the CHD population. In the United States, there were an estimated 1.4 million adults with CHD, of a total CHD population of 2.4 million in 2010 [12].

Although mortality continues to decline, CHD is never cured and is present as a chronic condition with cumulative complications over the life span, magnified since birth [13]. Therefore, the field of adult CHD cardiology has emerged, in order to manage not only young or middle-aged adults but also patients with CHD over 60 years old. Many adult patients are afflicted by residual hemodynamic lesions and also face additional challenges such as pregnancy, acquired heart disease, and non-cardiac pathology, necessitating integrated multi-disciplinary care at expert centers [14]. There is, consequently, an increasing need in educating physicians, nurses, psychologists, and other personnel in order to provide high-quality lifelong services to this rising population.

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Cardiac Defects

2

Basma Abdelkader Hammad and Michael A. Gatzoulis

2.1 Atrial Septal Defect

Atrial septal defect (ASD) is a communication between both atria that allows blood shunting across the defect. It's one of the common congenital defects with 6–10% as a single defect and in 30% associated with complex lesions [1]. There are several types based on the site of the defect at the interatrial septum (IAS); Fig. 2.1 demonstrates the anatomical location of the different types. Secundum ASD represents the most common type with a defect in the septum primum. Primum ASD is due to a defect in the endocardial cushions and accounts for a partial atrioventricular (AV) canal defect. Sinus venosus ASD is due to a defect in the atrial septum at the site of infolding of the vena cava; it could be superior at the superior vena cava (SVC) and is usually accompanied by partial anomalous pulmonary venous drainage with the right upper pulmonary veins entering into the right atrium at the site of the defect. Less common is inferior sinus venosus ASD at the site of entrance of the inferior vena cava (IVC). The least common type is coronary sinus defect where it opens into the left atrium [1].

2.1.1 Clinical Presentation

The clinical presentation depends on the size of the defect and degree of shunting. The degree of shunting varies based on the size of the defect, right ventricle (RV),

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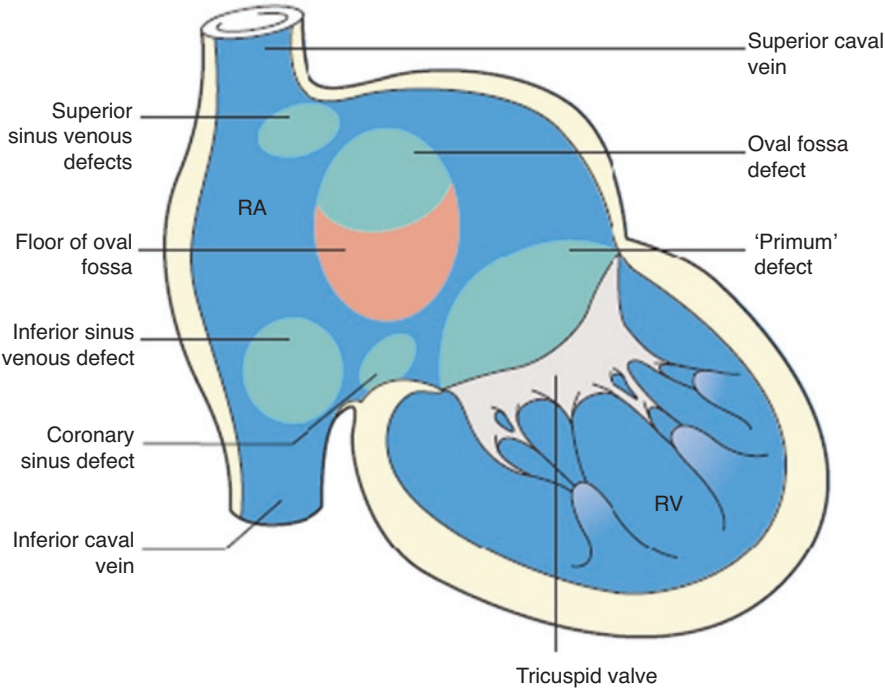

Fig. 2.1 Types of atrial septal defects

and pulmonary vasculature compliance. Most of the time it presents as an audible murmur in an asymptomatic child or the patient has been referred to a pediatrician for other various reasons. Patients with large defects >10 mm with a shunt Qp: Qs >1.5 tended to present with palpitations and exertional shortness of breath due to enlargement in the right-sided cardiac chambers (right atrium and right ventricle) [2].

In diagnosis, we should look for the size of the defect, degree of shunting, size and function of the right ventricle, pulmonary artery (PA) pressure, associated lesions, and suitability of percutaneous closure of the defect.

2.1.2 Management

The timing of intervention is based on the symptoms, degree of shunt, right ventricle dilatation, and pulmonary artery pressure.

Elevated pulmonary artery pressure with pulmonary vascular resistance (PVR) >5 Wood units in patients is considered contraindication for defect closure, and trial pulmonary hypertension therapy is recommended.

