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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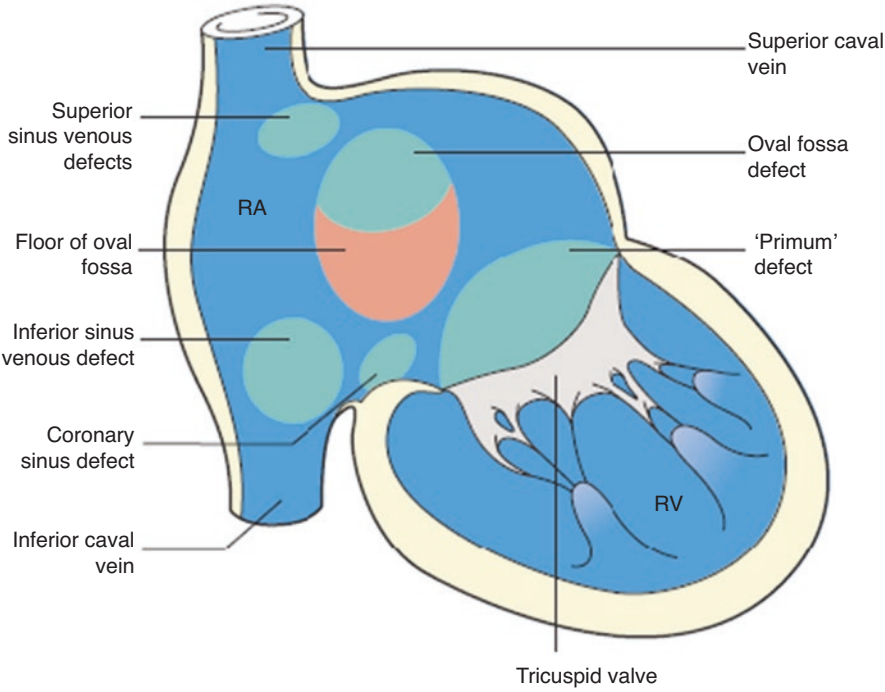

Fig. 2.1 Types of atrial septal defects

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

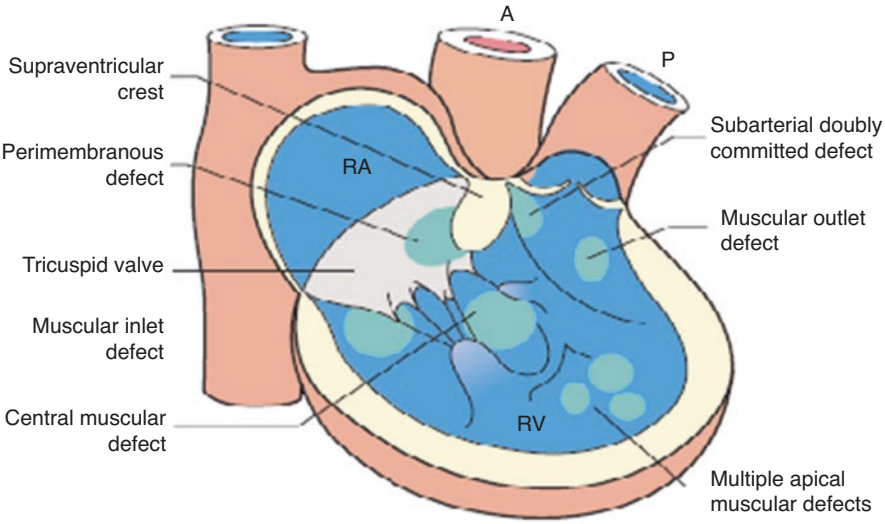

Fig. 2.2 Types of ventricular septal defects

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.

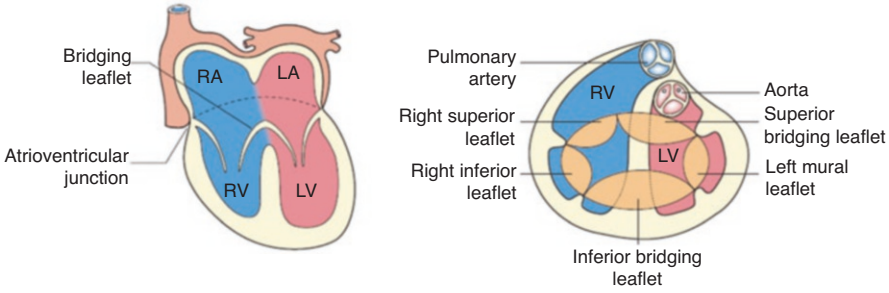

Fig. 2.3 Diagrammatic illustration of complete atrioventricular septal defect

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].

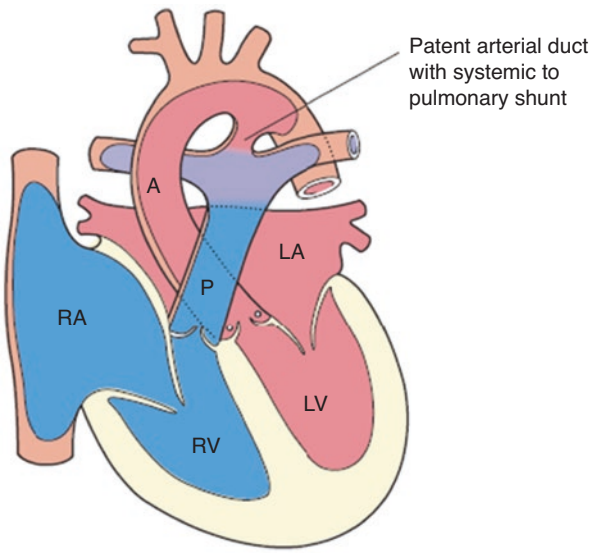

Fig. 2.4 Diagrammatic illustration of the patent ductus arteriosus

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.

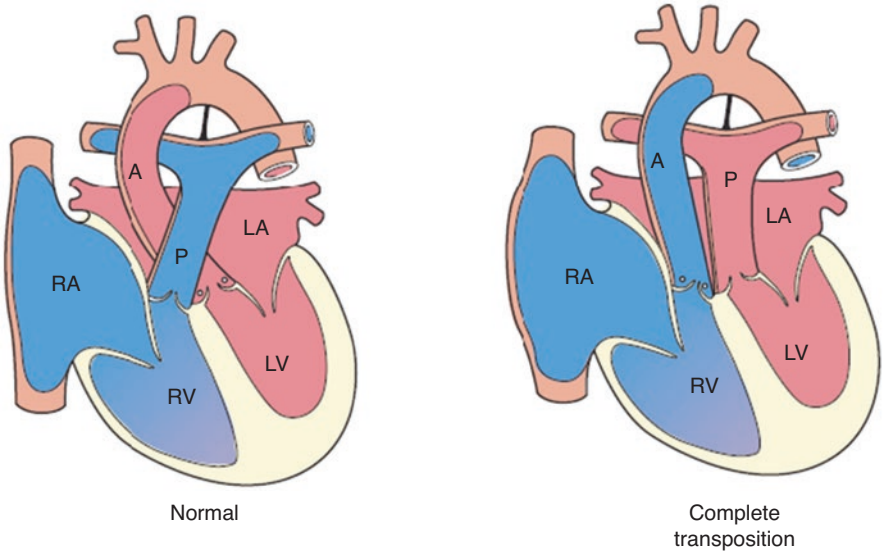

Fig. 2.5 Normal cardiac connection versus the transposition of the great arteries

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).

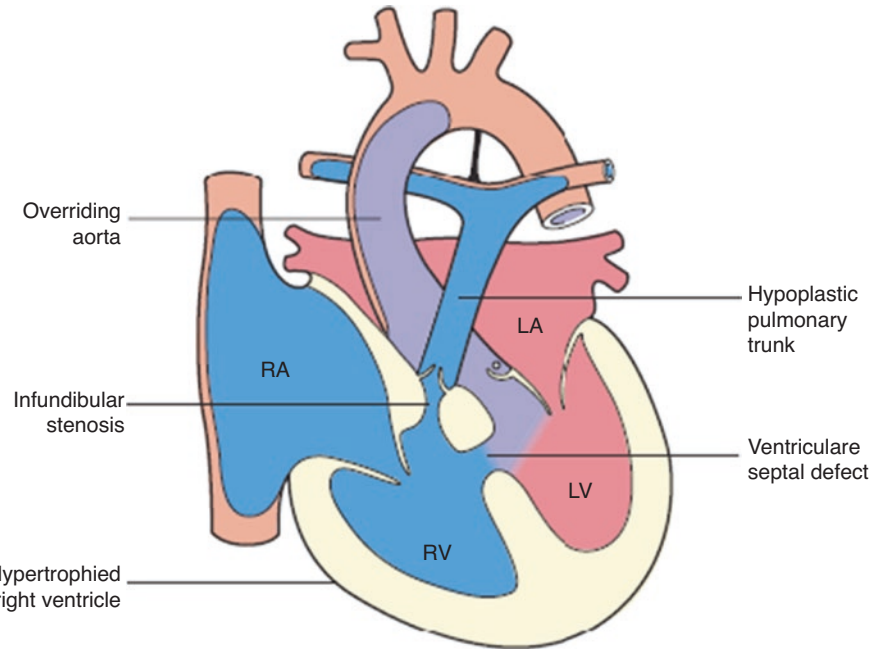

Fig. 2.6 Tetralogy of Fallot

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.

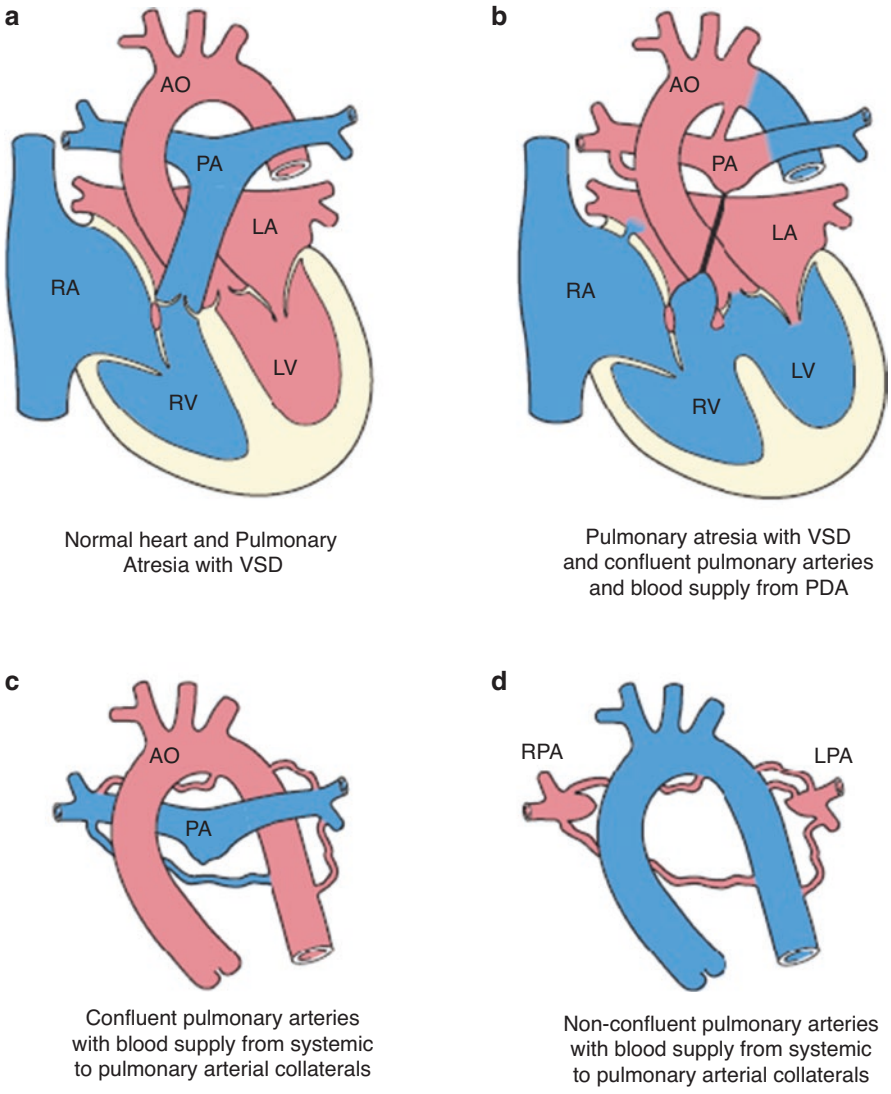

Fig. 2.7 Diagrammatic illustration of pulmonary atresia and pulmonary blood supply (**a**) Normal heart. (**b**) Pulmonary atresia with VSD. (**c**) Confluent pulmonary arteries with collaterals. (**d**) Non-confluent pulmonary arteries with collaterals

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

pulmonary vasculature. Those cases without obstruction to pulmonary blood flow have high flow to the lungs [32].

In tricuspid atresia, the tricuspid valve is replaced with fibrous tissue and the RV is rudimentary. In pulmonary atresia, pulmonary flow is dependent on the PDA/VSD flow or aortopulmonary collaterals.

2.13.1 Clinical Presentation

Clinical presentation in patients with single ventricle anatomy depends on the degree of the pulmonary blood flow.

If pulmonary blood flow is increased, cyanosis is mild and the presentation is similar to TGA with VSD. Signs and symptoms of CHF may be prominent.

If pulmonary blood flow is reduced, cyanosis is more severe and the presentation is similar to tetralogy of Fallot.

2.13.2 Surgical Procedure

The current modification is the total cavo-pulmonary connection (TCPC) with or without a fenestration. This consists of [33]:

An end-to-side anastomosis of the SVC to the top of the right pulmonary artery (Fig. 2.8). Flow from the SVC is directed toward the right pulmonary artery.

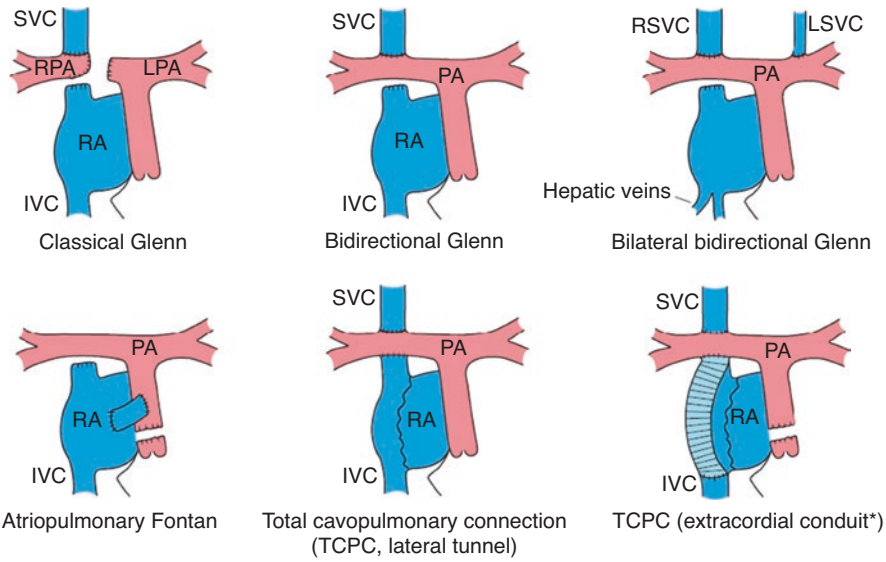

Fig. 2.8 Surgical palliation of a single ventricle

An end-to-side anastomosis of the cardiac end of the SVC to the underside of the right pulmonary artery, but offsets slightly from the SVC to right PA anastomosis to direct flow toward the left PA.
A tubular conduit from the orifice of the IVC to the orifice of the SVC. The conduit can be placed extra-cardiac (Fig. 2.8) or intra-cardiac (within the right atrium).

This can be done as a single- or two-stage procedure. If staged, a bidirectional Glenn anastomosis is performed first followed by completion of the Fontan. The advantages of the Fontan procedure are normal or near-normal arterial oxygen content and removal of the volume overload of the single ventricle.

2.13.3 Complications

Fontan circuit bypasses the sub-pulmonary ventricle, so it's considered passive circulation with possible complications related to this physiology including [34]:

Arrhythmias, mostly supraventricular
Intracardiac thrombus and thromboembolic events either pulmonary or systemic
Severe right atrial enlargement (earlier Fontan version)
Protein losing enteropathy
Pulmonary vein obstruction due to right atrial enlargement
Obstruction in the Fontan circuit
Progressive systemic ventricular dysfunction and heart failure
Progressive AV valve regurgitation
Persistent right-sided pleural effusion
Hepatic congestion and dysfunction
Cyanosis
Pulmonary arteriovenous malformations or systemic venous collaterals
Sinus or AV node dysfunction with the need for pacemaker placement
Re-operation for revision or obstruction in the Fontan circuit or ventricular outflow tract obstruction

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Part II

Transition from Pediatric to Adult Care

Transitional Care for Young People

3

Hajar Habibi

3.1 Background

Congenital heart disease (CHD) is the most common birth defect, with an estimated prevalence of 0.8% to 1.2% live births globally [1]. The community of CHD patients is increasing at a rapid pace, and patients are not only healthier but many now have an extended life expectancy [2]. This is due to the advances in diagnosis, medicine, surgical and endovascular treatments, and specialised nursing input that has altered the pattern of survival of CHD patients, and it is no longer considered a paediatric speciality [3, 4]. Ninety-five percent of young CHD patients will now transfer to adult care, and this new population will need continuous medical and surgical care and follow-up appointments. A planned and individualised transition process which is adapted to optimise care according to their needs is essential to support this transfer in care [5, 6].

The definition of the transition, stages of transition and lifestyle challenges faced by this group of patients will be discussed, all of which highlights the role of the congenital clinical nurse specialist to educate, empower and support patients to develop skills in self-care, independence and navigating adult services. Moreover, the important role of families/carers will be highlighted together with the key role peers can have and the input of the wider team. This input will be from parents/carers, relevant charities and the multidisciplinary team (MDT) such as consultants, CNSs and psychologists.

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3.2 What Is Transition?

Transition is a purposeful and planned movement or transfer of care of young adults with physical and/or medical conditions from paediatric care into adult services. The crucial role of nurses is to ensure that young adults with CHD are made aware of the need for regular follow-up because they are at high risk for complications as they approach adulthood [5].

Transition is a process that begins in early adolescence and is a planned process that usually starts at the age of 12, continuing through to early adulthood when health care management shifts from the family to patient-centred care [7]. On the other hand, the transfer is an event that occurs when the patient moves from a paediatric setting into an adult setting. The difficulty begins when transition and transfer from paediatric to adult health care is not planned, and in turn, the repercussions can be serious and may include hospital admissions, adverse health events and even death [8].

To facilitate a successful transition process, it is integral that team members from foetal to paediatric and adult congenital heart team including nurse specialists educate parents/cares and young adults about their diagnosis, management and preventative health measures. This includes lifestyle issues such as smoking, alcohol consumption, recreational drug use, sexuality, family planning, physical activity, driving and employment or specific to the patient population such as the importance of preventive measures and awareness of infective endocarditis. In this way, we can encourage independence and self-advocacy in adolescent and reinforce the importance of ongoing care [7]. This is achievable if all the team members involved in the care of the CHD patients work together to facilitate this process. Institutional support from all members of the paediatric health care team is essential in the preparation of the transition and transfer of care by having a direct line of contact with an adult specialist unit [5]. Multidisciplinary team input is needed in this process, including social workers, dietitians, physiotherapists, psychologists and counsellors [9] (Fig. 3.1).

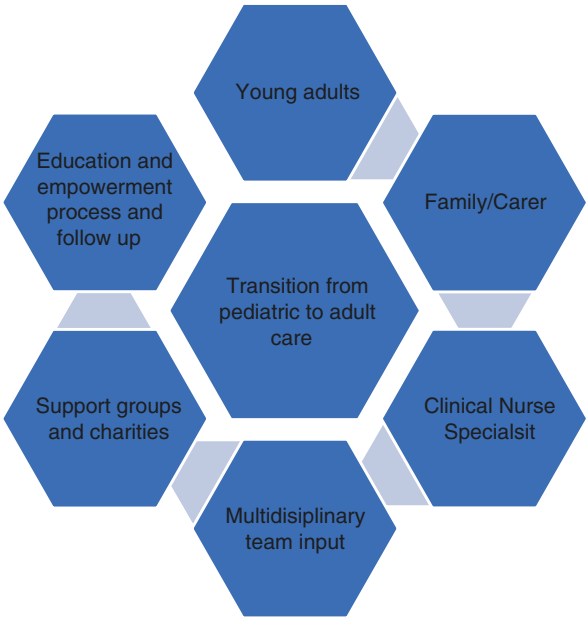

Fig. 3.1 The team approach to the transition and transfer of care. Copyright belong to the author (Hajar Habibi)

3.3 Process and Stage of Transition

The timing of transition should be flexible and dependent on the developmental readiness and health status of the patient and the capabilities of the adult setting.

During the transition, the young person's capability of assuming responsibility for their care should be assessed such as developing personal goals for self-care, approaching the adult health care team, living independently and managing their follow-up appointments. The transition process aims to empower young adults to take accountability and responsibility for their care; the transition outcome of young persons who require extensive assistance or caring activities might differ from those who do not have a physical or developmental special and additional needs; therefore, the process needs to be individually tailored, flexible and developmentally appropriate [10]. However, a target transfer age is useful for both staff and young patients in anticipating and preparing for transition which may differ across health care settings [11].

The transition process should start from the time of diagnosis informing parents of the lifelong care and follow-up (Fig. 3.2). The discussions on transition usually begin before the patient is 12 years old, in line with adolescent development, and continue until successful transfer to adult services which occurs at 16–18 years. However, the importance of tailoring this timeline to suit the individual and their needs has been greatly emphasised [12, 13]. This process is not complete when young adults enter the adult setting. It continues until they feel empowered to take responsibility for their health and lifestyle [13]. This is to ensure concepts of

Fig. 3.2 The lifelong care of CHD patients. Copyright belong to the author (Hajar Habibi)

autonomy, independence, self-determination and self-advocacy are integrated into the service of care. This should be continued during the later adolescent period (between ages 18 and 21 years), until the patient is indeed ready to transition [14].

Emphasis should be given to empowering adolescents and to supporting them in gaining control, increased awareness and self-management skills. The age and level at which transition input is provided should take into account factors such as emotional maturity, self-management skills or general assessment of transition readiness and developmental level, special and additional needs and not just their age [9, 15]. The CNS should use formal transition assessments and checklists such as a transition pathway for continuous nursing assessment. Both primary and tertiary care teams play a pivotal role in helping adolescents with CHD to transition to adult care providers. CHD patients with special and additional care needs and/or disabilities need the support of the wider team. Older adolescents getting ready for transfer into adult services also need information about how the condition may affect their career planning, housing, transportation and health care coverage/insurance [9, 16].

Young adults should be educated about their cardiac diagnosis, what symptoms to be aware of and when and whom to approach for help so they understand the need for regular surveillance and follow-up [17].

3.4 Patient Education and Empowerment

Studies show that if young adults and their families had a better understanding of their diagnosis, they will demonstrate a better understanding of the transition process compared to those with less knowledge about their diagnosis. This, in turn, highlights the role of the specialist nurse in health education and empowerment [5, 13].

Majority of adult congenital heart disease (ACHD) patients require lifelong specialist care and follow-up to achieve the best health outcomes. When the transition from paediatric to adult services occurs, the aim should be to provide education and

support to promote developing skills in self-care and independence [18]. Nurses can support and empower young adults with the knowledge to develop appropriate skills for self-care, make health care decisions and develop self-advocacy skills in preparation for transitioning into adult care [9].

Adolescence is a period of physical, psychological, social and emotional development. This period of change places CHD patients in a vulnerable position as their diagnosis will influence these factors, for example the onset of sexual maturity can lead to difficulty in accepting the physical aspect of their disease, in particular, scarring [19]. Due to the chronic nature of congenital heart disease, all young people should have access to ongoing support and advice from members of the wider multidisciplinary team including nurses. Lifestyle choices like body piercings and tattoos, skin and dental hygiene, signs and symptoms of infective endocarditis, nutrition and the use of substances such as tobacco, alcohol and drugs should all be continuously discussed by nurses [6].

This continuous assessment will enable health care professionals to measure the effectiveness of the care they provide. Both empowerment and illness perception are also related to the transition readiness of the patient [15, 20].

As part of the routine care, nurses should teach young adults about their diagnosis and anatomy, including red flags and warning signs so that they can recognise the signals that show that their condition may be worsening or it is time to seek medical advice [9].

Considering the current era of evolving technology, nurses should be utilising technology such as CHD-specific tailored app to provide a quick access to specific educational materials and past medical history. This will give them the autonomy to be in charge of their baseline information such as clinic letters, electrocardiogram (ECG) and echocardiogram (ECHO), and as a result, this facilitates the transition to adult care [21]. Every CHD patient can benefit from a personalised care plan as personalised approach may improve quality of life and patient satisfaction as patient education and empowerment may reduce unnecessary health care service utilisation and prevent morbidity and mortality in CHD [22].

During the COVID-19 pandemic, there have been many positive changes in the care of patients with CHD such as the use of telemedicine. Telemedicine has helped clinical staff to keep in touch with their patients, alleviating concerns and educating young adults on their condition. In future, it can be used to provide personalised care beyond the coronavirus pandemic [23].

3.5 Pregnancy, Contraception and Family Planning

The ever-growing number of ACHD patients, half of which are female and mostly at reproductive age, means issues regarding their reproductive health, which have become relativity timely with education and information sharing provided to CHD patients entering adulthood. Family planning should involve any patient with CHD regardless of gender or sexual orientation. Pre-conception counselling is relevant to address concerns on hereditability, the possibility of genetic counselling and testing.

For women, cardiac diseases can influence fertility and the choice between contraceptive options and pregnancy outcomes. Pregnancy is a major burden on the maternal cardiovascular system and may give rise to cardiac complications which should be discussed [24]. This discussion should start as early as possible and be re-emphasised during the transition process to highlight the importance of avoiding unplanned pregnancy [25].

Nurses can advise and approach the concept of family planning and contraception sensitively with the patient, in particular, on different types and methods of contraception for their CHD lesion to prevent unplanned pregnancy. Careful and timely consideration of contraception options for this group are critical bearing in mind the option of providing timely psychological support [26]. The concerns regarding body image can affect a young patient's self-esteem and have an impact on sexuality and sexual choices [6]. Nurses can provide patients with the opportunity to discuss issues of pregnancy and contraception and independent of parents, along with other sensitive and personal issues [27].

3.6 Exercise and Participation in Sports

The benefits of exercise and recognition of symptoms of overexertion should be intensively discussed during various stages of transition. During the transition period, nurses should discuss secondary complications of CHD such as coronary artery disease, myocardial infarction, diabetes mellitus, renal failure, hypertension and arrhythmias [9].

Regular physical exercise is associated with a lower risk of obesity and cardiovascular disease [28]. The majority of young adults with CHD can participate in physical activity and sports with minimal restrictions. Nurse as part of a wider multidisciplinary team should assess and support individual patients and advise on exercise and physical activity. Except for some cases with medically imposed restrictions of intensive physical exercise, most young CHD should be encouraged to be active during leisure time and to participate in physical exercise at school [6]. For example, a patient with coarctation of the aorta or significant ventricular dysfunction should be counselled to enjoy a wide range of recreational sports but limiting their involvement in competitive sport. If the young patients or parents need reassurance, objective assessment can be done by performing a formal exercise test to benchmark their current level of fitness [13, 29].

3.7 Preventing the Loss to Follow-up

It is estimated that 30% to 70% of CHD patients are lost to follow-up or experience lapses in care putting them at a greater risk of adverse health outcomes [6, 30]. Research shows there is a direct correlation between the level of a patient's knowledge and their compliance with follow-up appointments [5, 31]. Ongoing support of young patients highlights the rationale for routine follow-up appointments to show them how to navigate the adult system. Nurses can implement databases during the

transition to effectively track patient progress and post-transfer to the adult congenital service for prevention of loss to follow-up [13].

After the transfer to adult care, it is important to continue supporting patients as they approach adulthood so any gap in care is prevented and for consideration of mental and psychological well-being and quality of life [32].

The COVID-19 pandemic has changed the work patterns in health care settings, specifically affecting patients with CHD who require regular follow-ups. To minimise exposure to patients and staff during the pandemic, virtual and telephone-based consultations have been adapted. The pandemic has highlighted the need to develop new pathways of care and, in turn, has increased the collaboration between paediatric and adult services [23, 33].

3.8 Transition Information Day

Innovative methods of providing CHD transition care to young people and their families have been initiated widely across clinical practice. Transition information days provide the opportunity to provide transitional support from clinical teams alongside peer support. The transition day can be arranged as an informal day by inviting a multidisciplinary team including consultants, nurse specialists from foetal, paediatric and adult settings and psychologists and relevant charities. This encourages an open and safe sharing experience. Such a forum provides young adults to form peer groups. This process empowers young patients and promotes direct engagement reinforcing the importance of ongoing care [13]. The close collaboration between the multidisciplinary team from both paediatric and adult teams is necessary for a smooth process in transition day [34].

This forum not only helps families to vocalise and discuss their concerns and fears but is also an opportunity to educate the families to understand the importance of the supportive role they play in the young adult's transition process [9].

3.9 Charities Involvement

One of the ways to increase the knowledge of adolescents and young adults about transition is to involve representatives from charity organisations to introduce their services to CHD patients [13].

Involving charities can prove to be effective as they will be able to organise group meetings, web-based information/leaflets and peer support, all of which can reassure parents that there is a wider support available and young adults can spend time with peers to help them make autonomous decisions [35]. Charities also played a vital role in providing support and information to patients and families during the COVID-19 pandemic connecting them with the local support team to bridge the gaps that can exist between agencies by working in partnership with all parties and professional involved [36].

CNSs can put patients in touch with the different local, national and international services. This could be sources of information including handouts, Internet websites and charity support groups. Some young people may find face-to-face peer interactions a helpful source of support through meeting other young people with CHD to be able to share their experiences. Furthermore, charity events and trips could provide young people with a way of rebuilding their confidence, providing a safe space for them to be themselves, without feeling of being labelled as someone who had congenital heart disease [37].

3.10 Psychological Aspect of Transition and Challenges

A successful and smooth transition from adolescence to adulthood is important for improving psychosocial functioning and quality of life. Nurses should build a therapeutic relationship with young adults and their families to detect psychosocial distress and ask patients about specific challenges or difficult feelings that they may be experiencing in daily life [38].

Psychological challenges in CHD patients are common such as depression and anxiety, as a result of growing up with a complex health condition such as intrapersonal and emotional issues, body image functioning, educational difficulties and poor health behaviour [6]. Because of the complexity of most CHD and the need for lifelong follow-up, psychosocial or behavioural support may also be required in conjunction with medical care [9]. Many young people with CHD face future cardiac surgeries with trauma from previous medical interventions which can go on to affect compliance with medical treatment such as needle phobia which requires nurses to assess and to spend time with the young adult to help them to overcome needle-related distress [39].

The disruptions many young people face to education, employment and family planning as a result of their CHD diagnosis can cause further psychological distress. Nurses should routinely assess anxiety and depression in young adults with CHD and do a timely referral for appropriate psychological management [40].

The other important psychological aspect to consider is the COVID-19 pandemic adding an aspect of physical and mental isolation; COVID-19 has brought huge challenges and changes to the ways nurses see and provide information and support to young adults and their families/carers with a significant increase in patients seeking advice from the CNS team [41]. The pandemic has undoubtedly resulted in consequences for mental health and well-being. The young adults have experienced unexpected changes such as school closure, changes in family circumstances, remote learning with extended time at home and learning in a different environment, all of which could have a significant impact on young adults, mental well-being and ability to learn. Nurses should consider how these changes may affect young adults and work with the wider team to provide support. One of the ways we can overcome this is to do virtual clinics so we can stay in touch with patients and, if needed, refer them to the appropriate team [23].

3.11 Established Relationships with Paediatric Teams/ Involving Parents and Carers

Challenges to a successful transition include patient adherence, relocation and attachment. Young adults and their families are required to move from a family-centred paediatric model of care to an autonomous adult model. This is where self-management and self-advocacy skills are essential and where a nurse can develop a structured transition service based on the individual patient and family needs [6, 30].

The process of transition from paediatric to adult care can be challenging for young patients as they may have developed a firm bond with the paediatric team over the years. A well-planned transition process is a key to minimising anxiety for patients and their family in preparation for the transfer into adult settings [12]. This can be achieved by seeing the young adults in a joint transfer clinic with both the paediatric and adult teams present [13]. The development of close links and collaboration between the paediatric and adult services is beneficial to both services and ensures patients are not lost to follow-up [11]. Patients also need to be empowered by the paediatric centre to share long-term responsibility for their health and support parents and young people through this gradual shift in responsibility.

The most frequent barrier to the transition is reported to be the emotional attachment of both patients and parents to the paediatric team, and therefore, it is important to ensure parents continue to feel involved in care planning through the transition process [42].

3.12 Role of Family/Carers in the Transition

Parental involvement is an important factor in encouraging adolescents to increase their self-care and in supporting the young adult in communicating with the health care providers. The transfer to adult care is not only unavoidable but also appropriate for patients with inherited conditions [35]. Parents responsible for monitoring their child's medical care need to be secure and confident that the transfer is the best course of action and the patient will be supported during this period [35].

Parents should be involved to a higher degree as well-informed parents are better equipped to pass on their knowledge and experience to help young adults to gain self-management skills [43].

The family is the most important source of support for young adults with CHD, and their input must be respected and incorporated into all aspects of care [44]. It is challenging for the parents to shift roles and decrease their involvement, and many will worry about the future and how the young adult is going to navigate care in the adult setting [15].

The process initially involves parents/carers, with the inclusion of the child when they are capable of understanding. This process of the introductory stage usually starts at the age of 12 and allows young adults to develop an awareness of their heart lesions [13]. In every clinic, parents have multiple opportunities to impart transition skills, disease-based knowledge and the idea of maintaining lifelong cardiology

care and the need to be involved in every stage until the young adult has gained adequate and appropriate knowledge [2]. Although patients' independence is the key aspect of the transition process, preparing the family for this change should also be a focus of the transition education process [9].

3.13 Role of the ACHD CNS in Supporting Young Adults with CHD

Patient education is an important part of nursing and nurses should promote self-care and assist in patient's learning about their illness [35]. Chronic disease management places a higher emphasis on patients' education. Health education has been shown to have a positive impact on health-related quality of life [42].

Regardless of the severity of the disease, the educational aspect of care of CHD patients is fundamental [19]. Various researches in the field of congenital heart disease indicate that there is a lack of education and understanding among congenital heart disease patients [45, 46].

To provide high-quality care, it is important to understand patients' individual experience of care and the effectiveness. Patient experience is recognised as an aspect of high-quality care that highlights the importance of transition and continuity to help patients care for themselves. A key aspect to consider is to involve the young adults and their families in health care decisions as early as possible. Despite the movement towards patient-centred approaches, most disease management follows a dated model of decision-making where a patient receives instructions rather than participating in the management [47].

A key aim of transition is to empower adolescents to take ownership of their illness by providing them with all the information they require to successfully achieve this. This can be achieved by providing them with knowledge of their disease, how to recognize complications, the importance of making good lifestyle choices and attending for their follow-up care [35].

Nurses are the key to the smooth and successful transition and transfer of young adults. This can be achieved by providing continuous education from the time of diagnosis through to successful transfer, tailoring of education to individual needs and providing written and verbal information [6].

In a lot of cases, young adults feel ready to take over the responsibility for their health, but a lot of the time, adolescents overestimate their capacity or the parents do not acknowledge the adolescents' actual skills, underestimating their capacity. This is where nurses can provide specialist input assessing levels of knowledge and the readiness while providing tailored input and support [15].

3.14 Executive Summary

In summary, most children with CHD surviving into adulthood which is challenging for adolescence and young adults as they face other issues, mainly general health behaviour, uncertainty about the future, illness experience, pregnancy,

contraception, insurance and employment uncertainties. All of these factors can affect the social and psychological aspects of their daily life. The development of a transition and educational programme is therefore crucial to the successful transfer of care.

The transition period can be a difficult time for CHD patients and their families and/or carers. The key to a successful transition process is the provision of individualised care, effective communication, mutual trust and sustained partnership. Nurses play an integral role as part of a wider team in supporting a seamless and successful transition from paediatric to adult care.

Also requiring consideration is preparation for the patient's transition to adult life; education is an important aspect and there is a need to address sexuality, family planning and employment. All of these issues need to be addressed systematically and comprehensively to ensure that adolescents and young adults are prepared to take complete responsibility for their health.

Patient experience should be considered to improve services with the input from multidisciplinary team including relevant charities. Lapse of care is linked to an increase in the risk of complications which highlights the importance of developing transition programmes to provide education to adolescents with CHD. Nurses are in a pivotal position to initiate, tailor and continue education from the time of foetal diagnosis through to a successful transfer.

Parents should be involved in transition planning from the very beginning to facilitate their gradual handing over of responsibility to the adolescent. Nurses should ensure that patients are aware of the benefits of regular check-ups, understand the rationale for follow-up and empower them with the knowledge of what would be the potential problems arising with their individual lesions. This is essential to avoid lost to follow-up.

Nurses need to collaborate with the interdisciplinary and wider multidisciplinary team with expertise in the comprehensive psychosocial needs of adolescents as they move from paediatric-focused care to adult provider systems.

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4 Advancing High-Value Transitional Care: The Central Role of Nursing and Its Leadership

Serena Francesca Flocco and Rosario Caruso

4.1 Introduction

Congenital heart diseases (CHD) are the most prevalent and severe congenital disabilities, representing a major global health issue. Given a prevalence of 9.1 for 1000 live births, CHD are the leading cause of congenital disabilities associated with morbidity, mortality, and medical expenditures [1]. Currently, 1.3 million children are living with CHD over the globe, and roughly 90% of them survive into adulthood [1]. Therefore, adults with CHD (ACHD) represent a new growing population [1, 2]. It is estimated that almost three million adults are currently living with CHD, requiring a lifelong follow-up [3]. The period from childhood to adulthood (transition) is the most complex and delicate part of life for patients with CHD to establish a solid foundation for healthy growing [4, 5]. This population has several important peculiarities that have yet to be studied from different perspectives, considering clinical, prognostic, therapeutic, social, and psychological perspectives.

Adolescence is a crucial phase for shaping the personality, and adolescents with CHD have to face the consequences of their disease and the need to be adherent with their follow-up indications [6]. Moreover, during the transition into adulthood, adolescents with CHD could be exposed to many psychological issues involving the development of self-identity, self-esteem, and self-image [4]. Adolescents with CHD should acquire relevant knowledge about their disease to overcome frustration

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and anxiety and develop self-care strategies. In fact, in recent years, in addition to the medical problems of congenital heart patients, an increasing attention has been paid to their existential needs, which concern the possibility of studying, having a job, marrying and generating children, and carrying out a recreational activity and/or sports, essential conditions for feeling entirely similar for their peers [4, 6]. Therefore, the main problem is no longer simply to guarantee survival but a good quality of life. Knowing, therefore, how these people live, the experience and their perception of life, how they face moments of joy and difficulty, and their interpersonal and social relationships are of fundamental importance to guarantee them a personalized treatment path. Therefore, an appropriate approach is needed to help people with CHD meet their clinical and psychosocial needs during adolescence and adulthood [7]. In this scenario, to ensure an adequate treatment program, it is necessary to abandon the traditional care models focused on the "case" and adopt a holistic people-centered approach by acknowledging that "prevention is the best intervention," and nurses are pivotal professionals to achieve these goals.

Nurses are typically educated for holistic care. Professionals educated for this people-centered approach are better equipped to improve a patient's lifestyle choices and enhance determinants of health, the personal, economic, social, and environmental factors that influence health status. In this sense, the definition of Barbara Dossey et al. about holistic nursing might be insightful to understand the complexity of elements involved in holistic care: a "form of healing that gives attention to all aspects of an individual, including physical, mental, emotional, sexual, cultural, social, and spiritual" [8, 9]. Therefore, the holistic nursing assessment is a unique global patient assessment upon which the nursing profession is founded. The ACHD nurse coordinator has to provide integrated nursing care based on this approach by performing a comprehensive, holistic nursing assessment.

This chapter discusses issues related to holistic health care and nursing leadership in transitional care.

4.2 Focus on Holistic Care in Nursing

Holistic nursing is concerned with healing the whole person and emphasizes salutogenic approaches to improving health at each illness stage and/or clinical condition. As a result, the relationships between the mind, body, spirit, social/cultural, emotions, context, and environment represent holistic health. All of these factors contribute to the traits of any individual. As a result, this approach understands that a person is more than his or her sickness. Because patient care is multidimensional, the theoretical foundation of nursing has been organically holistic since the first nursing theorizations. In this regard, in 2006, the American Nurses Association (ANA) recognized holistic nursing as a specific nursing specialty with a predefined scope and standards of practice. This recognition is still poor in many countries globally, even within the same nursing community. Even the approach developed by Florence Nightingale might be considered holistic, considering that she encouraged holistic care by recognizing the importance of environment, touch, light, scents, music, and silent reflection in the therapy process [10].

Holistic nursing is a specialty practice that guides nurses in becoming therapeutic partners with individuals in human responses to aid the healing process and achieve completeness by drawing on nursing knowledge, nursing theories, wholeness, expertise, and intuition. Holistic nursing is concerned with safeguarding, promoting, and maximizing health and wellness. In this view, it is critical to aid healing, avoid sickness and damage, alleviate suffering, and assist individuals in finding peace, comfort, harmony, and balance through the diagnosis and treatment processes. Holistic nursing care is concentrated on the person's relationship and is healing-oriented, as opposed to disease- and cure-oriented. Holistic nursing views self-care, intentionality, presence, awareness, and therapeutic use of self as critical for facilitating healing and replicating wellness in others. All nursing practice, in some ways, must be comprehensive; all nursing practice may have a biopsychosocial perspective.

What makes holistic nursing practice a specific specialty is that there is a transparent philosophical background, a specific body of knowledge, and an advanced set of nursing skills applied to practice that recognizes the totality of the human being and then interconnectedness of the body, mind, emotion, spirit, energy, society, culture, relationships, context, and environment.

Holistic care is a complete caring concept that is seen to be at the center of nursing science and practice. Holistic treatment is based on the concept of holism. This idea emphasizes that the whole is more than the sum of its parts and that the mind and spirit impact the body. Holistic care is defined as the conduct that respects the full person and understands the connection of one's biological, social, psychological, and spiritual elements. Holistic care encompasses a variety of approaches, such as medicine, education, communication, self-help, and complementary treatment [11]. All aspects of patients and their influence on the treatment process are considered in holistic nursing, and the patients' ideas, feelings, cultures, opinions, and attitudes contribute to recovery, happiness, and contentment. Holistic care honors human dignity: in this style of care, the relationship between health care providers and patients is built on respect, relative openness, equality, and mutuality, and patients engage in decision-making [12].

Holistic care professionals view patients as full beings within their surroundings, and they understand that patients always include components provided by the body, mind, and spirit. Another feature of holistic care that leads to therapeutic consultation, hope, dignity, self-discipline, social progress, a sense of autonomy, vigor, and vitality is respecting the patient's position in the treatment process, having him/her participate in the process, and supporting self-care [12].

Holistic nursing care broadens caregivers' awareness of patients and their needs. Holistic treatment includes educating patients about self-care and assisting them in doing everyday chores on their own. Palliative care is a type of holistic treatment that enhances a patient's quality of life and their mental and physical well-being. Holistic care promotes patients' self-awareness and self-confidence while allowing nurses to better comprehend the consequences of a disease on a person's complete life and actual needs. It also promotes the balance of the mind, body, emotions, and spirit in a constantly changing environment [13].

The strategy presented in this chapter might serve as a catalyst for developing care plans for adolescents with CHD. When numerous components must be addressed in delivering care for promoting the well-being and healthy behaviors of adolescents and their families and their general health-related quality of life, the elements contained in the holistic approach work as a compass for nurses. The holistic approach must find practical applications in standardized evaluations and educational interventions, as standardization and personalization are two interrelated aspects of care. Patients expect and require personalization in their everyday experiences; standardization aims to reduce unwanted clinical variation, resulting in a more efficient, less expensive care and a more consistent patient experience and outcome; personalization is what patients expect and require in their everyday experiences. By creating a high-quality baseline in care planning, standardization sets the path for more personal encounters. Professionals will struggle to promote personalization in care delivery and overall patient experience without a uniform foundation. As a result, standardized core examinations and treatments for adolescents with CHD provide the path for practical holistic care in which nurses can adapt the standardized care techniques. In this regard, more study is needed to back up the best techniques of standardizing and customizing care delivery with data.

4.3 Holistic Needs of Transitional Care

Adolescence is the period of life between childhood and maturity (adulthood). It is a challenging growth stage marked by significant changes in physical maturity, sexuality, cognitive processes, emotional feelings, and interpersonal connections. In addition to these bodily changes, societal demands and expectations rise throughout adolescence. Adolescents must master various developmental tasks in order to form a personalized identity. Professor Robert Havighurst of the University of Chicago stated some years ago that phases in human development might best be understood in terms of developmental activities that are part of the natural transition [14]. Eleven developmental activities related to the teenage transition were identified by him. Each of the Havighurst duties may also be viewed as a component of the general sense of self that teenagers take with them as they enter early adulthood [14].