Percutaneous closure is the first option in secundum ASD but based on the availability of suitable rims in echocardiogram and size of the defect (max diameter

should not be more than 40 mm). Recently, a percutaneous approach adapted for sinus venosus with anomalous pulmonary venous drainage with suitable anatomy allows stenting SVC and redirecting pulmonary venous flow to the left atrium [2, 3].

Concomitant arrhythmia is commonly addressed before, during, or after the procedure based on the arrhythmia burden, degree of heart dilation, and response to medical therapy [4].

2.1.3 Complications

Arrhythmia (atrial flutter/fibrillation/atrial tachycardia)
Paradoxical embolization
Pulmonary hypertension
Right ventricle dilatation and dysfunction if left untreated due to volume overload

2.2 Ventricular Septal Defect

Ventricular septal defect (VSD) is the most common congenital anomaly with incidence of >20%. The interventricular septum (IVS) is composed of a small perimembranous part just beneath the aortic valve and a large muscular part that is divided into inlet, outlet, or trabecular portions [5].

There are different types of VSD based on anatomical location within the IVS (Fig. 2.2). They are perimembranous (supracristal), inlet (part of atrioventricular canal defects), muscular, and outlet (subpulmonic, doubly committed). The

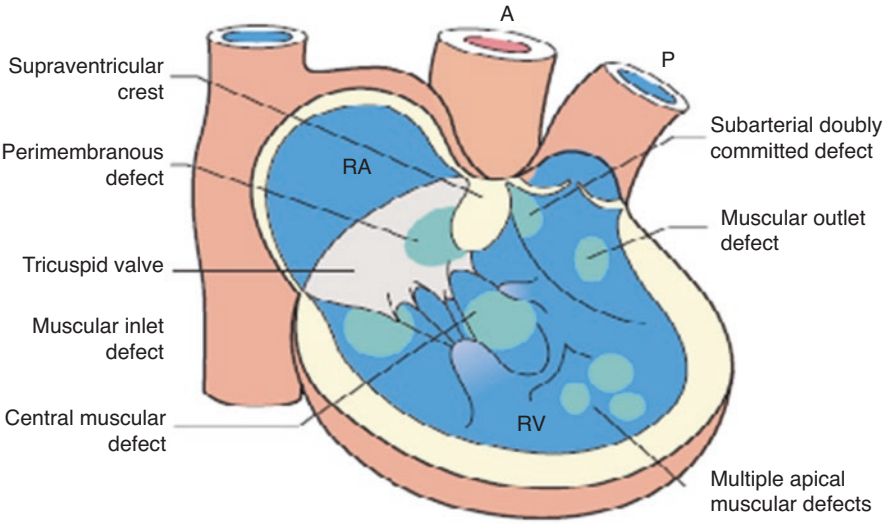

Fig. 2.2 Types of ventricular septal defects

perimembranous is the most common type that accounts for 40–60%. VSD can present as a single defect or can be associated with other complex anatomy as we will discuss later in cyanotic defects.

2.2.1 Clinical Presentation

The clinical presentation depends on the size of the defect, degree of shunting, and respective chamber dilatation.

Restrictive VSD in which the blood shunted Qp: Qs from the left to the right ventricle is less than <2.5 usually asymptomatic and picked up with audible murmur detected during examination for other non-cardiac illnesses.

Non-restrictive VSD with Qp: Qs >2.5 tended to present early with manifestation of heart failure, recurrent chest infections, and failure to thrive early in pediatric age or might develop symptoms of decompensation later in adult life with progressive enlargement of the left-sided cardiac chambers.

The common complications that might develop and warrant surveillance are progressive aortic valve regurgitation, and it tends to happen in perimembranous VSD when one of the aortic cusps prolapsed into the defect due to the Venturi-like effect of the shunted blood jet. The other common complications are infective endocarditis at the site of the blood jet with seedling of bacteria and development of vegetations on the right side and double-chambered right ventricle (RV) where the RV is divided into two chambers, one with low pressure and the other with high pressure with a hypertrophied muscle band as a sequel of the shunted blood direction [6–8].

In diagnosis, we should look for the size of the defect, pressure across the defect, degree of the shunt, left ventricular size, pulmonary artery pressure, and possible complications as to the degree of aortic valve regurgitation and the presence and degree of right ventricle outflow tract (RVOT) obstruction, if any.

2.2.2 Management

Treatment is based on the degree of shunt, patient symptoms, pulmonary artery pressure, and presence of complications [7, 9].

Restrictive VSD should be surgically repaired if the pulmonary artery pressure is normal with PVR <5 Wood units.

If PVR is >5 Wood units, the defect can be closed if pulmonary artery pressure showed a strong evidence of pulmonary reactivity with a response to pulmonary vasodilator (oxygen, nitric oxide).

Percutaneous closure of perimembranous VSD is considered an option nowadays based on the relation of the defect to the aortic valve cusps [5].

Untreated non-restrictive VSD with a high degree of shunting tended to develop increased pulmonary vascular resistance with eventual shunt reversal, development of cyanosis, and Eisenmenger syndrome.

Long-term follow-up is recommended for detection of complications in unrepaired patients, and repaired patients tended to develop arrhythmias due to cardiac chamber dilatation [7].

2.3 Atrioventricular Septal Defects

Atrioventricular septal defects (AVSD) are due to a defect in endocardial cushions, and it compromises a spectrum of presentation that can be like ASD, VSD, or both [10].

There are three types of AVSD: partial, intermediate, and complete (Fig. 2.3). The presentation is based on the site of the defect; in partial AVSD, there is a primum ASD and cleft left atrioventricular (AV) valve, and patient presentation is like ASD. In intermediate, it includes primum ASD, restrictive VSD, and two separate AV valves and presentation is like ASD. In complete type, there is a primum ASD, inlet non-restrictive VSD, and common AV valve, and presentation is like non-restrictive VSD and this type tended to develop pulmonary hypertension early in life if left untreated.

AVSD might be the single defect or associated with more complex lesions like tetralogy of Fallot (TOF).

AVSD tended to be more common in patients with trisomy 21 with an incidence of >75% for the complete type.

2.3.1 Clinical Presentation

The clinical presentation depends on the type of the defect, which signifies the degree of shunt, chamber dilatation, and pulmonary artery pressure.

In partial and intermediate type, the clinical presentation is like ASD. However, in partial ASD, the presentation might be earlier due to the added effect of the cleft left AV valve that invites more volume load on the left side of the heart.

In complete type, the clinical presentation is like non-restrictive VSD with manifestation of heart failure, recurrent chest infections, and failure to thrive.

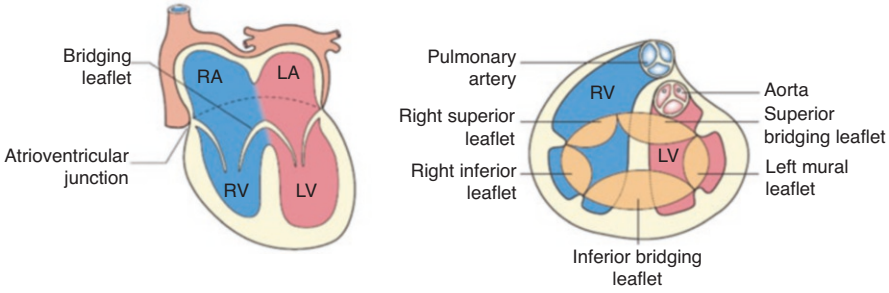

Fig. 2.3 Diagrammatic illustration of complete atrioventricular septal defect

The complications that might develop are arrhythmias (atrial flutter/fibrillation) due to increased size and elevated pressures in respective cardiac chambers. Around 3% of patients with AVSD complete type tended to develop complete heart block that warrants surveillance and close monitoring. Pulmonary hypertension and Eisenmenger syndrome tended to develop with trisomy 21 patients early in life. Left ventricle outflow tract obstruction is a common complication, and this develops due to the unwedging of the aortic valve. AVSD patients are at a risk of developing infective endocarditis, and proper emphasize on regular dental hygiene and avoiding tattoo, piercing, or any skin-invasive procedures are warranted [10].

2.3.2 Management

Treatment is based on the type of the defect and associated congenital lesions [11, 12].

Surgical repair is the standard treatment. For partial and intermediate types, treatment is like ASD with left AV valve repair. Complete type needs surgical repair with proper septation of the atria and ventricle along with reconstruction of the two separate AV valves.

One of the common late sequels is the recurrent regurgitation in both AV valves. This warrant proper surveillance, and most of the patients require second surgery for repair or replacement of the valves as the initial repair is not durable lifelong [13].

Another complication post surgical repair is AV valve stenosis that requires long-term follow-up to determine optimal timing for re-intervention prior to development of decompensation [14].

2.4 Left Ventricle Outflow Tract Obstructions

Left ventricle outflow tract obstruction encompasses subvalvular, valvular, and supravalvular aortic stenosis. It might be a single or multilevel obstruction.

Subvalvular aortic stenosis is an obstruction below the level of the aortic valve; it can be due to discrete membrane (most common) and fibromuscular ridge or tunnel like. It can be associated with another congenital anomaly due to anatomical configuration as in AVSD.

Valvular aortic stenosis including bicuspid aortic valve is by far the most common congenital defect with an incidence of 1–2% in normal population. In bicuspid aortic valve, there is a fusion between two cusps resulting in two unequal cusps with valve area narrowing and accelerated degeneration. Bicuspid aortic valve tended to be associated with aortopathy with dilatation and/or coarctation of the aorta.

Supravalvular aortic stenosis usually presents as part of Williams syndrome that involves neurodevelopmental delay and cardiac and facial defects, and this is due to 7q11.23 chromosome genetic deletion.

2.4.1 Clinical Presentation

The clinical presentation is variable based on the level and degree of obstruction and associated lesions [15, 16].