The developmental tasks of adolescence are listed below:

1. Since birth, no other phase in a person's life has seen such quick and dramatic bodily changes as early adolescence. As a result of this fast development, the early adolescent frequently gets preoccupied with his or her physique. The adolescent must come to terms with a new bodily sense of self.
2. The adolescent must adapt to new cognitive capacities. Adolescents have a dramatic rise in their ability to think about their surroundings, in addition to a quick increase in physical growth.
3. Adults perceive high school as a place where teenagers prepare for adult tasks and responsibilities and a place where they prepare for a higher education. Adolescents must acclimatize to higher cognitive demands in school. Regardless

of whether the teenagers have attained formal thought, school curricula are usually dominated by an increasingly abstract, challenging content. Demands for abstract thinking prior to obtaining that capacity may be frustrating since not all teenagers make the intellectual shift at the same time.

4. The adolescent must improve his or her speech skills. Adolescents must learn new linguistic abilities to accept increasingly complicated concepts and tasks as they progress cognitively, confront higher educational pressures, and prepare for adult activities. Their childhood language is no longer appropriate. Adolescents may look less capable because of their incapacity to articulate themselves meaningfully.
5. The adolescent must establish a sense of own identity. Prior to puberty, one's identity encompasses more than only one's parents. During adolescence, a child learns to perceive his or her individuality and independence from parents.
6. The adolescent must establish adult vocational goals. The adolescent must begin the process of focusing on the question, "What do you want to be when you grow up?" as part of the process of building personal identity.
7. Adolescents must develop emotional and psychological independence from their parents. Childhood is characterized by a significant reliance on one's parents. Adolescents may long maintain that safe, secure, supporting, and reliant connection. On the other hand, being an adult indicates a sense of independence, autonomy, and being one's own person. Adolescents may oscillate between a need for dependency and a wish to be self-sufficient. Adolescents may look hostile and unwilling to cooperate in an attempt to show their demand for independence and uniqueness.
8. The adolescent must develop stable and productive peer relationships. The ability of a teenager to establish friends and have an accepting peer group is a crucial determinant of how well the adolescent will adjust in other areas of social and psychological development. Although peer interaction is not unique to adolescence, peer interaction seems to hit a peak of importance during early and middle adolescence.
9. The adolescent must learn to manage her or his sexuality. With their growing physical and sexual maturity, adolescents must assimilate a set of views about what it means to be male or female into their personal identity. Their feeling of masculinity and femininity must be accommodated in their self-image. They must also include values around their sexual conduct.
10. The adolescent must adopt a personal value system. Adolescents embrace an integrated set of values and morals as they build increasingly complicated knowledge systems. Parents teach their kids a systematic set of principles of good and wrong and acceptable and unacceptable behavior during the early stages of moral formation. Eventually, the teenager must evaluate his or her parents' views, which frequently contradict those expressed by classmates and other elements of society. To resolve disagreements, the teenager restructures those views into a personal ideology.
11. The adolescent must develop increased impulse control and behavioral maturity. Most young people participate in one or more behaviors that put their phys-

ical, social, or scholastic well-being at risk as they transition to adulthood. Adolescents engage in risky actions to such an extent that risk-taking may be a typical developmental phase of adolescence. Risk-taking is more noticeable throughout early and middle adolescence. Adolescents gradually develop a set of behavioral self-controls that allow them to choose which acts are appropriate and adult like.

Adolescents with CHD must complete the same developmental activities as their healthy classmates. Having a cardiac problem and dealing with day-to-day life situations, on the other hand, are additional pressures. Adolescents with congenital heart disease must deal with the implications of their sickness and the necessity to follow their follow-up instructions. Furthermore, when they enter adulthood, CHD adolescents may be subjected to various psychological challenges relating to developing self-identity, self-esteem, and self-image [4, 6].

4.4 Nursing Leadership in Transitional Care

There is no common paradigm for CHD transition care, and existing programs are all very new to the area of congenital heart disease, with their own distinct characteristics. The "joint clinic model," "pediatrician-in-adult-care model," and "introductory model" are all "transfer models" since the emphasis is on transitioning the teenager from pediatrics to adult care, with little regard for the developmental process that is inherent in the move to maturity. The most functional is the "transition coordinator model," which focuses on the developmental process, accompanying the teenager through the transition to maturity and providing complete transitional care. In this concept, the transition does not end when the patient is moved but instead ensures continuity of care.

According to research on existing models for transitioning adolescents with congenital heart disease, the highest success is achieved when a transition coordinator is hired to guide adolescents and young people through this process (transition intervention for adolescents with congenital heart disease). The transition nursing coordinator is part of the interdisciplinary health care team caring for children with congenital heart disease and combines advanced training in assessment, diagnosis, and management of common pediatric health problems with advanced nursing expertise in patient education, counseling, and health promotion to meet the specialized needs of children and adolescent with congenital heart disease during the period of transition.

The transition nursing coordinator is in charge of facilitating the transition process for adolescents and young adults as he or she educates, supports, and accompanies and becomes a point of reference for them, evaluates any critical factors of each personal transition process, helps the patient to deal with them, and ensures their success, also thanks to the support of other health professionals. The transition nursing coordinator's primary function in the transition of teenagers out of pediatric care must be oriented on health education. Adolescent health education must be tackled

from several perspectives; the transition coordinator should offer seminars that improve group learning for teenagers with congenital heart disease, coordinate peer support within adolescent groups, and write up all health and educational material.

According to current research, a health care passport, which includes an in-depth description of the adolescent's cardiac anatomy and physiology, past cardiac medical history, the most recent diagnostic and imaging studies, and a list of current medications, is an essential component of a successful transition. Adolescents should be informed about their therapy, residual difficulties, contraception and pregnancy planning, considerations for infective endocarditis, and career and lifestyle challenges. Other medical histories, like growth and development, vaccines, allergies, and pertinent personal and family history, should also be included in the health care passport. During the transition phase, the transition coordinator should review and update this document at each visit. The health care passport is especially useful for young adults who travel or need to consult a specialist in a different department. It is important to confer with the teenager about the content of the passport and secure their permission to share medical information. A copy of this growing passport should be preserved in the adolescent's records and supplied to the adolescent with instructions on how to use it and the requirement for it to be updated regularly.

The activities and skills for which the transition nursing coordinator is responsible are as follows:

- Meet the young teenager aged 11–12 and her family to plan the transition process, providing them with a timeline of the development stages of the transition process.
- Educate the adolescent about his clinical condition and self-care.
- Prepare documentation (i.e., health passport), which includes a description of the clinical condition, medical and/or surgical procedure, medications, and potential future complications, as well as specific signs and symptoms for the clinical condition. This documentation must be updated at each follow-up visit.
- Document and update the self-care skills of the young adolescent, the self-management skills of drugs, and the level of knowledge.
- Periodic training seminars/program for young adolescents and their families.
- Support and reassure the family that the transition is a positive and necessary process for the adolescent to establish his or her identity and carry out tasks of his or her developmental stage.
- Collaborate and involve the multidisciplinary team (pediatric cardiologists and adult congenital cardiologists) in the implementation of the transition process.
- Stay up to date on any clinical changes in the adolescent.
- When the adolescent has reached chronological age (about 12 years), he or she evaluates the appropriate stage of development for the transition process and evaluates the effectiveness of the educational intervention.
- Inform and provide documented information to the adolescent and family regarding the transfer to the adult congenital cardiology unit.
- Become a reference point for the entire transition process and the subsequent transition to the adult congenital cardiology unit.

4.5 Conclusions

ACHD should educate themselves about their sickness to overcome frustration and worry and establish self-care practices. As a result, specific care is necessary to assist this group in meeting their medical and psychosocial needs throughout adolescence. The "transition clinic" (TC), a multidisciplinary standardized intervention to educate and assist individuals with CHD, is a critical component in providing treatment for patients from infancy to adulthood. The transition process is described as "the process through which adolescents and young adults with chronic childhood diseases prepare to take command of their lives and health in adulthood." Young people should be included in the techniques that assist them to build their self-efficacy, improve their self-management, and establish autonomy in an unfamiliar situation during this process. Nurses are uniquely positioned to assist young people in the tasks listed above. They are qualified to engage in transition readiness, examinations, and educational activities. They can also act as liaisons across health care systems, advocating for resource allocation and promoting collaboration.

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Building a Transition Program

5

Christina Sillman

5.1 Factors That Impact the Structure of a Transitional Program

There is a wide variety of adult congenital heart disease (ACHD) program structures, resources, and congenital heart care delivery internationally, which may create barriers to the development of the transitional program. Location of the ACHD program, available resources, physician support, administrative support, and governmental guidelines are all factors that may impact the choice of delivery method for transitional care.

Adult congenital heart disease program location varies depending on the health care structure of the country, resources available, and prevalence of adult congenital heart disease–trained cardiologists. ACHD programs may be housed within pediatric or adult hospitals/centers or in hospitals/centers that provide both adult and pediatric services. The gap between pediatric cardiology programs and adult congenital heart programs may be wide with large physical distances across state or province lines, barriers between private health insurance providers, or inherit obstacles within the national health care structure. Assessing for the gaps between the collaborative pediatric and adult congenital heart programs and the larger national health care structure is imperative when establishing a new transitional program in order to avoid lapse in care.

The clinical focus of the advanced practice registered nurse (APRN) may also impact the structure of the transitional program. The APRN leading the development of a transitional program may be resourced from the pediatric cardiology program and the ACHD program or as a separate institutional or governmental nurse. Ideally, the APRN will recruit nurses from both the pediatric and the adult CHD

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programs to collaborate in the development of the transitional program for collective investment in the program structure.

Resource allocation for the transitional program involves assessment of available administrative support, technological support, marketing and educational materials, medical record format (paper vs. electronic medical records), and space/location for care delivery. After identifying the barriers, the APRN can evaluate for the existing and needed resources to overcome the barrier and execute care. For example, adult congenital heart programs that will be recruiting transitional patients from rural low-income areas, delivery of care via virtual platforms such as video visits or video presentations may help to improve accessibility for patients. Therefore, assessing the existing and needed technology resources of the transitional program and the patients they will serve is crucial.

One of the most important starting points of building a transitional program is securing investment and support from institutional administration and the physician leaders associated with the pediatric and adult congenital heart programs. The APRN should prepare a presentation for administration that highlights the patient and programmatic benefits of a transitional program, the regional guidelines on transitional care, and the requested administrative support for resource allocation and transitional program financial investment. A separate presentation can be made to the physician groups to highlight the benefits and propose a programmatic structure. Physician investment, particularly the pediatric cardiologist, is essential for developing a streamlined referral process.

Once the access and administrative barriers are identified, hospital leadership and physician support are secured, and resources have been allocated; the nurse leader will work to identify the most appropriate model of transitional care delivery. The chosen model of transitional care should be best suited to prevent lapse in care with a seamless transfer from pediatric cardiology to adult congenital cardiology and comprehensive ongoing transition education.

5.2 Models of Transitional Programs

There have been several published models of transition described that identify the various approach to the transfer of care and transitional education of lifelong CHD care. Various models have their own benefits and disadvantages, but ultimately, the APRN will need to decide on a model that best fits the established framework of the congenital heart programs they are working within.

The European Society of Cardiology (ESC) global consensus statement on transition and transfer of adolescents with congenital heart disease described the following four models of transitional care [1].

The **joint clinic model** is a collaborative overlap model where the ACHD cardiologist meets the patient within the pediatric clinic and joins the pediatric cardiologist for the final visit prior to transfer to care. Transfer of provider occurs prior to physical transfer to clinic. The potential benefits of this model include demonstration of collaboration between providers for the patient and family, and

introduction of the ACHD provider prior to introduction of the ACHD clinic location, which may assist with wayfinding when physical transfer to the ACHD clinic occurs. The possible disadvantages of this model include coordination of provider scheduling, financial compensation for dual providers, and this model focuses on transfer of care without comprehensive education factored in.

The **introductory model** involves a pediatric cardiology–guided visit to the ACHD clinic to meet the providers and see the location of the ACHD clinic prior to transfer of care. The potential benefits of this model are that this can be achieved in a group setting (such as quarterly open houses at the ACHD clinic for all the pediatric cardiology transition patients approaching transfer), and the pediatric cardiology practice assists in wayfinding. The possible disadvantage to this model include that a lack of overlap between providers may lead to lapse in care, and this model focuses on transfer of care without comprehensive education factored in.

Pediatrician-in-adult-care model is a collaborative overlap model where the pediatric cardiologist continues seeing the patient within the ACHD clinic and gradually transfers care to the ACHD provider. The transfer of location occurs prior to the transfer of provider. The potential benefits to this model are that logistical barriers with location are overcome with the guidance of the pediatric cardiologist, and the patient and family observe the collaboration between the two providers. Possible disadvantages include barriers of physician privileging to practice in the adult setting, lack of a definitive transfer date for providers, which may lead to care delivery ambiguity, and this model focuses on transfer of care without comprehensive education factored in.

Transition coordinator model focuses on the educational and developmental process of transition to adulthood by a designated transition coordinator that provides continuity of care at both the pediatric and ACHD clinic and facilitates the transfer of care. The potential advantage of this model is that it comprehensively addresses both transition to adulthood through individualized assessment, education, and skill development in addition to providing continuity of a coordinator to facilitate the provider and clinic transfer process. The possible disadvantages include lack of a dedicated transition coordinator, limited resources, and administrative barriers between two separate facilities.

5.2.1 Methods of Transitional Education Delivery

Regardless of the model used to facilitate the transfer of care from pediatric cardiologists to ACHD cardiologists, or even if the patient must stay with the pediatric cardiologist due to regional scarcity of ACHD-trained cardiologists, there must be a comprehensive plan for the assessment of developmental status, autonomous self-care, and a clear transition curriculum for education and skill acquisition.

The mode of care delivery for transition assessment and educational curriculum may also vary to provide an individualized and developmentally appropriate medium. As early adopters of technology, adolescents and young adults live much

of their life on digital technology platforms. As clinicians, we must meet our patients where they are and provide individualized collaborative care [2].

This use of technology has exponentially increased with the necessary adjustments in society seen during the COVID-19 pandemic. The use of telemedicine is a rapidly evolving care delivery model that the medical community was thrust into using in the spring of 2020. In Canada, pre-pandemic virtual visits accounted for approximately 1% of clinical care, rising sharply to 70% at 4 weeks into the pandemic [3]. The use of digital technology for medical care delivery is likely to continue well past the needs of the COVID-19 pandemic and may be a preference among adolescents and young adults who grew accustomed to this medium for interacting within society.

Examples of digital formats for delivery of transitional assessment and education include:

- Independent study online or health app digital surveys
- Independent study online with educational modules
- Pre-recorded educational videos
- Group real-time virtual presentations
- Individual real-time video visits

Regardless of the digital format utilized, it is imperative to assess patient access to digital devices and high-quality internet connection, the digital security of health care information, and the patient's developmental/neurocognitive abilities to navigate digital mediums. Having diverse care delivery methods helps to ensure accessibility for all patients. Box 5.1 lists examples of existing digital transitional resources for patients.

In circumstances where digital delivery of care is not possible, the modes of care delivery include in-person clinic visits, in-person group visits, and paper-based educational information. In-person clinic visits can either be coordinated with physician visits or scheduled as separate APRN visits. Group in-person visits have the benefits of providing peer support but make individualized care delivery challenging due to patient privacy concerns. Paper-based educational information relies on the patient to autonomously navigate the information and relies on high health literacy and internal motivation.

Box 5.1 Online Transition Education Resources for Patients

https://iheartchange.org/—I Heart Change, international transitional project

https://www.cchaforlife.org/transition—Canadian Congenital Heart Alliance

https://www.rbht.nhs.uk/sites/nhs/files/Leaflets/CHD%20transition%20clinic.pdf—UK Royal Brompton transitional clinic

5.2.2 Timing of Transitional Care and Transfer of Care

To optimize outcomes and reduce lapses in care, transition should be an ongoing process that occurs over many years and transfer of care should be an event within the transition process. Both the AHA scientific statement [4] and European Society of Cardiology consensus statement [1] regarding transition state that transitional care should start in early adolescence at approximately 12 years of age. The initial phase of transition focuses on introduction of transition and the transitional process. Introduction to transition and lifelong care should be continually discussed with parents and child starting upon initial diagnosis and whenever discussing prognosis and expectations of care delivery in order to normalize the process from the onset of the congenital heart journey.

Beginning early-to-mid-adolescence (approximately 14 years of age), assessment of the patient's readiness for transition, health knowledge, and self-care abilities should occur. Assessment is an ongoing continuous process that serves as the foundation for counseling and education.

After the comprehensive assessment has been completed, the APRN will develop an interventional plan focused on education, counseling, self-care skill acquisition, peer support, and educational resource allocation.

At mid-adolescence (approximately 16 years of age), the topic of transfer to the ACHD clinic will begin with introduction of the ACHD program, providers, and clinic structure. This ongoing process is when care is most collaborative between the pediatric cardiology clinic and the ACHD clinic.

The culmination of the transition process is the transfer of care from pediatric cardiology to ACHD cardiology. This ideally would occur at end-adolescence (18–20 years of age).

The post-transition phase of the transitional process focuses on continuity of care, adjustments in care due to life changes (relocation due to schooling or employment, or health care insurance coverage), and continuing education, counseling, and autonomy promotion [1].

5.3 Structure of a Transition Clinic

The cornerstone of all nursing practice is the use of the nursing process. Developed in 1958 by Ida Jean Orlando, this holistic systematic approach involves five steps: assessment, diagnosis, planning, implementation, and evaluation [5]. A nurse-led transition clinic can mirror this process in a patient-centered approach that is goal-oriented with specific and measurable endpoints.

The initial step is the patient assessment. This process involves thorough patient interviewing in various domains to provide holistic assessment of the patient.

5.4 Domains of Assessment

5.4.1 Transition Readiness

Initially, assessment should focus on the readiness of the patient to begin the transitional process. Understanding their knowledge of transition, why it occurs, and how it can benefit them helps to establish expectations for the process. The American College of Physicians created a disease-specific transition tool kit with questionnaires that assist the clinician in assessing the congenital heart patient during the transitional process. The transition readiness self-assessment is aimed at children age 12–17 years and is an evaluation of the patient's understanding of transitional importance, medical confidence, CHD-related knowledge, and health care utilization [6].

Additional factors that impact transition readiness include emotional status, educational capacity, support systems, and health-related beliefs.

5.4.1.1 Mental Health

Assessment of any previously established mental health diagnosis and screening for undiagnosed mood disorders help to establish emotional readiness for transition. A review of the medical chart for past diagnoses and simple screening with tools, such as PHQ, GAD, PC-PTSD, help to quickly assess emotional readiness for transition [7–9].

5.4.1.2 Health Literacy

Health literacy is the "degree to which individuals have the ability to find, understand, and use information and services to inform health-related decisions and actions for themselves and others" [10]. Assessment of health literacy of the transitioning CHD patient helps tailor interventions with a patient-centered approach.

Two validated tools for assessment of health literacy include the single-item literacy screening [11] and newest vital sign [12].

5.4.1.3 Educational Preferences

Assessment for current educational level, educational challenges, and established individualized educational programs (IEP) through academic special education services helps to identify barriers to health education. Educational assessment includes preferred learning style and the identification of a preferred second learner.

5.4.1.4 Family and Social Support

Understanding the important family members involved in the patient's care and awareness of available social support through local and regional CHD support organizations help to identify the support system of the transitioning patient.

Assessment of the patient's family structure, cultural background/identity, primary household language, and socioeconomic status helps illuminate the context for the development of health-related beliefs and behaviors.

5.4.1.5 Health-Related Beliefs

There are well-known cultural health care disparities that exist across health care delivery systems. As we continually seek to provide culturally competent care to all our patients, this is particularly important during the transitional period as this experience helps lay the foundation of care throughout the lifespan of a congenital heart disease patient. Assessment and understanding of a patient's health-related beliefs are imperative to understanding their underlying motivation and behavioral patterns. Explanatory models are culturally determined beliefs that people hold about their health and illness, and assessment of these beliefs can be an important component of culturally competent practice [13].

The Brief Illness Perception Questionnaire (BriefIPQ) is a nine-item scale designed to quickly assess cognitive and emotional representations of patients' illness that has been validated in many disease states. The BriefIPQ demonstrated good predictive validity in patients recovering from a myocardial infarction related to functioning at 3 months follow-up, adherence to cardiac rehabilitation, and length of disability from work [14]. This type of assessment may be helpful in evaluating congenital heart patients' health-related beliefs that may cause barriers to adherence or place a patient at risk for lapses in care. Further research is needed on the use of assessment tools such as the BriefIPQ in congenital heart disease transitional care.

5.4.2 Health Knowledge

Understanding and knowledge about congenital heart disease and prior interventions may widely vary among patients. This understanding is often shaped by multiple factors including the degree of parental involvement, the patient's involvement in medical discussions, health literacy, and developmental and neurocognitive functioning. Assessing the current level of health knowledge helps direct educational interventions and goals. American College of Physicians transition tool "Health Knowledge Self-Assessment for Youth with Congenital Heart Disease" is aimed toward patients aged 12–17 years and specifically addresses the knowledge and skills in general health care and in understanding congenital heart disease [15].

CHD health knowledge assessments should include the name of their congenital heart defects, basic cardiac anatomy (both with a typical heart and with their congenital heart defects), and severity of the CHD. Additionally, knowledge of the number and type of surgical or catheter interventions, medication management, lifelong care needs, long-term sequelae of their CHD, and management of CHD are included in the health knowledge assessment.

5.4.3 Self-Care Assessment

The ability of a patient to autonomously manage their own health care needs is a developmental milestone that is impacted by parental relinquishing of management, neurocognitive deficits and developmental stage, and patient confidence. In

congenital heart disease, this transition from parental management to patient management should be well supported, and both the parents and patient should feel empowered to participate in this process. American College of Physicians tool "Self-Care Assessment for Young Adults with Congenital Heart Disease" is aimed toward young adults aged 18–29 years and addresses transitional skills for autonomous self-care as health care consumers [16].

The transitional APRN will need to assess the current degree of autonomy in order to develop interventions that foster and support patient empowerment. Some of this assessment is observational. Assess whether the patient or the parent is primarily asking questions, if the patient can independently answer questions, and whether the patient is the primary source of contact in subsequent interactions. Evaluation of self-care at home can be helpful toward understanding overall autonomy and is likely best achieved through interview. The ability to perform ADLs independently and manage schoolwork, finances, and employment responsibilities helps to inform the APRN on the patient's current capability to manage future health care–related items. Additionally, identification of barriers such as neurocognitive challenges that may limit autonomy is important when later establishing patient goals.

5.4.4 Care Preferences

Patient care preferences impact how they interact with the health care team. Assessment of preferences for communication (phone, secure messaging, email, in person), privacy (what items they would like to discuss privately), support (identification of preferred support persons), and comfort (what has worked well for them in the past) helps the care team to understand how best to deliver individualized care.

5.4.5 Future-Oriented Assessment

As a patient moves through transition an assessment of their plans for the future helps anticipate any additional care needs. If the patient plans to move to a different area due to schooling or work opportunities, this may impact transfer of care to the ACHD program. Additionally, discussion of how future life choices such as physically demanding career choices, rural living situations with limited access to health care, and family planning is impacted by their CHD. During adolescence and young adulthood, many are developing their long-term life goals, and a clear assessment of their understanding of how their CHD impacts these goals helps the patient make informed decisions.

Box 5.2 provides a sample comprehensive transition assessment incorporating the various domains of assessment and available tools.

Box 5.2 Transition Assessment Tools and Example Structure

Transition Readiness
- American College of Physicians Transition Readiness Self-Assessment for Youth with Congenital Heart Disease

Mental Health
- PHQ
- GAD
- PC-PTSD

Health Literacy
- Single-item literacy screener
- Newest vital sign

Education
Diagnosed with a learning disability or required special services in school?
Learning barriers: Reading, language, visual, hearing, emotional, cognitive
Learning preference: Listening, reading, demonstration, pictures, videos

Family Social Support
Family support:
Who do you live with?
Who supports you and how? Financially, emotionally, physically (transportation)?
What is the primary language in your household? Any secondary languages?
Do you have a cultural identity?
Do you have a religious preference?
Social support:
Do you know anyone else with CHD?
What CHD organizations have you interacted with?

Health-Related Beliefs
- BriefIPQ

Congenital Heart Disease Knowledge Assessment
- American College of Physicians Health Knowledge Self-Assessment for Youth with Congenital Heart Disease

Self-Care
- American College of Physicians Self-Care Assessment for Young Adults with Congenital Heart Disease

Care Preferences
1. What form of communication are you most comfortable with—Secure messaging, phone, virtual visits, in-person visits?
2. Do you have any specific fears or concerns regarding your care?
3. In the past, what comfort measures have worked best for you?

Future-Oriented Assessment
1. Do you have a life/career plan/goals?
2. Do you have a health care passport or health summary?
3. Are there any access to care concerns?
4. Are you aware of community resources?

[6–9, 11, 12, 14–16]

5.4.6 Diagnosis and Planning

After the completion of the multidimensional transitional assessment, the APRN can then analyze the data collected to establish nursing diagnoses and develop an individualized transition plan.

As defined by the North American Nursing Diagnosis Association International (NANDA-I), nursing diagnosis is defined as "a clinical judgment about individual, family, or community response to actual or protentional health problems/life processes. Nursing diagnosis provides the basis for selection of nursing interventions to achieve outcomes for which the nurse is accountable" [17]. Nursing diagnoses describe the problem, evaluate the etiology, and define the characteristics or the problem. Box 5.3 provides an example of CHD transitional nursing diagnoses.

After compiling the nursing diagnoses, the APRN can develop a plan for interventions, delivery of interventions, and collaborative goal setting with the patient.

Box 5.3 Example CHD Transitional Nursing Diagnoses

- Limited health literacy related to neurocognitive learning disabilities as evidenced by active IEP at school and the newest vital sign score of 2
- Impaired verbal communication related to primary household language different from the primary regional language as evidenced by the need for language interpreter
- At risk for social isolation related to lack of CHD peer support as evidenced by no knowledge of CHD peer organizations and lack of personal CHD peer relationships
- Lack of CHD-related knowledge related to parental dominance in provider communications as evidenced by inability to verbalize CHD diagnosis, past surgical history, or anticipated sequelae

5.4.7 Intervention Planning

The initial step in intervention planning involves prioritization of nursing diagnoses based upon severity and high-risk factors. The overall goal for transition is to avoid lapse in care; therefore, any nursing diagnosis that is high risk for causing a significant lapse in care should be prioritized first. Secondary prioritization should be given to any diagnoses within the transition readiness domain as any barrier to the transition process may make delivery of further care challenging. Further prioritization may be assigned based on severity of the nursing diagnosis or via patient preference.

There should be an initial conversation with the patient regarding the assessment, proposed interventions, expectations of the transitional process, and anticipated timeline.

5.4.8 Transitional APRN Interventions

Interventions should be specific and measurable and provide a clear timeline for implementation. Continuous reassessment of patient response to the established interventions creates highly individualized interventions.

For the transition readiness domain, the APRN must identify ways to overcome or mitigate barriers to transition readiness. Congenital heart disease knowledge

deficit interventions are focused on educating the patient. The APRN will develop an educational plan with consideration of the patient's health literacy, educational level, and preferred education delivery method, on a schedule that avoids overwhelm and promotes engagement.

Self-care interventions can utilize SMART goal setting. The acronym SMART stands for specific, measurable, attainable, realistic, and time related [18]. The goals should be set by the patient with the APRN serving as coach to help guide them through the process.

Care preference interventions involve creating the care delivery environment that supports the patient's preferences. Future-oriented interventions may involve further education, care coordination, and planning.

Examples of possible interventions for each domain are provided in Box 5.4.

Box 5.4 Examples of APRN Transitional Interventions

Transition domain	APRN interventions
Transition readiness	• Referral to mental health provider and patient establishes care within 1 month • Identification of a preferred secondary learner, assessment of secondary learner's health literacy and educational preferences. Clear and prominent documentation in the electronic medical record on the secondary learner's role • Provide language interpreter and interpret written material in primary language • Provide pamphlets or flyers for regional CHD peer organizations or institutional peer support groups/events • Patient will identify primary support persons and sign a communication consent form that will be scanned into the electronic medical record
Congenital heart disease knowledge	• APRN will spend 30 min of in-person visit reviewing typical cardiac anatomy/function in comparison to congenital heart defects and surgical interventions utilizing written materials, pictures, and videos. Patient "teach back" will occur at the subsequent visit and time will be allotted for questions
Self-care	• APRN assists the patient in developing a SMART goal to help promote autonomy with medication management: "John will download the pharmacy app on his phone and create an account before the next transition clinic visit."
Care preferences	• APRN assists patient in downloading the "my health online" app and plays a tutorial video on how to navigate the app. Patient will then send a message to APRN prior to next transition visit
Future oriented	• Social work or health coverage specialist to meet with patient to review insurance/health coverage and teach the patient how to navigate upcoming changes in health care coverage

5.4.9 Implementation

The implementation phase is when the APRN will be implementing the interventions set forth. This phase requires a clear communication between the APRN and patient to define the expectations, create an anticipated timeline, and establish an agreed-upon mode of delivery. A patient should leave every visit or interaction with an understanding of what their responsibility will be between visits. The implementation phase may take several weeks, months, or years depending on the identified barriers to care, knowledge deficits, autonomy, and risk for lapse in care. The APRN will continually monitor the patient for progress, adjust interventions as needed, and provide clear communication with the patient.

5.4.10 Evaluation

The APRN should create measurable goals to evaluate at the end of the transitional program. The ESC consensus described transition outcome goals that included improved quality of life, increased self-management, increased disease knowledge, maintaining continuity of care, appropriate health care consumption, disease control, access to care, interaction with adult providers, and interaction with peers. The authors described how these factors contribute to overall patient empowerment [1].

Acuña Mora et al. developed and validated a 15-item questionnaire designed to measure patient empowerment called the Gothenburg Young Persons Empowerment Scale (GYPES) that was validated with young patients, including patients with CHD [19]. Utilizing an objective evaluation tool such as the GYPES-CHD in a pre- and post-intervention questionnaire will help for evaluation of the transition program.

Examples of transitional care goals may include:

- Avoidance of lapse in CHD care during the transfer from pediatric cardiology to ACHD cardiology
- Improved patient post-intervention GYPES-CHD score indicating improved patient empowerment
- Patient completing the transition program and graduating with a comprehensive binder of resources and information within a 4-year timeframe

5.4.11 Future Nursing Research Opportunities

There are many opportunities for nursing research within the transition phase of congenital heart disease. We continue to strive to develop highly specific and sensitive validated assessment tools to aid in understanding the transitioning CHD patient. Existing validated tools mentioned in this chapter are often utilized but may be further improved upon specifically for the target population of adolescents and young adults with CHD.

As we engage in more virtual models of care delivery, we will need to understand their effectiveness compared to traditional in-person care delivery models. We additionally need further research on the effectiveness of the various models of virtual care delivery and usefulness of independent study programs.

One specific domain of assessment, health beliefs in CHD, has not been widely studied or explored and may help to better understand the variation in health beliefs and target supportive interventions appropriately.

Ultimately, we need further nursing research to support the autonomous role of the APRN within the development and delivery of transitional care for CHD patients.

5.5 Conclusion

The development of a transition program is a coordinated structured effort that works within the confines of institutional, regional, and systemic barriers and in an individually tailored manner. The structure of a transition program and the model of the program will depend upon the needs of the individual program.

A dedicated transitional APRN can develop and advocate for transitional services and champion the multidisciplinary team supporting the program. The nursing process of assessment, diagnosis, planning, implementation, and evaluation can serve as a framework for the development of the transitional program. Assessment involves evaluating the patient's transition readiness, health knowledge, self-care skills, care preferences, and future plans. This assessment is integral in clinical evaluation for nursing diagnoses, planning of interventions, and implementing the interventions. A strong objective evaluation of successful outcomes is helpful toward further evolution of the transitional program and helps with adaptation to future generations of transitioning congenital heart disease patients.

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Part III

The Role of the Advance Nurse Practitioner in the Care of ACHD Patients in Different Care Setting

Nursing Care in the Outpatient and Hospital Setting

6

Alexander Corless

6.1 Specialist Role

The role of the ACHD nurse in the outpatient and hospital setting will be dependent upon their locally defined scope of practice. It will be structured according to the professional title and the guidelines that underpin their role. The definition of an advanced nurse practitioner (ANP) can vary depending on the country of practice, the hospital or even the clinical discipline. In the United Kingdom, one is required to complete a post-registration, higher education qualification covering the four pillars of clinical practice: leadership, management, education and research [1]. A clinical nurse specialist (CNS), whilst not requiring such qualification, may fulfil a largesse of the same responsibilities in practice, but often with greater support from the ACHD medical specialists [2]. Whilst the practitioner's specific responsibilities may vary, the scope of practice and competency framework should be clearly defined by the practitioner's institution, to ensure patient safety and an appropriate utilisation of their skills [3].

In broader terms, the practitioner should be able to exercise higher levels of judgment and decision-making, each built upon the foundation of specialist education and clinical experience [4]. More specifically, the ACHD practitioner should have a reasonable understanding of congenital heart disease anatomy and physiology, the palliated circulations, potential disease progression and the common associated complications of their patient cohort.

Dependent upon the advanced nature of the role, the practitioner may work with some level of independence and even autonomy, but they must always have the access and capacity to feedback their patient assessment and care planning to the wider MDT. Furthermore, the practitioner must, in spite of their level of experience,

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remain conscious of the potential complexity of an ACHD patient and as such aware of their competency limitations. One notable discourse on the professional identity of the ANP sees them as medical substitutes, a lesser role and even a threat to the existing structure [3]. However, this belies the professional evolution of nursing role and how their innovative approach can contribute to a greater efficiency in healthcare practice [5]. Furthermore, the identity of the specialist nurse remains rooted in the foundations of nursing care, incorporating the compassionate approach to patient-centred care, whilst pursuing the highest level of clinical competency [3, 5].

There are many advantages to the ACHD specialist nurse role which broadly fall under two categories: support of the patient and support of the ward and outpatient clinical teams. One significant advantage relates to the relative constancy of the nurse specialist within an institution, especially in contrast to the medical team which is often more transient and sometimes non-specifically orientated to ACHD care, especially at the junior level. This steady presence allows for rapport building with not only patients but the other consistent body, the ACHD consultant group. However, in contrast to the ACHD consultant, the specialist nurse is accessible to the inpatient and outpatient alike. Furthermore, in institutions where there is no specialist ACHD clinical psychology support, the nurse may fulfil the role of proxy for non-acute psychological concerns relating to health anxiety, which is common in the ACHD cohort (see Sec 19).

The success of the ACHD nurse specialist, or indeed any specialist allied healthcare professional, is dependent upon the support of the medical team and reliant upon mutual respect. Furthermore, the extent to which these roles complement each other should be directly linked to an improvement in patient care.

6.2 Outpatient Setting

The familiarity and trust that are constructed through recurrent interactions are often cemented in the patients' awareness of the practitioners' specialist knowledge. Consequentially, they are frequently the preferred resource for outpatient support, advice and remote triage of symptom initiation or progression, above that of the primary care general practitioner (GP). This is generally facilitated by an outreach or helpline phone service, run by the specialist ACHD nursing team [6]. For this reason, the role should require the practitioner to be competent and professionally accredited with history taking and physical assessment training and fulfil the requirements under the locally devised competency framework [1]. Conversely, the practitioner can play a crucial role in contacting patients who fail to attend routine clinic visits or are at risk of being lost to follow-up, especially where complex familial, psychosocial, behavioural or addiction issues have been identified during transition. These can commonly represent potential barriers to engagement with healthcare services [7].

In this capacity, the practitioner may be the first contact for acute clinical deterioration, which may or may not pertain to the patient's cardiovascular condition, and thus a crucial first screen for appropriate healthcare management. They must be

Table 6.1 Cardiology red flags

• Chest pain
– OPQRST: **O**nset, **P**alliating/**P**rovoking, **Q**uality, **R**adiation, **S**ite & **T**iming. Associated symptoms. **Risk groups**: all. Higher risk: post-surgical, advanced valve disease notably supravalvular stenosis, known/high-risk CAD, palliated surgery involving coronary arteries (TGA arterial switch, PEARS prosthesis, supported Ross surgery)
• Shortness of breath/dyspnoea
– Rest/exertion, level of exertion, WHO functional class, positional; orthopnoea, paroxysmal nocturnal dyspnoea. Associated symptoms: cough, oedema or palpitations. Palliating/provoking factors: signs of peripheral/central cyanosis. **Risk groups**: all. Higher risk: heart failure, advanced valve disease/outflow tract obstruction, thromboembolic risk/history, arrhythmia history notably heart block and/or PPM/ICD in situ
• Haemoptysis
– Quantity/volume, consistency: blood-streaked sputum/frank blood/presence of clot. Associated symptoms: associated with activity. **Risk groups**: cyanotic patient/Eisenmenger's, post-repair PH, pulmonary AVMs, post-coarctation repair, anticoagulated
• Palpitations.
– Rest/exertion, descriptors: racing, pounding, fluttering, skipping. Regular/irregular, slow/fast, associated symptoms, palliating/provoking factors. **Risk groups**: arrhythmia history. TGA: senning/mustard repair, Fontan/univentricular, Ebstein's anomaly, Eisenmenger's
• Dizziness/light-headedness/pre-syncope/syncope
– Rest/exertion/postural, associated with palpitations, pre-syncope, aura, syncope, loss of consciousness. Hydration, diet, exposure to heat, infection. Alternative causes: disequilibrium, ataxia, vertigo, seizure, associated neurological dysfunction (TIA). **Risk groups**: arrhythmia history (as per palpitations group) notably SVT, non-sustained VT. Post-surgical, Eisenmenger's/PAH, advanced valve disease/outflow tract obstruction
• Oedema
– Location: lower limb/sacrum/abdominal/ascites. Severity: pedal/ankle/shin/thigh, weight increase. Fluid intake, diuretic regime, associated symptoms. **Risk groups**: heart failure (see Part 4, Sections 9 and 12; systemic RV, Eisenmenger's, Fontan circulation, post-surgical, arrhythmia history notably AF)
• Fevers/night sweats/high temperature
– Duration, general deterioration, weight and/or appetite loss, change to bowel habits, travel, obvious infection source (dental—pain, bleeding gums, oral abscess, recent dental work). Wound: surgical/trauma/post-procedure; redness, pain, swelling, pus, tracking. Recent tattoo, piercing or IVDU. **Risk groups**: infective endocarditis; previous infection, cyanotic conditions, prosthetic valves or prosthetic material up to 6 months post procedure/residual shunt, VSD, bicuspid aortic valve

*Unplanned pregnancy is a potential red-flag for all congenital lesions and should prompt referral to ACHD-obstetric consultant lead for assessment and care-planning [8]

alert to 'red flag' symptoms and have a pathway for escalation of care, which may include referral to a general practitioner, scheduling of an urgent specialist ACHD consultation, recommendation to attend accident and emergency or recommendation to call for emergency services (see Table 6.1). Conversely, a potential outcome of the phone assessment, and an important function of the nurse practitioner role, is patient reassurance and avoidance of admission, especially in a cohort with elevated incidence rate of health anxiety. Studies that have evaluated patient compliance with nursing teleconsultation advice have found that the level of reassurance is

significantly higher when advice pertains to escalation of care, compared with self-care [9]. Therefore, when escalation of care is not felt to be clinically required, reassurance should be supplemented with safety-netting advice in the event of worsening symptoms, and information should be provided on when the next routine outpatient assessment is scheduled. The spectrum of non-acute phone support might range from health promotion (see sec 17) and exercise guidance through to medication titration, should the practitioner have prescribing accreditation and have appropriate authorisation from their institution. Notably, outreach support may be sufficient in the titration of diuretic regimes in heart failure and anti-arrhythmia or rate-control medication in the presence of non-acute palpitations with known arrhythmia pathology, where competent to do so or with the support of a clinician.

Outreach phone support can also extend to providing advice to non-congenital centres caring for ACHD patients, who are known to the specialist institution. In such instances, it is important to take a comprehensive handover of the patient's condition and the locally devised care plan, utilising a standardised model for clinical handover to reduce the risk of inappropriate or delayed care, irrespective of whether their health concern relates to their ACHD condition [10, 11]. Appropriate support may be confined to providing background clinical information but may also require specialist advice on inpatient care, medication management, the need for updated cardiac imaging, general anaesthesia support or recommendation for transfer to an ACHD specialist unit [12]. Another notable nurse-led outpatient clinic format is a physiologist-supported clinic for interval assessment of mild lesions, including mild pulmonary stenosis, restrictive/small residual ventricular septal defects and repaired atrial septal defects, where a screening echo is performed prior to assessment. Furthermore, some specialist centres may have capacity for nurse-led urgent elective screening clinics for patients who contact their service with concerning symptoms [13]. Regardless of format, the clinical nurse should follow a structured format for history taking and assessment (see Table 6.2), take contemporaneous notes, summarise the findings, make a collaborative plan with their patient and root the consultation in a holistic approach [14].

Table 6.2 Outpatient review structure

Preparation:
• Review of past medical history/status on last review.
• Routine interval: TTE, cMRI, CPET/6MWT, PFT (consider up to date/provision/capacity/location/timing).
• Assess the need for chaperone/interpreter/carer dependent on potential psychosocial factors, age, language barriers and/or learning disabilities.
History taking:
• Lifestyle.
• Profession.
• Family/dependents/children.

Table 6.2 (continued)

• Stress/anxiety/depression/psychology support.
• Symptoms: OPQRST: **O**nset, **P**alliating/**P**rovoking, **Q**uality, **R**adiation, **S**ite & **T**iming.
– Chest pain.
– Dyspnoea.
– Palpitations.
– Dizziness/light-headedness.
– Peripheral oedema.
– Sleep: orthopnoea/paroxysmal nocturnal dyspnoea (PND).
• Exercise.
• Quality-of-life assessment.
• Other medical issues/diagnoses—including COVID history.
• Medications and allergies—including COVID vaccine status.
• Contraception/sexually active.
• Smoking/alcohol/illicit drug use.
• Dentist: last visit/recent treatment/infection, general dentition.
• Tattoos/piercings/skincare.
Assessment:
• Appearance: general well-being/personal care/habitus, work of breathing, speech, coordination, cognition.
• Mobilisation.
• Clinical observations: BP via right arm.
• Blue/pink.
• Height and weight: BMI.
• ECG.
• Limbs: oedema, clubbing (fingers and/or toes), temperature, hydration, splinter haemorrhages, stigmata, Osler's nodes, Janeway lesions, peripheral pulses, nicotine staining.
• Head/neck: central cyanosis/hydration (mucosa), acne, piercings, JVP, high-arch palette, neck webbing/syndromic features.
• Chest: habitus/malnutrition, excavatum/carinatum, scars, skin conditions, heave/thrill, heart sounds, respiratory sounds, accessory muscles, kyphosis/scoliosis.
• Abdomen: soft/distended, ascites, percussion, tenderness, palpable liver edge.
Care plan/outcome:
• Advocate yearly dental review.
• Avoidance of tattoos/piercings.
• Discuss IE risk – ± need for prophylactic antibiotic cover.
• Family planning/referral to ACHD cardiac obstetric pre-pregnancy counselling.
• Contraception: progesterone only unless low thrombosis risk.
• Exercise regime.
• Investigations—timing of MRI/TTE/CPET/CT.
• ± Referrals.
• Next review/discussion in MDT/admission.

6.3 Specific Lesions

Outpatient support should be tailored according to the clinical background and should acknowledge specific concerns pertaining to specific lesions. Appropriate preparation should be made to review the patient's history and most recent assessment and investigations ahead of review.

6.3.1 Valve and Outflow Tract Lesions

6.3.1.1 Left Heart

Congenital conditions of the left ventricular outflow tract, which include obstruction (LVOTO), sub-aortic stenosis and bicuspid aortic valve disease, may be associated with the effects consistent with an increased left ventricular afterload, hypertrophy, dilatation or failure (see Part 1). Furthermore, bicuspid aortic valve disease is associated with an elevated risk of aortic root dilation, dissection and acute aortic pathologies (see Part 1) [15].

In outreach support service, including teleconsultation, the symptoms that should concern the practitioner are progressive fatigue, exertional dyspnoea, chest pain, light-headedness, pre-syncope and, most concerningly, syncope [12]. Severe symptoms are often caused by a blunted blood pressure response on exertion, which is associated with a significant risk of mortality [16, 17]. In the post-adolescent group with known bicuspid valve stenosis under surveillance, it should be noted that symptoms can occur later in young people, and thus, particular attention should be made to subtle changes in exercise capacity and warrant a low threshold for escalation of care or urgent outpatient assessment and echocardiogram [12]. On ECG, evidence of progressive disease represented by increasing hypertrophy may be identified by elevated voltage in lateral precordial leads V5 and V6 [18]. Furthermore, ST segment changes and atrioventricular block may be apparent on ECG, particularly on ambulatory or exertional ECG monitoring. Serial exercise testing should be discussed with the clinician if there are concerns about vaguer symptoms such as worsening fatigue. Stress tests should be medically supervised if they are known to have moderate to severe disease, which is defined as those with peak velocity (V_{max}) >3 m/s across the aortic valve or aortic valve area (AVA) <0.8 cm on last echocardiogram [12, 19]. In severe aortic valve stenosis with a V_{max} >4 m/s on the last review, where the threshold for surgery has not been determined in ACHD MDT or whilst a patient is on the waiting list for aortic valve intervention, the British Society of Echocardiography recommends at least a 6 monthly repeat echocardiogram assessment [19]. The specialist nurse can be required to ensure such safety-netting assessment is in place.

Acute pulmonary oedema is a critical and late-stage finding in advanced outflow tract lesions, caused by a backpressure to the lungs caused by obstruction, raised left atrial pressure and reduced cardiac output. This finding may prompt urgent surgical referral or emergency inpatient management, so particular attention should be made

to signs including rapid-onset dyspnoea, progressive orthopnoea, wheezing, productive frothy sputum cough or the sensation of drowning [14].

Supravalvular stenosis may have coronary artery involvement, so exertional chest pain, whilst always a red flag, should be considered a significant source of concern and warrant urgent clinical assessment and potentially imaging.

Mitral lesions are most commonly detected and repaired in childhood, but in the presence of alarming symptoms, disease progression should be considered. Mitral valve regurgitation is heavily associated with atrial fibrillation, which may further accelerate disease progression, and as such, a new onset palpitations should prompt the practitioner to consider early rhythm assessment [20].

6.3.1.2 Right Heart

Isolated congenital pulmonary stenosis (PS) is a simple lesion that may be seen in nurse-led clinics, though mild PS (<30 mmHg) rarely deteriorates [12]. Moderate PS or secondary pulmonary regurgitation following ballooning needs lifelong follow-up due to the potential need for pulmonary valve intervention later in life. Furthermore, following Tetralogy of Fallot repair, many patients have residual, and sometimes progressive, PR [21]. Whilst asymptomatic for many years, this will lead to progressive RV dilatation and dysfunction and thus exercise intolerance, often from late adolescence onwards [22]. Owing to this concern, the practitioner should be alert to exertional fatigue, dyspnoea, chest pain and atrial arrhythmias. Although, it should be acknowledged that such symptoms often occur only in severe cases. On patient review, the practitioner should be mindful of right ventricular heave, a delayed P2 on auscultation and an ejection systolic murmur at the left sternal edge. On ECG, P-pulmonale, defined as a peaked p-wave greater than 2.5 mm in amplitude in the inferior leads or evidence of progressive RV hypertrophy and right axis deviation, will promote concern of increased right heart strain [18].

Ebstein's disease progression is characterised by a progressive right heart failure, and as such, concern is raised by peripheral congestion and oedema. Furthermore, the large atrialised portion of RV is a significate substrate for ventricular arrhythmia. Consequentially, this condition is associated with a sixfold increased risk of sudden cardiac death [23]. Palpitations should warrant concern and urgent escalation to the ACHD consultant and request be made for ambulatory ECG monitoring. In the presence of an atrial communication (PFO/ASD), right-to-left shunting may occur, resulting in cyanosis or exertional dyspnoea or even pre-syncope. The severity or acuteness of such findings would guide the need for expedited consultant review [12].

6.3.2 Septal Defects

Atrial septal defects (ASDs) are amongst the most common congenital lesions (see Part 1). Management and surveillance, under the guidance of the medical team, will be dependent on their repaired or unrepaired status. Unrepaired ASD cases can

present late and with associated comorbidities including AF (20% by 40 years) due to atrial dilatation, R-heart failure, LV dysfunction, HTN, paradoxical embolism, IE and in 10% progression to pulmonary hypertension (PH) [12, 15, 24]. Practitioners should be alert to exertional dyspnoea, pre-syncope or syncope and evidence of peripheral cyanosis that might indicate a reversal of shunt from right to left and thus PH. Pulmonary hypertension is a significant comorbidity with life-limiting implication, and as such, any red flags to indicate its development should prompt referral for echocardiogram and consultant review, with subsequent right heart catheter for diagnosis [25]. Outreach teleconsultation review of symptoms with unconfirmed cause should avoid diagnostic speculation to reduce patient distress and exacerbation of health anxiety [26].

On outpatient review, the practitioner should be alert to ejection systolic murmurs on auscultation or an RV heave. On ECG, there may be evidence of sinus node dysfunction, a prolonged PR or P-pulmonale [12, 18].

Restrictive VSD is a common lesion suitable for nurse-led follow-up, given the high threshold for repair in adulthood. Indication and, therefore, signifiers of concern on review, include evidence of infective endocarditis (IE) (see Table 6.1), ventricular dysfunction, arrhythmia or obstruction leading to the double-chambered right ventricle. Infective endocarditis has a mortality rate of up to 30% at 30 days, and thus, suspicion should be addressed with great urgency [27]. If clinical suspicion is raised through the description of symptoms including fevers, night sweats, fatigue and appetite or weight loss, in those with a predisposing congenital risk factor, then blood cultures should be arranged [28]. On face-to-face review, attention should be paid to the possible presence of Janeway lesions or Osler's nodes [29]. Three blood cultures should be taken 12 hr apart and prior to the commencement of antimicrobial therapy, as this will aid the successful detection of the specific bacteraemia in up to 98% of cases [30]. Referral for echocardiographic or CT imaging should be discussed with the IE and ACHD clinical group in the presence of high suspicion, and admission should be arranged upon the result of positive outpatient blood cultures to facilitate review. Confirmed cases of IE should furthermore be brought to the attention of the on-call CHD surgical team [28].

Larger unrepaired VSDs are rarer in the adult population, but they can also present with the reversal of shunt from left to right to right to left, and thus, the suspicion outlined under ASD lesions would also apply [12]. Given the close proximity of conduction tissue in the ventricular septum, both surgical and transcatheter VSD closures are associated with conduction and fascicular blocks [31, 32]. On review of the post-repair cohort, particular attention should be made to bradycardia on clinical observation and AV dissociation in the form of independent p-waves and QRS complexes on ECG, signifying complete heart block (CHB) [33]. CHB is associated with sudden cardiac death, and as such, urgent referral to the electrophysiology department and consideration for pacemaker insertion should be discussed with the clinical team.

6.3.3 Eisenmenger Syndrome

Eisenmenger's pulmonary hypertension patients should be under close routine surveillance in a congenital and PH specialist centre. Their complex medical requirements mean they are susceptible to mismanagement in a non-congenital centre and vulnerable to iatrogenic complications in all settings [12]. In the presence of major infection, peripheral vasodilation will cause a drop in systemic vascular resistance, which, in turn, leads to an increase in right-to-left shunting, resulting in progressive cyanosis and hypoxia. Outreach complaints of recurrent or systemic infection should be scrutinised and appropriate referrals be made, preferably to units with co-located specialist PH and ACHD support. On teleconsultation, symptoms suggestive of worsening hypoxia, including dyspnoea or exertional pre-syncope, should prompt urgent review, as such patients can be vulnerable to fatal syncope, especially on high exertion or physical stress [12]. Furthermore, patients in the critical care setting at a non-congenital centre are vulnerable to enthusiastic use of vasopressor therapy that might increase pulmonary vascular resistance and initiate a pulmonary hypertension crisis. Care should be taken to ensure there is direct communication with a congenital consultant cardiologist at the earliest opportunity.

Disease progression may present in the form of worsening right ventricular failure, so complaints of developing peripheral oedema or a drop in functional class should prompt a repeat assessment of cardiac function through echocardiogram, a neurohormonal hormone blood test, a 6-min walk assessment and clinic review [25, 34]. Patients are prone to compensatory haematological conditions including secondary erythrocytosis, with baseline haemoglobin (Hb) levels often >200 g/L [35]. Although hyperviscosity symptoms are often mild or even absent in this group, those with symptoms should be discouraged from venesection treatment, as the compensation aids adequate tissue oxygenation. If venesection is performed, simultaneous isovolumic fluid replacement should be administered to mitigate a potentially harmful drop in systemic blood flow [34, 35]. Furthermore, hyperviscosity contributes to the diathesis posed by both thrombus and haemorrhage, which coexists in the pulmonary circulation due to plexiform lesions and chaotic revascularisation. Additionally, the existence of a right-to-left shunt presents the risk of paradoxical emboli from venous thrombus causing stroke [36]. Consequently, one should be mindful of the symptoms of both pulmonary haemorrhage and neurological symptoms suggestive of cerebral infarct or abscess. There is additional risk posed by arrhythmias.

The incidence rate of Down syndrome in the Eisenmenger's cohort is possibly as high as 30% in the United Kingdom [37]. The spectrum of learning difficulties on this group therefore necessitates an effective channel of communication between the specialist and the community support team which might include their family, carer, support worker or community learning disability team. This is especially important in support of health promotion, which includes low-exertion exercise guidance, regular dental assessment, management of non-cardiac comorbidities and the communication of symptom progression and functional change. Lastly, non-cardiac procedures requiring mild sedation through to general anaesthesia will

require specialist congenital anaesthetic support, due to the significant mortality risk under anaesthesia, often quoted as high as 30% [38]. The specialist nurse can play a vital role in ensuring there is prompt and precise communication between the anaesthetist and relevant non-cardiac services.

6.3.4 Aortic Lesions

Coarctation of the aorta is often repaired in childhood with positive short-term and long-term outcomes [39]. However, multiple follow-up studies have also demonstrated a reduced median life expectancy compared with the general population [40], which may be attributable to hypertension, cardiovascular disease, re-coarctation or dissection [41, 42]. As the most significant complication, careful attention should be paid to the onset of severe, ripping or tearing pain on the anterior or posterior chest that can often radiate to the back, which should prompt urgent attendance to A&E for review of possible dissection [14]. Hypertension is associated with an increased risk in all-cause mortality, and thus, surveillance of blood pressure should be monitored intermittently, with awareness that a differential between pressures on the left and right arm may exist in the presence of a narrowing of the aorta, which is proximal to the left subclavian artery [43]. Secondly, in patients who have had a left subclavian flap repair, the left brachial arterial impulse may be absent. Consequentially, it is generally accepted that serial blood pressure monitoring, including 24 h monitors should be completed on the right arm. Additionally, on physical examination, one should simultaneously palpate the radial and femoral arteries to examine for radio-femoral delay, which may raise the suspicion of an obstruction or re-coarctation. Lastly, some connective tissue diseases, including Marfan syndrome, may be under surveillance in a congenital centre due to the risk of progressive aortic root dilation. Outreach concern largely relates to symptoms suggestive of dissection. All aortic lesions are best assessed radiologically with CT scan or MRI [44].

6.3.5 Transposition Complexes

The symptomatic concerns associated with transposition of the great arteries (TGA) are dependent upon the method of surgical repair (see Part 1). The most common form of repair, the arterial switch, can be associated with post-operative pulmonary artery stenosis, coronary artery stenosis and neo-aortic valve regurgitation [12]. Progressive development of a pulmonary artery lesion and valvular regurgitation should be adequately monitored through routine clinic surveillance under the medical team [13]. However, coronary artery stenosis, although rare to develop late in the post-repair cohort [15], should prompt concern on report of typical, central, crushing, chest pain which is provoked by exertion and alleviates on rest [14]. Non-invasive stress testing to include stress echo, stress MRI or cardiopulmonary exercise testing should be discussed with the medical team and considered before invasive coronary angiography.

Older TGA patients, whose surgical palliation occurred prior to the early 1990s, will commonly have had an atrial switch repair, namely Mustard or Senning surgeries (see Part 1). Within this cohort, there are a multitude of significant potential complications. Firstly, a systemic right ventricle, which is not morphologically designed for systemic pressures, is prone to accelerated rate of failure or dysfunction [45]. Consequently, the outreach practitioner should be alert to progressive dyspnoea, or in very-advanced- or late-stage failure, a new persistent frothy cough which may indicate pulmonary oedema. Furthermore, on ECG, a progressively dominant R-wave on V1 and dominant S-waves on V5 and V6 would signify right ventricular hypertrophy (RVH) or progressive strain [18]. Significantly, surgical baffle reconstruction of the atria creates substrate for arrhythmia, notably sinus node dysfunction or atrial flutter [15]. Careful attention should be paid to symptoms of either bradycardia or tachyarrhythmia, prompting an urgent 12-lead ECG or ambulatory ECG monitoring if symptoms are intermittent. Furthermore, owing to the systemic right ventricle and propensity for deterioration during prolonged fast atrial arrhythmias, the need for cardioversion and anticoagulation therapy should be expeditiously discussed with the ACHD clinician [46]. Lastly, there is 1 in 4 risks of developing a baffle leak, which may create either a L–R or R–L shunt, with the former potentiating further ventricular dysfunction and the latter cyanosis, which may worsen on exertion [12]. New exertional dyspnoea or pre-syncope and reduced oxygen saturation on clinical observation would necessitate expedited ACHD clinical review and appropriate imaging.

6.3.6 Tetralogy of Fallot/Pulmonary Atresia with VSD and MAPCA

Patients with unrepaired tetralogy of Fallot (TOF) are at a great risk of complications associated with cyanosis. As such, one should be mindful of the specialist care requirements as highlighted under the Eisenmenger's section. Furthermore, progressive aortic regurgitation and systemic hypertension create a greater risk of biventricular dysfunction, which would alert the practitioner to signs of progressive heart failure. Lastly, there is a risk of progressive aortic root dilatation akin to those with Marfan syndrome, so symptoms that suggest dissection should be cause for alarm.

Pulmonary atresia with a VSD is a complex cyanotic condition, which is associated with the formation of major aorto-pulmonary collateral arteries (MAPCAs), which provide systemic blood perfusion to segments or entire lobes of the lungs (see Part 1). Consequentially, there is a significant risk of pulmonary vascular disease or segmental pulmonary hypertension, which predisposes this group to great vulnerability during infection, general illness or arrhythmia, as noted in Eisenmenger's section. Furthermore, there is risk of progressive RV failure or stenosis on the MAPCA vessels. The complexity of this cohort ensures that in the presence of new-onset dyspnoea, arrhythmia, oedema, significant dehydration or systemic infection should prompt the practitioner to recommend urgent admission for inpatient assessment and management at a coronary care unit.

6.3.7 Univentricular Circulation/Fontan

The univentricular circulation is highly complex and can have a variety of malformations, but in the broadest terms, it refers to a lesion or lesions that result in a single functional ventricle, which, regardless of morphology, must provide the systemic cardiac output (see Part 1). As such, surgical palliation re-routes the inferior and superior systemic venous returns directly into the pulmonary arteries, referred to as total cavopulmonary connection or the Fontan circulation. The complexity of this condition requires this patient group to remain under regular surveillance in a specialist centre. As such, the triage nurse who encounters a Fontan patient with any red flag symptoms should consider urgent clinical review.

The lack of a right-sided pumping chamber means that systemic venous blood flow returns to the pulmonary circulation passively [15]. This concept is at the root of many specialist nursing precautions in both inpatient and outpatient management of the Fontan patient. Firstly, the pulmonary pressures are reliant on good peripheral vascular tone, which is supported by adequate hydration and patient activity. There should be lower threshold for concern when a Fontan patient presents or complains of conditions that might cause dehydration and, therefore, hypovolaemia, including gastrointestinal issues initiating diarrhoea or vomiting, heatstroke or infection [47]. Secondly, inactivity, weight gain or a steadily declining exercise capacity in this cohort is associated with worse outcomes including an increased risk of death [48]. As such, patients presenting in this manner should be provided with clear health promotion guidance and an appropriately devised exercise regime (see Sec 15 and 16). Moreover, they are at risk of a gastrointestinal condition called protein-losing enteropathy (PLE), which is defined as a low-serum total protein or elevated levels of faecal alpha-1 antitrypsin and is associated with diarrhoea and persistent or intermittent oedema [49]. This is a serious complication that is associated with poor prognosis which should be managed by an ACHD specialist clinician and at times may require inpatient management [13]. However, the nurse may support their PLE management through promotion of a high-protein, low-fat diet or make an appropriate referral to a dietician.

Passive systemic venous blood flow and right atrial blood stasis significantly increase the risk of spontaneous thrombus formation and thus catastrophic pulmonary emboli (PE). Whilst practice varies, this cohort is often on aspirin through childhood and adolescence and subsequently switched to formal anticoagulation in later life [13]. Though rare, Fontan patients that are lost to follow-up are at risk of not being adequately anticoagulated and thus a greater risk of PE. Therefore, a patient who contacts the service after a long period of disengagement should be screened to ensure medication concordance with anticoagulation therapy. If a specialist nurse is charged with organising a CT pulmonary angiogram, where there is the suspicion of PE, they should be mindful to stipulate the specialist requirement for dual-injection contrast delivery via the upper and lower limb cannulae to achieve bilateral pulmonary artery contrast flow. Standard protocol CT contrast delivery via an upper limb will preferentially supply contrast directly from the SVC to the right lung (85%) and may result in a falsely positive PE diagnosis [50].

A major complication of the palliated Fontan is venous congestion, which is associated with liver fibrosis, cirrhosis and hepatocellular carcinoma [13]. Consequently, the ACHD clinical nursing team is increasingly involved in the surveillance of Fontan-associated liver disease, through the arrangement of ultrasound and liver MRI, the performance of a FibroScan and the presentation of cases in specialist Fontan liver MDT, on at least a yearly basis. A new onset of abdominal ascites would prompt the need for repeat assessment and may necessitate inpatient hepatology assessment. As a consequence of portal hypertension and liver cirrhosis, this cohort are at risk of developing oesophageal varices and thus life-threatening ruptures and bleeds [51]. Haematemesis is a significant red flag, but so too, more subtly, is the presence of melaena [14].

A fenestrated Fontan, with a R–L shunt, and/or with pulmonary vascular disease or segmental pulmonary hypertension will have a degree of cyanosis with lower baseline peripheral oxygen saturation, and consequentially, one should be aware of concerns raised in the Eisenmenger's group. Equally, this cohort requires specialist anaesthetic support for GA procedures. Patients or tertiary centres who advise that a Fontan patient is scheduled for surgery in a non-ACHD cardiac setting should be advised that a congenital anaesthetist must be involved in the pre-operative planning, which the specialist nurse can facilitate. Lastly, Fontan patients are extremely vulnerable to atrial tachyarrhythmias, which have the capacity to rapidly become life-threatening [52]. Outpatients complaining of new-onset fast palpitations should be urgently reviewed and, if confirmed atrial flutter or atrial tachycardia, should be promptly admitted to a congenital specialist CCU for electrical cardioversion [13].

6.4 Inpatient Setting

6.4.1 Pre-assessment

A crucial pillar in the safe and effective care of the ACHD inpatient is a well-structured elective admission plan. In a patient group with complex care requirements, a comprehensive pre-assessment should be completed by a specialist nurse. The assessment should focus on the medical, physical, psychological and spiritual needs of the patient [5]. On review, the specialist should ensure that baseline observations, ECG, echocardiogram, blood tests and chest X-ray are available and should also consider the need for pulmonary function assessment (PFT), overnight oximetry, ambulatory ECG monitoring or referral to relevant non-cardiac specialists prior to admission [53].

There is an established association between congenital heart disease and neurodevelopmental morbidity. Learning disabilities caused by biological and environmental factors often coexist in the cohort of patients with genetic conditions such as Trisomy 21 and Noonan or Williams syndromes [54]. When planning an admission, the practitioner should acknowledge these specialist and holistic needs, often with the support of an established carer or guardian. Supplementary to this plan, the patient and carers should be supported in the production of a 'hospital passport',

which details a patient's likes, dislikes, communication aides and any reasonable adjustments that may be required on admission [55]. Furthermore, the practitioner should enlist the cooperation and support of a specialist learning disabilities nurse. Practically, one should consider the need for 1:1 bedside support and carer accommodation. For those requiring invasive procedures or surgery, modified or illustrative pain assessment charts can be employed to assist non-verbal communication. Needle phobia is a common problem in this cohort, and measures to reduce distress such as topical 5% lidocaine cream can be applied to access sites prior to phlebotomy or cannulation. In addition, in instances where repeat peripheral cannulation may be required over a long- to medium-term admission, one should consider the insertion of a medium-term line, such a PICC line, or even the retention of a central line post surgery.

Medication should be assessed to review the need for pausing, adjusting or bridging prescriptions ahead of surgery or cardiac procedures. Aspirin and P2Y inhibitors should be stopped up to 5 days prior to surgery unless there is significant stenosis, which may require the continuation of aspirin [53]. Anticoagulation should be paused and, in moderate risk cases, including a history of venous thromboembolic events in the last 6 months or valvular AF, should have bridging therapy with subcutaneous low-molecular-weight heparin. For those with moderate to high thromboembolic risk, including those with mechanical heart valves, early admission and IV unfractionated heparin may be required [56]. Angiotensin-converting enzyme inhibitors (ACEi) and angiotensin-II receptor antagonists may be ceased prior to surgery in some centres [53]. Importantly, pulmonary vasodilators for the treatment of pulmonary hypertension should not be stopped prior to surgery and often are instructed to be given immediately prior to anaesthesia to reduce the risk of rebound vasoconstriction or 'refractory pulmonary hypertensive crisis' [57]. Furthermore, pre-operative beta-blockers have been associated with the reduced incidence of post-operative AF. As such, they should not be ceased in advance of surgery, and initiation may be considered in those who are at high risk of atrial arrhythmias [58]. Lastly, the practitioner should initiate contact with non-cardiac specialists to review the safe optimisation of pre-operative medication regimes, specifically in haematological, respiratory, auto-immune and chronic pain conditions.

Infective endocarditis remains one of the most severe consequences of prosthetic cardiac intervention [59]. Moreover, undetected persisting odontogenic infections, as a result of poor dental hygiene and infrequent assessment, represent a significant risk for development of septicaemia and subsequent IE [60]. To compound the problem, those with CHD are at an increased risk. As such, practitioners must ensure those scheduled to have cardiac surgery or intervention should undergo a dental review and provide proof of examination prior to admission. The decision to treat outstanding dental lesions ahead of cardiac intervention will depend upon the bacterial risk the lesion poses and the urgency with which cardiac intervention is required. A 6-week period after invasive dental work should be considered when possible to reduce post-operative endocarditis risk. Those with learning disabilities represent a greater risk owing to difficulties maintaining oral hygiene, and as such,

consideration should be made to referral to a learning disabilities or special care sedation dentistry service.

It has become increasingly common for patients with Marfan syndrome or bicuspid aortic valve stenosis with aortic root dilation to be offered a personalised external aortic root support surgery (PEARS), to support the base of the aorta from further dilatation [61]. This personalised prosthesis requires the completion of a specialist CT to enable manufacture, which should be arranged in pre-assessment with a baseline renal profile blood test to ensure eGFR >30 mL/min/1.73 m^2, to reduce the risk of acute kidney injury on injection of CT contrast. Following PEARS prosthesis surgery, there is a commonly observed inflammatory response that presents as discontinuous episodes of fever but doesn't necessitate antibiotic therapy.

6.4.2 Admission

Ward-based cardiovascular care caters for medical admission (where CCU support is not required), surgical admission and recovery, cardiac intervention recovery and post-critical care recovery. The nurse-to-patient ratio will vary depending on the institution but will certainly exceed 1:2. Therefore, each patient will require an appropriately devised care plan that supports their safe and effective management. The practitioner can utilize their specialist knowledge to assist the ward team, whilst offering the wider ACHD MDT crucial updates on clinical progress. The specialist nurse may also serve to implement a wider MDT advice, raise concerns relating to clinical changes, discuss prioritisation with scheduling departments and bed managers and correspond with or make referrals to non-cardiac teams. Importantly, the specialist should seek to build a rapport with patients to facilitate comprehensive history taking, health promotion and emotional support.

6.4.3 Assessment

Initial assessment should take the individual patient's baseline parameters into account. In patients known to the institution, the measurements conducted in routine clinic surveillance should be acknowledged and recorded for reference. Routine clinical observations, including blood pressure, heart rate, oxygen saturations, respiratory rate, level of consciousness and temperature should be completed on a 4–6 h basis and incorporate an early warning scoring system, which prescribes a response to deterioration [5]. Appropriate modifications should be applied for patients with chronic, explained cyanosis, to prevent inappropriate action or escalation [62]. Alarm settings should be consciously set to reduce the risk of false or insignificant alarms that might initiate alarm fatigue [63]. Blood pressure measurements should be completed consistently on the right arm (see Sect. 6.3.4). An admission ECG should be presented for comparison to a baseline clinic trace and attention should be paid to common pre-existing findings, such as RBBB and axis deviation that can be present post ventriculotomy [64]. Although rare, those with

dextrocardia and situs inversus will require reversal of the precordial leads (V_1–V_6) to the right chest wall, to ensure correct interpretation [65]. A dry weight should be established for those with suspicion of or evidence of congestive heart failure.

Cardiac assessment must be completed on admission, routinely on each shift and in direct response to clinical deterioration. This can include a review of perfusion through completion of CAP refill assessment, peripheral temperature and palpation of peripheral and central pulses. End-organ function may be assessed through renal and liver function blood tests and urine output, which should exceed 0.5 kg/h. Attention should be paid to patient appearance, notably pallor, diaphoresis, oedema, ascites or signs of breathing discomfort, which may include the use of accessory respiratory muscles, nasal flaring and gasping. Chest auscultation should be conducted and adventitious cardiac and respiratory sounds documented. In patients who are sedate due to disability or clinical deterioration, a routine skin assessment should review potential pressure damage.

6.4.4 Oxygen

The inappropriate use of supplementary oxygen can cause patient discomfort, nasal irritation, reduced mobility and reliance. In light of the multiple lesions where peripheral oxygen saturations are reduced (<94%), the practitioners should ask their patients if they know their baseline saturation. For those with learning disabilities, where it may not be possible to appropriately communicate a baseline result, the practitioner should consult the clinic notes or the person's carer. Ward nurses should be educated on the mechanism for deoxygenated systemic venous blood bypassing the lungs in the presence of a right-to-left shunt, to avoid ineffectual attempts to drive peripheral saturation with oxygen delivery [15]. Conversely, supplementary oxygen should be considered to support relative hypoxaemia in the event of pulmonary oedema, pleural effusion and post-operative atelectasis, immediately post simple general anaesthesia recovery or for the management of acute respiratory distress. In such instances, it may be appropriate to conduct an arterial blood gas and escalate to the medical or surgical team [66]. In the case of patients with Eisenmenger physiology, desaturation may be driven by exertion, whereby a combination of increased cardiac output and high pulmonary vascular resistance (PVR) can increase the right-to-left shunt. Whilst there is some evidence to suggest ambulatory oxygen may provide symptomatic relief, it is not commonly prescribed [13]. Baseline oxygen saturations should return to normal upon rest and the decision to prescribe ambulatory oxygen be discussed with the congenital medical team.

6.4.5 Medical Admission

Medical admission may be driven by arrhythmia, heart failure, infection, inflammatory conditions or non-cardiac care. Admission for the assessment or management of suspected arrhythmia should first prompt the need for continuous telemetry

monitoring and daily 12-lead ECGs [53, 63]. High-risk groups, such as those with systemic right ventricles or univentricular circulations, should be reviewed promptly and escalation to CCU considered. Whilst there are a range of aetiologies that might precipitate arrhythmia, the most common reversible causes should be highlighted and investigated. Specifically, the practitioner should consider hypoxia, thyroid dysfunction, ischaemia, electrolyte imbalance, medication toxicity, reaction to beta-blocker or anti-arrhythmia medication, illicit drug use, pacemaker dysfunction or infective endocarditis [53]. As such, the specialist nurse should ensure blood tests to examine magnesium levels and renal, liver, bone and thyroid function are completed and toxicity levels and inflammatory markers considered. They should also support the medical team with the history-taking process. Furthermore, an echocardiogram should be considered to review the potential for progressive valve disease or changes in cardiac function, which may have precipitated or developed as a consequence of an arrhythmia [53, 67].

Atrial tachyarrhythmias are particularly common in groups with enlarged atria (see Sect. 6.3) and represent a significant risk factor for clinical deterioration. As such, the need for antiarrhythmic therapies, mechanical or chemical cardioversion or vagal manoeuvres should be considered with appropriate escalation to CCU (see sec 8) [13]. Care should be taken to scrutinise overnight telemetry traces to review the possible presence of pauses, prolonged PR intervals and bradyarrhythmia, which can be unintentionally dismissed as benign nocturnal sinus bradycardia [53]. Ward nurses should be alert to sudden changes in the heart rate, complaints of chest pain, palpitations, dizziness, light-headedness or altered levels of consciousness. For those with either a pacemaker or implantable cardioverter defibrillator device (ICD) in situ, the practitioner should alert the pacing physiologists and arrange device interrogation. Lastly, the practitioner should be alert to the need for anticoagulation dependent on an appropriate thromboembolic scoring system such as the CHA_2DS_2–VASc model and in agreement with the medical team [13, 68]. Ward nurses should pay attention to signs of bleeding through melaena, haematemesis or haemoptysis after the initiation of anticoagulation.

Congestive heart failure is one of the leading causes for medical admissions in the congenital heart disease population (see sec 9 and 12). Depending on the congenital lesion, the effect of right or left heart failure may present with peripheral or pulmonary congestion, respectively [13, 15]. On assessment, the ward nurse should record daily weights, a precise and contemporaneous fluid balance chart, and ensure regular renal function blood tests are taken [53]. The presence of oedema can be examined with review of the lower limbs, genitals and even the sacrum and assessed for signs of pitting. Chest auscultation should be performed and attention paid to crackles or reduced air entry. The use of diuretics should necessitate the strict vigilance of electrolytes, notably potassium, which is preferentially retained in the use of aldosterone antagonists and excreted with loop and thiazide diuretics. Furthermore, the use of thiazides and high-dose IV diuretics poses a significant risk to the development of an acute kidney injury. Consequently, the ward nurse or specialist should be alert to oliguria, an increase in oedema or an elevation in blood serum creatinine and alert the medical team who may consider a pause, switch or down titration of therapy, without ceasing diuretics entirely [69].

In some lesions with high venous pressure physiology, such as the univentricular population, the central gut oedema may inhibit the absorption of oral medications. In these instances, IV diuresis may be indicated, but additional medication for the management of arrhythmia or thromboembolic risk may also require review with alternative administration routes considered. Furthermore, this group is at risk of gross liver congestion, liver fibrosis and ascites and in advanced cases is at risk of spontaneous bacterial peritonitis, liver encephalopathy or development of hepatocellular carcinoma [13]. In the presence of ascites, liver function blood tests should be completed and referral made to a specialist hepatologist. In the same cohort, for those presenting with signs of PLE (see Sect. 6.3.7), the practitioner should consider a referral to the dietetics team and, where appropriate support, the completion of a food diary. Furthermore, all congestive heart failure patients should be counselled on low-sodium diet and foods that reduce the sensation of thirst, including chilled or frozen fruits like grapes or strawberries or sugar-free gum and hard candies [63]. Lastly, checking neurohormonal markers can provide another metric for severity of heart failure and provide a comparison point for previous or subsequent presentations.

The management of infective endocarditis (IE) may take place at ward level dependent on the stability of the patient, the necessity for surgery and the longevity of required antimicrobial therapy (see Sect. 6.3.2). For those with confirmed endocarditis, IV antimicrobial therapy may be administered via a tunnelled central line for up to 6 weeks, dependent on the causative organism [28, 53]. The bedside nurse is responsible for the prompt administration of therapy and in patients with an intracardiac shunt, and attention should be paid to infusions and administration lines to ensure there are no air bubbles to reduce the risk of paradoxical embolisation [70]. IV access sites should be assessed for signs of inflammation and phlebitis, access ports cleaned with 2% chlorhexidine wipes and dressings reviewed and changed on a 72 h basis. Surgical intervention may be required in severe cases, so the ward nurse should pay attention to cardiovascular status and signs of clinical deterioration, through changes in blood pressure, heart rate, fever and inflammatory blood markers. The specialist nurse should ensure that serial echocardiograms are arranged and the on-call congenital consultant and cardiothoracic surgeon are aware of the progress [28]. Lastly, the significant risk of embolic events should be considered, with periodic urinalysis performed for the presence of blood, and attention should be paid to neurological events suggestive of cerebral abscess or stroke [53].

6.4.6 Surgical Intervention

Admission for cardiac surgery requires careful pre-admission planning (see Sect. 6.4.1). For patients with univentricular circulation where adequate hydration is required to maintain peripheral vascular tone and pulmonary blood-flow, hypovolemia can lead to a drop in cardiac output. As such, where a patient is nil-by-mouth for an extended pre-operative period, IV fluids should be prescribed to reduce the risk of critical deterioration. Furthermore, strategies for preparing adults with learning disabilities or significant needle phobia should be agreed upon with the

consultant anaesthetist and might include the administration of an oral benzodiazepine prior to anaesthesia.

Post-cardiac surgery care should incorporate many of the standard principles of routine post-operative care, including haemodynamic surveillance, respiratory support, fluid management, pain control, wound care and psychological support. Owing to the risk of IE, post-operative temperature spikes >38 °C should prompt blood cultures and consideration for broad-spectrum antibiotics. Post-operative observations and cardiovascular assessments should be routinely performed and attention paid to a pericardial rub or dull respiratory bases on auscultation that might signify pericardial and pleural effusions, respectively. Steadily rising tachycardia and a decline in blood pressure should immediately raise the suspicion of tamponade [71]. Anti-inflammatories and low-dose diuretics should be considered for the treatment of mild-to-moderate effusions. Effective pain management will not only increase patient comfort but also enable deep breathing to aid the recovery of atelectasis and reduce the risk of lower respiratory tract infections. Following step-down from critical care, IV analgesia should be ceased in favour of oral therapies. Furthermore, paracetamol should be used ahead of weak opioids such as codeine, to promote activity and mobilisation. If codeine-based agents are used, laxatives should also be prescribed, and record should be made to bowel motions. Careful attention should be paid to the quality and intensity of chest pain and a cardiac cause refuted before it is presumed to be post-operative and, therefore, musculoskeletal in origin. Following surgeries that interfere with the aortic root, such as the supported Ross procedure, there is a risk of coronary artery compression. Whilst moderate elevation in troponin is common after cardiac surgery, significant elevation may signpost a major cardiac event [72]. In addition, daily ECGs should be performed and scrutinised for evidence of ST elevation, atrial arrhythmia or conduction blocks [18].

Ward nurses should also ensure that pro re nata anti-emetic therapies are prescribed, as needed, to manage post-operative or medication-initiated nausea. Adequate hydration and nutrition needs should be assessed and fluid status monitored. The sternotomy surgical wound should be assessed by the ward nurse on each shift and open to air after 48–72 h. The safe removal of mediastinal and pleural drains should be performed according to local policy and only after a defined period of minimal output. Following the removal, chest insertion sites should be covered to reduce the risk of infection and pneumothorax [5] and a chest radiograph performed. Temporary epicardial pacing wires are commonly inserted at the end of surgery to enable prospective support for post-operative conduction block or poor cardiac output. The removal of pacing wires will often be performed on day 3–4 and shortly prior to their post-operative echocardiogram to exclude the presence of tamponade. For the same reason, clotting factors should be within normal limits, anticoagulation paused and INR <2.5 [5].

6.4.7 Catheter Intervention

Prior to admission for catheter intervention, patients should be assessed for the requirement for critical care recovery. This is often advised in high-risk cases, including the cohort of patients with systemic right ventricles, cyanosis or univentricular circulation. In these instances, a timely request should be made for a critical care recovery bed and their prioritisation discussed with the bed management and critical matrons.

ASD device closure is one of the most common congenital catheter procedures. Despite a high safety rating, there exists a rare post-procedural risk of device embolisation. The consequence of embolisation can include atrial or ventricular arrhythmia, outflow tract obstruction or ischaemic events [73]. The ward and specialist nurse should ensure telemetry is employed for several hours into recovery and attention be paid to ECG changes, chest discomfort or dyspnoea. Complete device embolisation may require urgent surgical retrieval, which would necessitate the specialist nurses' support to promptly coordinate with the interventional cardiologists, congenital surgeons, theatre coordinators and critical care bed management teams [73]. A potential catastrophe ensuing risk post coarctation and pulmonary tract stenting is arterial dissection. The ward nurse should pay particular attention to complaints of a severe ripping pain in the chest that can radiate to the back, or haemoptysis, and escalate concerns to the medical and critical response teams [14].

Following step down from the recovery unit, supplementary oxygen is often still required to support a reduced respiratory effort following general anaesthesia [66]. The nurse should intermittently review femoral catheter access sites for signs of haematoma, bleeding or ischaemia. In the presence of haematoma or bleeding, manual pressure should be applied until haemostasis is attained [53]. The lower limb should be intermittently inspected for warmth, CAP refill and pulse pressure and consistency, to ensure sufficient peripheral perfusion [14]. Discharge advice for groin care should include the avoidance of taking a bath for up to 7 days to reduce the risk of infection and bleeding posed by maceration of the wound bed and vasodilation. Those discharged following transcatheter septal device closure should be reminded to continue an appropriate antiplatelet therapy regime, which can differ between centres, but should be given for up to 6 months whereupon the device is adjudged to have developed a complete endothelial layer [74]. Following the new insertion of a pacing system, patients should be assessed for dyspnoea, wound pain, swelling or discharge, arm swelling or fever. Furthermore, a subsequent pacing check should be completed prior to discharge and appropriate device guidance supplied by the pacing physiology team.

6.4.8 Psychological Support

Prolonged admission, cardiac surgery or new diagnoses are potential sources for psychological distress, particularly for younger patients. Whilst traumatic or life-changing effects of events such as cardiac arrest, urgent surgery or progression

towards palliative care may be obvious sources for distress, the practitioner should also be mindful of the many and often subtle psychological challenges that this patient population faces throughout admission and on a daily basis (see sec 19). The specialist nurse should utilise rapport-building strategies and time on the ward to discuss patient concerns, learn about their social challenges, discover the family and support network and acknowledge the acute stressors that may affect their inpatient journey and subsequent discharge. There is a well-established association between post-traumatic stress disorder (PTSD) and cardiac surgery or a prolonged ICU stay in childhood or infancy [75]. These issues may be triggered, exacerbated or even initiated on subsequent admissions in adulthood, especially if confronted with physical pain and discomfort or a potential change in lifestyle, exercise capacity or functional limitation as a consequence of disease progression. Care should be taken to show patience and compassion and to explore the availability and appropriate suitability of acute or long-term psychology service referral (see sec 19).

6.4.9 Discharge

The primary goal of discharge planning is to ensure that there is a smooth transition from acute inpatient support to community-based care. The strategy will be personalised according to each individual's needs and dependent on their level of independence but should incorporate some key characteristics. A comprehensive discharge plan may incorporate lifestyle, exercise, medication and wound care advice. In addition, the specialist should signpost organisations that provide information and support, which in the United Kingdom include the Somerville Foundation, the British Heart Foundation (BHF) or the Pulmonary Hypertension Association (PHA UK). The practitioner should also ensure that specific referrals are made to the relevant services. These may include but are not limited to cardiac or respiratory rehabilitation services, physiotherapy, occupational therapy, anticoagulation clinic, pacemaker physiologist, social worker, addiction services, psychology or counselling services or palliative care (see sec 20–22). Those who are newly commenced on anticoagulation should be educated on potential lifestyle considerations owing to their increased risk of bleeding [53]. Furthermore, those treated with warfarin may be supported in the procurement and use of a CoaguChek device for monitoring INR results at home [76]. In the United Kingdom, the specialist nurse can support an application for funding the device via the charitable organisation, the Somerville Foundation. Prior to discharge, the practitioner should ensure that the patient has an appropriately timed follow-up appointment with the ACHD cardiologist and cardiothoracic surgeon. Finally, the patient should be made aware of the safety net provided by the specialist outreach support service and educated on the potential, red flag symptoms associated with their condition or intervention (see Table 6.1).

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Nursing Care in the Cath Lab

7

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7.1 Cath Lab

The cardiac catheterization laboratory (cath lab) is a hospital area specialized in advanced cardiac procedures (Fig. 7.1). It is an essential tool for third-level centers of congenital heart disease (CHD) treatment.

In the past decades, invasive cardiology was often necessary to finalize or to confirm a diagnosis of CHD. This option made progressively uncommon once alternative and non-invasive tools like echocardiography, CT, and MRI become available for diagnostic purposes in cardiology. On the other hand, interventional cardiology progressively expanded the possibilities of percutaneous treatment of simple and complex CHD, from premature infants to elder patients. Therefore, the cath lab needs of a large pool of operators able to face different scenarios, and it may change several times in the same day.

7.1.1 Indication to Heart Catheterization

A cardiac catheterization can be indicated for diagnostic or interventional procedures [1, 2].

The most common indications to diagnostic studies are:

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Fig. 7.1 A cath lab room

- Right heart catheterization for pulmonary hypertension.
- Myocardial biopsy in cardiomyopathies or myocarditis or after heart transplant.
- Coronary angiography.
- Complex CHD with incomplete diagnosis or pending queries despite a full (when applicable) non-invasive assessment.

Cardiac catheterization allows the assessment of PVR, ventricular diastolic function (including constrictive and restrictive physiology), pressure gradients, shunt quantification, and evaluation of extracardiac vessels such as aortic pulmonary collateral arteries. In shunt lesions, catheterization including testing of vasoreactivity remains essential in the decision on defect closure.

The most frequent percutaneous interventions in ACHD patients are [3–5]:

- Closure of heart defects by device placement.
  - Atrial septal defects.
  - Ventricular septal defects.
  - Patent ductus arteriosus.
  - Fistulae embolization (artero-venous, veno-venous, artero-arterious).
- Balloon dilatation vessels (angioplasty).
- Balloon dilatation of heart valves (valvuloplasty).
- Arterial stent placement.
- Percutaneous heart valve replacement.

7.2 Role of a Cath Lab Nurse

Cath lab nurses are a vital part of the cath lab team. Usually there are two nurses for each operating room (OR). They have several tasks during the day, regarding the OR preparation, the patient preparation, the assistance to the patient and to the physician during the procedure, the assistance for hemostasis once completed the procedure, the patient mobilization, and post-operative monitoring. The nurses should have pre-fixed operative instructions, to achieve a clear distribution of tasks, to minimize the errors, and to save time and be efficient.

For these reasons, the nurses are the "pacemaker" of the OR, by monitoring the availability of the room for a given procedure, by preparing the patient for the procedure, and by assisting the physician by following a temporal and priority order.

They have many roles throughout the entire process, including to assist the physician as second or third operator during the procedure, when needed [6–9].

The daily activities of cath lab nurse are summarized as follows:

1. Daily room and equipment check (before starting)
2. Patient/procedure-based OR check
3. Procedure-specific tools check
4. Patient's check (see "patient preparation")
5. Patient's accommodation and monitorization
6. Assistance to anesthesiologists
7. Sterile trolley preparation
8. Assistance to the interventional cardiologist (dressing and catheter/devices deployment)
9. Drugs administration during the procedure
10. Second/third operator in the procedure, when needed
11. Hemostasis assistance after sheath removal
12. Patient's mobilization
13. Post-operative monitoring

7.3 Operative Room Characteristics

A cath lab room should be set up for patients with different features and needs. However, some tools can be considered essential in any procedure:

- Vital parameter monitoring system
- Syringe pumps
- Defibrillator
- Mechanical ventilator
- Oxygen supply
- Resuscitation trolley
- Temporary external pacemaker
- Coagulation analyzer (ACT machine)
- Anesthesiologist's trolley
- C-arm X-ray generator (biplane system recommended)
- Surgical lights
- Radiolucent surgical table
- Contrast injector
- Recording system (EKG and pressure)
- Overhead lead glass shield
- Sterile trolley
- Suction equipment

- Equipment cupboards
- Hemogasanalyzer
- Shelves, cabinets, or closet to collect catheters and devices

In addition to this minimal data set, additional tools and devices should be taken into account in the room organization:

- Echo machine for transesophageal echo and echo-assisted vascular access cannulation
- ECMO support or other mechanical support machines
- Electrical scalpel
- Radio-frequency machine

Given all the equipment listed and the operators needed in the room (from 4 to 8), the width of the room should be at least 100 m^2.

7.4 Room Preparation

Before to start the daily activity, the nurse should prepare the room based on the patient list. This includes many checks:

- ***Mechanical ventilator***. Pediatric patients usually perform the procedure sedated or in general anesthesia, while adult patients might undergo the procedure in local anesthesia. On the other hand, some procedures might be performed in general anesthesia irrespective of the age and the patient compliance. Finally, some procedures might suffer from complications requiring rescue anesthesiologic support. In conclusion, a mechanical ventilator should be ready for use in the room before starting the procedure. Based on the age and the weight of the patients, three different sizes of connector tubes are available:
  - Neonatal: 10 mm smoothbore breathing system with a neonatal electrostatic filter HME and, in this case, a L connector to reduce dead space and a 0.5 L latex-free breathing balloon.
  - Pediatric: 15 mm smoothbore breathing system with a pediatric electrostatic filter HME, a pediatric catheter mount, and a 1 L latex-free breathing balloon.
  - Adult: 22 mm smoothbore breathing system with an electrostatic filter HME, a catheter mount, and a 2 L latex-free breathing balloon.