Subvalvular obstruction usually progresses over time and associates with a degree of aortic valve regurgitation. Clinical presentation could vary from shortness of breath to manifestation of heart failure, and low cardiac output symptoms (chest pain, dyspnea, syncope) depend on the degree of obstruction.

Valvular obstruction tends to progress overtime with a range of symptoms (dyspnea, chest pain, palpitation, syncope); however, some children are born with critical stenosis with manifestation of heart failure and failure to thrive.

Supravalvular obstruction tends to manifest early in life as being part of a syndrome with facial feature defects and neurodevelopmental delay [17]. They could be associated with other peripheral pulmonary stenosis, systemic arterial hypertension, or coronary ostial narrowing.

2.4.2 Management

Treatment is based on the level and degree of obstruction.

In subvalvular obstruction, surgical resection is recommended in symptomatic patients with a systolic gradient across the left ventricle outflow tract (LVOT) of >50 mmHg or in the setting of progressive aortic regurgitation (AR; more than mild). However, obstruction tends to recur later in life and warrants long-term follow-up to determine optimal timing for re-intervention.

Valvular obstruction requires treatment in the setting of severe (indexed aortic valve area <0.6 cm^2) and symptomatic aortic stenosis [18] or severe asymptomatic with reduced left ventricle systolic function (LVEF) <55%. In the setting of aortic dilatation, prophylactic surgery should be considered with aortic root diameter >55 mm to avoid dissection.

In congenital critical aortic stenosis, balloon dilatation is recommended to alleviate obstruction.

Valve replacement can be in the form of mechanical valve, biological valve, or Ross procedure in which the pulmonary valve implanted at the aortic position and pulmonary valve is replaced with homograft.

In supravalvular obstruction, surgical repair is recommended with a peak gradient >50 mmHg.

Patients with left ventricle outflow tract obstruction (LVOTO) require long-term follow-up for the possible complications that might develop post repair including recurrent subaortic membrane, progressive aortic valve regurgitation, morbidity related to Ross procedure with progressive valve degeneration and aortic root dilatation. Injury to AV node might happen at the time of surgery or in re-sternotomy, and those patients require permanent pacemaker implantation [19].

2.5 Coarctation of the Aorta

Coarctation is stenosis in the aortic arch usually at or just distal to the site of the patent ductus arteriosus (PDA). It accounts for 7% of congenital defects. This happens due to the narrowing implied by contraction of ductal tissue post closure of the PDA. It could be as the single lesion or associated with other anomalies. It is also associated with hypoplasia of the aortic arch or main branches and aneurysm formation [20].

Associated lesions that warrant surveillance are:

- Bicuspid aortic valve (with or without aortic stenosis), up to 85%
- Patent ductus, VSD
- Bronchial collateral network
- Anomalies of head and neck vessels, 5%
- Intracerebral berry aneurysms, 5%
- Proximal arteriopathy (ascending aorta, etc.)
- Multiple left heart obstructive lesions
- Shone's complex syndrome (multilevel left heart obstruction)
- Turner's syndrome (25% have coarctation)

2.5.1 Clinical Presentation

Presentation depends on the degree of stenosis, site, and status of collaterals.

In infants, they usually develop shock and heart failure following duct closure (ductal shock). Patients with good collaterals present late in life with audible murmur, early-onset systemic hypertension, and absent femoral pulse and rarely with cerebrovascular accidents.

2.5.2 Management

Surgical repair or catheter-based treatment is recommended at the time of diagnosis to reduce the rate of complications; there are different surgical techniques:

- Direct end-to-end anastomosis (common in childhood but interposition graft may be necessary in adulthood).
- Subclavian flap repair, by augmentation using the left subclavian artery (more often in children). Following surgery, the left radial pulses may be weak and left arm blood pressure artificially low.
- Patch grafting (has now been abandoned due to an increased risk of aneurysm formation).

Catheter-based treatment in the form of primary stenting is becoming a standard treatment for adults with native coarctation or in re-coarctation. This is a relatively safe and highly effective technique, although long-term follow-up data is not yet available. Its use should be confined to tertiary referral centers.

2.5.3 Complications

Long-term surveillance with annual follow-up is indicated due to the likelihood for developing complications in repaired patients.

- Early onset of systemic hypertension even in repaired patients
- Early onset of atherosclerosis, the commonest cause of death
- Re-coarctation
- Aneurysm formation at the site of repair
- Progression of concomitant bicuspid valve disease or other associated lesions

2.6 Patent Ductus Arteriosus

Patent arterial duct or patent ductus arteriosus (PDA) is a vessel communication connecting the proximal left pulmonary artery to the descending aorta just distal to the left subclavian artery (Fig. 2.4). It accounts for 12% of congenital heart lesions. It is a common lesion in premature infants (0.8%) and with maternal rubella [21].