A CO_2 detector is available for all the systems described. This is particularly helpful in case of ventilation by laryngeal mask, given the hard control of the device stability/efficacy. In addition, a too long connection system may give a too high death space reflecting on a progressive increase of pCO_2 and consequent respiratory acidosis. On the other hand, a too short connector may find it difficult to manage the C-arm X-ray, especially for lateral or near lateral projections. The machine should be calibrated before the first patient of the day and after any change of connector's length.

- In addition, the availability of masks and tubes should be checked:
  - Laryngeal mask: There are different measures from 1 to 6.
  - Endotracheal tube: There are different measures (from 3 to 8 mm) and two kinds of tubes (cuffed or not cuffed).

The choice of kind of ventilation and size and type of the device depend on the patient's size, airway anatomy characteristics, the expected duration of the anesthesia, the type of procedure planned, and the airway approach (nose or mouth). Despite some algorithms being available to predict the mask or the tube size for each patient, the choice of the device is demanded by the anesthesiologist after the patient's airway anatomy evaluation.

- ***Defibrillator***. Early defibrillation is crucial for survival after a malignant arrhythmia, mostly in compromised patients. The readiness to defibrillation is the major determinant of the success of resuscitative attempts. Therefore, a biphasic defibrillator should be available and ready to use in all the single operating room and in the pre-/post-operative rooms. Two sizes of pads are available: pediatric (for <8 years or < 25 kg children) and adult. The pads might be placed before starting the procedure (in very-high-risk patients) or just when the maneuver is indicated. Despite the preventive application of the electrodes might save some seconds in case of malignant arrhythmias, the presence of the pads on the chest may impact on the X-ray images and on the X-ray dose delivered so that the preventive application of the pads should be carefully evaluated. The position of the pads is on the right side of the chest, just below the collarbone, on the left side below the nipple for ventricular arrhythmias, in the middle of the sternum, and on the back face to face in atrial arrhythmias. The nurses must check battery status and verify the presence of working (and not expired) tools before starting the activity.
- ***Emergency pacing equipment***. In case of acute and complete AV block during procedure, a temporary pacing is needed. The pacing might be performed by using the defibrillator pads and the dedicated function, when available on the machine, or by placing a dedicated stimulating catheter (usually from a venous access in the right ventricle) and a dedicated connecting cable to an external pacemaker. The nurses have to check battery status and verify the presence of a working tool before starting the activity.
- Drugs and other ***emergency materials*** in the resuscitation trolley and in the ***anesthesiologist's trolley***. Restoration of the missing materials and drugs is made daily; expiration dates should be checked every month.
- ***Oxygen supply***. There are different types of oxygen supports:
  - Nasal cannula: It can be neonatal, pediatric, or adult.
  - Oxygen mask: It can be pediatric (there are different measures depending on the age of the patient) or adult.
  - Reservoir oxygen mask: It can be only adult.
  - General anesthesia.

There are two kinds of oxygen supplies: an oxygen cylinder (useful for patient handling) and terminal units in the walls, to ensure continuous oxygen flow to the respirators and to the flowmeters. The availability of oxygen supply, especially in the cylinder, should be verified periodically.

- ***Suction equipment***. There are many sizes of sterile endotracheal suction catheters, from 1.7 mm (size 5) to 8 mm (size 24). A suction system should be available in any operative room and in the pre-/post-operative area.
- ***Coagulation analyzer***. The anticoagulation monitoring during interventional procedures should be periodically verified, to prevent intravascular thrombosis. Usually, intravenous heparin is administered once vascular accesses are obtained, at the dose of 50–100 IU/kg. However, response to heparin might be unpredictable, so the effectiveness of the anticoagulation should be tested 15–30 min after the first administration and before any interventional maneuvers and before removal of vascular accesses. In very sick patients, the coagulation might be impaired, so the use of heparin might be more careful and more frequently monitored. Nurses should run the quality check of the device daily, while a detailed quality check should be performed weekly.
- ***Infusion pumps***. Nurses should verify the presence of at least two functioning pumps and check their battery status.

7.5 Procedure's Management

7.5.1 Patient Preparation

During the patient's admission in the room, one of the two nurses checks patient details and completes the pre-procedure checklist:

- To provide patient with information and ensure patient has signed the consent form and fasting for at least 8 h.
- To exclude pregnancy status when the patient is a woman in fertile age verifying β-HCG levels.
- To check blood supply availability, when applicable.
- To check allergies and, if the patient has an ascertain or suspect contrast medium allergy, the application of the most appropriate pre-medication protocols.
- To verify, together with the doctor, the blood test results (in particular, INR, CRP, and creatinine) and appropriate medical therapy management (antithrombotic therapy, antibiotic therapy, hydration, etc.). Usually, anticoagulant like warfarin and NOAC should be withdrawn 3–5 days before the procedure and replaced with heparin to have an easier control of the anticoagulation during the procedure.

Furthermore, nurses should assess anxiety levels and create a friendly and warm environment.

Nurses invite the patient to empty the bladder and accompany him or her inside the cath lab. They ascertain the patient is wearing an exam gown, surgical cap, sandals, and identification bracelet.

Once accompanied the patient in the operating room, nurses have two different tasks: one of them will take care of the patient and the other one will prepare the sterile operating field.

The first nurse (circulating nurse) lays the patient in operatory bed and takes off the exam gown and sandals.

When the patient is adult, circulating nurse must:

- Remove every jewel.
- Check trichotomy of the areas where the catheters will be inserted: the groin area (left and right) and wrists (usually right).
- Place electrodes and record an electrocardiogram.
- Place oximeter.
- Make sure peripheral access works fast and place an intravenous infusion.
- Complete intra-procedural checklist.

Regarding the correct positioning, adult patient needs to stay supine with the arm in line with the body or upon the head if the access is femoral (depending on the kind of procedure planned). In the case of radial approach, the arm involved needs to lay upon a rigid support with a roll that exposes the wrist (Fig. 7.2).

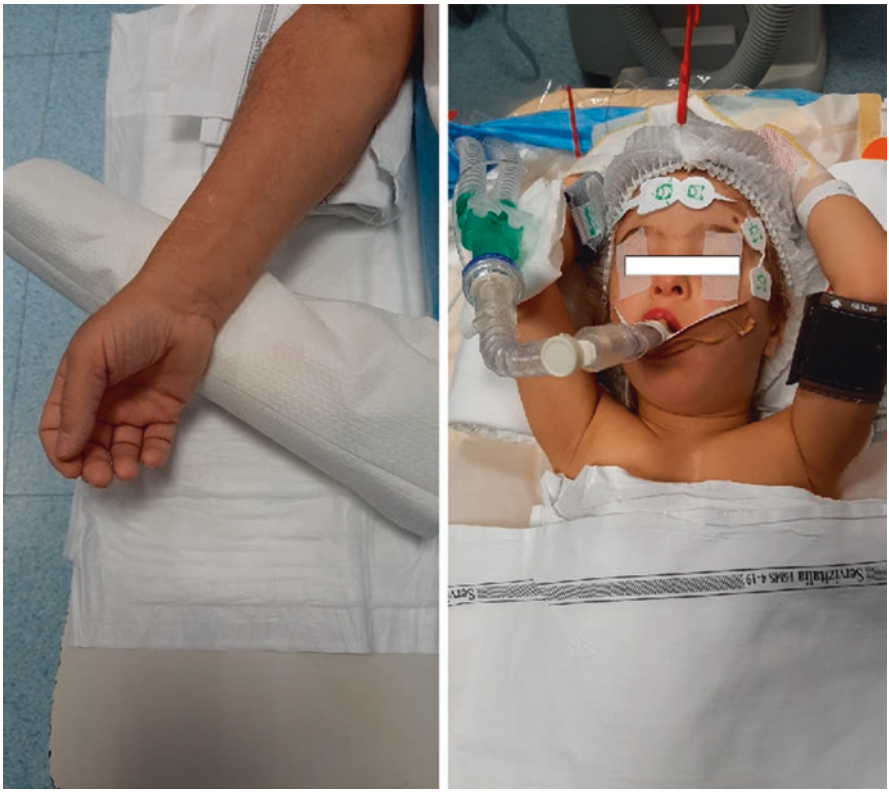

Fig. 7.2 On the left, the arm positioning in the case of radial approach. On the right, correct positioning in a pediatric patient

When the patient is neonatal or pediatric, circulating nurse must:

- Place electrodes and record an electrocardiogram.
- Place oximeter.
- Place pressure cuff.
- Assist the anesthesiologist in the positioning of a laryngeal mask or an endotracheal tube.
- Make sure peripheral access works and collaborate with the anesthetist in placing intravenous infusion and administering medications
- Complete intra-procedural checklist.

An important job of the nurse in pediatric procedures is to ensure the correct positioning. The patient needs to lay supine on the cath lab bed. If the procedure needs lateral views, the arms should be raised upon the head, tied together, and fixed on the bed, in order to minimize the X-ray exposure (Fig. 7.2). If the procedure needs just a posteroanterior views, the arms can be left in line with the body.

In neonate and small infants, a lining roll is applied below the pelvic zone to get more superficial and straighten the femoral vessels.

While the first nurse prepares the patient for the procedure, the second nurse (scrub nurse) collects the required tools and prepares the sterile trolley with appropriate equipment.

7.5.2 **Sterile Trolley Preparation** (Fig. 7.3)

In the case of a diagnostic intervention, materials to be used are:

- Catheterization sterile kit
- Gauze
- Syringe: 5, 10, 20 mL
- Needles: 18, 16 G

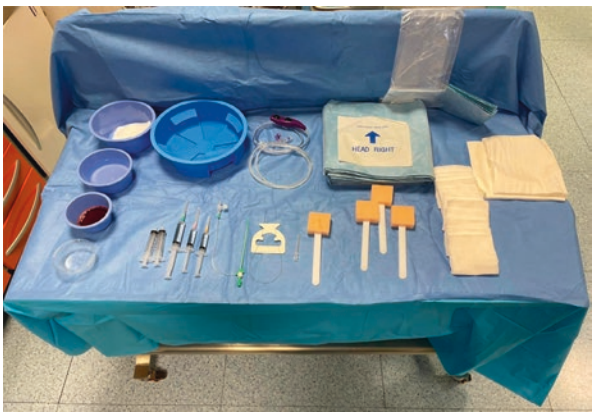

Fig. 7.3 Sterile trolley preparation

- Scalpel
- Angiographic kit
- Guidewire
- Sheath
- Catheters
- Heparinized saline (5000 IU heparin in a 500 mL saline solution)
- Lidocaine for local anesthesia

7.5.3 Cath Lab Tools

The ***sheaths*** are essential to perform the exam. These allow the introduction of catheters and guidewires into the vessel and warrant the hemostasis during the procedure. The caliper of the sheath is measured in French (Fr). Different from the catheters, the size in Fr (1/3 mm) is referred to the internal caliper of the sheath (in a 4 Fr sheath, a 4 Fr catheter can be introduced and so on). The caliper ranges from 3 to 18 Fr. In addition, the sheath may have different lengths, ranging from 5 to 45 cm. Most frequently, a 7 or 11 cm long sheath is used for routine procedures. The sheath's choice depends on the size of the patient, the vascular access used, and the kind of planned procedure. Larger sheaths allow the use of larger catheter but are exposed to a higher risk of vascular complications. If the venous catheterization usually warrants a higher compliance also in small babies, arterial cannulation may be difficult in smaller children and in patient facing several percutaneous or surgical procedures. Therefore, the principle is to use the smallest sheath (and catheter) possible for a given procedure. In special conditions, longer sheaths (long sheaths and Mullins sheaths) or hydrophilic sheaths may be used to allow a safe and effective vascular access or to perform interventional procedures.

In conclusion, despite the choice of the sheath being standardized for a lot of procedures (i.e., 5 or 6 Fr sheath for arterial access and 6 or 7 Fr sheath for venous access), the sheath choice is demanded to the physician, based on the intended procedure.

There are many ***catheters*** available for heart catheterization. They differ in kind (diagnostic catheters or guiding catheters), type, and size. There are two categories:

- **Diagnostic catheters**: They are used for diagnostic purposes. These catheters can be used to inject the contrast medium into large or small vessels/chambers or to measure the pressure (and pressure gradients) at any site. The catheters can be single (end) hole (coronary catheters) or multi-holes (pigtail, NIH, and MPA-II). There are many kinds and sizes of catheters. The size of diagnostic catheters ranges between 3 and 6 Fr. The most frequently used types are Judkins left (JL) and Judkins right (JR) coronary catheter series, pigtail, multipurpose I (end hole) and II (multiple holes), Amplatz left (AL) and Amplatz right (AR) coronary catheter series, left and right coronary bypass catheter (LCB and RCB) families, and internal mammary (IM) catheter series. Usually made of plastic materials (polyurethane, polyethylene, Teflon, or nylon), they might have fine metal or plastic

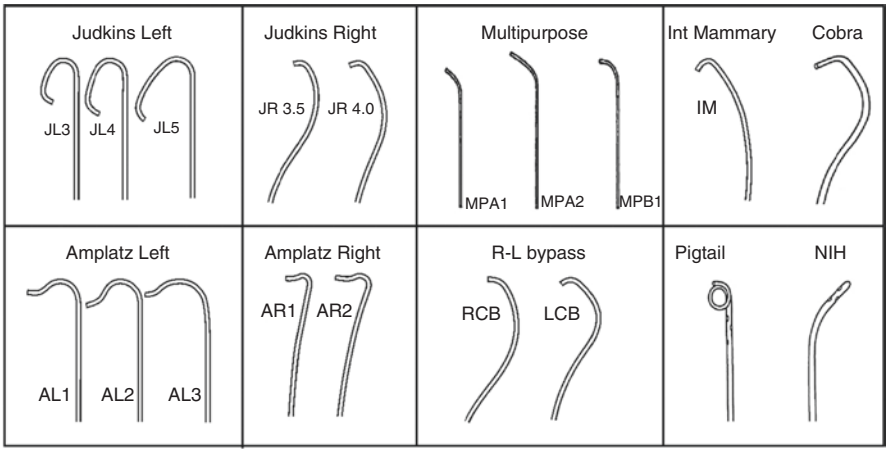

Fig. 7.4 Diagnostic catheters

strands incorporated into the wall, which enables the catheter tip to be responsive to gentle rotation of the shaft (Fig. 7.4).

- **Guiding (interventional) catheters**: Specifically designed for interventional coronary procedures, they can be used for different diagnostic and interventional purposes in CHD. The caliper ranges from 5 Fr to 8 Fr, and the available shapes and the sizes are the same for diagnostic JL, JR, AL, AR, RCB–LCB, and IM catheters. Compared to diagnostic catheters, they have a larger lumen and thinner but stiffer (armed) walls to provide an adequate support for the balloon catheters, stents, and device delivery. Some of them have shorter and more flexible tips to decrease catheter-induced trauma.

The catheters are usually inserted and maneuvered by using a **guidewire**. There are several types of guidewires. They can be clustered by size, length, stiffness, and coating (hydrophilic coating or non-coated guidewires). Roughly, larger guidewires and stiffer guidewires are more supportive, while smaller and floppier guidewires have a higher crossability and trackability. A detailed description of all the guidewires and their indication for use is far from the purpose of this chapter. In the following paragraphs, we will summarize the basic characteristics of guidewires.

- ***Size***: The guidewire's size is measured in inches. The most used guidewires range from 0.014″ to 0.038″. The 0.014″ guidewires are known as coronary guidewires, because they are routinely used for coronary interventions, given the high effectiveness to cross distal coronary vessels and the low risk of wall injury. Given the low profile, the support capability is low. It can be increased by using guiding catheters or long sheaths.

  Larger guidewires are available for a different kind of procedure. Commercially available sizes are 0.018″, 0.021″, 0.025″, 0.032″, 0.035″, and 0.038″.

The largest part of diagnostic catheters fits with guidewires up to 0.035″ or 0.038″, so these sizes are the most frequently used for diagnostic purposes.

Balloon catheters and pre-mounted stents fit with 0.014″–0.018″ or 0.035″, so these sizes are the most used for interventional purposes.

- ***Length***: The standard length of guidewire is 150–180 cm. Longer guidewires are available to allow the change of catheters leaving the guidewire in place or for interventional purposes. Exchange guidewire length ranges between 260 and 300 cm.
- ***Stiffness***: There is a large range of stiffness for a guidewire. Floppier guidewires warrant a higher trackability, in particular, in stenotic or tortuous vessels, but are lacking in support. Once it crossed the lesion, a low-profile catheter can be advanced on the guidewire to change the guidewire and continue the procedure. Stiffer guidewires warrant a higher support for larger catheters and interventional purposes. The scale of guidewire stiffness is Rosen wire, Amplatz Stiff, Amplatz ExtraStiff, Amplatz SuperStiff, Amplatz UltraStiff, Backup Meier, and Lunderquist ExtraStiff.
- ***Tips***: There are different guidewire tip shapes: straight and "j"- and "u"-shaped (or "angled") tip. Standard coronary guidewires have a j-shaped tip; standard 0.035″ guidewires have a u-shaped tip. The aim is to combine smoothness and atraumatic. Some guidewires may have special tips to be accommodated in specific sites or anatomies (i.e., spiral tip in Safari® guidewire to be placed in the left ventricle).
- ***Coating***: Standard and stiff guidewires are usually uncoated. Sometimes, in the case of difficult vascular accesses and challenging anatomies (stenosis, kinking, tortuous vessels), a hydrophilic coating may improve the smoothness. The coating may involve the entire guidewire or just the first centimeters (5–15 cm) behind the tip. Hydrophilic coating needs periodical wetting to maintain their properties.

7.5.4 Interventional Procedures

About 70–75% of the procedures performed in ACHD cath lab have an interventional purpose [2, 10]. Despite the detailed description of the single procedures, which is beyond the scope of this chapter, in this paragraph, we will describe the most frequently used tools and devices used in cath lab for CHD treatment.

Interventional procedures are performed by using special catheters (balloon catheters, cutting balloon catheters, Rashkind catheters) and devices (stents, plugs, coils, septal occluders, duct occluders, heart valves sutured in stents, etc.). In addition, these tools require adequate or dedicated guidewires and delivery sheaths to be used. These tools may be in stock in the cath lab or should be specially ordered for the procedure. For these reasons, a material check of available tools and devices should be performed before starting the procedure.

- ***Balloon catheters***: There is a large availability of balloon catheters on the market. They can be divided by size, compliance, guiding system, length, etc.

  The smallest balloons are coronary balloons. The diameter ranges between 1.5 and 5 mm. The length ranges between 8 and 20–30 mm. To save a low profile, they will need a 0.014″ guidewire and are delivered with a monorail system (see below). Usually, a guiding catheter is needed (5–6 Fr) or a 3–4 Fr sheath. Small balloons (for valvuloplasty or angioplasty of small vessels) range between 4 and 10–12 mm; the balloon length is 20–40 mm. Currently, these balloons are over-the-wire catheters. Low-profile balloon (requiring a 3–5 Fr sheath) requires a 0.014″–0.018″ guidewire; balloons tracked on a 0.035″ guidewire usually needs larger sheaths (5–7 Fr). The shaft length ranges between 60 and 135 cm. Large vascular balloon ranges between 10–12 and 20–35 mm in diameter. The length of the balloon is between 20 and 60 mm; the shaft length ranges between 80 and 135 cm. These balloons need a 0.035″ exchange guidewire, and the shaft diameter depends on the balloon width, usually starting from 7 Fr for 10–12 mm in diameter to 12–16 Fr for larger balloon diameters.

  Regardless of the size, the balloon can be compliant and non-compliant. Compliant balloons may change the effective diameter along with the balloon length, they are less traumatic, and they are mostly used in applications that require the balloon to fully conform to or occlude the anatomy. Semi-compliant balloons are commonly made of Pebax or higher durometer polyurethanes. These balloons are used in applications that require medium to high pressures but need a more compliant than a non-compliant balloon and more flexibility to ease delivery. Non-compliant (high-pressure) balloons are commonly made of polyester or nylon. These balloons are used for applications in which the balloon needs to expand to a specific diameter and exert high pressure to open a blockage or to dilate the vessel.

  The balloons can be over the wire and monorail. Over-the-wire system means that the guidewire is inside the balloon from the tip to the distal hole. It warrants a higher control of the balloon and of the guidewires; on the other hand, an exchange guidewire is needed and the shaft size is usually larger. The monorail system consists of a balloon catheter in which the guidewire passes through the balloon itself, exits the catheter proximal to the balloon, and runs alongside its small shaft. It offers distinct advantages like low profile, easy contrast injections, and rapid balloon exchanges. This system is almost exclusively used for coronary treatment because it needs a guiding catheter or a long sheath because of the low support and low stability of the coronary guidewire–monorail system.

  Finally, the balloons can be bare or drug coated. The balloons can be pre-mounted three or more blades (cutting balloon), to improve the compliance of the most resistant stenoses or a stent (pre-mounted or manually crimped). The balloons have a cylindrical shape with two shoulders (proximal and distal) and the transition between the shoulders, and the effective balloon area is usually identified by two radio opaque markers on the shaft. However, sometimes the balloons may have special shapes (for example, the hourglass shape of Toray Balloon® for mitral valvuloplasty).

The balloons can be inflated manually with a Luer lock syringe or with an indeflator. Manual inflation can be used with large balloons to reach low pressures (2–4 atm); the indeflator is indicated for small balloons and to achieve a pre-fixed and controlled pressure. The balloons have a nominal pressure of inflation and a rated pressure. Nominal pressure usually corresponds to the nominal diameter of the balloon, and higher pressures lead to a balloon overexpansion.

All these information (minimal catheter/sheath diameter, guidewire accepted, nominal and rated pressure, diameters, length, etc.) can be easily found on the balloon package.

- ***Stent***: Vascular stents are frequently used in cath lab. The stent can be classified by its diameter (small, medium, large), length, cell design, and coverage.

  Coronary stents are usually pre-mounted on coronary balloons. The diameter range between 2.5 and 5 mm and the length between 8 and 30–40 mm. These stents fit in a 4 Fr sheath or in a 5–6 Fr guiding catheter, so is also indicated in neonates for rescue procedures.

  Small peripheral stents are usually pre-mounted on 4–10 mm balloon catheters accepting 0.014–0.018″ or 0.035″ guidewires. The stent length ranges between 12 and 30–36 mm:

  Large (12–18 mm), XL (15–22 mm), and XXL (18–32 mm). These stents may be designed with closed, semi-open, and open cells. The (crimped) length can range between 15–22 and 60 mm. Given the large diameter range, the expanded stent length will be related to the expanded diameter of the stent. Usually, these stents have to be manually crimped on a 0.035″ guidewire catheter balloon according to target diameter and stent length. On the other hand, some stents are available in a pre-mounted format. All the stents should be delivered by using sheath or long sheath (Mullins sheaths or similar) to cross the lesion to be stented covered by the sheath. The sheath size depends on the size of the balloon–stent system. It is indicated on the package of pre-mounted stent, while it should be tested with dedicated or custom-made covers after manual crimping. Usually, L-sized stents need 8 to 11 Fr Mullins sheath, XL 9–14 Fr Mullins sheath, and XXL 11–18 Fr Mullins sheath.

  Stents can be bare or covered. A covered stent allows to cover potential or already-evident vascular injuries like aneurisms, dissections, aberrant vessels, vessel/prosthesis rupture, etc. On the other hand, a covered stent may cover side branches, so its use should be carefully evaluated. In addition, they need a Mullins sheath 1–2 Fr larger than the corresponding bare stent.
- ***Devices***: Devices are often used to treat CHD without surgery. Atrial septal defects, ventricular septal defect, and patent ductus arteriosus can be closed with a dedicated device with a high effectiveness rate and low complication rate. They are deployed with a dedicated kit, including delivery sheath, delivery cable, and device. Atrial septal occluder, ventricular septal occluder, and duct occluder devices are available with different design and delivery systems. Feasibility and device type depend on the underlying anatomy; the devices range from very-small-profile devices (i.e., Piccolo® device for PDA closure in premature babies) to bulky devices.

In addition, specific devices (plug, flow, reducer, coils, etc.) are available for different lesions, like vessel embolization, aneurism closure/embolization, etc. These devices can be deployed through diagnostic catheters, guiding catheters, or Mullins sheaths.

- ***Valved stents/valvular devices***: Recently, a device for treatment of all the heart valves is available. In CHD, the most common procedure is the percutaneous pulmonary valve replacement. However, percutaneous tricuspid or mitral valve replacement or treatment and percutaneous aortic valve replacement are emerging procedures in CHD cath lab, too. Frequently, these tools are fully equipped, containing dedicated guidewires and delivery system, and/or pre-mounted. In other cases, common Mullins sheaths and extrastiff guidewires can be used.

7.5.5 Nurses During the Procedure

Once the sterile trolley has been prepared, scrub nurse can collaborate with the cardiac interventionist during the procedure. Nurse should:

- Ensure all catheters and materials are flushed with heparinized saline to prevent colts and air embolism.
- Disinfect the skin in the procedure area.
- Cover the patient with sterile technique.
- Prepare the contrast medium injection line.
- Help the cardiac interventionist in the administration of local anesthetic and prepare materials when he finds the artery.
- Pass the equipment when the procedure starts.
- Maintain a sterile field to prevent cross infections.
- Dispose of sharps and blood-contaminated equipment safely.
- Monitor patient's vitals.

The circulating nurse, during the procedure, should continue to fill in the procedure checklist: identify all the team members and confirm again the patient's identity and kind of procedure. The nurse supplying the scrub staff with sterile equipment and monitoring patient vital signs and administering fluid and medications if necessary should provide emergency intervention if patients experience bleeding or serious side effects from the procedure. The nurse must have continuous feedback with the patient, monitoring verbal and non-verbal signs of pain.

7.5.6 Post-procedural Monitoring

In the post-procedure, the scrub nurse removes needles and scalpel to avoid the risk of accidental cutting. Then, the nurse removes the introducer and starts a local compression of the artery or vein to prevent bleeding and to achieve an adequate hemostasis.

- When the introducer is placed in the radial artery (Fig. 7.5), the scrub nurse applies a little gauze roll and an elastic compressive bandage and checks pulse. The nurse communicates to the patient to stay in bed for a few hours, don't drink for an hour, and don't move the wrist for 6 h.
- When the introducer is placed in the femoral artery (Fig. 7.6), the scrub nurse should control the ACT by a blood sample from the introducer. Based on the result, the cardiac interventionist can decide to administer protamine, and then after 30 min, the nurse has to repeat the ACT: if the result is on the range, the nurse has to make a 15-min vigorous manual compression to obtain hemostasis and then apply a big gauze roll and an elastic compressive bandage. After checking the pulse, the nurse communicates to the patient to stay in bed for 12 h without moving the leg and don't drink any beverage for an hour.
- When the introducer was placed in the femoral vein, the scrub nurse should make manual compression for about 10 min and then apply a big gauze roll and an elastic compressive bandage. The nurse communicates to the patient to stay in bed for 6 h, don't move the leg, and don't drink any beverage for an hour.

In the meantime, the circulating nurse continues to monitor patient's vital parameters; records an electrocardiogram (when indicated); monitors symptoms like chest pain, pain around the vascular access, or any other discomfort; and reports the monitoring the checklist.

Finally, the nurses help the patient to pass in the bed and give him or her further nursing instructions that include details on medications, activities, and dietary restrictions following the procedure. Patient is re-admitted in the ward for the continuation of treatment or in intensive care, following physician's instructions.

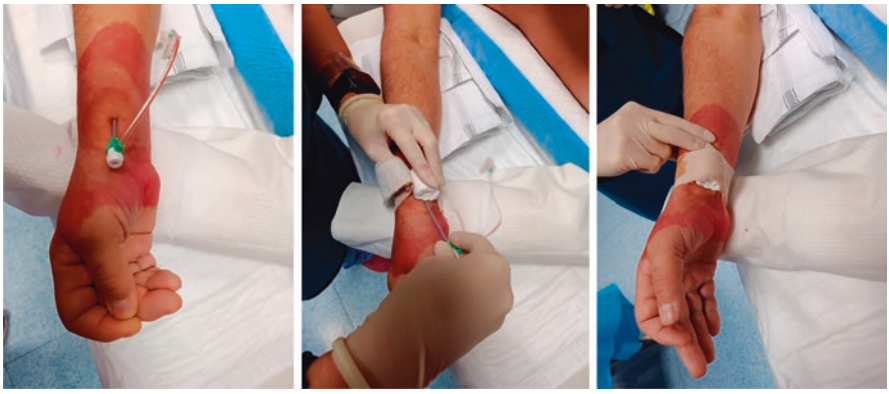

Fig. 7.5 Sheath removal from radial artery and application of the compressive bending

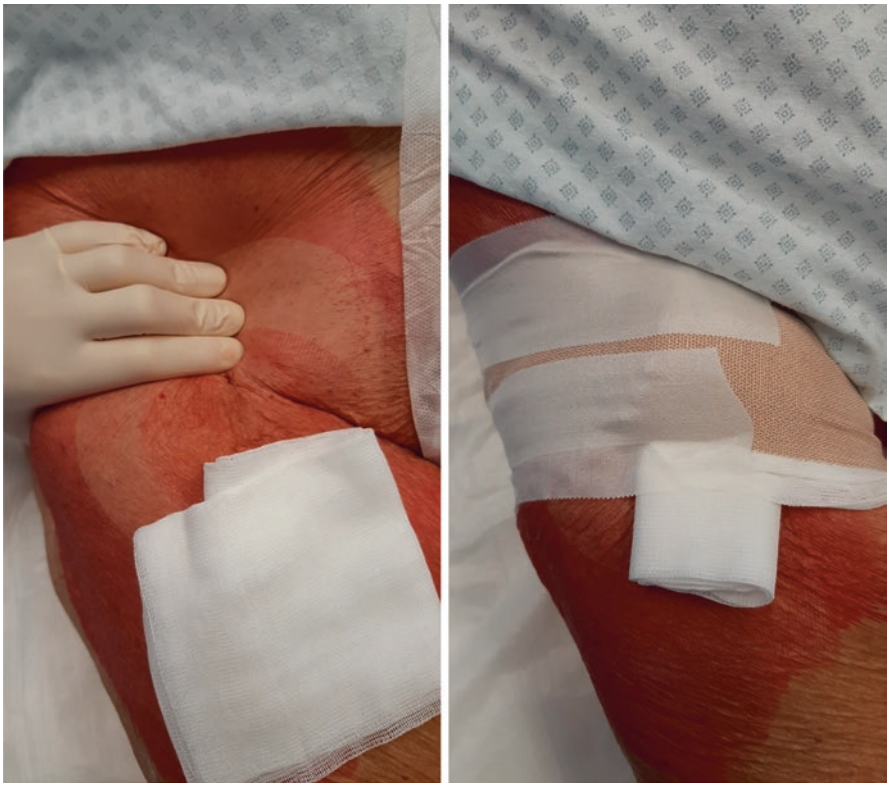

Fig. 7.6 Manual compression of the femoral artery and compressive bandage

7.6 Tools Management

Catheters and device stock should be constantly monitored to have an adequate amount of material. Guidewires, sheaths, and catheters should be fully available in the cath lab, as well as any other lifesaving tool or devices helpful in emergency or rescue procedures. The list of high priority material should be agreed between nurses and physician and constantly monitored. Devices for elective procedures may be ordered and delivered for a specific procedure. Despite a full stocking of all the tools (also potentially) eligible seems to be easier and more appropriate, the large gamma of device sizes and the availability of different types of devices for the same lesion require large stocking area and are at risk of product expiration before use. Thus, this choice can be uneconomic and be too much scattering, especially in centers managing less than 500 procedures per year. However, the delivery *on call* of "special material" should take into account the technical time for packaging shipment that sometimes can require 2 or 3 working days for the delivery. Therefore, an appropriate planning of these procedure should be timely managed.

The digitalization of warehouse and of the tools in stock helps to manage the tools monitoring.

At the end of the procedure, all the material and devices used needs to be registered in the informatic system scanning the barcode. In this way, the material is linked to the patient card and updates the amount of the stock for a given product. When the number of pieces available falls below the pre-fixed minimum amount, the dedicated operator provides to request of supply. The gamma of product and the minimum stock amount should be periodically revised according to the current needs and to the availability of new or more adequate tools.

Finally, the expiration dates of the goods in stock should be checked periodically.

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Nursing Care in the Critical Setting

8

Emily Dong, Tara Dade, and Anitra Romfh

8.1 Pediatric vs. Adult ICU

Due to the surgical and medical advancements in the field of congenital heart disease, there are now more adults living with congenital heart disease than children, and the complexity of the underlying congenital heart defect (CHD) of these adult patients is increasing [1]. Furthermore, the majority of these advancements are palliative, leaving a large majority of moderate-to-complex patients who will require further interventions in adulthood. As a sequela of their underlying disease, many experience arrhythmias, heart failure, pulmonary hypertension, endocarditis, and thrombotic events [1]. Care for these complications is often provided in an ICU setting. To add further to the complexity, there are often comorbidities that have to be taken into consideration [2]. Given these multifactorial issues, various aspects of the patient's profile must be considered in this heterogenous group when deciding to admit to a pediatric or adult cardiac intensive care unit (CICU).

ACHD patients in the ICU setting require a multidisciplinary team of experts that understand their complex anatomy, physiology, comorbidities, and psychosocial needs [1]. They require access to specialists in congenital cardiac surgery, catheterization, electrophysiology, and advanced heart failure therapies and consultation from other adult subspecialities for comorbidities such as gastroenterology, pulmonology, nephrology, and palliative care, to name a few. Other disciplines such as nursing, advanced practice providers, technicians, physical therapy, occupational therapy, and social work are also vital in the care of this population in an ICU setting. There is some data that shows improved outcomes for patients undergoing

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Table 8.1 Differences in caring for adults in a pediatric vs. adult setting [2] (Reprinted/adapted by permission from Springer Nature: Springer, Intensive Care of the Adult with Congenital Heart Disease by Eduardo da Cruz, Duncan Macrae, Gary Webb. © 2019)

Pediatric CICU	Adult CICU
Familiarity of CHD anatomy, physiology, diagnostic tools, and palliative procedures in terms of surgical, interventional, and medical interventions	Familiarity of caring for adults in the ICU setting including management of arrhythmias (especially atrial fibrillation), thromboembolic events, myocardial infarction, ICU delirium, glycemic control, and sedation needs (especially while intubated)
Availability of consult services that are familiar with CHD patients	Availability of internal medicine specialists to manage comorbidities
Familiarity of right-sided heart disease and heart failure	Familiarity of cardiac rehab and resources for early mobilization
Expertise in managing cyanotic heart disease	Environment designed for adult patients including support services and placement options such as acute rehab or skilled nursing facilities
Long-term relationships/trust with patients and families	Heart failure–focused nutrition counseling with widely available low-sodium diet options
Familiarity and understanding of childhood experiences of the CHD patient that can lead to psychosocial issues such as PTSD	Equipped to handle the pregnant patient requiring ICU level care
Familiarity of chromosomal abnormalities associated with certain CHD lesions and their implications on other organ systems	

repeat cardiac surgery in a pediatric setting vs. an adult hospital; however, the data is limited, emphasizing that a one-size-fits-all setting is nearly impossible for the heterogenous group [2]. Instead, one must assess the strengths and weaknesses of each location in determining the ideal setting for the patient. Where there is an educational gap, it is essential that training is ongoing to overcome the deficits. Table 8.1 highlights the important differences in strengths in the pediatric vs. adult setting:

Key Points

- ICU care of adults with congenital heart disease is complex and heterogeneous. Decision of ideal ICU location should take into consideration the patient's anatomy, complexity, comorbidities, and psychosocial factors.
- Care must include providers with specialized training in adult congenital heart disease and ideally one that is ACHA accredited.
- Ongoing training in the care of an ACHD patient must be provided for nursing and advanced practice providers in the ICU setting.

8.2 Hemodynamic Monitoring

Physical exam remains the most important tool in assessing patient hemodynamics. However, as technology has advanced, there are now supportive monitoring devices to complement the physical exam to determine patient stability and anticipate the

trajectory of the patient's clinical course. Many factors affect preload, afterload, cardiac output/index, oxygen delivery, and contractility of the ventricle [2]. A combination of non-invasive and invasive monitoring allows for a complete assessment of key clinical data and allows one to determine the following:

- Cardiac output (CO): The amount of blood pumped by the heart per minute. Calculated by multiplying the heart rate (HR) and stroke volume (SV). It is expressed in liters (L) per minute. Different means of calculating CO include thermodilution and Fick's principle, which can be determined with invasive cardiac monitoring.
- Cardiac index (CI): The CO indexed to the patient's size. Calculated by dividing the CO by the patient's body surface area (BSA).
- Tissue oxygen delivery (DO2): Cardiac output × 1.34 × hbg concentration × oxygen saturation.

8.2.1 Non-invasive Monitoring

Physical exam (PE): There are various key aspects of the PE that are helpful in the ICU when assessing the ACHD patient. On the skin exam, one should note any prior surgical scars (sternotomy, thoracotomy). On the cardiac exam, one should assess for changes in the location and quality of a murmur and the presence of a new murmur or extra heart sound. The regular first and second heart sounds are denoted S1 and S2. An extra heart sound just before S1 is denoted as an S4 (often indicative of a stiff ventricle), and an extra heart sound after S2 is denoted as an S3 (often heard in acute systolic heart failure). The quality of the heart sounds is also important (i.e., distant heart sounds as in a patient with a pericardial effusion or a loud pulmonary component of the second heart sound as is present in a patient with pulmonary hypertension). For the vascular assessment, one should assess the pulses and their quality (for example, bounding pulses with sepsis or acute aortic regurgitation or faint as in the patient in cardiogenic shock).

On the pulmonary exam, it is equally important to assess for the presence of rales or decreased breath sounds, which may indicate the presence of pulmonary edema or a pleural effusion, respectively. The inspiratory and expiratory length can be assessed, with prolonged expiration seen with air trapping as in chronic obstructive pulmonary disease (COPD). On the extremity exam, peripheral edema may be a result of heart failure, renal failure, or low oncotic pressure resulting in the so-called third-spacing. In recumbent patients, edema is often located at the sacrum due to the effect of gravity. The PE is also vital in determining perfusion. Signs of poor perfusion include cool extremities, decreased peripheral pulses, decreased urine output, and altered mental status.

ECG: All ACHD patients admitted to the ICU should be placed on continuous telemetry. This allows for both real-time and retrospective analysis of the patient's rate and rhythm. In addition, all patients should have a recent baseline ECG available for review. For surgical patients, a postoperative ECG should be done upon arrival to the ICU and as needed for any concerning changes on telemetry [2]. Most

postoperative patients will also have epicardial pacing wires placed in the operative room. An atrial ECG can be utilized to magnify the p-waves to determine atrial activity. The absence of a magnified p-wave could indicate a junctional rhythm or complete heart block (CHB). Aortic valve surgery, mitral valve surgery, patients with L-transposition of the great arteries (L-TGA; with or without surgery), and reoperation for patients with a history of atrioventricular canal are at an increased risk for CHB. Patients with a prior history of atrial arrhythmias and tricuspid valve surgery (especially Ebstein's anomaly) are at an increased risk of atrial arrhythmias. Patients with a prior history of ventricular arrhythmias or frequent premature ventricular contractions (PVCs), prior incision into the ventricle or large right ventricular outflow patches (tetralogy of Fallot [TOF]), and ventricular enlargement and/or dysfunction are at increased risk of ventricular arrhythmias [3]. All arrhythmias have the potential to decrease cardiac output.

Oxygen saturation probe: An oxygen saturation probe is any easy way to estimate oxygen saturation non-invasively, though it has its limitations. The reliability of the O2 probe is decreased when oxygen saturations are less than 80%; thus, it is not accurate for the chronically cyanotic ACHD patient [2]. It is also important to know the expected O2 saturation range for your population. A Fontan patient will not have an O2 saturation of 100% due to the coronary venous return into the systemic ventricle. Patients with Eisenmenger syndrome or other complete mixing lesion may have saturations around 85% or less. If the Eisenmenger syndrome is due to a patent ductus arteriosus (PDA), then it is important to place the O2 saturation probe on the toes, as they can have higher oxygen saturations in the fingers than toes due to the fact that the PDA is often distal to the subclavian arteries. ACHD patients can have adult comorbidities such as sleep apnea or lung disease that can also decrease their O2 saturation. These should be considered if other monitoring parameters indicate a normal CO.

Oscillometry: Non-invasive measurement of blood pressure by utilizing a blood pressure cuff. This requires pulsatile flow and correct sizing of the cuff. Too small can falsely elevate the blood pressure reading, and too large can falsely lower the blood pressure reading. This modality of blood pressure monitoring is inadequate in patients with low cardiac output, arrhythmias, vasoconstriction, and significant edema [3]. Given this, critically ill and postoperative patients require invasive blood pressure monitoring with an arterial line in addition to oscillometry. Placement of the blood pressure cuff requires knowledge of the prior surgical history. For example, in patients with a history of tetralogy of Fallot may have had an initial classic Blalock–Taussig shunt as part of the initial palliation (involving sacrificing the subclavian artery to provide pulmonary blood flow), resulting in an inaccurate BP reading on the same side as the thoracotomy scar.

Near-infrared spectroscopy (NIRS): This determines regional tissue oxygen and blood flow of the brain (cerebral NIRS [ScO2]) and organ tissues (somatic NIRS [StO2]). In intraoperative and postoperative patients, the kidney is the main organ monitored with the somatic NIRS. Normal values vary greatly depending on the device utilized and the patient population; however, the generally accepted range of ScO2 is 60–80%. Normal StO2 values vary even more than cerebral NIRS due to

differences in patient age, weight, site of monitoring, and distance of monitor to the organ being measured. Due to this, the difference in ScO2 and StO2 (StO2–ScO2) can be utilized in determining trends in CO. The difference in healthy patients is ~10–20% [4]. As CO is compromised, the percent difference decreases to preserve cerebral perfusion. Given the high variability in normal NIRS values, trends in the levels at each site as well and the difference between the two sites are an important clinical tool to use in conjunction with the other monitoring parameters discussed in this section.

8.2.2 Invasive Monitoring

It is important to make a daily assessment of the need for invasive catheters as they are a risk for infection and thromboembolism. Patient anatomy needs to be taken into consideration when placing and using of a line. Special considerations should be taken for Glenn and Fontan patients as a thromboembolism could be especially catastrophic. Additionally, many patients have chronic venous obstruction from prior invasive lines and multiple catheterizations so obtaining access can be challenging [5]. Air vigilance is vital for patients with intracardiac shunts as they are at risk for thromboembolism into the system circulation. Invasive monitoring allows for important diagnostic laboratory monitoring, and some examples are:

Blood gas analysis: Both arterial blood gas (ABG) and venous blood gas (VBG) analysis can be performed using blood samples from a peripheral draw or the central venous line (VBG) and arterial line (ABG). The presence of a base deficit could indicate lactic acidosis and poor CO [6]. In a postoperative patient, it could indicate low cardiac output syndrome (LCOS) from myocardial dysfunction or the need for volume resuscitation because the patient went into surgery mildly hypovolemic (especially if the patient is operated on later in the day and has had nothing by mouth after midnight the night before).

Mixed venous saturation (MVO2): Oxygen saturation from direct sampling of the blood in the pulmonary arteries using a pulmonary artery catheter. This reflects true mixing of the venous blood from the entire body (as opposed to the central venous oxygen saturation, which is preferential to venous blood from the head and upper extremities as it is usually drawn from the superior vena cava). This value indicates whether the CO and tissue delivery are enough to meet the metabolic demands of the body.

Central venous oxygen saturation: Sampling blood from the central venous line can be a surrogate to determine the MVO2 and thus tissue consumption (with the limitations discussed previously).

Lactate: Lactate levels increase when there is impaired tissue oxygenation, which can occur in a low cardiac output state. Postoperative and critically ill patients are at risk for hyperglycemia, which, in turn, can increase lactate levels due to ongoing glycolysis following the blood draw [7]. This creates a mixed picture for assessing the cause of lactic acidosis and underscores the importance of glycemic control in this patient population.

8.2.2.1 Types of Invasive Monitors

Arterial line: Invasive blood pressure monitoring that gives systolic, diastolic, and mean arterial pressure values. Arterial blood gases can be drawn from this catheter.

Central venous catheter (CVC): Often accessed via the internal jugular vein, this CVC provides central venous pressure monitoring which indicates volume status and assessment of preload (when used in conjunction with other clinical data) [6]. Also this provides access for vasopressor and inotropic support, large volume, rapid fluid resuscitation, and laboratory information such as the VBG and central venous oxygen saturation. The waveform provides additional important information, such as cannon a waves that can indicate complete heart block [2].

Right atrial line: Surgically placed directly in the right atrium for pressure monitoring. This line is often used in surgeries on right-sided structures, especially tricuspid valve repairs. It is also used in patients with single ventricles to decrease the risk of thrombus in the Glenn/Fontan pathway.