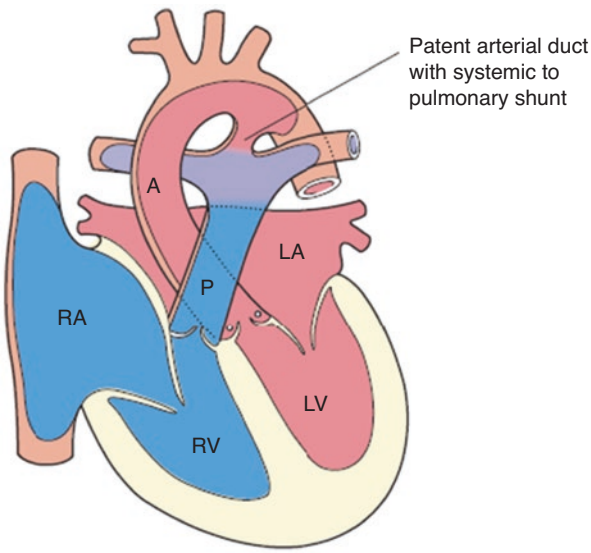

Fig. 2.4 Diagrammatic illustration of the patent ductus arteriosus

2.7 Clinical Presentation

This depends on the size of the duct and its hemodynamic impact.

Silent: Tiny PDA detected only by non-clinical means (usually echo); no heart murmurs audible.

Small: Audible long-ejection or continuous murmur, radiating to the back. Causes negligible hemodynamic change. Normal peripheral pulses and normal left atrial and left ventricular size without any pulmonary hypertension.

Moderate: Wide, bouncy peripheral pulses (as with important aortic regurgitation). Audible, continuous murmur. Causes enlargement of the left atrium and left ventricle and some degree of pulmonary hypertension (usually reversible).

Large: Usually in adults without Eisenmenger physiology. Signs of pulmonary hypertension. Continuous murmur is absent. Causes differential cyanosis (lower body saturations are lower than right arm saturations) and toe clubbing.

Patients with moderate to large shunts tended to present early in life with manifestation of heart failure, failure to thrive, and recurrent chest infection due to excessive blood flow.

2.7.1 Management

PDA closure in adults should be considered in the following situations:

- The presence of a PDA, except for (a) the silent tiny duct and (b) the presence of severe, irreversible pulmonary vascular disease
- The occurrence of an episode of endarteritis, irrespective of the size of the PDA

If pulmonary hypertension is present (pulmonary arterial pressure > two-thirds of the systemic arterial pressure or pulmonary arteriolar resistance exceeding two-thirds of the systemic arteriolar resistance), there must be a net left-to-right shunt of 1.5:1 or more or evidence of pulmonary artery reactivity with reversibility studies.

Device closure is the preferred method for the majority of PDAs in most centers today.

Surgical closure should be reserved for patients with PDAs too large for device closure. Very occasionally, ductal anatomy may be so distorted (ductal aneurysm or post-endarteritis) as to making device closure undesirable.

2.7.2 Complications

Closure device embolization
Residual pulmonary hypertension
Aneurysm formation and calcifications
Left ventricle dilatation

Arrhythmia
Endocarditis

2.8 Transposition of the Great Arteries (D-TGA)

Transposition of the great arteries is the most common congenital cyanotic heart disease in neonates. The estimated incidence is 1 in 3500–5000 live births, with a male-to-female ratio of 1.5 to 3.2:1. In d-TGA, there is atrioventricular concordance and ventriculoarterial discordance, i.e., the right atrium is connected to the right ventricle (RV) that gives rise to the aorta and the left atrium is connected to the left ventricle that gives rise to the pulmonary artery. This anatomical orientation is incompatible with life with two parallel circulations, unless there is a shunt with blood mixing between both circulations (Fig. 2.5). The shunt could be small PFO/ASD, PDA, or VSD. 50–60% of d-TGA has VSD.

2.8.1 Clinical Presentations

It is the most common cyanotic heart disease in the neonates. The presentation depends on the size of shunt and degree of mixing. Neonates are pink at birth and gradually develop progressive cyanosis once the duct is closed. The survival before surgical repair depends on the degree of shunt. Atrial septostomy (Blalock–Hanlon atrial septectomy or Rashkind balloon atrial septostomy) is emergency septectomy at the atrial level to promote blood mixing if there is no adequate natural shunt.

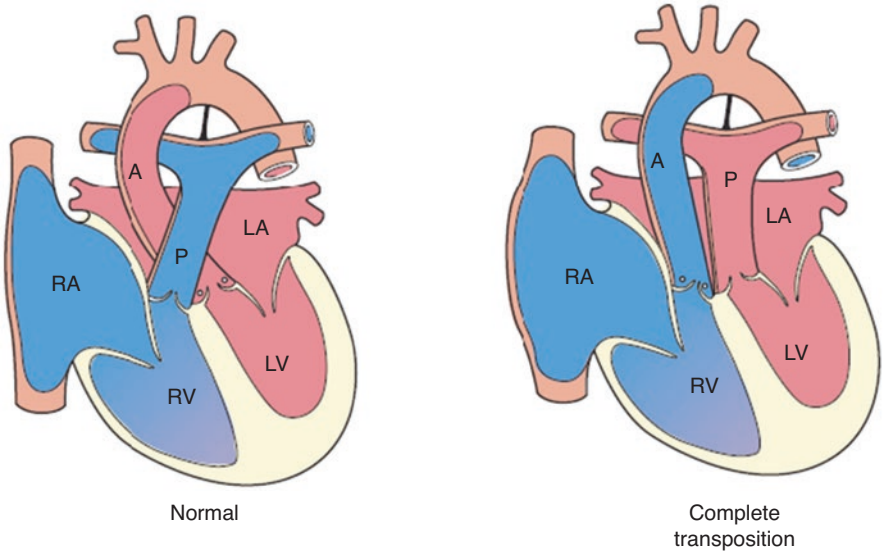

Fig. 2.5 Normal cardiac connection versus the transposition of the great arteries