Left atrial line: Similar to the right atrial line, this surgically placed line provides direct left atrial pressure monitoring. It is often used in surgeries on left-sided structures—aortic valve, mitral valve, or concern for coronary artery perfusion since diastolic elevation and thus increased left atrial pressures are often the first sign of ischemia.

Pulmonary arterial line: This is a catheter placed directly into the pulmonary artery. Sampling from this catheter provides an MVO2 value. A true estimate of CO can be assessed using thermodilution or Fick (if an intracardiac shunt or significant valvular regurgitation is present, only the Fick method should be used).

Key Points

- Both invasive and non-invasive monitoring is needed in the critically ill adult with congenital heart disease; however, the physical exam remains the most important indicator of the patient's clinical status.
- No one single data point will provide the full clinical picture. All data should be reviewed and interpreted by the clinician in order to have a proper assessment of the patient's hemodynamics.
- Patient anatomy needs to be taken into consideration when determining what type of monitor is appropriate. Chronic venous obstruction is often an issue in this patient population.
- The need for invasive monitoring should be assessed daily so as to decrease the risk of infection. Air vigilance is vital in patients with intracardiac shunts.

8.3 Fluid Resuscitation

As with many other aspects of care for the adult with congenital heart disease, fluid resuscitation and stabilizing hemodynamics are complex and personalized. Understanding the anatomy, physiology, and prior cardiac interventions of the patient is key in managing their fluid balance. The goal of fluid resuscitation is to maintain systemic perfusion without causing significant peripheral or pulmonary

edema. Data from a prior cardiac catheterization is helpful in managing fluid resuscitation and should be taken in conjunction with bedside data.

Hypotension and volume depletion are common in postoperative patients for a variety of reasons:

- Intravascular volume depletion from being NPO prior to surgery
- Bleeding
- Low cardiac output syndrome (LCOS)

During surgery, there is a high risk of bleeding as most ACHD patients have had prior surgery. Adhesion formation alters the vascular anatomy. Patients with chronic cyanosis are at particular risk of excessive bleeding after surgery due to rheologic abnormalities. Chest tube management is important in assessing the amount of bleeding in the postoperative period. The bedside nurse should regularly assess the output including patency of the chest tube, amount of drainage, and quality of the drainage. Chest tube drainage in a child is based on norms for size (mL/kg/h); however, in an adult, the norms are based on total volume, not weight based. The acceptable amount is ultimately determined by the surgeon, but the typical amount is no more than 200 mL/h in the first hour or 1000 mL in the first 24 h [2]. Blood pressure management is also important in decreasing bleeding, especially in those with suture lines on the aorta. In these patients afterload reduction is important in managing bleeding. CBC and coagulation factors should be checked regularly and blood products given as clinically indicated.

Low cardiac output syndrome (LCOS) typically occurs 9–12 h postoperatively and can be caused by a variety of factors including ischemia from aortic crossclamp and circulatory arrest, third spacing due to the inflammatory process caused by cardiopulmonary bypass, and cardiac tamponade. Signs of early shock include tachycardia, cool extremities, decreased peripheral pulses, decreased urine output, decreased mentation, and prolonged capillary refill. Late signs of shock include bradycardia, hypotension, decreased oxygen saturations, and metabolic acidosis. In this setting, fluid resuscitation is indicated, knowing that diuresis will be needed once hemodynamically stable. Of note, third spacing is not as apparent in adults as there is more surface area to "hide" fluid so daily weights are essential to assessing volume status and volume overload as a result of resuscitation in an adult. There is a wide variation in practice in terms of fluid resuscitation with crystalloids vs. colloids and no ideal type of fluid exists. The benefit of colloids is that less volume is typically needed (1:3 ratio), and the benefit of crystalloids is that they are inexpensive and widely available [5].

8.3.1 Special Considerations

Pre-existing ventricular dysfunction: Patients with right ventricular enlargement or ventricular dysfunction have a fine balance between maintaining preload and worsening failure and hepatic congestion from volume overload. Patients with chronic pulmonary outflow obstruction can develop a diastolic dysfunction with restrictive

physiology, and large volume resuscitation can cause heart failure as well [8]. After each intervention, a reassessment of patient hemodynamics should be made and adjust the treatment as needed. Similarly, patients with left ventricular dysfunction often require inotropic support with volume resuscitation in order to maintain CO and prevent pulmonary edema.

Pulmonary hypertension: Pulmonary hypertension can develop as a consequence of many different forms of congenital heart disease (both repaired and unrepaired). This can occur in ~15–30% of patients with ACHD [9]. Both PVR and SVR must be managed and balanced when replacing fluids. Pulmonary blood flow is improved with a lower PVR, whereas systemic blood flow is augmented with a lower SVR. At a minimum, patients should be maintained on their outpatient pulmonary vasodilator therapies (abrupt discontinuation can cause rebound pulmonary hypertension and a PH crisis), and they may require escalation of these therapies in conjunction with vasopressors for systemic perfusion [10].

Single ventricle: Patients with Fontan circulation are highly dependent on preload and thus sensitive to changes in preload. Their blood flow is in series and dependent on a single pumping chamber. This population requires higher venous pressures to maintain forward flow, so hypovolemia and/or decreased vascular tone can lead to significant hypotension [11]. Special consideration should be taken when using vasoconstrictors because increasing afterload could decrease forward flow through the circuit. Hypervolemia is also an issue because it can decrease the pressure gradient between the systemic and venous systems and thus compromise CO [5]. Finding the balance of adequate preload, venous congestion, and CO is very difficult and requires expertise on the physiology and careful watch on subtle changes in clinical status.

Key Points

- Fluid resuscitation of the critically ill patient with ACHD is common, complex, and highly individualized. Common causes are dehydration, bleeding, LCOS, and acute heart failure.
- Understanding the patient's anatomy, physiology, and baseline hemodynamics is critical in managing fluid resuscitation and volume status. Every intervention has the potential to significantly alter hemodynamics so vigilance should be taken into reassessment after intervention. Volume resuscitation should be judicious.
- This patient population often requires inotropic, pulmonary vasodilator and/or systemic vasodilator therapies during fluid resuscitation depending on their underlying congenital heart disease.
- In the postoperative patient, chest tube management is key in assessment of clinically significant bleeding and hypovolemia.

8.4 Ventilation and Sedation

ACHD patients are at risk for reduced pulmonary function owing to various developmental factors such as airway disease, abnormal pulmonary arterial vasculature, pulmonary vein stenosis, and pulmonary parenchymal hypoplasia. They are also

prone to acquired abnormalities as a result of prior surgeries. Factors such as chest wall restriction from prior thoracotomy scars, diaphragmatic weakness from phrenic nerve damage, or barotrauma from prior prolonged mechanical ventilation contribute negatively to lung function. Other factors such as obesity, scoliosis, obstructive sleep apnea, asthma, left heart failure, and pulmonary hypertension can also contribute to impaired lung performance. Still other factors may include direct toxins to the lungs from the environment or more commonly from cigarette smoking or vaping. As such, looking for these historical clues in the past medical history can help the bedside practitioner understand the expected duration of mechanical ventilation or respiratory support (either post-surgery or during a medical admission).

Measuring lung function through pulmonary function tests (PFTs) is important whether it is in the form of spirometry or lung volumes. It has been shown that inspiratory muscle dysfunction and restrictive lung disease are predictive of exercise intolerance in ACHD patients [12, 13]. Further, in adult patients with heart failure, inspiratory muscle dysfunction can be impaired in both systolic and diastolic heart failure [14]. Thus, reviewing the preoperative assessment of lung function by either formal PFTs or routine cardiopulmonary exercise testing is extremely useful for understanding the ventilation physiology of the patient in the ICU.

In addition to the factors that affect ventilation, adults with CHD often have factors that influence their oxygenation. These can be grouped into intracardiac vs. extracardiac ventilation–perfusion (V/Q) mismatch. For example, the baseline saturations for an Eisenmenger patient may be 70–80% on room air due to bidirectional shunting, representing an example of an intracardiac V/Q mismatch. These saturations will not improve with exogenous oxygen administration. Other patients with chronic lung disease may have saturations 88–93%, representing desaturation of the extracardiac type. The saturation in these patients will improve with oxygen administration, since the impairment is at the lung tissue level where oxygen is delivered. Some patients may have secondary erythrocytosis due to chronic hypoxemia, a clue that their baseline saturations are not in the normal range. This is important in the critically ill patient when setting targets for extubation from mechanical ventilation or setting acceptable alarm targets for bedside saturation monitoring.

Patients in respiratory distress may require positive pressure ventilation (PPV). This may be achieved through non-invasive means such as continuous positive airway pressure (CPAP) or via endotracheal intubation. The means of respiratory support has effects on cardiac output, pulmonary vascular resistance, and preload. These effects can differ with respect to systemic (pumping to the body) or pulmonary (pumping to the lungs) ventricles. For example, in patients with LV dysfunction, the increased intrathoracic pressure from PPV decreases the transmural pressure gradient and LV wall stress, which, in turn, decreases afterload to the ventricle. In contrast, PPV has the opposite effect on the RV due to the increased intrathoracic pressure and decreased preload. Thus, a patient with a Fontan circulation or pulmonary hypertension will likely have worse hemodynamics with PPV, and the goals of management may be to prioritize weaning of PPV in these situations.

Sedation and pain management are important in ICU patients. For adults being cared for in a pediatric ICU setting, care must be taken not to utilize weight-based

dosing of sedation/analgesia in continuous infusions as they can result in overdose and delayed clearance. Muscle paralysis is rarely used with sedation and analgesia in the adult patients unless respiratory compliance on the ventilator is a large issue. Adults can easily become disoriented with too much sedation and analgesia and ICU delirium can result. Risk factors for ICU delirium include advanced age, baseline cognitive impairment, comorbid conditions, type of surgery, and duration of sedation [15]. As such, baseline cognitive assessments should be carefully done during the preoperative exam to aid in management in the ICU. Sedation and pain management should be weaned as quickly as tolerated. Additional factors that help adult patients in establishing orientation in the ICU include providing patients with their baseline eyewear, maintaining consistent sleep–wake cycles, and allowing family or friends to accompany patients at the bedside.

Key Points

- Baseline assessment of saturations and pulmonary function in the outpatient setting are important to understanding the expected clinical dynamics behavior in the ICU setting.
- Positive pressure ventilation helps systemic ventricular dysfunction but can impair subpulmonary ventricular function.
- Sedation and pain management given in continuous infusion should NOT be weight based.
- Understanding the risk factors for delirium is as important as the non-pharmacologic measures needed to orient patients in the ICU.

8.5 Arrhythmia Management

Arrythmias in ACHD patients result from substrate from their structural heart abnormalities, hemodynamic changes, and prior cardiac surgical scars that put them at a greater risk of developing arrhythmias. Symptomatology ranges from asymptomatic and benign to poorly tolerated with hemodynamic consequences that can lead to sudden cardiac death. Arrhythmias remain an ongoing challenge to manage and are one of the most common causes of hospital admission in the ACHD patient. Regardless of their reason for admission to the ICU, nurses should be aware of the patient's arrhythmia history including notable arrhythmias on prior event recorders or electrophysiology studies.

All patients should have a 12-lead EKG and be placed on telemetry upon admission into the ICU. Telemetry is a useful tool to monitor patient vitals and identify abnormal heart rhythms in real time, which aids in determination of hemodynamic stability when arrhythmias are present. Arrhythmias after congenital heart surgery are a known complication [16]. Bedside nurses should be trained to identify common and life-threatening arrhythmias early, enabling a timely response to any required intervention. Treatment is guided by hemodynamic stability and correcting reversible causes. The defibrillator should be stored nearby, and nurses should be trained for synchronized cardioversion and defibrillation use if the patient is hemodynamically unstable.

Sudden cardiac death (SCD) remains one of the leading causes of death in the ACHD patient, and risk stratification for arrhythmia risk should be performed prior to any procedure, especially cardiac surgery. Complex congenital heart lesions including Eisenmenger syndrome, cc-TGA, left-sided outflow obstructions, and repaired tetralogy of Fallot have the highest risk for SCD [17] though many SCD victims remain unrecognized [18]. Using additional clinical tools such as QRS duration, ventricular dysfunction, history of ventricular arrhythmias, and gender can improve risk stratification of SCD [19]. Any concerning arrhythmias on telemetry should be followed up with a 12-lead EKG. Table 8.2 reviews common causes of postoperative arrhythmias. Patients will often come out of the operating room with temporary pacing wires that may be connected to an external pacemaker if necessary.

Tachycardia in the adult patient is a heart rate greater than 100 beats per minute and should prompt concern for a tachyarrhythmia. The most common tachyarrhythmia in ACHD patients with severe congenital heart disease is supraventricular tachycardia (SVT) with a 50% lifetime risk by the time they reach 65 years of age [20]. After congenital heart surgery, SVT can be a major cause of morbidity and mortality. Atrial tachycardia can be found with a fixed heart rate despite adjustments in volume, ventilation, and pressor support. If the 12-lead EKG cannot confirm suspected SVT, obtaining an atrial tracing using the temporary pacing wires can confirm the diagnosis. With hemodynamically stable SVT, overdrive pacing can be done at the bedside using temporary pacing wires with continuous EKG monitoring [21]. Careful observation should be made as atrial fibrillation can occur as a consequence of overdrive pacing which can lead to hemodynamic instability. Patients in SVT experience the greatest drop in blood pressure in the first 10–30 s and normalize by 60 s [22].

Junctional ectopic tachycardia (JET) is another arrhythmia that can occur post congenital cardiac surgery and is typically seen within the first 48 hours after surgery, more commonly in younger patients. This uncommon arrhythmia can be potentially life-threatening and is typically treated with amiodarone [16].

Atrial fibrillation is the most common arrhythmia in the general adult population with an incidence of ~1% of the US population [23] and can also occur in the adult

Table 8.2 Causes of postoperative arrhythmias

Hemodynamic changes
Fluid shifts from surgery
Pericardial effusion
Inflammation
Prior surgical scar
Ischemia
Medications
Electrolyte imbalances
Cyanosis
Vasoactive infusions and inotropes

congenital heart patient at a much younger age. Non-cardiac risk factors that increase the risk for development of atrial fibrillation include obstructive sleep apnea, diabetes, and age, in addition to cardiac risk factors of atrial scarring, anatomic structure, chronically remodeled atrium from hemodynamic stress, hypertension, and residual shunts [24].

Management of tachyarrhythmias in the **hemodynamically unstable patient** involves **immediate electrical cardioversion** as the mainstay of treatment. Patients with complete transposition of the great arteries who have undergone a Mustard or Senning procedure, Ebstein's anomaly, tetralogy of Fallot, and single-ventricle patients repaired by a Fontan are examples of patients at high risk for developing acute, atrial tachycardia. Patients who depend on an atrial kick to maintain adequate cardiac output will decompensate quickly if left in a sustained atrial tachycardia. Early return to sinus rhythm decreases the risk of thrombus formation, improves cardiac output, and prevents unwanted atrial remodeling [20].

Management of stable **chronic atrial fibrillation** is focused on anticoagulation and either rate or rhythm control. Standard management for thrombus prevention if onset of atrial flutter or atrial fibrillation is unknown or greater than 48 hours is anticoagulation for 3 weeks prior to and 4 weeks after cardioversion [24]. If the duration is unknown or greater than 48 hours and the **patient is unable to tolerate the arrhythmia for this period of time**, anticoagulation should be initiated immediately and the patient should undergo a **transesophageal echo** to exclude a thrombus in the atrium or appendage. As with other cases of cardioversion, the duration of anticoagulation should be continued for a period of at least 4 weeks afterward, to account for the risk of thrombus formation due to initial electromechanical delay post-cardioversion [24].

After this 4-week period, continuation of chronic anticoagulation is determined based on the CHA2DS2-VASc score. The CHA2DS2-VASc [25] clinical scoring model is used to stratify the risk of stroke from atrial fibrillation. Atrial fibrillation can lead to thrombus formation, typically in the left atrial appendage. Systemic embolization can cause a stroke. Thus, the following risk calculator can guide clinicians on options for thromboprophylaxis against stroke.

Congestive heart failure
Hypertension
Age (>65 years +1 point, >75 years +2 points)
Diabetes
VAsculopathy
Sex (female +1 point)
Clot/stroke/TIA/embolism

Patients with a CHA2D2-VASc score of 0 do not require any treatment, while those with a score of 1 can be treated with aspirin alone. A score of 2 is considered high risk and carries an annual stroke risk of 2.2%. These patients should be treated with anticoagulation depending on the clinical scenario [24].

Rate control for atrial fibrillation can be achieved with medications like calcium channel blockers, beta blockers, or digoxin. Rhythm control in the perioperative period is commonly achieved with amiodarone. It can be given intravenously or orally [26]. The basis for anticoagulation with chemical cardioversion using an antiarrhythmic is the same as that used for electrical cardioversion. Namely, anticoagulation should be used if the duration of arrhythmia is greater than 48 hours or unknown [25]. Baseline thyroid function tests, liver function tests, and pulmonary function tests should be performed prior to administration as chronic amiodarone use can lead to toxicity. Sotalol is another class III antiarrhythmic agent used to treat these atrial tachyarrhythmias, and like all class III agents, it can cause QTc prolongation. Upon initiation or uptitration of sotalol, serial 12-lead EKGs should be done to assess for QTc prolongation as long QT syndrome can potentially lead to torsades de pointes and sudden cardiac death [19].

Non-sustained ventricular tachycardia (NSVT) is a common tachyarrhythmia after adult congenital heart surgery. ACHD patients with prior right ventricular outflow intervention or prior ventriculotomy (often found with tetralogy of Fallot (TOF) or older ventricular septal defect repairs) may have a history of NSVT, which predisposes them to be at a higher risk of developing NSVT post-surgery [16]. In the ICU, hypokalemia is associated with ventricular tachycardia postoperatively and aggressive potassium repletion should be considered when giving higher-dose diuretics. These patients are often placed on a beta blocker postoperatively for a brief period to decrease sympathetic input that may contribute to ventricular tachycardia (VT). VT can deteriorate to ventricular fibrillation and sudden cardiac arrest. Particularly in the pediatric ICU, all bedside nurses should undergo ACLS training so immediate defibrillation and activation of ACLS protocol can be performed.

The presence of left bundle branch block (LBBB) should be noted, particularly after subaortic membrane resection and aortic valve surgery or in a cardiomyopathy [27]. This should be distinguished from a new LBBB that occurs without an obvious cause, which may represent a STEMI equivalent (especially if the ST segment and the T-wave are concordant in their displacement). It should be noted that the presence of LBBB is overall rare in patients with CHD; in contrast, the presence of RBBB is far more common.

Inotropes and vasopressors often used in the ICU can be proarrhythmic. Endocarditis increases risk of AV block if there is abscess formation around the aortic root. When arrhythmias are refractory to ICU management, a qualified electrophysiology (EP) consult is warranted for consideration of alternative drug therapies, PM, AICD, DCCV, or catheter or surgical ablation.

Key Points

- Arrhythmias are common in the ACHD patient.
- Bedside nurses should be trained to identify life-threatening arrhythmias quickly and certified in the advanced cardiac life support (ACLS) algorithm.
- Single-ventricle patients left in a sustained atrial arrhythmia can have deleterious effects leading to hemodynamic instability.

8.6 Anticoagulation

Anticoagulation is used in various settings for ACHD patients. Anticoagulation is often used in the setting of known thrombus, in guarding against the formation of thrombus and subsequent thromboembolism (e.g., arrhythmia), or in preventing thrombus formation on a mechanical valve. In contrast, antiplatelet agents are used to prevent platelet adhesion. The remainder of this section will focus on the main considerations for anticoagulation in these three settings.

Of note, many patients confuse anticoagulation and antiplatelet therapies, using the term "blood thinner" inadvertently for both. Therefore, it is important to distinguish anticoagulation (warfarin, direct oral anticoagulants, heparin) and antiplatelet therapy (aspirin, clopidogrel, prasugrel, for example). Often, the bedside provider is an important source of this education.

In the setting of known thrombus, anticoagulation should be started right away without a delay in getting to steady-state levels. Thus, intravenous heparin infusions (ideally with a bolus) and therapeutic Lovenox at a dose of 1 mg/kg are good options in the ICU that have a quick onset and can be easily stopped or reversed if needed.

Regarding risk stratification for guarding against thrombus formation in the setting of arrhythmia, the aforementioned CHAD2S-VASC score is often used. Although developed for use in non-congenital heart patients, it is often used as a baseline guide for congenital heart patients. In the ICU, there are often extenuating circumstances that increase bleeding risk (such as the postoperative setting or shock with liver dysfunction), and thus, the need for anticoagulation therapy needs to be weighed against the bleeding risk. The HAS-BLED score is often helpful to stratify the bleeding risk in adult patients. The following factors comprise the HAS-BLED score [28]:

Hypertension (uncontrolled, ≥160 mmHg systolic)
Abnormal renal (dialysis, transplant, Cr >2.26 mg/dL) or liver function (cirrhosis, bilirubin >2× normal with AST/ALT/alkaline phosphatase >3× normal)
Stoke history
Prior major **B**leeding or predisposition to bleeding
Labile INR (unstable or high INRs)
Elderly (age > 65)
Drugs/alcohol (≥8 drinks/week)/medications that predispose to bleeding such as antiplatelet medications or NSAIDs

Once the need for anticoagulation with either a known thrombus or stroke prevention in arrhythmia is determined, heparin (unfractionated) or low-molecular-weight heparin (LMWH) is often used. This is due to the need for short-acting agents that can be quickly stopped or reversed based on clinical changes in the ICU. Once stable, patients can be transitioned to warfarin, should long-term anticoagulation be needed. One can also consider direct oral anticoagulants (DOACs), but not all are readily reversible in the acute setting and do not have clinically established serum monitoring [22].

Anticoagulation is also used for surgically placed prosthetic heart valves. The type of valve and its position determine the need for anticoagulation and the rapidity of reaching the anticoagulation goal level. For example, with a mechanical mitral valve, anticoagulation may be initiated post-surgery with unfractionated heparin to allow quick attainment of a therapeutic PTT level. The quick attainment is desired due to the relatively low velocity flow across a mitral valve. The heparin is continued as a "bridge" to maintain therapeutic anticoagulation while warfarin is introduced, since it takes several half-lives of warfarin to achieve the therapeutic target. Warfarin levels are monitored using an international normalized ratio (INR) with a goal of 3. A mechanical valve in the aortic position, on the other hand, does not require a "bridge" with unfractionated heparin as the velocity of flow across an aortic valve is greater and is thus less likely to develop thrombus over the ensuing days while warfarin is being titrated to the therapeutic target of 2.5. In summary, both mechanical mitral and aortic valves require long-term anticoagulation, but only a mechanical mitral valve requires a heparin bridge [29].

In general, bioprosthetic pulmonary and aortic valves do not require anticoagulation and patients can be initiated on antiplatelet medication in the postoperative period once the chest tube drainage is serosanguinous and there are no other significant bleeding risks. In contrast, bioprosthetic valves in the mitral or tricuspid position require short-term anticoagulation with warfarin. Table 8.3 gives a pathway for anticoagulation and antiplatelet therapy management in the postoperative setting for various types of valves [24].

Table 8.3 Postoperative anticoagulation for prosthetic heart valves

Prosthetic Heart Valve Placement: Post-Surgical Anticoagulation Pathway

Inclusion Criteria:
Patients undergoing prosthetic valve placement or replacement

Exclusion Criteria:
Presence of other risk factors for thromboembolism (i.e. prior thromboembolism, chronic atrial fibrillation, LV systolic dysfunction, hypercoagulable conditions) or bleeding (i.e. bleeding diathesis, on multiple blood thinning agents)

Patients with prosthetic valves undergoing other surgical procedures

Develop individualized management plan with CVICU, CV surgery subspecialty team input

What type of prosthetic valve does the patient have?

Valve Type	Bioprosthetic Pulmonary	Bioprosthetic Aortic	Bioprosthetic Tricuspid or Mitral	Mechanical Aortic (with and without Composite)*	Mechanical Mitral
Bridge with heparin?	No	No	No	No	Yes
INR goal for Vitamin K antagonist (VKA)	NA	NA	2.5	2.5	3
VKA Duration of Therapy	NA	NA	3 months	Lifelong	Lifelong
Initiate Aspirin (75-100 mg) on..	POD 1	POD 1	After discharge (by cardiologist)	After discharge (by cardiologist)	After discharge (by cardiologist)

Anticoagulation Tips
- Initiate VKA +/– heparin bridge if indicated on POD 1 unless patient has ongoing bleeding
- Heparin bridge goals: HAL 0.3–0.5 or PTT 60-80; following the lab test with the higher result to minimize bleeding risk; discontinue heparin once INR reaches 2.0
- Per AHA/ACC guideline, target a single INR (with acceptable range+/– 0.5 INR units) If patient has more than one indication for Coumadin, use the highest INR goal
- Ok to remove temporary wires as long as INR <2, otherwise either hold heparin infusion 4 hours before and after wire removal OR cut wires rather than remove them

*** Mechanical On-X aortic heart valve**
(On-X Life Technologies Inc., Austin, Texas)

Warfarin dosing is targeted to an INR of 2.5 (range 2.0 to 3.0) for the first 3 months after surgery [3]A lower INR target of 1.5 to 2.0 (in conjunction with aspirin 81 mg daily) may be considered for long-term management, beginning 3 months after surgery.

Key Points
- The use of anticoagulation is often balanced against the bleeding risk.
- Anticoagulation is used in the setting of acute thrombus, in the setting of arrhythmia to prevent stroke, and for mechanical mitral and aortic valves.
- CHAD2S-VASC2 scoring helps determine the risk of stroke with atrial arrhythmia.
- HAS-BLED scoring is helpful to stratify the bleeding risk.

8.6.1 Endocarditis

Infective endocarditis (IE) is more common in the ACHD patient compared to the general population. A high degree of clinical suspicion must be present to make the diagnosis. The higher incidence in the ACHD population is often due to the presence of prosthetic valves, residual shunts near the site of prior patch material, bidirectional shunting of intracardiac shunts, annuloplasty rings, intracardiac leads, and a history of endocarditis. Patients may present with sustained bacteremia or fungemia, valvulitis, or peripheral emboli. Patients with "right-sided" or subpulmonary endocarditis may present with lesions on the subpulmonary valves (such as tricuspid or pulmonary valves). In both cases, peripheral emboli will often be found in the lung. Patients with "left-sided" or systemic endocarditis have lesions on the systemic valves (such as mitral and aortic), and this is most commonly due to oral flora. The diagnosis is established according to the modified Duke criteria, which include blood culture evidence of infection (three sets of blood cultures should be obtained), valvular vegetations or evidence that valvular vegetations have embolized, or evidence of an intracardiac abscess. The etiology can be broken into native valve IE, prosthetic valve IE, and IE from injection drug use.

Diagnosis of valvular lesions can be made by transthoracic echo, but transesophageal echo may be required if the transthoracic echo is indeterminate. A gated CTA should also be used in right-sided endocarditis if the pulmonary valve is unable to be seen by echocardiography of either type. This can also help establish the presence of peripheral emboli in the lungs. Once the diagnosis is made, antibiotic therapy duration and the need for surgery are important to determine. Consideration of hardware removal (such as indwelling pacemakers) requires additional consideration as they often sustain continued infection. Other considerations like removal of AV fistulas or eventual closure of VSDs require further consideration [30].

Key Points
- Signs/symptoms of endocarditis are nonspecific but include marantic symptoms like weight loss, decreased appetite, fatigue, and persistent fever.
- Physical exam may include fevers, new heart murmurs, splinter hemorrhages under the nail bed, Osler nodes on the pads of the fingers and toes, Janeway lesions (erythematous macules on the palms and soles of the feet), Roth spots (on the retina), evidence of systemic emboli (brain, kidney, spleen if systemic

spread), congestive heart failure, and respiratory symptoms if multiple pulmonary emboli.

- Transthoracic echo (TTE) is first line in diagnosis along with three sets of blood cultures, but the TTE can be negative in up to 70% of patients with definite endocarditis. Additional evaluation includes transesophageal echo (TEE) if TTE is negative, gated CT especially in pulmonary valve endocarditis (can be difficult to image by TTE/TEE and added benefit of establishing evidence of distal emboli in the lung); neuro exam, head CT and ophthalmologic exam to look for systemic emboli evidence for systemic vegetations.
- Indications for urgent or emergent surgery include persistent vegetation after systemic embolization, embolic events after completion of antimicrobial therapy, size of vegetation >10 mm on a systemic valve, increase in vegetation size despite antimicrobial therapy, significant valvular dysfunction, valve perforation or rupture, new heart block or large abscess or extension despite antimicrobial therapy [30].
- CTA chest gated for right-sided endocarditis may be helpful to visualize pulmonary valve abscess or establish the presence of peripheral emboli in the lungs.

8.6.2 Pain Management

Pain is a subjective and complex experience in the ICU setting. The pain that patients experience in the cardiac ICU can come from a variety of factors including body positioning/manipulation, prolonged immobility, surgical trauma, intubation, chest tube positioning, localized muscle weakness causing overuse of compensating muscles, and other physical manifestations of psychological stress. These experiences can be a great source of stress and have long-lasting and significant psychological effects on patients long after hospital discharge.

Nurses play a crucial role in optimizing pain control with routine assessments, documentation, and implementation of medications and other treatments in a timely manner. Pain assessment should take into consideration non-verbal cues and can also manifest as fatigue, weakness, or behavioral changes. Assessment of pain in the non-verbal patient continues to pose a challenge for both nurses and clinicians. Currently there is no universal pain assessment tool for non-verbal or cognitively impaired patients [31], and these patients are at a higher risk for inadequate pain control. Non-verbal pain scales such as the behavioral pain scale can be used to quantify pain which can guide therapies. For patients who can communicate their pain severity, the self-reported patient assessment is the most reliable and considered the gold standard. The most commonly used self-reported assessment in adults uses a numeric rating scale (0–10).

Nursing pearls for pain management:

- Provide a laminated card that includes the number scale and description of pain intensity (0, no pain; 1–3, mild pain; 4–6, moderate pain; 7–10, severe pain).
- Allow for adequate time for patient to respond during assessment.

- Educate patient on goal of pain management to optimize for comfort (numeric rating scale close to 3 but can vary by patient).
- Educate patient about breakthrough pain and provide and communicate measures to relieve pain before it becomes severe.
- Include pain assessment during team rounds.

Inadequate pain control can lead to increases in blood pressure, heart rate, respiratory rate, and hypoxemia. These physiologic variables are not considered a valid indicator for pain assessment as they can be nonspecific and should be used with caution, especially in the cardiac patient where there may be confounding variables that affect vital signs [32]. Inadequate pain control may also lead to long-term chronic pain syndromes [33–35].

Initial pain localization after cardiac surgery most commonly falls along the sternal or thoracotomy incision line and chest tube insertion sites. In the subsequent postoperative days, pain is often a combination of musculoskeletal and pleuritic chest pain due to the presence of chest tubes. Multimodal pharmacologic therapy has become more widespread than opioids to include NSAIDs, acetaminophen, and gabapentinoids. NSAIDs should be used with caution in patients with renal insufficiency, and acetaminophen should not exceed 2 g per day in the patient with liver cirrhosis. Initial pharmacologic treatments include the use of opioids at the *lowest effective dose* to limit adverse side effects. High-dose opioids can lead to increased risk of ventilator-associated pneumonia, respiratory depression, prolonged intubation, delirium, hypotension, pruritus, nausea, constipation, vomiting, postoperative ileus, and urinary retention, which also leads to increased length of ICU stay. Use of opioids increases risk of delirium in the ICU [36] and especially with use of benzodiazepines. Gabapentin is often the preferred treatment for neuropathic pain given its rapid onset, no QT prolongation, and no delirium or daytime drowsiness. Patient-controlled analgesia (PCA) can be considered after extubation with transition to an oral regimen once the patient can tolerate oral intake. In the early postoperative period, opioids delivered through a PCA provide better analgesia than conventional parenteral opioid regimens and also result in better patient satisfaction [37]. Paravertebral blocks can also be considered for pain control post-cardiac surgery [38]. ACHD patients with pre-existing chronic pain from prior cardiac surgeries or other non-cardiac issues may experience exacerbation of baseline pain due to hyperalgesia or allodynia. For patients who are on chronic pain therapies prior to hospitalization, management may require a multidisciplinary approach with the use of a pain team consultation.

Nonpharmacologic modalities of pain control are often low cost and can be used as adjunctive therapy. These modalities, including massage therapy and music therapy, have demonstrated decreased self-reported pain scores and anxiety, in addition to decreased need for pain meds [39]. Massage therapy includes areas of the hands, feet, back, neck, and shoulders and is best optimized by tailoring the massage to the individual's needs. Other non-pharmacologic treatments include cold therapy, relaxation techniques, meditation, and emotional support through the presence of family/friends.

Suboptimal pain management can result in delayed mobilization, impaired rehabilitation, and increased hospital length of stay. Acknowledging and respecting reported pain levels promote a healthy and trusting therapeutic relationship between the team and patient. Many adult congenital heart patients have underlying anxiety and PTSD that may play a role in their perceived pain. Some patients report heightened anxiety during periods where there is loss of control (i.e., being under anesthesia/intubated, inability to talk or communicate, fear of the unknown). Identifying triggers of anxiety and collaborating with the patient on a pain management plan that includes nonpharmacologic modalities prior to surgery may help lessen perceived pain postoperatively. This patient-centered personalized care plan gives some of the control back to the patient and is an important part of maintaining their dignity. Patient satisfaction despite report of high pain levels is dependent on communication, listening, and the attitude of the health care team [40].

Key Points
- Establishing a collaborative pain plan prior to surgery is ideal.
- Nonpharmacologic modalities to reduce pain should be considered.
- Use the lowest effective dose of pain medication to limit adverse side effects.

8.6.3 Cardiac Rehabilitation

Mobilization is key to recovery for adults after cardiac surgery. Once the patient is clinically stable, the sooner they are out of bed, the quicker their recovery. Early rehabilitation decreases risk of deconditioning and improves overall outcomes [2]. Rehab starts in the ICU and continues throughout the hospitalization and often in an outpatient rehabilitation center. Pain management is closely tied to rehabilitation as the patient needs to be comfortable enough to participate, but not too sedated from being overmedicated. Physical and occupational therapy should be consulted for every patient. Figure 8.1 gives an example of a rehabilitation chart that can be posted

Postoperative Day (POD)	POD 0	POD 1	POD 2	POD 3
Date				
Out of Bed (OOB) to Chair Daily during meals MAXIMUM SITTING TIME PER		Goal: Up in chiri 2x Time Done / If not why?	Goal: Up in chair 3x Time Done / If not why?	Goal: Up in chair 3x Time Done / If not why?

Fig. 8.1 Sample postoperative mobilization protocol

SESSION: 1.5 HOURS				
Ambulation Progressive Goals PT/OT Evaluation Goal POD 1 Actual Date:	Goal: Dangle at edge of bed 5-6 hours post extubation Time Done | If not done why?	Goal: Walk 2x Time of walk | If not done why?	Goal: Walk 3 x Time of walk | If not done why?	Goal: Walk 3 to 4x Time of walk | If not done why?

Fig. 8.1 (continued)

at the patient's bedside in the ICU. It should be reviewed daily on multidisciplinary rounds with patient participation. Prior to discharge, patients should be referred to outpatient cardiac rehabilitation to help with return to preoperative activity.

8.6.4 Nutrition

Optimal nutrition in the post-cardiac surgical patient or heart failure patient in the ICU setting is an important component of recovery and can affect patient outcomes. Most ACHD patients are in the ICU postoperatively for a few days before being transitioned to a lower acuity clinical setting. For more prolonged stays, patients can be in a catabolic or anorexic state and are at a greater risk of developing malnutrition. Malnutrition can lead to skeletal muscle wasting, infections (i.e., pneumonia), poor wound healing, and muscle weakness, which can prolong hospital length of stay. Nurses play an active role in identifying, managing, and advocating optimal nutritional goals.

Postoperatively, early advancement of enteral feeds assists in wound healing, return of gut motility, and improved immune function with less systemic inflammation. Enteral intake is preferred over parenteral nutrition due to infectious complications that can occur with indwelling lines and lack of intestinal stimulation. Furthermore, those patients with enteral vs. parenteral nutrition have been shown to have a decreased ICU length of stay [41].

Patients should be evaluated for their ability to swallow prior to oral intake initiation by using a nursing bedside swallow screen. In addition, patient voice quality should be assessed for range, weakness, or hoarseness to rule out vocal cord dysfunction [42]. Choking on saliva or frequent coughing or throat clearing may point to aspiration risk with oral intake. Nurses should have a low threshold for a formal bedside swallow study if there are concerns. Vocal cord irritation from intubation and recurrent laryngeal nerve injury are potential causes of swallowing difficulties. Those that undergo extensive neck dissection or aortic arch reconstruction are at a

higher risk of recurrent laryngeal nerve injury or irritation. In addition, patients with prolonged supine positioning (ECMO, balloon pump, groin lines) are at risk for silent aspiration. For patients with dysphagia or altered mental status, a nasogastric (NG) tube can be utilized short term. It is important to confirm correct positioning of the NG tube prior to every administration as movement and coughing can dislodge the tip of the tube.

In circumstances where enteral nutrition is not well tolerated, parental nutrition can be given as the primary source of calories where all nutrients are delivered intravenously. This formula consists of a mixture of protein, carbohydrates, lipids, and electrolytes. As previously mentioned, potential complications of parenteral nutrition include infection, dehydration, hyperglycemia, hypoglycemia, thrombosis, and micronutrient deficiencies.

Postoperative cardiac surgical patients who stay in the ICU for 3 or more days are at a high risk for inadequate nutrition therapy [43]. Metabolic rate increases in the setting of an acute stress response such as surgery, sepsis, or chronic illness, which, in turn, leads to an increase in endogenous glucose production and breakdown of protein stores in muscles and organs. Critically ill patients require 25–35 kcal/kg/day to maintain adequate nutrition subdivided into 50% carbohydrates, 20% protein, and 30% lipids [2]. Whether enteral or parenteral, interruption of feeds often occurs in the ICU and efforts should be made to minimize interruptions when possible and maximize nutrition ahead of time if long NPO times are necessary. Interruptions include postoperative dysphagia and vocal cord paralysis, vomiting, nausea, GERD, constipation, fasting for procedures/studies, and missing orders or documentation.

ACHD patients hospitalized for heart failure may also present with cardiac cachexia. They may have unintended weight loss or low BMI despite high caloric intake, leading to increased mortality. These patients have a higher metabolism and develop inadequate absorption of enteral calories due to gut edema that leads to lean muscle wasting [44]. There are studies underway looking at ways to optimize nutrition for this population; however, there are currently no formal recommendations for protein intake. A caloric intake of 35 kcal/kg/day has been shown to be safe and effective in increasing muscle mass in heart failure patients [45]. There is no specific recommendation for micronutrients, although it is known that loop diuretics increase loss of water-soluble micronutrients. Many heart failure patients are on ACE-I or ARBs, which can cause zinc deficiency [46]. Proinflammatory foods such as sugars and saturated fats should be avoided, while anti-inflammatory foods such as oily fish, olive oil, garlic, and turmeric may be helpful [47]. There had previously been controversy on administration of enteral feeds for patients on vasopressors in the ICU. Recent data suggests that early enteral feeds in patients on vasopressors are associated with reduced mortality [48].

Laboratory markers such as albumin, prealbumin, and protein can be used in the nutritional assessment. Fontan patients with protein-losing enteropathy are commonly found to have low protein stores due to malabsorption in the gut. These ICU patients may require albumin infusions and a diet high in protein and medium-chain triglycerides. Following cardiac surgery, a low salt diet (2–3 g of sodium) is

recommended to reduce volume overload that occurs post cardiopulmonary bypass. Fluid accumulation occurs due to exogenous fluids used in the pump prime and cardioplegia and as a result of renal dysfunction that causes delayed clearance of sodium and water [49]. Most adult hospitals have a low-sodium, heart-healthy menu that patients are encouraged to order from.

Hyperglycemia is a common occurrence in the ICU setting, even in the absence of pre-existing diabetes mellitus, and is associated with increased morbidity and mortality in the adult population [45]. Postoperative glycemic control aids in wound healing. Tight glucose control reduces morbidity with a lesser impact on mortality due to hypoglycemia. Insulin therapy in critically ill patients should be initiated for glucose levels >180 mg/dL with a target of 140–180 mg/dL while avoiding hypoglycemia [50, 51]. Continuous infusions of IV insulin enable quick titrations to be made and can achieve target glucose levels in 4–8 h. Hyperglycemic order sets that include glucose measurements and sliding scale orders are effective to ensure timely dose adjustments for insulin when necessary in the ICU. For diabetic patients, extra care must be taken to ensure adequate titration of insulin to ensure appropriate glucose levels.

Key Points

- Nurses are optimal champions to promote good nutrition for patients.
- Enteral nutrition should be considered as early as possible when deemed safe.
- Patients may have difficulty swallowing post cardiac surgery for a number of reasons and should have a low threshold for formal bedside swallow study if there are concerns.

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Part IV

Advanced Heart Failure in ACHD

Management of Acute Heart Failure

9

Paolo Ferrero

9.1 Introduction

Heart failure (HF) is a major cause of morbidity and mortality in patients with congenital heart disease (CHD) [1, 2]. Similar to HF secondary to acquired diseases, hospitalization for heart failure is an independent risk factor; indeed, a 3-year post-discharge mortality of CHD patients with HF ranges between 25 and 35% [3]. According to this background, we can predict a growing impact of patients with CHD on the healthcare system prompting a timely organization.

Management of HF has significantly improved in the last decade due to the abundance of established evidence-based treatments; however, adult patients with CHD are a peculiar subset due to the heterogeneity of the anatomy and circulatory models [4, 5].

Heart failure is defined as a clinical syndrome characterized by the inability of the cardiovascular system to grant an output matching with the metabolic requirements. Although this general definition still holds true, many different HF profiles have been defined over the last decade: acute, chronic, worsening decompensated, HF with reduced EF (HFrEF), HF with preserved EF (HFpEF), and HF with mid-range HF (HFmEF). This apparently undue plethora of definitions has been devised in order to better phenotyping patients with HF aiming to fill a still significant gap in terms of morbidity and mortality. In particular, the distinction between acute and chronic HF has been almost replaced by a more clinically efficient categorization of heart failure in chronic vs decompensated. From the operative's point of view, chronic HF patients are at a lower risk and managed as outpatients, while those with 'acute' decompensated HF usually require hospitalization and have a higher mortality [5].

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Since care of an ACHD patient is a relatively new discipline, so far, interventions and therapies have been largely extrapolated from models of acquired heart disease. Heart failure is a typical example of this attitude; indeed, most of the terminologies, metrics, and therapies applied in ACHD patients are derived from the bulk of evidence established in HF patients without CHD.

Given this background, some questions arise whether the general definition of HF is also valid in the subset of patients with CHD, whether the same classifications are meaningful, and if evidences about therapies are transferable.

In the following paragraphs, we will quantify the size of the problem of ACHD patients with acute HF, and we will cover the principles of management, particularly focusing on pathophysiologic unicity of CHD that has to be taken into account when transferring evidences into practice in this subset of patients.

9.2 Epidemiology of Heart Failure in CHD

The overall incidence of HF in patients with CHD is about 1/1000 [1]. The global risk of developing HF in ACHD patients varies according to the specific anatomy, achieving 40% for more complex anatomies. Patients deemed to be at a higher risk are those with systemic right ventricle, ranging between 20% in those with transposition of great arteries (TGA) post atrial switch and 30% in those with congenitally corrected TGA. Patients who had undergone univentricular palliation have a particularly high risk of HF, up to 40%, due to the peculiar physiology of Fontan circulation [6, 7].

Mortality risk also depends on initial clinical presentation, and patients with acute decompensated heart failure have a fivefold risk of death as compared to chronic but not hospitalized patients [1]. This data reproduces a similar trend observed in HF patients with acquired heart disease.

9.3 Pathophysiology

9.3.1 General Concepts and Circulatory Models

Etiopathogenesis of HF in patients with CHD recognizes different layers of causative factors. HF may be the result of intrinsic myocardial architectural factors, mainly under genetic control, of the cumulative effect of hemodynamic residual burden after surgical repair and of perfusion defects. Beside these specific patient-related factors, there are variables related to the surgical injuries themselves, which add additional layers of complexity and heterogeneity [8] (Table 9.1). All these factors shape the process of myocardial remodeling, which is counteracted by the mechanisms of adaptation of the cardiovascular system.