2.8.2 Management

Different surgical repairs have been considered including:

Atrial switch (Mustard/Senning) [22]: This considered physiological repair more than anatomical one. In atrial switch, the venous flow is redirected where systemic venous caval flow baffled to the left ventricle and pumped into the pulmonary circulation via the pulmonary artery. The pulmonary venous flow is redirected to the RV and pumped into the systemic circulation via the aorta. The drawback of this surgical repair is that the right ventricle is acting as the system RV facing the systemic circulation, which is not a situation the RV is adapted to (high pressure circulation). Hence, over time, it hypertrophied and eventually fails [23].

Arterial switch [24]: This considered anatomical repair in which the great vessels are transacted and connected to the respective ventricles. Coronary arteries also reimplanted into the neo-aorta. The drawback of this repair is that the pulmonary artery is stretched to be anterior to the aorta that invites stenosis at the site of anastomosis or peripheral branch stenosis. The neo-aorta tends to dilate with neo-aortic valve leak. The reimplanted coronaries might develop ostial narrowing, compression, and ischemic consequence in the myocardium.

Rastelli operation: This considered anatomical repair in which the aorta is tunneled to the left ventricle via VSD and pulmonary artery connected to the RV via external conduit. This repair is considered in patients with d-TGA who are not candidate for arterial switch due to the presence of valvular/subvalvular pulmonary stenosis, or VSD. The drawbacks of this repair are that the conduit requires revision due to degeneration (stenosis/regurgitation) [25], risk of infective endocarditis, and LVOTO.

2.8.3 Complications

Atrial switch:

- Systemic RV failure
- Systemic AV (tricuspid) valve regurgitation
- Bradycardia, heart block, arrhythmias due atrial scarring

Arterial switch:

- Right ventricular outflow tract obstruction
- Peripheral pulmonary artery stenosis
- Neo-aorta dilatation
- Neo-aortic valve regurgitation
- Coronary ostial occlusion and myocardial ischemia

Rastelli:

- Right ventricle to pulmonary artery (RV-P) conduit degeneration (stenosis/regurgitation)
- LVOTO
- Residual VSD

Regular follow-up and surveillance for possible complications are mandatory in repaired d-TGA patients. Unrepaired patients are unlikely to survive with 90% mortality by 1 year.

2.9 Tetralogy of Fallot and Right Ventricular Outflow Tract Disorders

The main pathology in the TOF is anterior and cephalad deviation of the ventricular septum resulting in a non-restrictive VSD; an overriding aorta (but <50%); right ventricular outflow tract obstruction (RVOTO), which may be infundibular, valvar, or (usually) a combination of both, with or without supravalvar or branch pulmonary artery stenosis; and consequent right ventricular hypertrophy (Fig. 2.6).

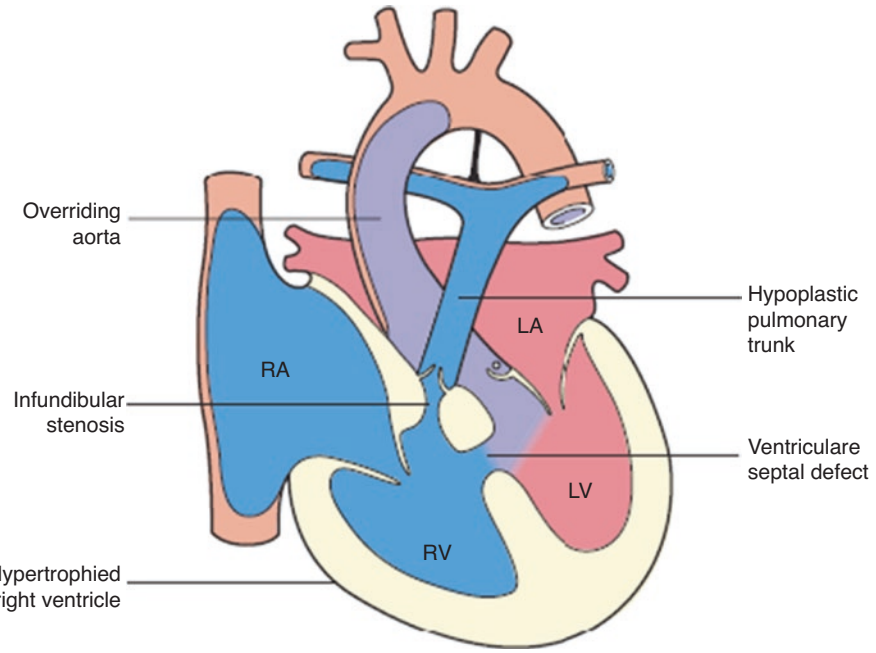

Fig. 2.6 Tetralogy of Fallot

TOF is the most common congenital cyanotic heart disease with an incidence of 10% or more. Approximately 15% of patients with tetralogy of Fallot have a deletion of chromosome 22q11.