From the pathophysiologic point of view, we have to distinguish two main models of circulation: biventricular and univentricular. In patients with biventricular circulation, two separate ventricles grant the systemic and pulmonary output. In this

Table 9.1 Summary of the main etiologic factors involved in HF development in CHD

Etiologic factor	Mechanism
Myocardial architecture	Failure of a thinner two-layer RV systemic ventricle
Pressure overload	Hypertrophy and fibrosis leading to diastolic dysfunction, systolic dysfunction at later stages
Volume overload	Dilatation and systolic dysfunction
Perfusion defects	Endothelial dysfunction. Supply–demand mismatch (systemic RV supplied by the right coronary)
Pulmonary arterial hypertension	Increased RV afterload, RV dysfunction
Cyanosis	Endothelial dysfunction, fibrosis
Surgical injury	Coronary injury, myocardial scar, inadequate myocardial protection
Passive pulmonary circulation (Fontan)	Passive flow through the lung without subpulmonary ventricle leading to venous congestion and systemic ventricle underfilling

scenario, all factors mentioned in Table 9.1, namely, a morphologic RV as a systemic ventricle, the presence of significant pressure or volume overload, the effect of surgical injuries, and so on, contribute to the hemodynamic burden to the ventricles, which ends up with HF. In patients with univentricular circulation, all these issues hold true, but there is an additional peculiar hallmark in this subset of patients due to their specific anatomy. Fontan intervention was contemplated for the first time in the earlier 1970s as a definitive palliation of univentricular circulation. It has undergone several modifications, but the principle remains to that of connecting the systemic venous drainage directly to the pulmonary arteries, bypassing the subpulmonary ventricle. This apparently simple procedure has tremendous consequences, as the central venous pressure has to increase to a non-physiologic level in order to allow the blood to passively flow through the lungs. This circuit, by definition, realizes a condition of systemic congestion, ventricular underfilling, and low output and can acutely fail whenever external detrimental factors intervene (Fig. 9.1) (also see the following paragraphs).

9.3.2 Phenotypes of HF in CHD

As mentioned in the introduction, the definition and nomenclature of HF have evolved over the years, recognizing different phenotypes. In the guidelines published in the last decade, clinical presentation has been dichotomized into HF with reduced or preserved EF (HFrEF and HFpEF). This classification assumes an EF cutoff of 40% and is based on the concept that most tested therapies have demonstrated a positive effect in terms of morbidity and mortality in patients with reduced EF [6].

Both categories of patients may present with chronic ambulatory or acute decompensated HF. This dichotomization can be retained in the field of CHD as we can recognize pathophysiologic models in ACHD patients with HF fulfilling these definitions. However, in congenital patients, the boundary of 40% can either apply to a

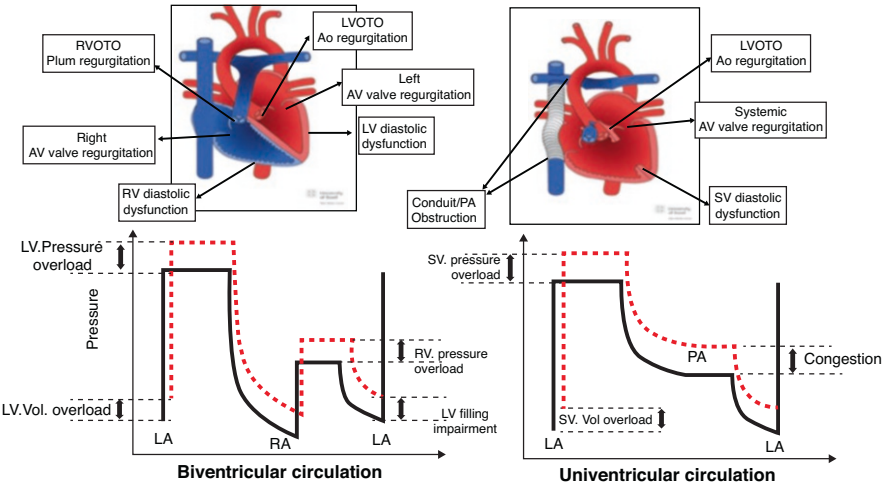

Fig. 9.1 Depiction of biventricular and univentricular circulation. Dotted red lines illustrate the hemodynamic burden secondary to residual defects and putative etiologic factors. RV, right ventricle; LV, left ventricle; SV, single ventricle; Ao, aorta; PA, pulmonary arteries; AV, atrioventricular valve

morphological right or left systemic ventricle, adding a peculiar additional element of complexity to the classification. Regardless of the EF cutoff, the common background of the vast majority of non-congenital adult patients with HF is dominated by a normal or reduced cardiac output, while in the field of congenital defect, a not infrequently 'high output' HF may occur. The term 'high output' HF is again imported from the acquired pathology, whereas in a normal series circulation, the peripheral resistances fall or are partially bypassed by arteriovenous channels, eventually leading to an exaggerated increase of the stroke volume, which, in turn, cause overload and finally failure of the ventricles. In the context of congenital defects, the definition of 'high output' actually applies to a situation where there is an amount of blood that 'recirculates' through the lungs without participating to the oxygen exchange with the metabolically active tissues. This 'overcirculation' occurs in many congenital heart disease characterized by a systemic to pulmonary shunt and, similar to the aforementioned scenario, causes ventricular overload and eventually failure.

9.3.3 Main Differences of HF in Acquired Vs Congenital Disease

There are several differences between HF in acquired model of disease as compared to CHD. These differences can be recognized at different levels: etiology, pathophysiology, functional assessment, and prognosis (Table 9.2).

We have already discussed that HF in patients with CHD have a far more heterogeneous etiopathogenesis recognizing multiple hits, as compared with acquired heart disease. As a consequence, clinical course and natural history are less predictable. Different from acquired heart disease, most congenital defects involve the

Table 9.2 Conceptual differences between HF in acquired vs congenital heart disease

	Acquired heart disease	Congenital heart disease
Etiology	Ischemic, genetic, valvular	Multifactorial[a]
Ventricle usually involved	Left ventricle	Right ventricle
Clinical course	Remitting	Progressive
Functional evaluation	NYHA	NYHA not reliable
Sudden death risk	5–10%/year	0.1–2%/year
Associated AV valve regurgitation	Mitral valve, functional	Tricuspid valve, functional or structural
Neurohormonal activation	Proved	Conflictual data
BNP	Associated with prognosis	Associated with prognosis
Beta-blocker efficacy	Proved	Few evidences
RAAS antagonism efficacy	Proved	Few evidences
Sacubitril–valsartan efficacy	Proved	Few evidences
Digoxin	Proved	Few evidences

RAAS renin–angiotensin–aldosterone system, *BNP* brain natriuretic peptide
[a]Ischemic, chronic hypoxia, pathologic load conditions, poor surgical protection, persisting shunts (see text for details)

right AV valve and/or right ventricular outflow tract; therefore, the right ventricle is most often involved [9].

A major concern in ACHD patients' follow-up is the stratification of sudden death risk. The incidence rate of sudden death in patients with congenital substrates, even including those with ventricular dysfunction, is significantly lower as compared with ischemic or primitive cardiomyopathy. This observation makes stratification of arrhythmic risk challenging and tailored [10].

Neurohormonal hyperactivation, leading to overexpression of molecules that exert deleterious effect on the myocardium, has been largely accepted as a critical pathophysiologic mechanism in HF [11]. Despite the extreme anatomic heterogeneity, data are consistently showing the whole cohort of ACHD patients with HF display a sign of neurohormonal activation as demonstrated by an increased level of renin, angiotensin, aldosterone, and brain natriuretic peptide (BNP), likewise in patients without CHD [12]. This finding not necessarily translates in a comparable efficacy of neurohormonal antagonism–targeted therapy. Indeed, evidences about efficacy of beta-blockers and renin–angiotensin–aldosterone system (RAAS) inhibition are scant and conflictual [8].

9.4 Management

9.4.1 General Principles

The cornerstones of treatment of ACHD patients with acute or worsening HF do not differ from those with non-congenital heart disease: respiratory distress relief and hypoxia correction, fluid overload correction, systemic vasodilatation, and

contractility enhancement. These principles are conceptually based on Frank–Starling curve manipulation.

Patients presenting in the acute setting usually require prompt intervention using oxygen supplementation, non-invasive ventilation and intravenous medications (Tables 9.3 and 9.4).

After controlling the acute phase, the therapy is downgraded from intravenous to oral agents. During the subacute phase, the treatment is directed to the same objectives of the acute treatment: maintain euvolemic state and counteract neurohormonal activation. Although there are no solid proofs of the efficacy of ACE inhibitors, angiotensin receptor blockers, and mineralocorticoid antagonist on mortality, in clinical practice, these agents are commonly used in congested ACHD patients, assuming that RAAS overactivation results in a detrimental increase of afterload and fluid retention, which begets further congestion [13].

Table 9.3 Principles of treatment according to clinical presentation

	Acute phase	Subacute phase
Respiratory distress/hypoxia	Oxygen NIV	
Congestion	IV diuretics IV vasodilators	Oral diuretics RAAS inhibitors
Low output	IV vasodilators IV inotropes	RAAS inhibitors
Low output and congestion	IV vasodilators IV inotropes/inodilators IV diuretics	RAAS inhibitors Oral diuretics

NIV non-invasive ventilation, *IV* intravenous

Table 9.4 Doses of principal intravenous agent used in AHF

Intravenous medication	Dose
Diuretics	
Furosemide	20–500 mg bolus Infusion: 5–40 mg/h
Torasemide	10–200 mg bolus Infusion: 5–20 mg/h
Vasodilators	
Nitroglycerin	20–400 mcg/min
Nitroprusside	0.3–5 mcg/kg/min
Inotropes	
Dobutamine	2–20 mcg/kg/min
Dopamine	2–10 mcg/kg/min
Epinephrine	0.05–1 mcg/kg/min
Norepinephrine	0.1–1 mcg /kg/min
Inodilators	
Milrinone	0.1–0.8 mcg/kg/min
Enoximone	1.25–7.5 mcg/kg/min
Levosimendan	0.5–2 mcg/kg/min

The long-term use of beta-blockers, RAAS antagonists, and more recently sacubitril–valsartan, which are the cornerstone of chronic treatment of HF patients with reduced EF, is not supported by solid evidences in ACHD patients. However, these medications are commonly recommended in selected patients according to the limited available evidences and to a pathophysiology-tailored approach.

9.4.2 Practical Management Multistep Algorithm of Acute HF in CHD

The algorithm depicted in Fig. 9.2 illustrates the crucial steps guiding the management of acute decompensation of patients with CHD. The main steps can be summarized as follows (Fig. 9.2):

First step. It includes risk assessment, in order to establish the proper clinical environment where to treat the patient (ward vs intensive care). This initial triage can be initially done by assessing skin perfusion, mental status, peripheral pulses, and heart and respiratory rate. There are few congenital anatomies where peripheral pulses are ubiquitously absent such as aortic coarctation with aberrant right subclavian artery. Desaturation may be preexisting and due to intracardiac right to left shunts; therefore, not necessarily it is secondary to respiratory failure. Arterial gas analysis in hypoxic patients is useful to differentiate a chronic from acute condition.

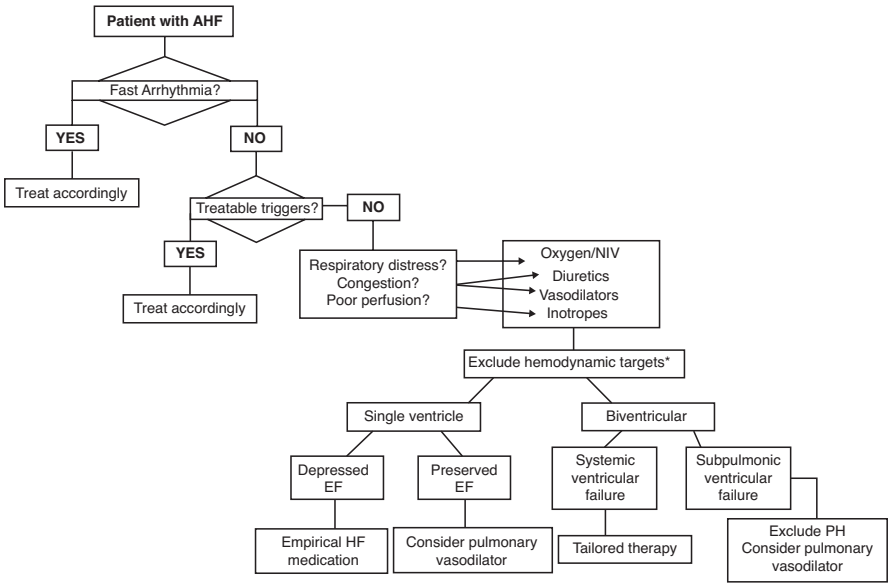

Fig. 9.2 Practical algorithm of acute HF management. *EF* ejection fraction, *NIV* non-invasive ventilation

Second step. It aims to exclude a potentially life-threatening condition that can precipitate acute decompensation, such as arrhythmias and atrioventricular blocks. Even supraventricular arrhythmias that are usually deemed to be benign may significantly impair cardiac output in patients with CHD and need aggressive treatment. Other treatable conditions that can precipitate acute HF, which can be easily ruled out, are acute anemia and infections.

Third step. The preliminary treatment of acute HD is directed at correcting the main hemodynamic abnormality: congestion, low output, and low output and congestion (Fig. 9.2).

Despite the lack of specific evidences on ACHD patients, in a selected subset of patients, we are reasonably authorized to assume the analogue models of diseases well known in the acquired pathology. This is the case of patients with HFrEF with morphologic left ventricle associated or not with AV valve regurgitation that, as widely demonstrated in non-congenital patients, should be treated with beta-blockers and RAAS inhibitors. Likewise, patients with HFpEF may benefit from filling optimization by using a beta-blocker.

On the other hand, there are several pathophysiologic peculiarities that have to be taken into account. Patients post Mustard or Senning interventions have an abnormal atrial compliance that affects ventricular filling. In this scenario, aggressive diuresis and vasodilatation can have detrimental effects on ventricular preload. A similar caution in using systemic vasodilators needs to be applied for patients with LVOTO or RV under pressure (Table 9.5).

Table 9.5 Medical management of HF according to anatomic category

Category	Pathophysiology	Therapy	Caution
TOF PA/VSD PA/IVS Ebstein	Pulmonary valve regurgitation Conduit dysfunction Tricuspid regurgitation PAH Subpulmonary RV failure	Diuretics Aldosterone antagonists Beta-blockers Pulmonary vasodilators	Avoid massive Systemic vasodilatation
CTGA Atrial switch Single-ventricle palliation	Systemic RV dysfunction Tricuspid regurgitation Preserved vent. Function Depressed vent. Function	Diuretics Aldosterone antagonists **Beta-blockers** ACEi/ARB (TR) Supportive therapy Diuretics/ aldosterone antagonist Pulmonary vasodilators Beta-blockers ACE inhibitors/ ARB Digoxin	Avoid massive diuresis and systemic vasodilatation. Avoid massive systemic vasodilatation. Avoid aggressive diuresis[a] Avoid aggressive diuresis[a]

Table 9.5 (continued)

Category	Pathophysiology	Therapy	Caution
Left-sided lesions	Left AV valve regurgitation HFpEF HFrEF	Beta-blockers ACE inhibitors/ ARB Digoxin Sacubitril–valsartan	Avoid massive systemic vasodilatation.

[a]In cyanotic patients

TOF tetralogy of Fallot, *PA* pulmonary atresia, *VSD* ventricular septal defect, *IVS* intact ventricular septum, *ACEi* angiotensin-converting enzyme inhibitor, *ARB* angiotensin receptor blockers, *TR* tricuspid regurgitation, *HFpEF* heart failure with preserved ejection fraction, *HFrEF* heart failure with reduced ejection fraction.

9.5 Key Learning Points

- Incidence rate of HF in ACHD is expected to increase.
- The risk of major adverse events in patients with HF increases overtime and depends on the clinical presentation (acute vs chronic), the anatomic complexity, the presence of a morphologic right systemic ventricle, and the presence of single-ventricle circulation.
- A stepwise approach to AHF follows similar stabilization priorities in ACHD as compared with non-congenital patients.
- In the absence of solid evidences about medical therapies and interventions in this particular population, we can assume some of the concepts proved in other pathologic models. However, we should be cautious in straightaway transferring evidences from acquired heart disease to CHD.
- Treatment should be always tailored to the pathophysiology assumed for any individual patient.

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Mechanical Circulatory Support Strategies

10

Mauro Cotza

10.1 Introduction and Definition

The advances in surgical and medical treatment of children born with congenital heart defects (CHD) have led to improving their survival: most reach adulthood despite complexity, with the emergence of a new cohort of young adults known as adults with congenital heart disease (ACHD).

The dark side of these advances is the rise of a subgroup of them affected by heart abnormalities, which evolves to heart failure (HF), the leading cause of mortality and morbidity in this subset [1].

Management of HF is challenging in these patients and becomes an early hard challenge, given their structural variations and the associated physiological consequences.

Heart transplantation is of limited utility in this group because of either donor shortage or associated comorbidities that make these patients ineligible for transplantation.

Primary or secondary pulmonary vascular disease, high levels of allo-sensitization due to previous surgery and transfusions, and comorbidities secondary to volume or pressure overload (such as liver cirrhosis) constitute causes for delisting [2].

MCS devices have evolved as an alternative treatment modality to conventional medical and electric (cardiac resynchronization therapy [CRT]) strategies in supporting the failing myocardium and overcoming chronic or acute cardiocirculatory failure of this population.

Unfortunately, they are often used less frequently than in those with a structurally normal heart because of the anatomical and physiological variations and anyway require an adequate knowledge on supporting the hearts of ACHD patients to reduce mortality and morbidity [3].

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S. F. Flocco et al. (eds.), *Guide for Advanced Nursing Care of the Adult with Congenital Heart Disease*, https://doi.org/10.1007/978-3-031-07598-8_10

10.2 Indications and Classification

ACHD may suffer the same causes of heart failure as the general population, but it's rare that they develop an ischemic myocardial disease while heart failure is generally associated to anatomical and physiological conditions related directly or indirectly to their primitive heart disease; we can cite, as an example:

- Systemic right ventricle (post atrial switch surgery or in cc-TGA).
- Single ventricle (right or left) after palliation (Fontan).
- Valve disease following surgical correction (CAVC, Fallot, pulmonary atresia).
- Left ventricle non compaction (LVNC).
- Myocardial ventricular fibrosis (late post-op Fallot).

As for normal structured heart, MCS in ACHD can be classified as a bridge to decision, bridge to recovery (BTR), bridge to bridge (BTB; to subsequent long-term MCS), and bridge to transplant (BTT) or as destination/alternative to transplant therapy (DT/ATT).

Furtherly, MCS can be used over the short, medium, or long term; according to duration we can distinguish these as follows:

- Class 1 devices: percutaneous axial flow pumps (Abiomed Impella), intra-aortic balloon pumps (IABP), external centrifugal pumps (Centrimag), extracorporeal membrane oxygenation (ECMO).
- Class 2 devices: membrane pumps (Berlin Heart EXCOR), external centrifugal pumps (Centrimag), extracorporeal membrane oxygenation (ECMO).
- Class 3 devices (extended support time):
- BTR: membrane pumps with portable driver (Berlin heart EXCOR), implantable pulsatile pumps (IVAD, P-VAD), implantable axial/centrifugal pump (Abbott Heartmate3, Jarvik 2000).
- BTT: membrane pumps with portable driver (Berlin heart EXCOR), implantable pulsatile pumps (IVAD, P-VAD), implantable axial/centrifugal pump (Abbott Heartmate3, Jarvik 2000), total artificial heart (Syncardia TAH).
- DT/ATT: implantable pulsatile pumps (IVAD, P-VAD), implantable axial/centrifugal pump (Abbott Heartmate3, Jarvik 2000), total artificial heart (Syncardia TAH).

The clinical status of the patient is discriminant for VAD choice within the various classes.

In general, patients can be classified into four clinical categories:

1. *Elective chronic hemodynamic deterioration* in patients already on the waiting list for heart transplant (HTX).
2. *Permanent refractory heart failure* in patients with contraindications to HTX.
3. *Urgent acute hemodynamic deterioration* in patients already on the waiting list for HTX.
4. *Emerging acute cardiogenic shock* in patients not on the waiting list for HTX (AMI, myocarditis, post-CPB).

For elective patients and for patients with contraindications to transplantation (categories 1 and 2), the choice will fall preferably on class 3 devices, and to guarantee the best quality of life at home, intracorporeal devices will be privileged. In this case, the therapeutic purposes will be BTT and DT.

The patients belonging to these two categories have hemodynamic signs of chronic decompensation (wedge>20 mmHg, mean systolic pressure < 60 mmHg), hyponatremia (serum Na < 135 mmol/L), and signs of multiorgan failure. In these cases, intolerance to ACE inhibitors is an additional criterion for indication to VAD.

Clinical category 3 includes all patients already on the waiting list in which decompensation degenerates acutely: these patients need hospitalization in the intensive care unit (ICU) and should be considered for a VAD only after failure of maximal medical therapy, even possibly associated with the use of IABP. This category preferred a class 2 VAD (such as BTT, or BTR or BTB) for short to medium support and, in case of HTX delay, is indicated to switch into a VAD class 3 for BTT.

In the patient of category 4, given the acute clinical picture, the space of the various therapeutic options is necessarily reduced: in these cases, easy-to-implant, readily available devices are to be preferred. Class 1 VADs are the ones that better fit the purpose: among these, we remember ECMO, centrifugal pumps and, more recently, a new generation of percutaneous axial flow pump systems (Impella), sometimes acting in combination with ECMO (ECPELLA).

In all cases, these are extracorporeal devices, with continuous flow, usable as BTR or as BTB toward a higher-class VAD, since they can perform their function for periods not very long.

The Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) scale is another helpful tool to stratify HF patients with ACHD requiring MCS [4] and to analyze risk factors and outcomes [5].

Using the INTERMACS scale, patients are classified based on the severity of symptoms and the trajectory of decline over time, which helps prognosticate those with advanced HF receiving MCS [6].

10.3 MCS Modalities

MCS is aimed to support or replace heart workload depending on the residual function of the ventricle that is failing.

MCS can replace right or left or both ventricles and, in specific cases, can also provide ventilatory support.

MCS may consists in an extracorporeal life support device (ECLS) or a left or right ventricular assist device (L-VAD, R-VAD) that, connected before ventricular inlet and after ventricular outlet, unloads the failing ventricle, thereby reducing myocardial oxygen demand, enhancing recovery, and provides adequate cardiac output (Fig. 10.1).

Respiratory failure secondary to heart failure or the presence of refractory pulmonary hypertension may suggest supporting lung function as well: in this case, extracorporeal membrane oxygenation (ECMO) acts like a cardiopulmonary support, vicariant of both functions (Fig. 10.2).

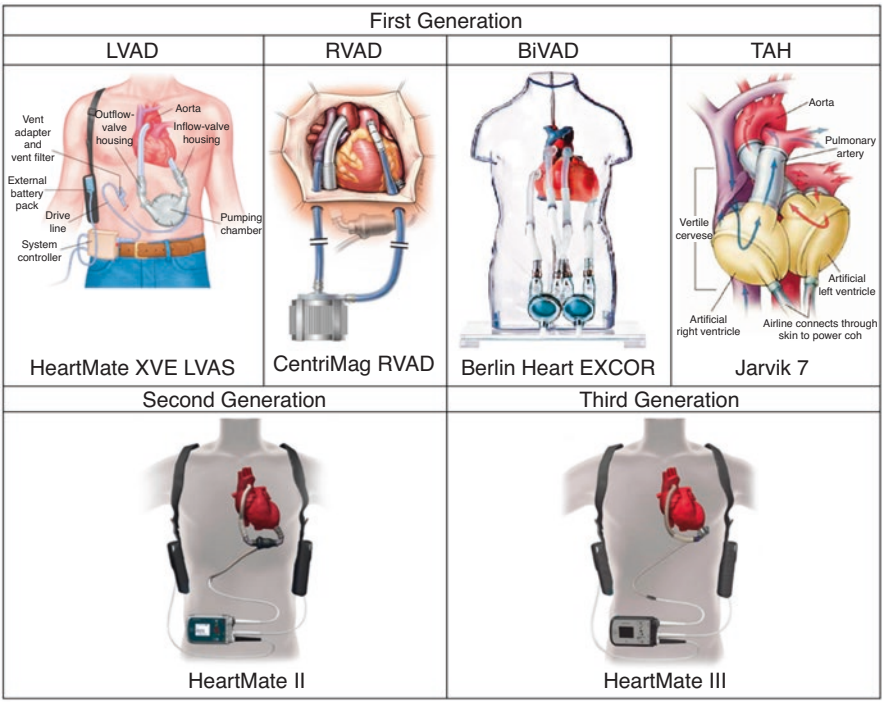

Fig. 10.1 MCS generation strategies

10.3.1 ECMO

Extracorporeal membrane oxygenation (ECMO) is a world-recognized leading strategy mainly used in acute heart failure or in those cases in which an early recovery is expected, generally in days or weeks.

ECMO in acute ACHD is generally im plemented after surgery or in case of acute decompensation in fragile patients waiting for heart transplantation, if eligible.

Extracorporeal life support organization (ELSO) (6-ELSO) reports an overall survival to discharge of 43% (Table 10.1).

Anyway, survival in this cohort is strongly affected by the etiology (Table 10.2).

ECMO is obtained through a complex system made of:

- An extracorporeal line (plastic tubes) that receives blood from a venous access.
- A centrifugal pump that propels blood to a membrane oxygenator where thermal homeostasis and gas exchange occur and, finally, returns blood to an arterial access (Fig. 10.3).

ECMO in adults can be established either percutaneously or by an open technique (cut down). Cannulation can be in the neck (internal jugular vein and the common carotid artery, rarely in adults unless no other viable option), groin

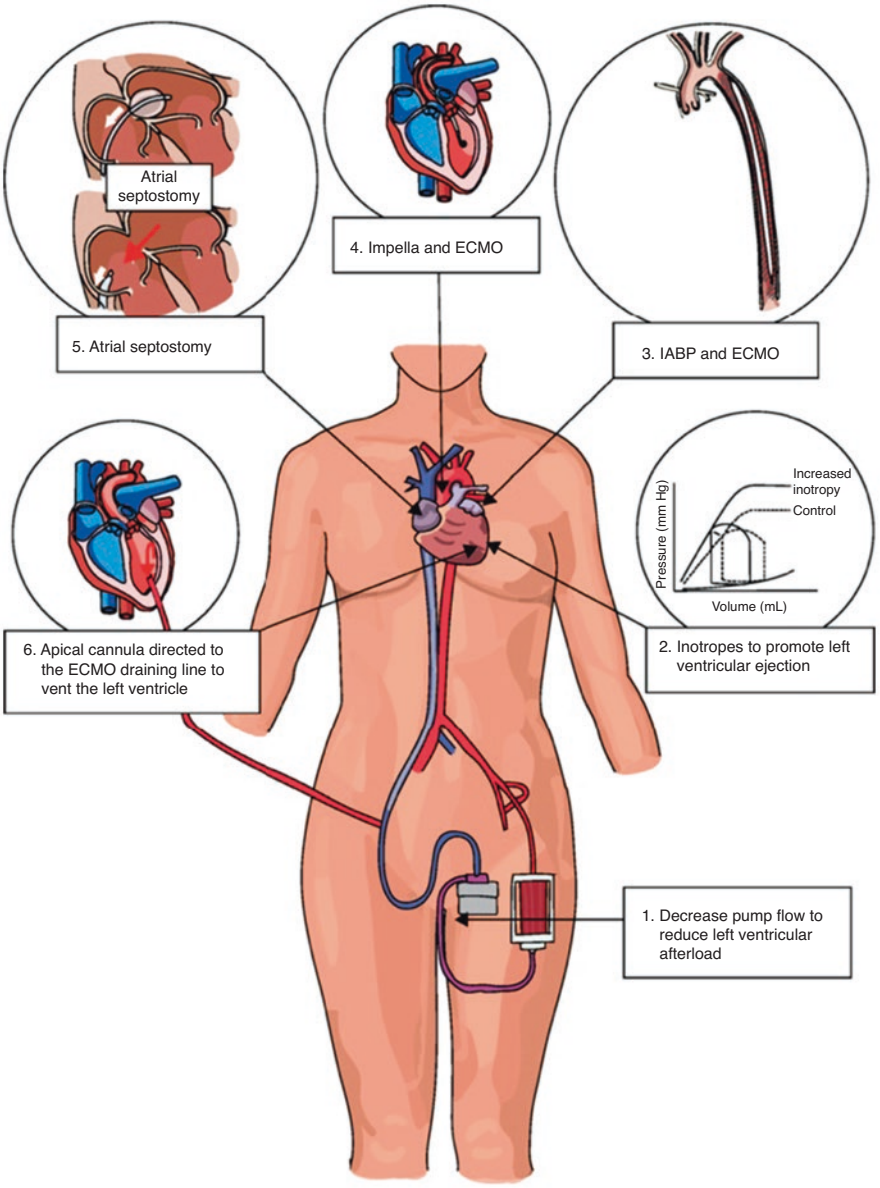

Fig. 10.2 ECMO and ECMO configurations

(femoral vessels, the most used), or central (right atrial appendage and the aorta; after cardiac surgery, femoral vessels are not viable). Patients with venous anomalies and prior cavo-pulmonary anastomosis require special attention regarding cannulation strategies [7].

Table 10.1 Adult cardiac runs by diagnosis (by ELSO International summary of statistics, October 2021)

	Total runs	Avg run time	Longest run time	Survived	% Survived
Congenital defect	185	171	846	80	43%
Cardiac arrest	414	111	1013	159	38%
Cardiogenic shock	7421	169	4783	3354	45%
Cardiomyopathy	178	185	2116	83	46%
Myocarditis	151	202	1370	110	72%
Other	13,479	156	6359	6352	47%

Note some runs are missing primary diagnoses

Table. 10.2 Adult cardiac runs by congenital cardiac diagnosis (by ELSO International summary of statistics, October 2021)

	Total Runs	Avg Run Time	Longest Run Time	Survived	% Survived
Left to right shunt (ASD/VSD/PDA/AV canal/AVSD/ECD)	64	186	766	24	37%
Left-sided obstructive (aortic stenosis/mitral stenosis/coarctation)	59	157	618	24	40%
Hypoplastic left heart	3	186	312	2	66%
Right-sided obstructive (pulmonary stenosis/pulmonary or tricuspid atresia)	8	190	340	2	25%
Cyanotic incr. Pulmonary flow (truncus asteriosis/TGA/TGV)	5	248	846	3	60%
Cyanotic decr. Pulmonary flow (TOF/DORV/Ebsteins)	12	140	281	10	83%
Other	34	163	840	15	44%

10.3.2 VAD

Ventricular assist devices (VADs) are *"mechanical pumps that take over the function of the damaged ventricle and restore normal hemodynamics and end-organ blood flow"* [8].

Advantages of VADs over ECMO (given specific indications) include a smaller priming volume, fewer anticoagulation and blood products, a reduction in the infection rate and sensitization [9], the fact that patients can be extubated, and early mobilization and, in predetermined condition, discharge at home.

In the more recent years, VAD has acquired a role in creating the conditions of eligibility for transplantation in those patients excluded for temporary criteria as in postcapillary pulmonary hypertension [10], oncological therapy with a favorable prognosis, and patients who experience long transplant wait times and serve as DT when there are contraindications to transplantation [11].

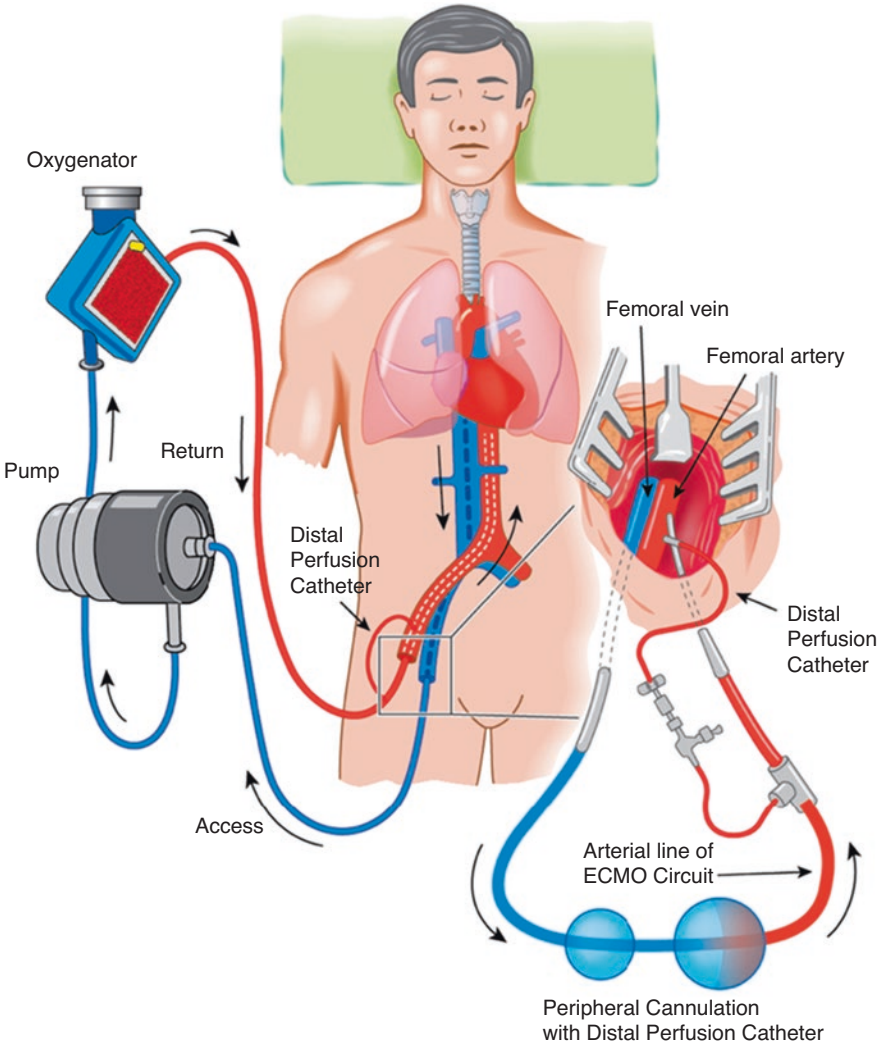

Fig. 10.3 ECMO circuits and components

In adults, contraindications (and so exclusion criteria to VAD) are generally considered, as well as severe neurologic impairment, a barrier to adequate anticoagulation, congenital anomalies with poor prognosis, and major chromosomal aberrations.

From the technical point of view, VAD implantation in CHD is different than in the structurally normal heart: anatomic considerations related to prior complex repairs, abnormal size and location of the aorta, orientation of the ventricular chambers, heart position in the chest, and in those patients with systemic right ventricle, devices designed for a morphologic left ventricle.

But there are also comorbidities of the hemostatic system, malnutrition, and end-organ dysfunction that make VAD implantation technically challenging. Other considerations include the thickness of the ventricles, semilunar valve regurgitation, and intracardiac shunts [12].

At least epicardial or transesophageal echocardiography should be used to assist with cannula placement. The RV free wall and diaphragm surface are both alternative options to best orient the inflow cannula toward the tricuspid valve. Regardless of the epicardial entrance point, implantation of devices into a systemic RV may require resection of trabeculae to provide unobstructed inflow. To avoid right-sided issues with damage to the liver or bowel or chamber compression upon closing the chest, the device may be placed back to front, closer to the midline, or through the right chest. Non-sternotomy approaches and non-aortic outflow graft positions may be reliable alternatives.

Some authors described the creation of a VAD-driven subpulmonic chamber following takedown of a Fontan circuit [13]. Others have described total artificial heart (TAH) implantation in Fontan patients requiring the creation of neo right atria, which can be problematic due to the lack of normal compliance [14].

An important change in VAD technology is the introduction of continuous-flow (CF) VADs. These can be implanted intracorporeally, which also allows for outpatient care. Over the past few decades, there has been a major shift in use from pulsatile VAD to CFVAD, and studies have reported superior survival rates for CFVAD vs pulsatile left ventricular assist devises (L-VAD) [15].

10.3.3 TAH

The TAH (Total Artificial Heart, SynCardia Systems, Tucson, AZ, USA) consists of right- and left-sided pumps and can be used as BTT or as DT. TAH pumps can be configured in a variety of ways to addresses various physiologic (1 or 2 ventricles) and anatomical variations peculiar to CHD. The TAH provides optimal hemodynamic support in CHD patients with residual lesions compared with a VAD or biventricular assist device (BiVAD) alone; however, implantation of TAH is much more challenging than VAD implantation (Fig. 10.4a–c), especially in CHD patients considering the wide range of anatomical variations. One of the major advances in the TAH implantation is the development of a patient-specific virtual implantation based on cross-sectional imaging studies [16].

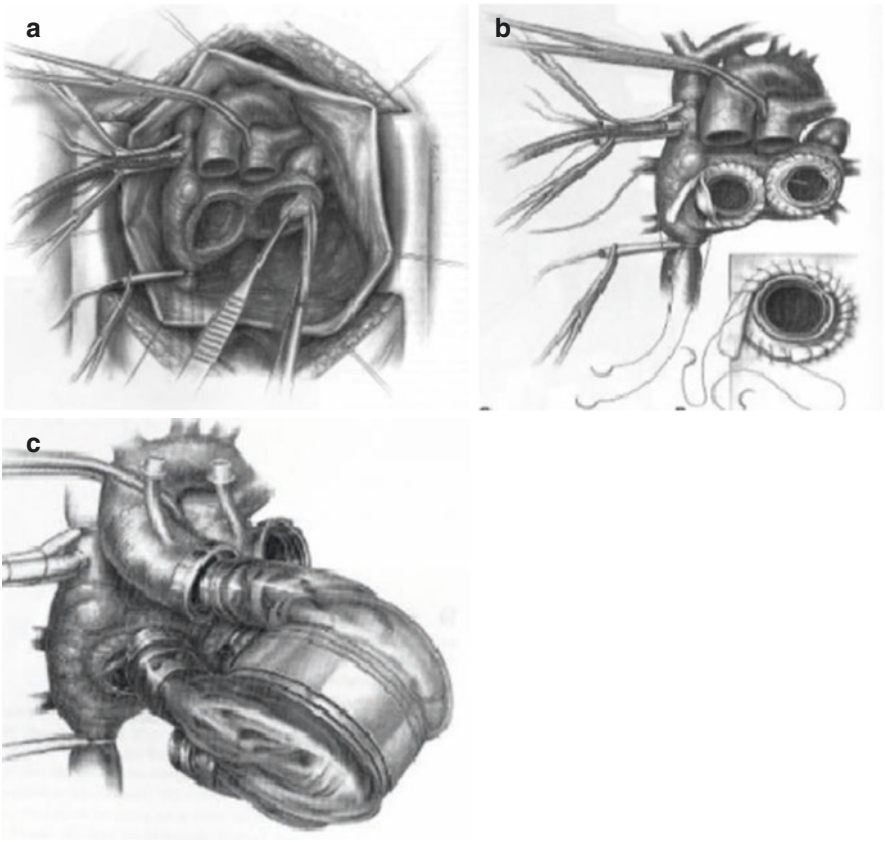

Fig. 10.4 (**a**) TAH cardiectomy. (**b**) TAH flange sutures. (**c**) TAH VAD connection

10.3.4 Impella and Tandem Heart

Percutaneous axial flow pumps (Fig. 10.5a, b) are another significant technological innovation in VADs, and these are increasingly being used as short-term support in patients with critical cardiogenic shock and postcardiotomy failure for both left and right ventricles [17, 18]. Tandem Heart (TandemLife, Pittsburgh, PA, USA) is a percutaneous VAD (Fig. 10.6) that uses cannulas placed in the left atrium with a transeptal approach to facilitate the direct unloading of the left heart [19].

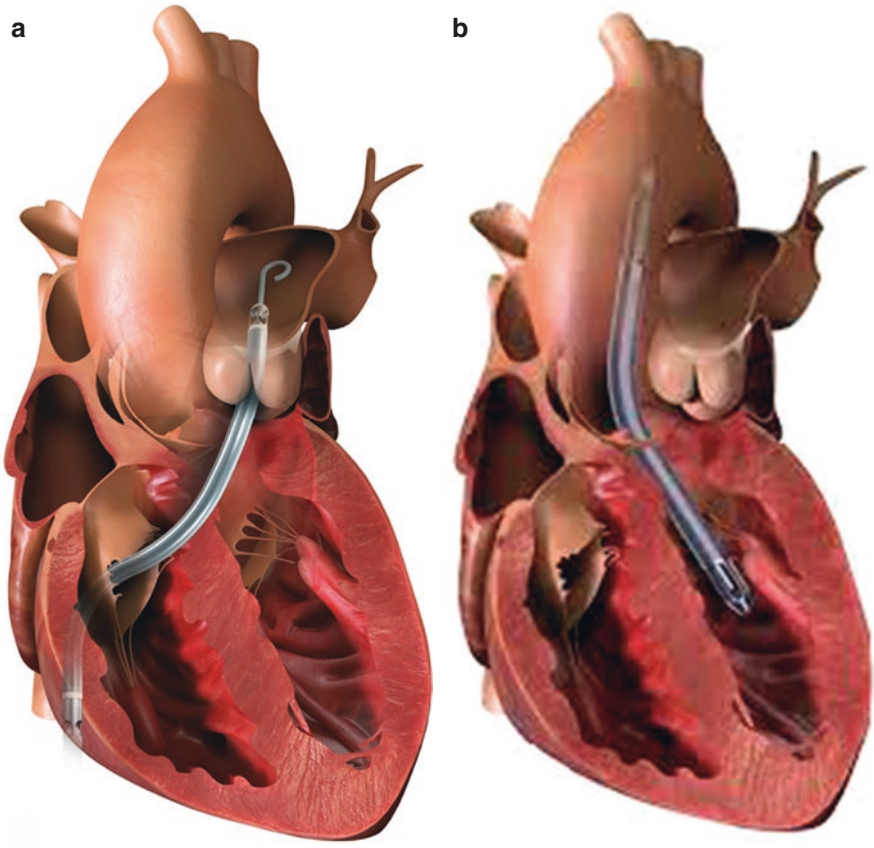

Fig. 10.5 (**a**) Impella RP axial flow pump. (**b**) Impella CP axial flow pump

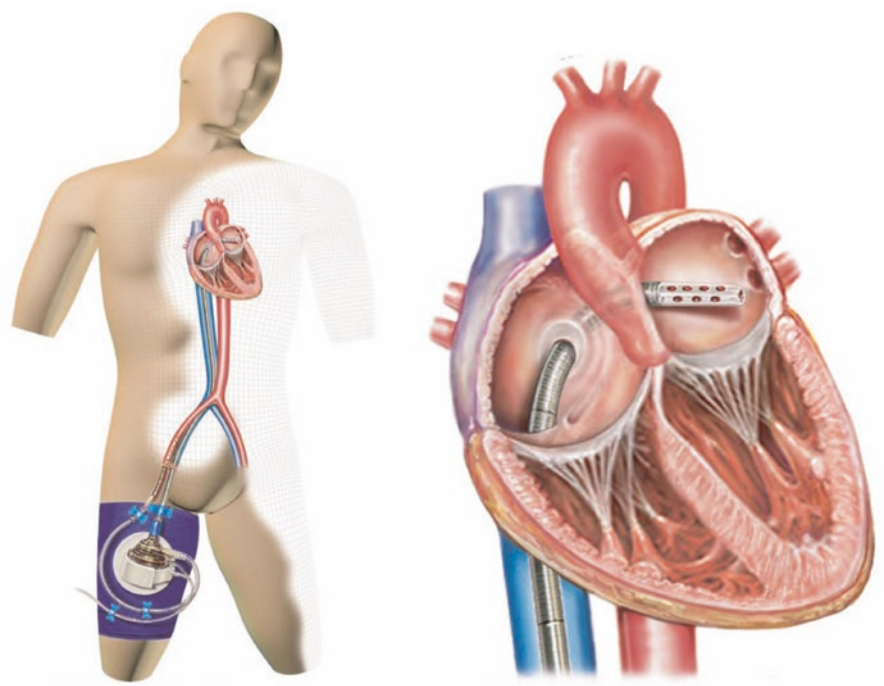

Fig. 10.6 Tandem Heart percutaneous L-VAD

10.4 MCS Reports and Outcomes in ACHD Patients with Biventricular Physiology

MCS is challenging in ACHD patients with biventricular physiology because of ventricular morphology, residual lesions, systemic venous abnormalities, limited vascular access, previous multiple surgeries (sternotomies, thoracotomies), pulmonary hypertension, aortopulmonary collaterals, coagulopathy, and end-organ dysfunction.

There are limited data on postoperative ECMO in ACHD patients and ECMO survival. The underlying cardiac anatomy, along with the associated comorbidities, complicates the decision to use ECMO in ACHD patients [20]. A study from the Mayo Clinic showed a survival rate at discharge of 46% in high-risk ACHD patients requiring postcardiotomy ECMO support [21].

A systematic review of durable MCS in teenagers and ACHD revealed the frequent utilization of MCS in patients with ACHD. Short-term survival rates in published series are approximately 70%, and the use of durable MCS as a BTT was 77% [22].

In an INTERMACS analysis by VanderPluym et al., 21% of cases were successfully bridged to transplant and 51% were alive with the devices. The survival rate was the same for non-ACHD patients [23].

The SynCardia TAH has now been implanted in over 50 patients <21 years of age and in more than 20 patients with ACHD [24, 25].

The use of the EXCOR heart (Berlin Heart, The Woodlands, TX, USA) is limited in ACHD patients.

VAD to support a failing systemic RV with previous Senning or Mustard operation for TGA or ccTGA as a BTT is growing. Studies have shown that implantation of a VAD in a failing systemic RV can be used as a bridge to heart transplantation [26–28].

Complex ACHD does not contraindicate the use of MCS and should be considered early as a treatment option. INTERMACS scores have trended toward lower acuity, suggesting implantation earlier during advanced HF, similar to those with acquired heart disease.

L-VADs have shown to be promising, with similar survival benefits as in non-ACHD patients. The role of VADs as DT is increasing. The use of VADs to support a failing systemic RV is encouraging. Virtual implantation techniques using cross-sectional imaging may significantly alter device selection in ACHD patients.

10.5 MCS in IN ACHD Patients with Single-Ventricle Physiology

The most important factor predicting the anticipated survival for any single-ventricle patient requiring MCS is the stage of palliation [29]. A patient's current anatomy will affect their response to MCS [30]. These patients typically have altered mediastinal anatomy due to numerous previous sternotomies, are chronically anticoagulated, and have hepatic and renal dysfunction related to long-standing single-ventricle physiology and diastolic dysfunction with preserved systolic function [31, 32].

ECMO support in failing Fontan circulation is challenging and carries high mortality and morbidity rates. A survival to discharge rate of 35% was reported by Rood et al. in Fontan failures supported by venoarterial ECMO [33]. ECMO can be used in an acute setting as a bridge to decision or bridge to recovery. The stage of Fontan failure and patient selection determine the success of ECMO support in Fontan failure. The use of ECMO in advanced stages of Fontan failure is associated with even higher mortality rates due to the associated end-organ damage.

There is growing evidence that VAD support can be successful in selected patients with a failing Fontan circulation [34, 35].

Patients with isolated or predominant ventricular systolic dysfunction are likely to benefit from VAD alone, but such cases among patients with a failing Fontan circulation are rare. A VAD will surely not help the clinical situation if the end-diastolic pressure of the systemic ventricle is not high (at least >12 mmHg).

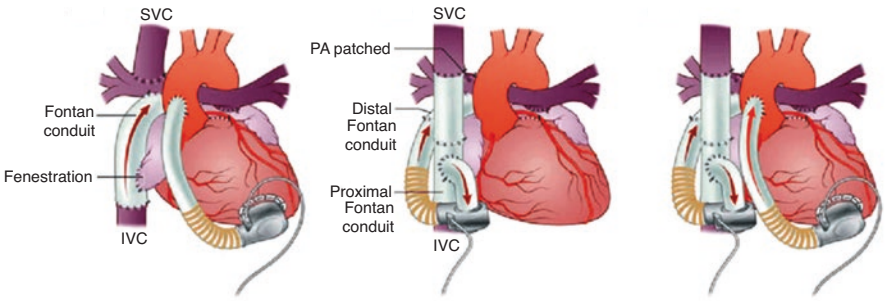

Fig. 10.7 Different configurations for R-VAD on Fontan circulation: Continuous flow (with courtesy by Buratto E)

Fontan failure patients with marked end-organ dysfunction, protein-losing enteropathy, and/or plastic bronchitis who are otherwise not good transplant candidates may benefit from TAH.

Fontan failure may be independent of ventricular function and often is driven by elevated pulmonary vascular resistance or pressure: VAD may or may not help in these patients, and cavo-pulmonary support may be necessary.

There are substantial research efforts into developing MCS for the failing Fontan circulation. Some authors showed that the use of an Impella for cavo-pulmonary assist in the failing classic Fontan lowers the Fontan pressure, attenuates systemic venous congestion, and augments systemic oxygen delivery [36].

Derk et al. showed the feasibility of an axial pump (Jarvik 2000) to restore baseline hemodynamics and cardiac output in Fontan circulation in a pig model [37]. Many other authors have studied possible alternative to use L-VAD as a subpneumonic artificial ventricle (Fig. 10.7).

Improvements in palliation of single-ventricle physiology have given rise to an increase in the number of single-ventricle patients susceptible to HF later in life, which necessitates the use of some form of MCS.

Supporting the failing myocardium in single-ventricle patients shows the feasibility of MCS in patients with a failing single ventricle, but MCS outcomes are better in early stages of failure than in the advanced stage.

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Heart Transplantation: The Challenging Journey of an ACHD

11

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11.1 Congenital Heart Disease and Heart Failure

Congenital heart diseases (CHD) are among the most common inborn defects, with an estimated incidence of about 1 of 100 live newborns.

Cardiac malformations are currently treated at pediatric age, with an overall early surgical mortality inferior to 3–4% [1], while, prior to the advent of surgery, less than 20% of children with congenital heart malformations survived to adult life. Currently, most deaths for CHD occur in grown-up age. In fact, during the last two decades, the extraordinary advances in cardiac surgery, intensive care, and noninvasive diagnosis have drastically modified the natural history of almost all CHDs. Approximately 90% of neonates born with cardiovascular anomalies can expect to reach adulthood, and, with continued improvement in surgical technique, it is going to increase further in the near future. This improvement has selected a growing number of newborns and infants who have survived through adolescence until adulthood. This population constitutes a new medical community that has been named in several ways, such as grown-up congenital heart (GUCH), or adult congenital heart disease (ACHD) patients, that may require lifelong cardiac and noncardiac-specific services [2].

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It is well-known that CHD acts as a systemic pathology that involves several complications as further operations, arrhythmias, complications in other organs, increased risk of heart failure, and premature death if managed inappropriately. Major concern is expressed for patients with complex CHD. These more complex patients are obviously vulnerable to additional acquired comorbidities that impact both their cardiac and medical care, including hypertension, pulmonary, renal, liver, myocardial, and coronary artery disease [3]. It is estimated that about 55% of ACHD population is at medium to high risk (defined as those at significant risk for premature death, reoperation, and complications) and thus needs to be seen regularly in a regional dedicated center and followed lifelong. These patients include those with single ventricle physiology but also transposition of great arteries (TGA) variants, complex Ebstein's anomaly, tetralogy of Fallot, pulmonary vascular disease, and complex septal defects.

11.2 Cardiac Surgery for End-Stage Heart Failure in ACHD

In the long term, the development of HF is common in patients with congenital heart disease, especially those with complex abnormalities and single ventricle physiology. Reduced exercise tolerance and fatigue are prevalent even among those patients who consider themselves asymptomatic. It has been reported that HF develops in about 22% of patients with congenitally corrected TGA, in 32% of patients with TGA after atrial switch procedures, and up to 40% of patients after Fontan palliation.

Currently, transplantation is the treatment of last resort in ACHD with ESHF. This is because ACHD patients present multiple unique surgical and medical challenges related to transplantation owing to their complex anatomy, multiple prior palliative and/or corrective procedures, frequently increased pulmonary vascular resistance derived from longstanding congestive heart failure or cyanosis, liver insufficiency, and overall debilitated conditions. For these reasons, CHD is a significant risk factor for increased 1-year mortality in heart transplant recipients. Despite an increased early mortality after transplant in ACHD patients, recent reports demonstrates that those who survive after the first posttransplant year have an excellent long-term prognosis [4–6].

11.3 Heart Transplant and Listing

Heart transplant is considered the gold standard treatment for ESHF with progressive symptoms despite optimal medical and conventional surgical therapies. However, in the past and now, the number of patients suffering from ESHF and listed for cardiac transplant is greater than donated organs. This is particularly magnified for CHD patients due to their limited number at list [7].

Donors are people who received a diagnosis of brain death. Brain death is a prerequisite for organ donation, and it is defined as the irreversible injury to the whole brain and impairment of brain functions.

There is a second preliminary requirement for the donation: a dead person can be considered as donor if he signed an advance directive of treatment, or his relatives must agree to organ donation.

The process of transplantation begins when the patient is sent to the Cardiology Department by his attending physician because of a worsening of the clinic condition.

If the criteria for transplant have been satisfied, several blood tests are done for blood group analysis, crossmatch, and lymphocytotoxic antibodies.

To stay on the waiting list, patients must undergo quarterly, semiannual, or annual health checks regular medical supervision.

When a heart organ becomes available, the prospective recipient has to reach the hospital fast in order to perform diagnostic tests and to verify the immunological compatibility; if the donor heart is suitable, the transplant begins.

11.4 Transplant Eligibility and Listing

11.4.1 When Is a Heart Transplant Performed?

The waiting time for transplant is very variable according to the medical state of the patient, in fact an "Annual activity report from the UK showed a median waiting time of 1487 days for the non-urgent list and 32 days for the urgent list" [8].

Commonly, heart transplantation is considered when:

- Heart function declines to the point that patient's survival is threatened, despite optimal therapeutic approaches (i.e., drugs of last generation).
- Malignant arrhythmias unresponsive to medical treatment are present.

However, the uniqueness of CHD physiology led to dedicated indications:

- New York Heart Association functional class IV and HF is not amenable to palliative or corrective surgery
- Severe symptomatic cyanotic heart disease not amenable to palliation.
- Post-Fontan procedure with refractory HF, or Fontan's specific complications as persistent protein-losing enteropathy and/or plastic bronchitis despite optimal medical and surgical therapy
- Pulmonary hypertension with the potential risk of developing fixed, irreversible elevation of pulmonary vascular resistance (PVR) that could preclude heart transplantation in the future

Contraindications must be taken into account in judging whether a patient is likely to benefit from heart transplantation since contraindications impact short- and long-term patient and cardiac graft survival.

Absolute Contraindications

- Systemic illness with a life expectancy <2 years despite heart transplantation
- Active substance abuse (alcohol, drug, tobacco)
- Clinically severe symptomatic cerebrovascular disease
- Major psychopathy causing inability to comply with drug therapy
- Irreversible pulmonary vascular hypertension defined as irreversible PVR >3 Wood unit. In these cases, a combined heart-lung transplantation may be considered as an alternative
- Current diagnosis of cancer or history of cancer with inauspicious prognosis
- Multiple extracardiac organ dysfunction.

Relative Contraindications

- Age >70 years old. It is an eligibility criterion because an increased age intensifies the risk of posttransplant death. However, the median HTx recipient age is increasing, and several recent studies show the same morbidity and survival in younger patients compared with older patients.
- Irreversible other single organ dysfunction as renal and hepatic failure. However, a combined transplantation can be considered.
- Frailty, defined by the presence of cachexia, slow walking speed, and decreasing strength, because it increases morbidity and mortality.
- Global severe vasculopathy.
- Obesity. If body mass index (BMI) is elevated, postoperative chances of survival are reduced. In particular, BMI >35 increases risk of complications (i.e., rejection, infections, etc.).
- Pulmonary hypertension (PH): it can cause right ventricular failure and lead to mortality in postoperative phase.
- Psychosocial disorders and addiction. A careful evaluation before HTx is recommended to ensure optimal medication understanding and compliance. As will be explained later in this chapter, transplant patients are forced to take immunosuppressants.
- Severe osteoporosis.
- Severe chronic obstructive pulmonary disease.
- Insulin-dependent diabetes with organ damage.
- HIV infection [9].

Heart transplantation evaluation and operation in ACHD should be performed only at centers with established medical, surgical, and nursing experience in both adult CHD and heart transplantation.

Listing criteria and pretransplantation management have to be tailored to ACHD in order to increase survival to transplantation. The correct timing to referral has to

be considered the main prognostic factor. Listing may occur too late, and it is likely that we need to list patients with CHD sooner than the development of end organs dysfunction or deterioration of health and nutritional status.

Use of mechanical circulatory support (MCS) as a bridge to transplantation (including as a therapy for reversal of high PVR) has been demonstrated to be associated with a higher likelihood of survival on the waiting list for patients without CHD; this subset of patients is already restrictively applied, even if slightly increasing. The unfavorable anatomy and the high risk of bleeding in ACHD patients represent strong limitations in their use as well as the need of further analysis to identify those CHDs amenable to effective mechanical cardiac support [10].

Operatively, transplant checklist provides blood tests (including cell blood count, electrolytes, BNP, renal and hepatic function) and instrumental evaluation for the multidisciplinary assessment, where the nurse plays a crucial role of planning and even performing.

Before the insertion into transplant list, patient and his family are informed about the importance of the immediate willingness to be hospitalized if there was an available heart for transplant.

11.4.2 Crossmatch

The organ allocation is based on patient and donor blood group (AB0) and human leukocyte antigen (HLA) test. HLA typing allows to evaluate "mismatches" (i.e., the HLA diversity between donor and recipient) which might be recognized by donor, causing reject.

- *Positive crossmatch*➔ patient and donor are immunologically incompatible: patient is discharged.
- *Negative crossmatch*➔ there is immunological compatibility between patient and donor: the transplant starts.

11.5 Heart Transplant Surgery: Nursing Management

11.5.1 Preoperative Phase

In the preoperative phase, nurse must evaluate patient's psychological state involving him and his family or caregiver in the therapeutic project. They have to be informed about risks and benefits of the surgery and potential difficulties encountered during rehabilitation.

The rehabilitation treatment begins with patient admission: nurse educates patients to correctly perform pulmonary rehabilitation exercises, active mobilization of extremities, and pain control techniques.

11.5.2 Heart Transplant

If the donor heart is suitable, recipient is transferred to the operating room. In order to minimize the duration of cardiopulmonary bypass and the graft ischemic time, the team has to synchronize organ retrieval and graft implantation. The donated heart is storage cooled (+4 °C) to keep it vital for 4–5 h [11].

Briefly, in general anesthesia and mechanical ventilation, after median sternotomy, cardiopulmonary bypass is established through ascending aorta and bicaval cannulation. Then, cardiotomy is performed and the donor graft sutured orthotopically.

Recipient cardiectomy is performed at the same time the donor heart arrives in the operating room: the great vessels are divided; the superior vena cava is transected at the cavo-atrial junction; a cuff of inferior vena cava is prepared (alternatively a right atrial cuff is prepared with the orifices of the two vena cavas); and the left atrial cuff is prepared leaving a large tissue with the orifices of the four pulmonary veins.

Orthotopic heart transplantation (Fig. 11.1) can be performed by a bicaval anastomosis technique or by biatrial anastomosis technique.

Surgical sequences for the bicaval anastomosis technique:

1. Left atrial anastomosis.
2. Superior and inferior vena cava anastomosis.
3. Pulmonary artery and aortic anastomosis [12].

Surgical sequences for the biatrial anastomosis technique:

1. Left atrial anastomosis.
2. Right atrial anastomosis.
3. Pulmonary artery and aortic anastomosis.

After implantation, inotropic and chronotropic drugs (such as dopamine, epinephrine, norepinephrine, and isoprenaline) are frequently required for stabilizing the patient's hemodynamic.

In the operative room, there are two nurses with different roles: the circulating nurse and the ant scrub nurse.

Circulating nurse's duties are:

- Evaluation and management of anxiety and pain before the sedation.
- Placing peripheral venous and urinary catheter.
- Helping the anesthetist during intubation and vascular accesses.
- Managing the external defibrillator, because especially during sternotomy, there may be ventricular dysrhythmia.
- Ensuring that vital signs and invasive measurements are always accurately reported to transport monitor (data are not always reliable). Data must be interpreted and linked to the patient's status. Monitoring allows to understand patient's

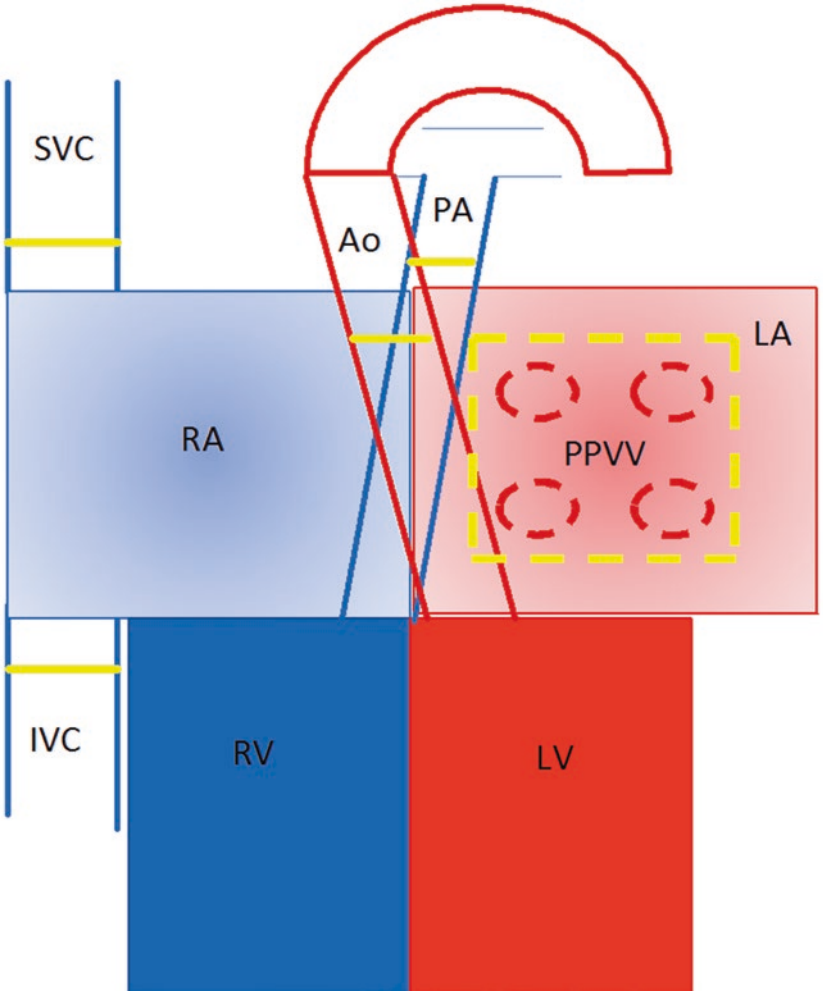

Fig. 11.1 Cartoon showing the orthotopic heart transplant bicaval technique. Legend: Ao aorta, *IVC* inferior vena cava, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *PPVV* pulmonary veins, *RA* right atrium, *SVC* superior vena cava

clinical condition, early identify pathological status, and check the efficacy of medical treatment.
- Monitoring the patient's core temperature.
- Ensuring that sterile procedures are followed during surgery.
- Ensuring that necessary equipment and medications are available for the operating room team.

Another important role for the nurse in the operative room is the "scrub nurse" specialized in cardiac surgery. In this case, the nurse is an expert in operative

nursing and works closely with surgeons and other surgical staff to ensure a smooth procedure, minimize complications, and continuously assist with maintaining a sterile field. They have to know and follow the surgeon in every single step of the operation and be organized for the surgery and possible complications, anticipating or solving them and plan for the next step. They have the important role in preparing the operating table and the surgical instruments for the operation.

As transplant can be performed 24/24 h, 7 days a week, they work both during the day shift during the typical work week and "on call" during the evenings, weekends, holidays, and nights [13].

11.6 Postoperative Phase in the Intensive Care Unit

After leaving the operation room, the patient is transferred in the intensive care unit (ICU), still sedated and intubated, with several invasive lines to be continuously monitored during the following period. In the ICU, hemodynamic parameters have to be monitored: invasive blood pressure (IBP), central venous pressure (CVP), pulmonary artery pressures (PAP), if needed, cardiac rhythm, urine output, central temperature, oxygen saturation, hemogasanalysis evaluation, and chest tube output.

Neurological and cardiovascular assessments are mandatory.

Neurological status: Ramsay Sedation Scale to monitor level of sedation; responsiveness to pain and/or to the call, and pupils' observation (miosis, mydriasis, anisocoria, reaction to light).

Cardiovascular function: rhythm, heart rate (100–120 bpm), cardiac output, and urine output (achieving an adequate renal perfusion).

The monitoring is based on:

- *Neurological status* at waking up (i.e., mental confusion, orientation).
- *Cardiac rhythm*. ECG in continuous to understand the heart electrical activity. Placing three electrodes (the yellow one to the left shoulder, the red one to right shoulder, and the black/green to the lower part of chest). Electrodes shall not interfere with the possible placement of paddles to shock. Epicardiac pacing wires (atrial and ventricular) are usually positioned and connected to an external pacemaker. The identification of rhythm disturbances (i.e., atrial fibrillation, ventricular fibrillation, ventricular runs, asystole, pulseless of V-tach) needs to be treated.
- *Invasive blood pressure* (IBP). The monitoring system registers systolic pressure, diastolic pressure, and mean blood pressure. It is of paramount importance controlling the functionality of continuous arterial infusion (extremely slow) which affords to maintain the patency of the artery. The nurse must prepare the circuit to monitor IBP by washing and filling it with saline solution; then the nurse connects the transducer to the arterial line. The circuit must be free of air bubbles because they can cause ictus and cardiac arrest. To zero the transducer, it is necessary to put it in contact with ambient air, and this procedure allows the team to have a referral point to monitor IBP. Then, stop the communication between ambient air and transducer. The transducer must be placed at the heart level.

- *Arterial oxygen saturation*. Oximeter sensor is placed peripherally (such as on a fingertip). Saturation usually needs to be maintained >96%.
- *Cardiac output* (CO). Monitored indirectly through arterial pressure and urinary output.
- *Central venous pressure* (CVP): central venous catheter allows to measure the central venous pressure and to have available, reliable, and secure venous access for drugs, solutions, or total parental nutrition. CVP is monitored, thanks to a second transducer, using the same circuit used to monitor IBP. Normal range of CVP values: *4–8 mmHg.* It is influenced by respiratory function; in fact, CVP may decrease with spontaneous inspiration, while it may increase with mechanical positive pressure ventilation.
  - *CVP increases* in case of hypervolemia, cardiac tamponade, high pulmonary resistance or artery pressure, right heart failure.
  - *CVP decreases* in case of hypovolemia or venous vasodilatation.
- *Core temperature* (normothermia: 36 C°).
- *Quantity and quality of urine output*, monitored through the urinary catheter.
- *Chest tube output*: usually in aspiration, it is useful for the evaluation of postoperative bleeding that should be caused by surgical complications requiring surgical revision or discoagulopathy requiring blood derivates as fibrinogen or plasma.
- *Hydroelectrolytic balance*: it is fundamental maintaining euvolemia, avoiding hypovolemia and hypervolemia.
- *Blood gases analysis*: to assess the metabolic and respiratory status, electrolyte, and hemoglobin. Performing arterial blood gas analysis every 3 h or when needed in case of hemodynamic compromise or complicated postoperative course.

Respiratory function: arterial blood gas analysis is the gold standard to evaluate the partial pressure of oxygen (PaO_2); it also provides information about partial pressure of carbon dioxide ($PaCO_2$), pH, bicarbonates, electrolytes, hemoglobin, glucose, and lactate.

Normal range of blood gas analysis:

- *pH: 7.35–7.45,*
- *PaO_2: > 60 mmHg.*
- *$PaCO_2$: 35–45 mmHg.*
- *HCO_3^-: 22–26 mEq/L.*
- *SpO_2: 95%.*

PaO_2 value decreases in case of pneumonia, parenchymal lung disease, hypoventilation, atelectasis, and pulmonary edema.

The monitoring is based on:

- Evaluating the correct tube placement and its fixing.
- Evaluating the alteration of acid-base balance and the respiratory gas exchanges through pulse oximetry or blood gas analysis; both provide information about oxygen saturation. The first method is noninvasive because pulse oximetry is monitored

by placing the oximeter's sensors on the fingertip [14]. The value obtained by pulse oximeter is not always reliable, because of hypotension, hypoperfusion, and hypothermia, and other factors can influence the peripheral perfusion.
- Bronchoaspiration when needed (i.e., worsening tachypnea and oxygen saturation, cyanosis).
- Checking ventilator function, avoiding hypercarbia, hypoxia, and acidosis. Acidosis increases high inspiratory pressure and auto-PEEP which increases the venous return; it also makes the ventricular dysfunction and pulmonary arterial pressure worse.

After extubating, due to pulmonary rehabilitation, recipients could achieve a proper restoring of lung ventilation; the use of respiratory aids, the control of bed posture, and respiratory exercises allow pulmonary rehabilitation.

In addition, basing on a holistic evaluation of the patient's health, other systems and organs are assessed:

Renal function: usually renal dysfunction is secondary to HF because of the correlation between renal perfusion and blood volume and blood pressure. In this setting, surgery can worsen renal damage. Quantity and quality of urine output and electrolytes (Na^+, Cl^-, Ca^+, P^+) are fundamentals [15].

Infectious prophylaxis: antibiotics are administrated because of the risk of opportunistic infections secondary to induced immunosuppression.

Gastrointestinal system: perioperative nutrition must reduce immunological, cognitive, and muscle complications after surgery ensuring protein intake. The recovery of overall bodily functions improves postoperative recovery [16].

Initially, parenteral nutrition (PN) is an effective approach when oral or enteral nutrition (EN) would be unsafe or insufficient [17].

Before administrating EN, monitoring gastric stasis is mandatory because of paralytic ileus, "'second-line' complications" [18].

Orally nutrition is resumed as soon as the gastrointestinal system works properly, monitoring gastric stasis (if it is >100–200 ml, the meal should be delayed) and removing the nasogastric tube on medical advice.

Mobility: sternotomy, intubation, and anesthetics drugs compromise mobility. Patient must progressively reach the sitting and standing position; the next goals will be to stand and walk (with or without mobility aids) and to become as soon as possible autonomous.

Integumentary system:

- Taking care of surgical wound.
- Evaluating and reducing the risk of developing pressure ulcers.
- Preventing dehydration.

Aims in postoperative phase:

- Early extubating.
- Improving graft function and preventing ischemia through inotropic and chronotropic support.
- Achieving a balance between preventing rejection and preventing infection.

In cardiac ICU, patients are supported to maintain blood pressure (thanks to intravenous infusion, such as fluid and volume replacement and inotropic agents) and to avoid immune system response against new organ, thanks to antirejection therapy. Due to median sternotomy, patient could feel pain which is reducible with analgesic drugs.

Standard precautions for infection prevention and prophylaxis must be followed due to infection predisposition in these immune-suppressed patients (see in the Ward Unit section).

11.6.1 Vasoactive Medications

The administration of inotropes allows to optimize heart function, in particular ventricular preload, contractility, and afterload.

In case of vasodilatory shock, vasopressors (norepinephrine, phenylephrine) are administrated to maintain the optimal mean arterial pressure.

11.6.2 Nitric Oxide

Nurses have the role of managing the inhaled nitric oxide (iNO) device. Inhaled nitric oxide is a pulmonary vasodilator which allows to relax vascular smooth muscle; it is used to reduce the right ventricular afterload. NO reduces pulmonary artery pressure and increases cardiac output [19].

11.6.3 Extracorporeal Membrane Oxygenation

Some transplanted patients need to be hemodynamically supported by the extracorporeal membrane oxygenation (ECMO). ECMO in a venoarterial configuration consists in a cardiopulmonary support where deoxygenated blood is drained through a venous cannula to an oxygenator for gas exchange and then pumped into the arterial system through another cannula.

ECMO support can be established in operative room when weaning off cardiopulmonary bypass is not feasible (postcardiotomy support) or in ICU due to hemodynamic instability with not or few responses to maximal inotropic support.

For the nurse, ECMO management includes the evaluation of cannulas insertion sites, taking care of their correct positioning, and monitoring of its correct functioning. The anticoagulation therapy is fundamental for its functioning, but on the other hand, it can cause coagulopathy, embolism, thrombosis, or hemorrhage [20].

11.7 Postoperative Phase in Transplant Unit Ward

When homeostasis and hemodynamic stability are achieved, patient is extubated (6–24 h after surgery), weaned off inotrope support, and transferred to the dedicated ward, called transplant unit. The role of this ward is to achieve psycho-functional health of the patient continuing to monitor multiorgan correct functioning.

The inpatient unit should have single-bed rooms with isolation capability. These rooms must comply with the standard of safety and comfort of patients in a tertiary care hospital facility. Medical and nursing staff coverage should be available 24 h a day, including holidays. Emergency cart with drugs for resuscitation should be always available [21].

The transplant patient is subjected to IVP and CVP monitoring in the first days; then if not needed, vascular catheters are removed.

Chest tubes are removed when drainage is minimal, and there is no pneumothorax risk, after medical evaluation.

Blood tests should be performed three times a week:

- Hemato-chemical and urine tests to check hemachrome, renal and liver functioning, electrolytes levels, and possible drug toxicity.
- Microbiological tests to evaluate possible infections and to assess activation or reactivation of viruses (as Epstein-Barr virus and cytomegalovirus) or parasites (Toxoplasma gondii).
- Hematological levels of immunosuppressors.

In this period, rejection monitoring is performed including both noninvasive and invasive procedures.

Noninvasive evaluations include:

- Clinical visits excluding signs of heart failure.
- Chest X-ray to avoid pleural effusion.
- EKG to evaluate voltages and rhythm disturbances.
- Transthoracic echocardiography to evaluate biventricular functioning, valvular performances, and pressures.

The gold-standard procedure to monitor acute rejection is the *endomyocardial biopsy* (*EMB*), that is, an invasive procedure. It consists in taking a sample on right-sided myocardial interventricular septum through jugular or femoral vein for a histological and microscopical evaluation (Figs. 11.2, 11.3, and 11.4). Rejection monitoring calendar includes one EMB per week during the first post-HTx month, then one EMB every 2 weeks during the second and the third post-HTx months, and one EMB per month for the first post-HTx year.

The nurse must monitor vitals (neurological status, blood pressure and volume, pulse rate, respiratory function) and can be the scrub nurse during EMB procedures.

In case of signs of moderate or severe rejection, high doses of corticosteroids are necessary.

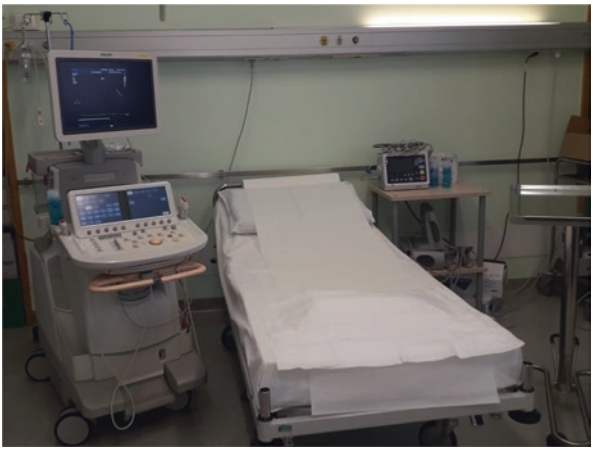

Fig. 11.2 Picture showing the setting for performing the 2D-echocardiographic guided endomyocardial biopsy

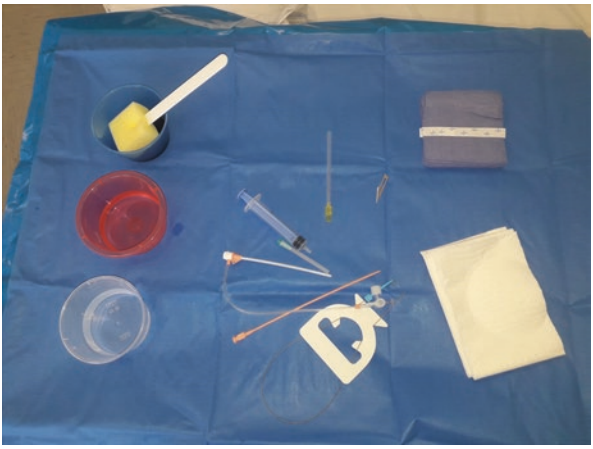

Fig. 11.3 Picture showing the kit for disinfection and introduction (internal jugular vein access) for the following endomyocardial biopsy

Fig. 11.4 Picture showing the biotome which is used to achieve the endomyocardial biopsy

11.8 Complications during Hospital Stay

In the heart-transplanted population, the mortality risk is very high during the post-transplantation first year. The main causes of morbidity and mortality are:

- *Rejection*: immune system attacks foreign tissue (transplanted heart) because of considering an enemy (host vs. graft disease). Sometimes, rejection episodes are without symptoms. The treatment, based on corticosteroids, depends on grading, hemodynamic, severity of rejections, symptoms, and timing post-HT. Rejection is classified as:
  - Hyperacute: it usually leads to death because of anti-HLA class I antibodies against the donor. This reaction has almost eliminated thanks to routine performance of donor-recipient cross-matching at the time of transplantation.
  - Acute cellular: nowadays, it remains common (especially in the first 3–6 months post-HTx).
  - Antibody-mediated: it is less common, but it causes graft dysfunction as well.

    What to do: monitoring of hypotension, tachycardia, fever, vomiting, and decreased urine output is present. It is very important to educate patients about early identification of rejection in order to act quickly before serious heart damage.
- *Infections*: due to immunosuppressive therapy, patients are extremely weak and prone to infection, caused by pathogens, especially multidrug-resistant organisms (MDROs), which are very difficult to treat because of the lack of effective medication.

  What to do: avoiding the contamination between different patients through standard precautions that include hand hygiene (using soap and water or alcohol-based solutions) and wearing of appropriate protective equipment (gloves, surgical mask or eye/face protection, gowns) during procedures and activities. If patients are potentially or definitely infected, nurses have to use additional precautions (such as isolation).
- *Hemodynamic complications.*
  - *Bleeding*: less common than in ICU. It occurs with drowsiness, bradypnea, bloodless or blood-abundant drains, tachycardia, hypotension, cyanosis, and low hemoglobin. It is caused by surgical complications.

    What to do: monitoring heart rate, blood pressure, drains, hemoglobin value (normally hemoglobin is >12 g/dl, but it might decrease because of blood loss during surgery). It requires administrating blood components and, less common, surgical revision.
  - *Hemodynamic changes* (cardiac tamponade, cardiogenic shock) *and vasoplegia* (hypotension and low systemic vascular resistance); these consequences are due to heart altered contractility or conductivity.

    What to do: monitoring tachycardia, excessive blood pressure lowering, cold and cyanotic extremities, metabolic acidosis, and confusion of mind. It could require surgical revision.

  - *Arrhythmias* (severe atrial fibrillation, ventricular fibrillation, ventricular runs, asystole, pulseless of V-tach, etc.): it is caused by hydro electrolyte imbalance or abnormality of the electrical conduction of the heart.

    What to do: monitoring heart rate, re-equilibrating electrolytes (magnesium, potassium, calcium, etc.), placing the defibrillator near the patient's bed, anti-arrhythmic drugs, and pacemaker implantation.
- *Respiratory failure*: it could be related to sternotomy, accumulation of mucus in airways and bronchi, immobility, difficulty and pain in expectoration or ineffective expectoration, and respiratory tract infection.
- *What to do*: monitoring blood saturation, respiratory rate (it may manifest with tachypnea), and mental alertness and identifying early respiratory acidosis through arterial blood gas (A.B.G.) measurement. Secretion aspirations and performing postural drainage are recommended.
- *Wound dehiscence*: wound of sternotomy would be facing dehiscence (reopening of the wound) because of infection, ischemia, diabetes, cortisone, obesity, and malnutrition.

  What to do: checking peri-wound skin, cleaning it, and removing excess moisture; checking the suture and the wound and identifying signs of local (pain, redness, heat, and swelling) and systemic (fever, chills, discomfort, hypotension, tachycardia, decreased urine output) infection; change the bandage according to protocols and reducing bioburden and providing protein diet.
- *Pain*: a bad pain control slows down the healing. A good pain control is necessary through painkillers and complementary techniques (controlled breathing, desensitization, etc.). Assessing pain through specific scale (visual analogue scale (VAS), numeric pain rating scale (NRS), and critical-care pain observation tool (CPOT) are recommended.
- *Hydro electrolyte imbalance, acute kidney injury, and anemia*: caused by arterial hypotension and venous congestion. The risk is increased by diabetes mellitus and BMI >40.
- *Asthenia*: it is caused by surgery, hypermetabolism, and fasting.
- What to do: providing protein diet and promoting the progressive recovery.
- *Deep-vein thrombosis* due to complete immobility. Nurses have to encourage patient mobility and independence to administrate heparin or to help patient to wear anti-thrombus elastic stockings.
- *Constipation*: due to surgery, immobility, drug therapy, and fasting. It is necessary to encourage the patient to eat (preferring a diet high in protein and calories) and drink water once gastrointestinal function is established. Following the indications about the proper management of the nasogastric tube in order to avoid suction and pulmonary infections and obstruction.
- *Psychosocial depression, family caregiver burden, noncompliance associated with drug side effects.* The creation of mutual aid and personalized interview with nurses and doctors might be useful.
- *Diabetes*: the administration of cortisone increases blood sugar.
- *What to do*: monitoring plasma glucose level (fasting <100 mg/dl), introducing foods that reduce blood sugar (vegetables, cereal high in fiber, etc.), and limiting high sugar-containing foods.

- *Education and counseling*: during the inpatient ward, they are two essential planned interactive learning processes designed to support and enable patient to manage their life with their new graft and the immune suppressive therapy and optimize their health and well-being. They are fundamentals for all the posttransplantation management after discharge.

11.9 Clinical and Home Rehabilitation

11.9.1 Posttransplantation Management

The return home usually occurs if hemodynamic and general health conditions permit it. Heart-transplanted recipients can come home only if they reacquire their previous mobility and self-sufficiency.

Before the return home, it is essential to plan a route of secondary prevention based on education to new lifestyles' physical and psychological rehabilitation because HTx leads to metabolic and physiological consequences.

Moderately intense physical activity can improve heart function and blood pressure. Thanks to long-term endurance training, parasympathetic activity is increased, and sympathetic activity is decreased in the human heart at rest.

Transplanted patients return home not earlier than 15 days, and they have to pay more attention on food (avoiding raw foods) and on lifestyle (they have to avoid crowded places and to meet sick people). Length of hospitalization depends on various factors, including the patient's clinical response but also his education and learning of new lifestyles.

Patient's duties after discharge:

- Checking the patient's weight daily.
- Patient and his caregiver are instructed on blood pressure monitoring.
- Regularly taking immunosuppressive drugs.
- Paying attention to feed, drug therapy, and social life (avoiding crowded places and ill people).

Immunosuppression. Maintenance immunosuppression is the necessary prerequisite for avoiding cardiac rejection; in fact, following organ transplant, patients' immune system will try to reject the new organ. Patients take different kinds of medications: calcineurin inhibitor drugs – CNIs (mainstay of immunotherapy) as cyclosporine and tacrolimus, antimetabolites (drug that inhibits enzymatic reaction in order to interfere with metabolic pathways) as mycophenolic acid or azathioprine, and corticosteroids. Antirejection treatment is based on these three classes of drugs: CNI + antimetabolite + corticosteroid.

Patient is called to submit to routine surveillance check: he goes back to the hospital to be submitted to clinical visit, endomyocardial biopsy, and blood tests during the first 12 months post-HTx. Endomyocardial biopsy is the gold standard for rejection diagnosis.

Goals of Out-of-Office Monitoring:

- Self-sufficiency.
- Improving the compliance with drug treatment.
- Showing the consequences of immunosuppressive therapy.
- Making the patient able to recognize flu and symptoms of rejection.
- Motor and respiratory rehabilitation.
- Explain the need for follow-up.

11.9.2 Follow-Up

During follow-up, the team evaluates patient's clinical status through blood tests and physical examination.

Instrumental evaluation as EKG, transthoracic echocardiography, and chest X-ray will be taken to understand the performance of the transplanted heart.

In addition, after the first year of HTx and later then every 2 years, it is necessary to assess coronary disease (cardiac allograft vasculopathy – CAV) to avoid or treat chronic rejection through coronary angiography or indirect procedures as echo-stress or scintigraphy.

Side Effects of Antirejection Therapy

- *Infection and pneumonia*: it can lead to death because the patient is immunocompromised. Infections are caused by bacterial agents (*Listeria* and *Nocardia*), viral agents (*Cytomegalovirus*, Epstein-Barr) and others (fungi and parasites) [22].
- *Osteoporosis.*
- *Nephrotoxicity and abnormal liver function.*
- *Systemic hypertension*: if uncontrolled and long-lasting, it can damage heart and blood vessels. Nurse has to educate patient about self-measurement of blood pressure and the recognition of symptoms of hypertension (headache, stunning, blurred vision).
- *Bone-marrow suppression*: monitoring the levels of CBCs, RBCs, and blood platelets. Erythropoietin may be administrated in case of anemia.
- *Cancer.*
- *Atherosclerosis and diabetes.*
- *Death.* Mortality and morbidity are higher in ACHD patients than in patient who does not suffer from CHD [23].

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Part V

Adult Congenital Heart Disease and Pregnancy

Congenital Heart Disease During Pregnancy

12

Silvia Favilli and Gaia Spaziani

12.1 Introduction

Although maternal mortality has substantially decreased in the last decades in western countries (12/1,000,000 live births following the WHO report), it remains 20-fold higher in low-income countries (230/1,000,000), and cardiovascular diseases are the main cause of non-direct mortality. CHD is the most common condition encountered in women with preexisting heart disease, and numbers are expected to rise due to the increased survival until the adulthood of neonates born with even complex CHD. The risk of complications is particularly high in women with uncorrected or palliated CHD [1].

Maternal age, obesity, and arterial hypertension are additional risk factors for maternal morbidity and mortality. Most CHD may accommodate to hemodynamic changes imposed by pregnancy; however, cardiovascular complications may occur, especially in patients with moderate to complex CHD, requiring a prepregnancy careful assessment of risk and structured management during gestation, delivery, and postpartum. Pregnancy is still contraindicated in a small number of women because of the excess of maternal and fetal mortality and morbidity.

The need for a multidisciplinary, well-defined approach to pregnancy by a dedicated cardio-obstetrics team has been underlined by recent international Guidelines [2–4]. Ideally, the team should evaluate patients before pregnancy, providing them targeted counselling, optimizing if needed cardiac status, and planning the management of gestation and delivery. The essential role of a specialized nurse is also highlighted.

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Due to the great heterogeneity of CHD population, assessment of pregnancy-related risk and individualized counselling should begin in early adolescence and be part of the "transition" program of care [5].

12.2 Hemodynamic Changes During Pregnancy and Delivery

Complex hemodynamic changes occur during pregnancy, delivery, and postpartum. Cardiac output (CO) increases during pregnancy (to 30%–50%) until the 32nd week of gestation. This is mainly driven by an increase in plasma volume (due to the activation of the renin-angiotensin-aldosterone by progesterone and estrogens), associated with less red cell mass growth, leading to hemodilution and physiological anemia of pregnancy. In the later pregnancy, compression of the inferior vena cava by the pregnant uterus may occur in the supine position, reducing venous return and affecting CO; when needed, a lateral left-sided position is therefore recommended. Systemic vascular resistance falls 30–40% during the second trimester. Heart rate increases by 10–20 beats per minute in the second part of pregnancy, reaching a maximum in the third trimester. During labor and delivery, heart rate further increases *due to effort and pain*, together with the release into the general circulation of an amount of 300–500 ml of blood at every uterine contraction. In the postpartum period, the increase of CO is mostly driven by the autotransfusion from the uteroplacental district. This additional blood volume is usually balanced by a similar (300–500 ml) blood loss during delivery. However, the volume overload may adversely affect the heart in women with underlying CHD when this loss is small. The increased diuresis contributes to intravascular volume normalization in the first 24–48 h after delivery, and the return to prepregnancy values may require several weeks.

12.3 Assessment of Maternal and Fetal Risk and Prediction of Outcome

Several score systems have been developed to assess maternal pregnancy-related risks. Among them, the modified World Health Organization (mWHO) score system, which is based on the type of the underlying cardiac condition, is actually considered the best tool for the initial risk stratification (Table 12.1). The risk assessment may be further improved/refined using other score models: CARPREG (1 e 2) and ZAHARA, the last of which was specifically defined for CHD. These models include other parameters in addition to the type of cardiac lesion (e.g., heart failure, cyanosis, cardiac medications, history of arrhythmias, etc.); however, their capability to predict pregnancy outcome is dependent on the type of populations on which they are used [6, 7].

However, in clinical practice, the predictive capability of mWHO classification exceeds that of the other stratification scores [8–10].

Special programs of care are needed in high-risk conditions.