2.9.1 Clinical Presentation

The clinical presentation varies depending on the degree of right ventricular outflow tract obstruction. With mild obstruction, the presentation is of increased pulmonary blood flow with dyspnea and minimal cyanosis, the so-called "pink tetralogy" or "a cyanotic Fallot." Most children, however, have significant RVOTO with consequent right-to-left shunt and cyanosis.

2.9.2 Management

2.9.2.1 Palliative Surgery

The aim is to increase pulmonary blood flow and this can be one of the following:

Blalock–Taussig shunt (classic or modified subclavian artery to pulmonary artery end-to-side shunt or interposition graft)
Waterston shunt (ascending aorta to right pulmonary artery shunt)
Potts shunt (descending aorta to left pulmonary artery shunt)

2.9.2.2 Corrective Surgery

The corrective surgery involves closing the ventricular septal defect with a Dacron patch and relieving the RVOTO. The latter may involve resection of the infundibular muscle and insertion of a right ventricular outflow tract or transannular patch.

2.9.3 Complications

Pulmonary Regurgitation (PR) Significant pulmonary regurgitation (PR) is almost always encountered when the transannular patch repair technique has been employed and warrants surveillance. PR is usually well tolerated if mild to moderate [26, 27].

Severe chronic pulmonary regurgitation, however, may lead to symptomatic RV dilatation and dysfunction. The severity of pulmonary regurgitation and its deleterious long-term effects are augmented by coexisting proximal or distal pulmonary artery stenosis or pulmonary artery hypertension.

Right ventricle dilatation and dysfunction [28, 29] as consequences to significant PR and/or residual RVOTO or peripheral pulmonary stenosis. This causes tricuspid regurgitation that adds on to the RV volume and function deterioration.

2.10 Residual Right Ventricular Outflow Tract Obstruction

Aneurysmal Dilation of the RVOT This is relatively common in patients with previous pericardial transannular patch repair and significant pulmonary regurgitation. This area acts as a focus for sustained ventricular tachycardia.

Residual Ventricular Septal Defect (VSD) Residual VSDs can be due to either partial patch dehiscence or failure of complete closure at the time of surgery.

Aortic Regurgitation (AR) with or without Aortic Root Dilation AR can be due to damage to the aortic valve during VSD closure or secondary to an intrinsic aortic root abnormality.

Left Ventricular Dysfunction [30] It can be due to chronic volume overload from residual shunt, AR, or inadequate myocardial protection during surgery.

Arrhythmia in the form of atrial ore ventricular tachycardia.

2.11 Endocarditis

2.11.1 Sudden Cardiac Death

Post TOF repair patients need annual follow-up for the residual lesions and the likelihood for complications and re-intervention.

2.12 Pulmonary Atresia

Pulmonary atresia accounts for 1–2% of congenital heart disease. A small percentage (10–20%) of individuals with tetralogy of Fallot have pulmonary atresia rather than outflow tract obstruction. There is the absence of a communication between the right ventricular cavity and the pulmonary trunk. This lack of communication can be subvalvular with muscular blockage of the outflow tract (most common) or at the valve [31].

Pulmonary blood flow could be unifocal in which the intrapulmonary arteries are connected to unobstructed, confluent pulmonary arteries with blood supply from a patent duct (Fig. 2.7).

It can be multifocal, which occurs in the majority of time (85%), when different parts of a lung are supplied from more than one source. The branch pulmonary arteries are confluent, but usually hypoplastic since blood supply is from systemic to pulmonary arterial collaterals.

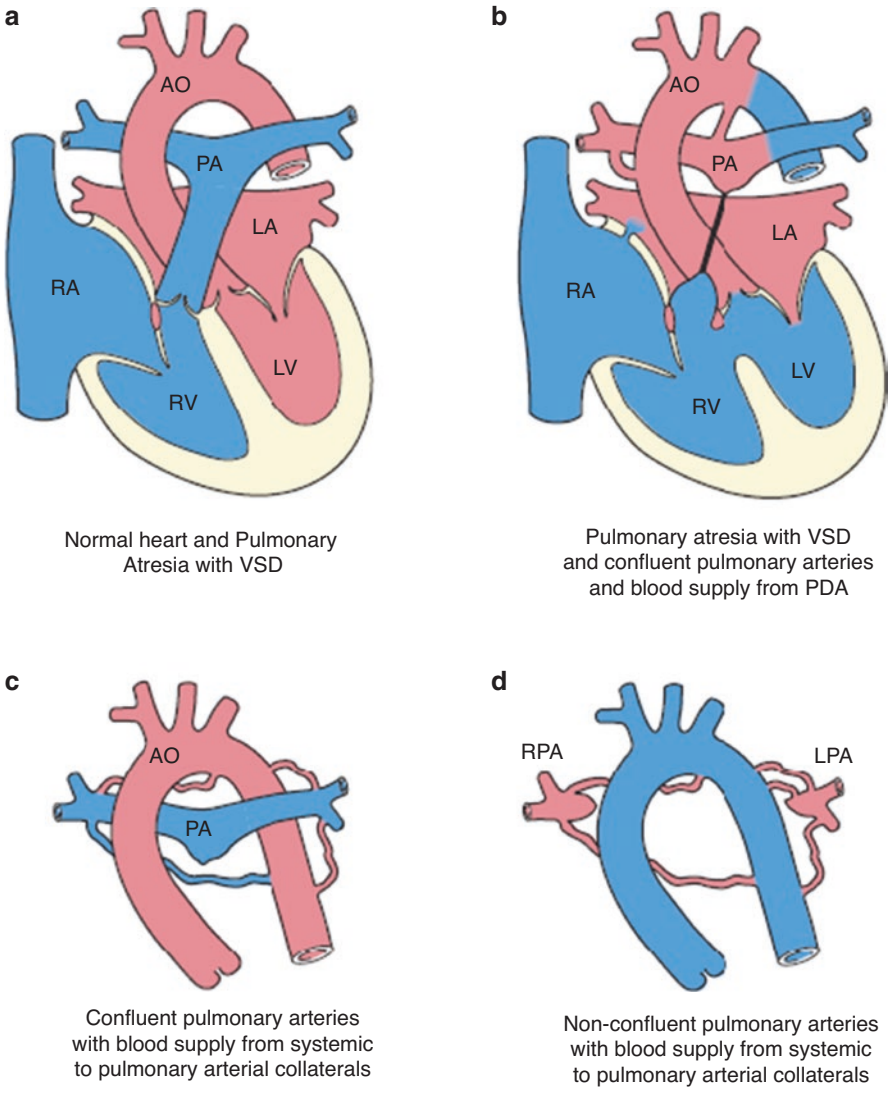

Fig. 2.7 Diagrammatic illustration of pulmonary atresia and pulmonary blood supply (**a**) Normal heart. (**b**) Pulmonary atresia with VSD. (**c**) Confluent pulmonary arteries with collaterals. (**d**) Non-confluent pulmonary arteries with collaterals

2.12.1 Clinical Presentation

It depends on the degree of the pulmonary blood flow. It can be cyanosis (inadequate flow), failure to thrive, recurrent chest infection, and heart failure (excess pulmonary flow).

2.12.2 Management

Survival is unlikely without surgery. The goal of traditional surgery is to close the VSD and to reconstruct the RVOT and pulmonary vasculature. How this is achieved depends upon the anatomy [31].

When there are collaterals or hypoplastic pulmonary arteries, a single-stage repair is not possible. A palliative surgery is done to promote growth of the pulmonary vasculature. Three options are possible.

- A systemic to pulmonary artery shunt.
- Right ventricular outflow tract reconstruction leaving the VSD open or at least fenestrated. This provides for a more uniform enlargement of the pulmonary arteries.
- A central-to-side shunt (hypoplastic pulmonary trunk attached to the side of the ascending aorta). This must be the correct size to promote growth without subjecting the lungs to excessive arterial pressure.

2.12.3 Complications

The long-term sequelae vary depending upon the type of surgical palliation or repair. The need for re-operation is about 10–15% over 20 years. Replacement of the pulmonary conduit is a recurring issue (freedom from re-operation at 10 years is about 55% and at 20 years is 32%).

Residual RVOTO
Residual VSD
Arrhythmia
Conduit endocarditis
Hemoptysis due to residual collaterals

2.13 Single Ventricle and Fontan Circulation

Fontan circulation (total cavo-pulmonary connection [TCPC]) is a palliative surgery for patients who are not candidate for biventricular repair as in double inlet left ventricle (DILV), tricuspid atresia, or pulmonary atresia with intact interventricular septum (IVS). In such palliation, the superior vena cava (SVC) flow is directed to the right pulmonary artery and the inferior vena cava (IVC) flow is tunneled via the right atrium or external conduit to the SVC bypassing the sub-pulmonary ventricle. In DILV, both AV valves are connected to a single ventricular cavity. This main ventricle is connected to a rudimentary chamber through a bulboventricular foramen. One great artery arises from the ventricle and the other from the rudimentary chamber. The single ventricle is left type in 80% of cases. Transposition of the great arteries occurs in 85% of cases with the most common form being “double-inlet left ventricle with L-TGA” (aorta arising from the rudimentary chamber). Pulmonary stenosis or atresia is present in about half the cases, providing some protection to the