Table 12.1 Modified WHO Classification of maternal pregnancy-related risk (Modified from Regitz Zagrosek et al. 2018)

mWHO class I	mWHO class II	mWHO class II-III	mWHO class III	mWHO class IV
Mild pulmonary stenosis	Unoperated ASD or VSD	Mild LV systolic dysfunction (EF > 45%)	Moderate LV systolic dysfunction (EF 30–45%)	Pulmonary arterial hypertension
Small PDA	Repaired ToF	HCM	Previous PPC (without residual ventricular impairment)	Previous PPC (with any residual ventricular impairment)
Mild MVP	Turner syndrome (without aortic dilatation)	Mild mitral stenosis, moderate aortic stenosis	Mechanical valves	Severe systemic ventricular dysfunction or NYHA class III-IV
"Simple" defects (ASD, VSD, PDA) after successful correction	Supraventricular arrhythmias	Marfan syndrome or other HTAD without aortic dilatation	Systemic RV with good or mildly decreased systolic function	Systemic RV with moderate or severe ventricular dysfunction
Anomalous venous drainage after successful correction		Bicuspid aortic valve (BAV) with aortic diameter < 45 mm	Fontan circulation (if uncomplicated)	Fontan circulation (with any complication)
Isolated supraventricular or ventricular beats		Repaired aortic coarctation	Unrepaired cyanotic CHD Any complex CHD	Severe re-coarctation
		Atrioventricular septal defect	Ventricular tachycardia	Vascular Ehlers Danlos
			Moderate mitral stenosis, severe aortic stenosis (if asymptomatic)	Severe symptomatic aortic stenosis, severe mitral stenosis
			Moderated aortic dilatation (40–45 mm in Marfan or other HTAD; 45–50 mm in BAV	Severe aortic dilatation (>45 mm in Marfan/other HTAD; >50 in BAV)

PDA patent ductus arteriosus, *MVP* mitral valve prolapse, *ASD* atrial septal defect, *VSD* ventricular septal defect, *ToF* Tetralogy of Fallot, *LV* left ventricle, *HCM* hypertrophic cardiomyopathy; *HTAD* hereditary toracic aortic disease, *PPC* peripartum cardiomyopathy, *RV* right ventricle, *CHD* congenital heart disease

12.3.1 Pulmonary Hypertension

PH secondary or associated with CHD recognizes multiple causes, and prognosis varies depending on the underlying subset. Pregnancy is still considered at high risk, particularly in patients with pulmonary arterial hypertension (PAH) and Eisenmenger syndrome, in whom maternal mortality remains prohibitive in the modern era, despite medical progress and new targeted therapies [11]. The highest risk occurs in the postpartum, which accounts for 2/3 of maternal deaths due to abrupt hemodynamic changes. Therefore, careful monitoring of clinical and hemodynamic parameters is mandatory, at least for the first week postpartum, and a close follow-up should be continued for the first months after delivery.

With the improvement in care and the availability of new targeted drugs for PAH, maternal mortality has decreased from nearly 40% in the late 1990s to 12% in the current years. Endothelin inhibitors must be discontinued due to fetal teratogenic effects, but both phosphodiesterase-5 inhibitors and intravenous prostanoids are commonly employed during pregnancy and may probably improve the final outcome. Although maternal mortality seems related to the severity of PAH, there is no "safe" threshold for pregnancy. Therefore, women with CHD-related PAH should be advised against pregnancy and should be offered termination in case of pregnancy. Besides maternal mortality and morbidity, also fetal risks should be taken into account. There is a significant proportion of preterm births (58% in the current years) and fetal loss. In Eisenmenger patients, fetal outcome is related to cyanosis too, and fetal prognosis is poor when maternal oxygen saturation is less than 85% [12].

12.3.2 Fontan Circulation (Total Cavopulmonary Connection)

Since 1968, Fontan operation (in current years, total venous pulmonary connection) represents the surgical strategy to palliate single ventricle CHD. The surgical treatment places the systemic and pulmonary circulation in series, with the single ventricle pumping into the aorta. As pulmonary flow is passive, driven by venous return, low pulmonary resistances are an essential prerequisite; the hallmark of the Fontan circulation is increased venous pressure and low cardiac output. If a fenestration of the circuit is provided, there is a relief of the systemic venous hypertension and an increase of the CO, but at the expense of a decreased oxygen saturation.

Women with Fontan circulation may not accommodate the significant hemodynamic changes which occur during pregnancy; particularly, the inability to increase CO and decrease pulmonary vascular resistance may lead to heart failure (HF) [13]. Therefore, this condition is considered at high risk in case of pregnancy, and a preconception counselling and careful prepregnancy assessment are required [14]. Women should be aware also of the high fetal risks, with a nearly 50% risk of miscarriage and 60% of preterm deliveries.

Following the mWHO score system, women with Fontan circulation are classified in class III. However, the ACOG bulletin [15] places only women with

uncomplicated Fontan in class III, while in case of severe complications, they should be considered class IV; although the bulletin does not specify which complications contraindicate pregnancy, women with severe ventricular dysfunction, protein-losing enteropathy or plastic bronchitis, and complex arrhythmias or advances liver disease should be definitely advised against pregnancy.

Due to the high maternal and fetal risks, pregnancy in women with Fontan circulation should be followed by an experienced multidisciplinary team in a tertiary level center [16]. Careful monitoring of hemodynamic conditions and oxygen saturation is required; in patients with a fenestration, a lower oxygen saturation during pregnancy is usually secondary to decreasing systemic resistances. Reduction of physical activity is usually recommended in the later stages of pregnancy. Anticipated delivery of gestation (after 36 weeks of gestation) should be provided in case of mother decompensation.

Hemorrhage is a frequent complication, occurring in 25% of patients, both before and postpartum. The increased risk of thromboembolism in pregnant women with Fontan should be therefore balanced with the risk of hemorrhage. Usually, low-molecular-weight heparin (LMWH) at the therapeutic dose is recommended in patients at higher thromboembolic risk, while aspirin or low-dose LMWH is preferred in the other cases [16].

12.3.3 Hereditary Aortopathies

Hereditary aortopathies represent a large spectrum of diseases in which pregnancy may be at risk. This is especially true for women with Marfan syndrome (MS), Loeys-Dietz syndrome (LDS), or vascular Ehlers-Danlos syndrome (EDS); aortic dissection is the most threatening complication, but in some cases also, bicuspid aortic valve (BAV) syndrome may be at increased concern when associated with marked dilation of the ascending aorta. Prophylactic surgery should be considered when aortic dilation exceeds thresholds defined for different conditions.

In a recent national study [17], aortic dissection represented one of the main cardiovascular causes of maternal death. Most dissections occurred in the third trimester (40%) and postpartum (35%); relevant prepregnancy morbidity (connective disease) was present in 40% of women.

Pregnancy leads to aortic structural and functional changes: increase of the size, secondary to hemodynamic changes (an increase of circulating volume and CO), and weakening of the vessel wall (due to reticulin fragmentation and loss of elastic fibers organization) [18]. These changes are based on complications during gestation and delivery. No maternal deaths were reported in a recent series from the multicenter Registry ROPAC, but 2% of women presented an acute aortic dissection during pregnancy [19].

Following the European Guidelines [3] pregnancy should be discouraged in women with MS when the aortic diameter exceeds 45 mm. However, the aortic dimension is not the unique parameter to be taken into account, such as a family history of dissection, progression of aortic dilation, and gene mutation may affect

prognosis. In general, in female patients with aortic dilation, a pathogenic variant is related to a worse prognosis. Women with EDS, who present a more aggressive vascular course and in whom a 12% pregnancy-related mortality is reported due to arterial or uterine rupture, are advised against pregnancy even in the absence of aortic dilation. In the case of pregnancy, delivery should not be delayed after 36–38 weeks of gestation.

The risk of dissection seems as high as 100-fold in women with Turner syndrome (TS). Pregnancies are rare because of frequent infertility but may sometimes occur in women with mosaic TS.

In all women with aortopathy, careful management during pregnancy is recommended, and periodic consultations are advised to assess aortic diameter; beta-blockers therapy may be used in selected cases, while surgery should be considered only in case of rapid progression of aortic dilation, exceeding 50 mm. Strict control of blood pressure is advised to prevent values exceeding 140/90 mmHg. The progression of aortic diameter after pregnancy is not completely defined and requires a regular postpartum follow-up.

12.3.4 Congenital Heart Diseases with Systemic Right Ventricle

CHD with systemic right ventricle (SRV) includes congenitally corrected transposition of great arteries (CCTGA) and transposition of great arteries (TGA) after atrial switch procedure. In both conditions, the right ventricle acts as a systemic ventricle, and there is concern about its ability to tolerate hemodynamic changes during pregnancy.

Although data from the literature are scarce, a favorable outcome of pregnancy in women with SRV has been reported by a recent prospective study performed on the basis of the ROPAC Registry [20]. Nevertheless, nearly one-tenth of women enrolled in the study required hospitalization for HF, more often in the second and third trimester or within 6 months after delivery. Prepregnancy clinical signs of HF or right ventricle systolic dysfunction (Ejection Fraction-EF- < 40%) were the main predictors of adverse maternal outcome. Prognosis was poorer in women with TGA and atrial switch than in women with CCTGA.

Hypertensive disorders during pregnancy may also occur in patients with SRV, requiring a careful assessment of blood pressure during gestation and postpartum [21, 22].

A postpartum dilation of the SRV in women with a “normal” prepregnancy SRV has been reported, especially after the second or third gestation. Together with the observation that HF may present in the postpartum, these above data highlight the need for careful monitoring throughout pregnancy and delivery.

12.3.5 Cyanotic Congenital Heart Diseases

Out of Eisenmenger syndrome, cyanosis may be found both in CHD in natural history and after surgery. CHD in natural history include, for example, Ebstein anomaly, single ventricle with pulmonary stenosis, and pulmonary atresia with ventricular

septal defect and major aortopulmonary collaterals. Some of these women may have few symptoms, especially in their younger age, and may wish to become pregnant. However, cyanosis exposes both the mother and the fetus to serious complications during gestation and after delivery. Maternal mortality and morbidity seem to be reduced in the most recent series [23], probably due to the better management by multidisciplinary teams with expertise in high-risk pregnancies. Nevertheless, maternal complications (HF, thrombosis, arrhythmias, and endocarditis) are reported in more than 15% of cyanotic women during pregnancies [3]. Thromboembolic events remain the most common maternal complication. Anticoagulation (with LMWH) is warranted, especially in the postpartum, when the risk of thromboembolism is threefold increased.

12.4 The "Pregnancy Heart Team"

Both European and American Guidelines underline the need for a multidisciplinary team ("Pregnancy Heart Team") to optimize the management of pregnancy in women with cardiovascular diseases, especially in high-risk conditions; reduce maternal mortality; and provide a comprehensive strategy for management of cardiovascular diseases during pregnancy [3, 24–26]. The multidisciplinary team should include at least/at minimum a cardiologist, a gynecologist/obstetrician, and an anesthetist, all with expertise in the management of high-risk pregnancies and adult CHD, but other experts are usually involved (e.g., geneticists, pediatricians, neonatologists, cardiothoracic surgeons). Moreover, a specialist nurse plays a crucial role in coordinating the program defined by the team and in the relationship with the pregnant woman.

The main tasks of the Pregnancy Heart Team should be:

- A comprehensive evaluation of the woman with CHD *before* pregnancy to define the pregnancy-related risks based on the risk stratification scores. Maternal morbidity and mortality in case of pregnancy, fetal outcome, and the possibility of genetic transmission should be discussed with the patient. Preconception counselling should include discussion about functional status. Ideally, maternal conditions should be optimized before gestation. The need for correction of CHD (in patients in natural history) or of residual defects after previous surgery should be analyzed.
- A thorough prepregnancy cardiologic examination, including ECG, echocardiography, and effort test, is recommended. Cardiopulmonary effort test (CPET) is of paramount importance in order to predict patient tolerance of the hemodynamic changes occurring during pregnancy, labor, and delivery; CPET, which has been related to prognosis in adults with CHD, is also able to predict pregnancy outcome and maternal and fetal complications [27].
- Serial monitoring of cardiac condition team-based care during pregnancy should be provided.
- In all pregnant women with moderate or elevated complexity CHD, an individualized plan for delivery should be defined in advance. This plan (including loca-

tion and mode of delivery, need for hemodynamic monitoring, possible therapies, postpartum observation) must be written in the patient's record and available for all healthcare providers involved in the team.

– Monitoring of the postpartum period. Some high-risk conditions require hospitalization and high intensive care at least for the first week after delivery. This is the case of CHD-related PAH, in which most complications occur in the postpartum period.

In addition, the multidisciplinary team should provide contraception advice, individualized on the basis of the underlying cardiac lesions and the woman's needs. In the rare conditions in which pregnancy remains contraindicated, the efficacy and safety of contraception are of paramount importance, and, in the case of pregnancy, the possibility of termination should be offered.

It's important to remind that information about reproductive health and possible pregnancy-related risks should begin early in female patients with CHD, as part of the *transition* program. Implications of their CHD on a future pregnancy should be discussed during adolescence, as soon as the young patients have reached adequate maturity. Together with counselling about contraception, this "pre-counselling" should be planned by the cardiologic—an obstetric team of the Pediatric Hospital or (where available) of the Transition Clinic, focusing on the individual risks (or the absence of risks!) but also on the possible risks for the future offspring [28–31] (Fig. 12.1).

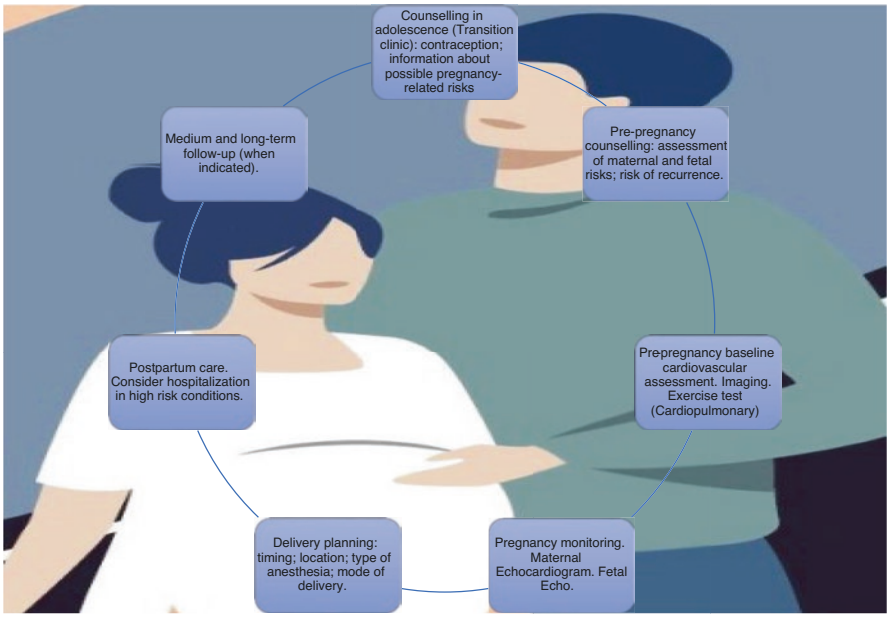

Fig. 12.1 The role of the Pregnancy Heart Team

12.5 Medications

According to ROPAC Registry, cardiac medications are used by at least one-third of women with preexisting cardiovascular disease [32]. Some medications which are commonly used for the treatment of cardiovascular diseases are contraindicated in pregnancy. Problems may be related to a possible change of pharmacokinetics during gestation and, on the other side, to potential harm for the fetus. During pregnancy, the absorption of drugs may be reduced because of nausea and vomiting, and the use of anti-acid to reduce these common symptoms may decrease the drug bioavailability. Physiological hemodilution of pregnancy (see paragraph 14.2) affects the volume distribution of drugs, while the upregulation of the enzymatic systems may modify metabolism (e.g., cytochrome P450), which occurs during pregnancy, and excretion is affected by the increased glomerular filtration rate.

Since 2015, the previous classification of drugs in five (ABCDX) categories was abandoned because it was considered potentially confusing and inadequate to define the real relationship between the use of different medications and the possible fetal risk. The Pregnancy and Lactation Labeling Rule differentiates recommendations regarding the use of medications during pregnancy and labor from those for the lactation period.

An exhaustive discussion about the use of cardiovascular drugs in pregnancy is beyond the scope of this chapter. We will briefly consider the main problems in pregnant women with CHD.

Supraventricular arrhythmias (especially atrial tachycardia) are not uncommon during pregnancy in women with CHD, driven by chamber dilation and hormonal effects. Also atrial fibrillation may eventually occur. CO may be significantly impaired because of tachyarrhythmia, and sinus rhythm should be restored as soon as possible. The use of antiarrhythmic drugs should be avoided during the first trimester and eventually used at lowest doses because of teratogenic risks. In the case of arrhythmias leading to hemodynamic impairment, prompt electric cardioversion is recommended and is considered safe for the fetus. Intra-atrial reentrant tachycardia is the most common mechanism of supraventricular arrhythmias in patients with CHD. Beta-blockers are the medications that have been more extensively used; they are usually considered safe in pregnancy, although neonatal hypoglycemia and reduced fetal growth have been reported; β1 selective drugs as metoprolol, together with digoxin and eventually verapamil, represent the first-line therapy. In atrial fibrillation, a rate control may be a possible option when the restoration of sinus rhythm is not obtained. Flecainide is commonly recommended in pregnancy both for maternal and fetal arrhythmias, but it should be used with caution in case of structural heart abnormalities and ventricular dysfunction. Ventricular arrhythmias are rare but may possibly occur in women with CHD. Treatment is based on beta-blockers or lidocaine as first-line therapy; sotalol or flecainide might be eventually used, while amiodarone is considered only as “last-line” therapy when all the other treatments fail.

Hypertension is a common complication of pregnancy in the general population, and the risk is somewhat increased in women with CHD (e.g., aortic coarctation).

Careful monitoring is required in order to avoid secondary maternal hypotension and consequent fetal damage. First-line drugs to control pressure values in pregnancy include beta-blockers (metoprolol or oral labetalol), calcium antagonists (nifedipine), and α-methyl dopa. Diuretics should be used with caution for the potential decrease in placental perfusion. Doses of hypertensive drugs may require an adjustment in later pregnancy because of the physiological decrease in systemic pressure values in the second trimester. Ace inhibitors and angiotensin receptor blockers, which are widely used also in patients with CHD, are contraindicated in pregnancy as they may lead to severe adverse effects on the fetus; however, ace inhibitors can be assumed during lactation [32]. The need for a withdrawal of medications during pregnancy and the potential maternal risks should be discussed in the preconception counselling. On the contrary, Carvedilol, an α/β-blocker, may be used safely in pregnancy and has not been related to impaired fetal growth.

A subgroup of CHD patients is commonly treated with antiplatelet drugs or anticoagulants. The risk of thromboembolic events is notably raised in pregnancy due to the hypercoagulable state, and this risk further increases in high-risk conditions, such as cyanotic CHD and Eisenmenger syndrome. Moreover, anticoagulants are needed in women with mechanical heart valves. The vitamin K antagonist warfarin carries well-known fetal toxicity (embryopathy, miscarriage, stillbirth, hemorrhage) and should be avoided whenever possible. More recently, these adverse effects have been particularly related to higher doses (>5 mg/day). Therefore, in all women taking >5 mg of warfarin, the treatment should be switched to LMWH or unfractionated heparin, which does not cross the placenta, at least for the first trimester, when the risk of embryopathy is higher. Close monitoring is required, especially in women with mechanical prostheses, to avoid thrombotic complications, as pregnancy per se is associated with a hypercoagulable state. In any case, warfarin must be stopped and changed to LMWH or unfractionated heparin at 36 weeks of gestation to avoid fetal hemorrhage at the moment of delivery.

A list of drugs that are commonly used during pregnancy is reported in Table 12.2.

Table 12.2 Drugs which are considered safe in pregnancy (modified from Halpern et al. 2019)

Heart failure	Arrhythmias	Hypertension	Anticoagulants	Pulmonary hypertension
Metoprolol	Adenosine	Labetalol	Unfractionated heparin	Epoprostenol
Carvedilol	Metoprolol	Nifedipine	Enoxaparin	Sildenafil
Furosemide	Digoxin	α-Methyldopa (oral)		
Dopamine	Lidocaine	Furosemide		
Dobutamine				
Norepinephrine				

12.6 Cardiovascular Complications Pregnancy-Related

Patients with CHD are at higher risk for cardiovascular events throughout gestation and at delivery. Due to the stress imposed by pregnancy on the cardiovascular system, pregnant women with CHD experience HF, arrhythmias, myocardial infarction and thromboembolic events (stroke, pulmonary embolism), and obstetric complications fetal events. The rate of cardiac complications is related to the complexity of the underlying lesion [33].

HF represents the most common severe complication affecting women with preexisting heart disease and is a relevant cause of in-hospital death [34]. Although cardiomyopathies account for most cases, HF also complicates pregnancy in women with CHD, especially if associated with PH, or in patients with uncorrected lesions [1]. Women with prepregnancy HF, EF of the systemic ventricle <40%, NYHA class >II, or mWHO class IV are also at major risk for developing HF.

A significant risk of systemic hypertension is reported in women with aortic coarctation, with a peak in blood pressure expected between 3 and 8 days after delivery requiring a scheduled control [35].

In CHD patients with systemic hypertension, acute HF with preserved EF (≥50%) may occur during pregnancy and in the postpartum, mainly related to diastolic dysfunction secondary to the increased afterload.

In women with uncorrected CHD, a high rate (25%) of hospital admissions for cardiac complications has been reported. With HF, ventricular tachycardia is common in severe CHD in natural history. Death mainly occurs in the postpartum, highlighting the need for a strict postpartum follow-up in this subset of patients [1]. Uncorrected severe CHD, often complicated by cyanosis, are less common in western countries but in more recent years may be increasingly observed due to migration from emerging countries.

Arrhythmias and HF may present in a relevant number of pregnant women with SRV [20]. In these patients, prepregnancy signs of HF or an SRV-EF less than 40% were predictors of major complications during pregnancy. Maternal HF negatively impacts fetal hemodynamic: fetal mortality, impaired fetal growth, and premature birth are reported in one-third of cases in ZAHARA registry [21].

Aortic stenosis, mainly associated with BAV, is relatively rare in women of childbearing age. HF is one of the most common presentations and the main complication during pregnancy. Although pregnancy has been reported to be safe in the majority of women with aortic coarctation [36], HF may present in a minority (3%), probably related to the severity of the obstruction. Atrial or ventricular arrhythmias and stroke may also occur [37].

Arrhythmias, especially atrial arrhythmias, are not rare in CHD patients during pregnancy. In a large nationwide study [33], higher arrhythmia rates were reported in women with moderate and complex CHD compared with women with “simple” defects. Apart from premature supraventricular complexes, the most common form is represented by reentrant intra-atrial tachycardia, mainly secondary to postincisional scars. The hemodynamic effects are related to the type of underlying congenital cardiac malformation (e.g., CHD with SRV). In all conditions at risk for

hemodynamic impairment, elective cardioversion can be safely performed; however, careful monitoring of fetal cardiac function is recommended.

Also, the risk of stroke is reported to be higher in women with moderate or complex CHD.

The risk of deep vein thrombosis and pulmonary thromboembolism (PE) is increased during pregnancy and the puerperium and represents a significant cause of maternal morbidity and mortality [3]; the risk is further increased in cyanotic CHD. As symptoms and signs of PE (dyspnea, tachycardia, chest pain, syncope) may be misleading in pregnancy, a high level of suspicion is recommended, especially in high-risk conditions. Specific investigations should be performed in suspected cases (D-dimer testing, computed tomography pulmonary angiography and/or ventilation perfusion lung scanning). *For treatment, see section* "Medical therapy in pregnancy".

An additional risk should be considered in cyanotic CHD with PH (Eisenmenger syndrome) where right-to-left shunting may predispose to paradoxical embolism.

12.7 Delivery Planning

A comprehensive plan for delivery should be planned by the Pregnancy Heart Team at the very beginning of the third trimester and be easily available for all the health professionals involved in care. A preterm delivery (because of spontaneous labor or medical induction) is not uncommon in women with CHD. In all cases, delivery should not be delayed after 41 weeks of gestation.

Vaginal delivery is preferred for most CHD, caesarian section (CS) being reserved for obstetric indications. CS is, however, indicated for a small subgroup of women with high-risk CHD (mWHO class IV) as PAH, critical mitral or aortic stenosis, or severe aortopathies. In some conditions, vaginal delivery with an assisted second stage to avoid prolonged pushing can be a reasonable option. Delivery is associated with greater cardiac work due to the increase in systemic pressure, heart rate, and circulatory volume (see paragraph 14.2). Careful monitoring of the fluid balance (amount of blood loss versus "autotransfusion" at every uterus contraction) is essential in all CHD at risk of decompensation. Pain control during labor can mitigate the increase in systemic pressure. However, spinal anesthesia leads to venous dilation, reduced blood return to the heart, and hypotension and should be avoided in patients with hemodynamic instability (e.g., women with PH). In women who need anticoagulation (e.g., in the presence of mechanical prosthesis), the risk of intrapartum hemorrhage must be considered.

12.8 Follow-Up after Delivery (Postpartum and Long-Term Follow-Up)

The postpartum usually does not require a specific follow-up for women in class mWHO I-II, in the absence of other potential risk factors (gestational diabetes, hypertension, etc.).

On the contrary, the important volume shifts occurring after delivery may rapidly decompensate high-risk conditions (class mWHO III-IV), and admission to intensive care unit should be planned for women in class mWHO IV at least for the first 72 postpartum hours.

Patients with PAH (even if not severe) require close monitoring for the first several days after delivery because acute right ventricular overload may lead to right ventricle decompensation, and most maternal deaths occur within the first postpartum months.

Venous thromboembolism risk reaches its peak in the postpartum (8–12 weeks after delivery), and prophylaxis is strongly recommended, especially in women with PAH or cyanotic CHD.

Volume overload should be anticipated and eventually treated in moderate and severe valve lesions; a postpartum monitoring of volume overload and diuresis is warranted, especially in severe mitral and aortic stenosis patients.

Most of the readmissions to the hospital occur within the first postpartum weeks, primarily because of HF. In all high-risk women, a postpartum visit with the cardiologist should be planned within 15 days from the delivery.

In pregnant women with hereditary aortopathies, aortic dissection, although rare, is relatively more common than in the age-matched nonpregnant women (see paragraph 14.3).

Pregnancy has been reported to accelerate the structural valve deterioration of biological prostheses, increasing the likelihood of anticipated reoperation. The risk is probably lower for the new biological models, including stentless porcine valves, but the data need to be confirmed in larger studies [38]. All these data reinforce the indication to strict monitoring in the first days/weeks after delivery and a planned follow-up at least for the first 6 months.

Long-term cardiac outcomes after pregnancy in women with CHD have not been extensively explored and are still unknown for most conditions. In a large retrospective study, conducted in two large referral centers and involving over 700 women with CHD, parous patients were more likely to have anticipated cardiac surgery vs. nulliparous ones; in the overall CHD population, previous pregnancy was associated with more long-term cardiac events, including death, stroke, and HF [39].

Finally, associated complications such as systemic hypertension or gestational diabetes, which may also present in women with CHD, must be considered independent cardiovascular risk factors in the long-term follow-up.

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13 Contraception, Family Planning, and Pregnancy Management: The Nurse Specialist Role

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13.1 Family Planning and Contraception Counseling

Following advances in the medical and surgical treatment of congenital heart disease (CHD), more than 90% of children born with CHD will survive to their childbearing years. Among these, an estimated 9–15% have complex CHD [1]. While many will tolerate pregnancy well, patients with complex congenital heart defects or advanced physiologic stages incur an increased risk for cardiac and obstetrical complications with pregnancy [2]. Therefore, pregnancies in the ACHD population must be planned to allow for identification and treatment of cardiac problems prior to conception; however, a 2019 study found approximately 54% of pregnancies among women with CHD are unintended [3]. Along with high rates of unplanned pregnancy in this population, studies have found that a significant proportion of ACHD patients are either not using contraception at all despite being sexually active or are not using a form of contraception that is recommended for their cardiac condition [4–6]. The following section discusses the importance of contraceptive counseling for women with CHD in their reproductive years, as well as the role of the advanced practice nurse (APRN) in the coordination of timely and comprehensive contraception counseling for all ACHD patients capable of pregnancy.

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13.1.1 Contraception Counseling

The APRN role in contraception counseling is centered around assessing patient values and preferences, identifying potential barriers to effective contraceptive use, and recommending contraception that is clinically suitable for the patient. Contraception counseling should begin at the first visit to the ACHD clinic and should be reviewed at regular intervals beginning in adolescence [7]. Adolescents have been identified by the WHO as a demographic group in need of improved access to family planning resources. In some places, cultural or religious factors may restrict access to contraception for unmarried adolescents, or they may avoid seeking contraception due to fear of judgment, stigma, or concern about lack of confidentiality [8]. While involving parents or guardians in the discussion about sexual activity and contraception is important, adolescents should be given the option to keep their reproductive choices confidential [9]. The law varies regionally, but in many places, the APRN can assure the adolescent patient that they have a right to confidentiality and that reproductive health services are available without parental consent [6].

The APRN may initiate a family planning conversation by asking questions that encourage the patient to share their timeline for pregnancy, as well as their priorities for contraception [10]. Research has shown that women are more satisfied with their contraceptive choice if their provider employed the techniques of the shared decision-making model (SDM). The SDM is a communication framework that emphasizes patient-centered care and collaborative decision-making between patients and providers. In the context of contraception counseling, the APRN should focus the conversation around patient preferences, allowing the discussion to flow back and forth in an interactive way, rather than unilaterally dictating the plan [10]. While considering the patient's values and preferences, the APRN should also assess for any potential barriers to effective contraceptive use.

13.1.2 Barriers to Contraceptive Use

For the general population, common challenges to the safe and effective use of contraceptives include difficulty accessing clinics where contraception is available due to a lack of local facilities, limited transportation, and the financial cost of services. The APRN practicing in the United States should be aware of disparities in access to family planning resources. Black and Hispanic women, women of lower socioeconomic status, women aged 18–24, and those with lower levels of education are less likely to have access to contraception and are more likely to experience unintended pregnancy [11]. Social workers play a key role in connecting patients with low-cost clinics and affordable transportation.

Other potential considerations include patient disabilities. Persons with intellectual or physical disabilities have reproductive rights and are entitled to

family planning and contraceptive care. For those with intellectual disabilities, conversations about reproductive health may need to involve a guardian. Patients with memory problems may have difficulty using contraceptive methods that require taking a daily pill or changing the patch or ring on a schedule. Setting reminders on a phone or device may help with consistency, though other methods that do not require as much participation on the part of the patient may be preferred, such as the DMPA injection, IUD, or subdermal implant. For persons with certain physical disabilities, consider that insertion of the vaginal ring or the use of a condom may present challenges due to the body mechanics involved [8].

Underprepared providers are a significant barrier to contraceptive counseling in the ACHD clinic setting. A 2010 paper by Vigl et al. found that nearly two-thirds of young women with CHD did not receive contraceptive or pregnancy counseling from their cardiologist and suggested this was due to the provider's lack of knowledge on the subject [4]. Similar findings were published by Brown et al., who surveyed pediatric and adult cardiology fellows and found they did not routinely discuss preconception counseling or contraception with their female patients, and the majority did not feel they had been adequately trained to provide counseling on that subject [12]. This represents a significant gap in care, as evidence suggests that a low proportion of ACHD patients receive preconception counseling related to their heart condition from any source [5, 13]. Consistent, high-quality contraceptive counseling from cardiology providers is an unmet need in the ACHD population.

13.1.3 Contraceptive Options

Published guidelines assist with determining the appropriate contraceptive choice while accounting for cardiac risk level. Published in 1996, the World Health Organization Medical Eligibility Criteria for Contraceptive Use has been a critical source of guidance regarding contraceptive use for women with cardiovascular disease [14]. In 2006, Thorne et al. published the modified WHO Classification of Maternal Cardiovascular Risk (mWHO) to delineate these risks more definitively for women with congenital heart disease [15]. The mWHO classification is widely used by ACHD providers to estimate contraceptive and pregnancy risks in women with CHD.

Advanced practice nurses and ACHD cardiologists typically do not prescribe contraception, but they are responsible for making recommendations to help guide the OB/GYN or primary care provider. Therefore, it is necessary for the APRN to understand the contraceptive options available. While there is a wide variety of contraceptives on the market today, the choices are fewer for patients with moderate to complex CHD. Table 13.1 provides a tiered system for contraception effectiveness and their associated safety concerns [2, 3, 16–18].

Table 13.1 Contraceptive tier and safety concerns

Efficacy	Method	Safety considerations
Tier 1: Most effective (1% or less chance of failure with typical use) < 1 pregnancy per 100 women in 1 year	IUDs	• Theoretical risk of endocarditis with implantation in persons with history of bacterial endocarditis or with prosthetic heart valves. • Possible vasovagal response during insertion may cause hemodynamic compromise in some with complex CHD.
	Male/female sterilization	• General anesthesia required for most common types of female sterilization. Unsafe for patients at high anesthesia risk (pulmonary hypertension, etc.).
	Implant	• No specific safety considerations for ACHD population.
Tier 2: Less effective (6–12% chance of failure per year with typical use) 4–7 pregnancies per 100 women in 1 year	Combined hormonal contraceptives (pill, patch, ring, injection)	• Avoid CHCs for patients with elevated clotting risk, including unrepaired cyanotic defects, Fontan circulation, the presence of mechanical valve, atrial enlargement or unrepaired atrial septal defect, pulmonary hypertension, atrial fibrillation or flutter or intra-atrial reentrant tachycardia, history of thrombotic events, or significant ventricular dysfunction. • Contraindicated with preexisting hypertension.
	Progestin-only contraceptives (pill, injection)	• Contraindicated with severe decompensated hepatic cirrhosis or liver tumors, use with caution in those with advanced Fontan-associated liver disease (FALD). • Risk for hyperkalemia with drospirenone.
Tier 3: Least effective (18–28% failure rate per year with typical use) > 13 pregnancies per 100 women in 1 year	Condoms, sponge, withdrawal, spermicides, diaphragm	• No device-related safety concerns. • Patients with increased pregnancy risk should be counseled to use a more effective method (tier 1 or 2).
	Emergency contraception	• Possible acute fluid retention with the use of levonorgestrel. • See above for IUD safety considerations.

13.2 Tier 1: Most Effective (1% or Less Chance of Failure with Typical Use)

13.2.1 Intrauterine Devices (IUDs)

Levonorgestrel (LNG)-emitting devices: *Mirena, Liletta, Kyleena, Skyla*
Copper IUD: Paragard

IUDs are among the safest, most effective, and most popular forms of reversible contraception used worldwide [16]. The device is comprised of a polyethylene frame with added copper or levonorgestrel (LNG). The copper IUD (Paragard in the US) is FDA approved for 10 years of use, though the data reflects continued efficacy for 12 years and beyond [16]. The copper IUD is the only form of long-acting reversible contraception that is hormone-free, and it can also double as emergency contraception if implanted within 5 days of unprotected intercourse. Both copper and LNG IUDs prevent fertilization of the ovum by altering conditions within the uterus, making it difficult for sperm to penetrate and survive. LNG-emitting IUDs cause thickening of cervical mucus and changes to the endometrial lining of the uterus, and systemic uptake of the LNG may cause suppression of ovulation [16]. Through these mechanisms, IUDs are very effective at preventing pregnancy.

There are two common concerns regarding IUDs in the congenital heart population; firstly, there is a theoretical increased risk of endocarditis with the implantation of an IUD, particularly in women with a history of bacterial endocarditis or prosthetic heart valves, though little supporting data on this subject has been documented [16]. The WHO Medical Eligibility Criteria recommends administration of prophylactic antibiotics prior to IUD insertion for certain higher-risk patients, but the ACC/AHA guidelines do not recommend SBE prophylaxis for this procedure [8, 19–21].

Another concern regarding IUD usage in the cardiac population is the possibility of vasovagal response during insertion that could lead to syncope or bradycardia, particularly in sensitive groups such as those with complex CHD (Fontan circulation, pulmonary hypertension/Eisenmenger's syndrome, cyanosis) therefore consider insertion in an inpatient setting with pain control [3, 18, 19]. In summary, IUDs are highly effective at preventing pregnancy, generally well tolerated, and safe for most ACHD patients.

13.2.2 Progestin Subdermal Implant

Etonogestrel: Nexplanon, Implanon
Levonorgestrel: Norplant (no longer in production), Jadelle, Sino-implant

The implant consists of one (Nexplanon) or two (Sino-implant, Jadelle) flexible rods containing progestin that are implanted under the skin of the upper arm. The etonogestrel implant is approved for use for 3 years by the FDA, though studies have shown continued efficacy up to 5 years after implantation. The Sino-implant

and Jadelle are considered to be at peak effectiveness for up to 4 years, though efficacy wanes more quickly for women weighing more than 70 kg. As with the IUD, a benefit of the implant is the lack of failure due to user error. It primarily works by suppressing ovulation, and after removal of the implant the return to fertility is swift, within approximately 1 month [17]. A drawback of this method is that it must be placed and removed by a provider in a medical setting, which can make the cost of the implant higher. There are no specific concerns for the congenital heart disease population associated with the implant.

13.2.3 Female and Male Sterilization

The most invasive form of contraception is sterilization. Female sterilization entails blocking or severing the fallopian tubes to prevent sperm and eggs from meeting and is the only form of contraception that is considered a permanent end to fertility [8]. The most common approach is via laparotomy, and the risks associated with general anesthesia and abdominal insufflation may be prohibitive for some patients. The Essure device, which is inserted into the fallopian tubes via the cervix with local anesthesia and sedation, was recalled in the United States in 2018. Male sterilization, a minimally invasive procedure that involves blocking the vas deferens, is highly effective, quick, and safe but may warrant a conversation with female patients regarding the need for alternate contraception if they have multiple sexual partners. Sterilization is a reasonable choice for many ACHD patients who may undertake general anesthesia (16) with tolerable risk.

13.3 Tier 2: Less Effective (6–12% Chance of Failure per Year with Typical Use)

13.3.1 Progestin-Only Injectables

Depot Medroxyprogesterone Acetate (DMPA) subQ or IM, DepoProvera
Norethisterone Enanthate (NET-EN)

Injectable progestins prevent pregnancy by inhibiting ovulation and have a 6% failure rate with typical use [21]. The most frequently used agent is intramuscular DMPA, commonly known as the "Depo shot." The NET-EN formulation is approved for use in many countries around the world but not currently in the United States. The DMPA injection is administered every 3 months, and the NET-EN is given every 2 months; patients must be able to return to a healthcare facility at regularly scheduled intervals for injections to ensure the efficacy of their contraception. A consideration when recommending this method is the potential for fluid retention with the use of the progestin-only injectables; use caution when recommending this method to patients with heart failure [19]. Unlike other methods of contraception, the injectables may cause prolonged return to

fertility of up to 6–10 months [22]. Overall, progestin-only contraceptives are a reasonable option for some ACHD patients, and they have little to no effect on blood pressure, coagulation, or lipid levels [21].

13.3.2 Progestin-Only Pills

Norethindrone

Drospirenone

Desogestrel, Levonorgestrel, and Other Progestins Available in Countries Outside of the United States

Progestin-only pills contain no estrogen and therefore are an option for women who prefer to take a pill but for whom combined oral contraceptives are contraindicated. The progestin-only pills work primarily by suppressing ovulation, increasing the viscosity of cervical mucus, and promoting changes in the endometrial lining of the uterus that discourage implantation [16]. When compared to CHCs, the POPs carry no increased risk for thromboembolism but have the same failure rate (9%), though they must be taken at the same time every day [16, 21]. The POPs are contraindicated (MEC category 3) for those with severe hepatic cirrhosis or liver tumors and therefore may not be a safe choice for patients with advanced Fontan-associated liver disease, though these patients ideally would not be using POPs in favor of more reliable forms of contraception [14, 16]. For patients who are at risk for hyperkalemia, be aware that drospirenone has anti-mineralocorticoid activity comparable to spironolactone, and serum potassium levels should be monitored if drospirenone is used concurrently with ACE inhibitors, angiotensin-II receptor antagonists, aldosterone agonists, heparin, potassium-sparing diuretics, or frequent use of NSAIDs [16].

13.3.3 Combined Hormonal Contraceptives

Combined Oral Contraceptives: Various Formulations

Transdermal Patches: Evra/Xulane

Vaginal Ring: NuvaRing

Injection: Cyclofem, Mesigyna (Not Available in the United States)

The combined hormonal contraceptives include the pill, the transdermal patches, the vaginal ring, and the combined hormonal injectables (not available in the United States). The progestins provide most of the contraceptive effect through suppression of ovulation and changes to the cervical mucus, and estrogen helps with control of the menstrual cycle. They all have a typical use 1-year failure rate of 9% [16]. The main concern with combined hormonal contraceptives is the increased risk for clotting secondary to the estrogen content. Estrogen may also increase blood pressure and should be used with caution in patients with preexisting hypertension [14, 18]. The patch and the ring may not be suitable options for persons with some physical disabilities.

Women with some forms of CHD are counseled to avoid estrogen-containing products due to concerns about increased clotting risk. To determine who should avoid estrogen, the APRN should understand the biochemical role of estrogen as it pertains to clotting. Estrogen (usually as ethyl estradiol) is a sex hormone found in combined hormonal contraceptives such as combined oral contraceptives, injections, and the patch and ring. Estrogen promotes coagulation by stimulating the production of clotting factors while simultaneously hampering the body's inherent antithrombotic mechanisms [17]. It is widely accepted that the risk of venous thromboembolism in women using combined hormonal contraceptives is elevated and has been estimated to be approximately twofold higher compared to women not on CHCs [14, 17, 23]. The risk for arterial thrombosis, specifically myocardial infarction and stroke, is estimated to be 1.6-fold higher in CHC users, with the most risk associated with higher doses of estrogen [23]. Other risk factors for thrombosis include current history of smoking, obesity, migraines, hypertension, known thrombogenic mutations (i.e., Factor V Leiden), and increasing age (35+). While limited data is available directly examining the safety of CHCs for women with congenital heart disease, it is reasonable to assume that the increased thrombosis risk found in the general population also applies to women with CHD [3]. Certain conditions that are found among CHD patients add additional risk for clotting and should be carefully considered. It is reasonable to avoid CHCs if the patient has an unrepaired cyanotic defect or Fontan circulation, a mechanical valve, atrial enlargement or unrepaired atrial septal defect, pulmonary arterial hypertension, atrial fibrillation/flutter or intra-atrial reentrant tachycardia, history of thromboembolic events, or significant ventricular dysfunction [3, 17, 21].

Counseling patients to stop CHCs may be complicated, particularly when patients derive other benefits from estrogen-containing contraceptives beyond pregnancy prevention such as improvement in menorrhagia or dysmenorrhea, or if they are comfortable with their CHC and reluctant to consider switching to an unfamiliar alternative. It is important to remember that thrombosis risk associated with CHCs is still lower than the potential risk for thrombosis with pregnancy, so CHCs may be the best choice in cases where contraceptive options are very limited or other options not available [3]. For patients taking warfarin, there is no clear evidence that the anticoagulant effects of warfarin effectively neutralize the increased risk of clotting with concurrent use of CHCs [19].

13.3.4 Emergency Contraception

Copper IUD

Levonorgestrel (Plan B)

Ulipristal Acetate

Emergency contraception is generally safe for women with congenital heart disease (MEC 1–2), though acute fluid retention should be considered with the use of levonorgestrel, and the insertion of the IUD comes with the small potential risk of syncope and bacterial endocarditis [19].

13.4 Tier 3: Least Effective (18–28% Failure Rate per Year with Typical Use)

13.4.1 Barrier Methods, Spermicide, Diaphragm, Sponge, Fertility Awareness, and Withdrawal

These include barrier methods such as the female and male condom, which prevent sperm from being released into the vagina while also protecting against sexually transmitted infections. The use of the diaphragm or the sponge involves inserting the device into the vagina to block or incapacitate sperm before it reaches the uterus, usually combined with spermicide, a chemical that kills sperm. These contraceptive methods have waned in popularity in recent years but are still available, though they may require a prescription. Fertility awareness or rhythm methods are ways of planning sexual intercourse during less fertile times of a woman's menstrual cycle. Withdrawal entails removing the penis from the vagina before ejaculation occurs to avoid depositing sperm into the vagina [16]. These methods carry no cardiac contraindications, aside from the increased risk for pregnancy compared to more effective methods. Women with high-risk congenital heart lesions should be counseled against the use of these methods alone, due to the high rate of failure with typical use. Ideally, ACHD patients should use a highly effective method along with a barrier method to prevent pregnancy and protect against sexually transmitted infections.

13.5 Prepregnancy Counseling

Patient counseling prior to pursuing a planned pregnancy is an important phase of preconception counseling and a necessary step in the process of formulating a delivery care plan. This phase of care involves a thorough assessment of the anatomic, physiological, socioeconomic, and psychosocial state of the patient considering pregnancy. The APRN's role includes a detailed medical history-taking, physical exam, evaluation of current medications, maternal risk estimation, referrals to an obstetrician and other members of the pregnancy heart team, and genetic counseling as appropriate. These tasks are ideally undertaken before conception occurs but should also proceed if the patient arrives already pregnant.

The information collected during the preconception counseling phase, in combination with the prepregnancy workup, will help to estimate the degree of cardiac, obstetric, and fetal risk with pregnancy and should be reevaluated at every patient visit. Multiple risk assessment models including CARPREG (CARdiac disease in PREGnancy), ZAHARA (Zwangerschap bij Aangeboren HARtAfwijking), and ROPAC (Registry Of Pregnancy And Cardiac disease) pertain to maternal morbidity and mortality risk in the ACHD population, but the modified World Health Organization classification of maternal cardiovascular risk is the most widely accepted in the ACHD community [18, 24, 25].

While most women with mild CHD are able to carry a pregnancy without difficulty, women with moderate to complex congenital heart disease may face

significant challenges, and in some cases the timing of pregnancy is an important consideration. For example, women with valvular disease such as pulmonary valve disease in Tetralogy of Fallot or bicuspid aortic valve with severe valvular dysfunction will face multiple surgical or transcatheter valve replacements in their lifetime. Timing of valve replacements should be taken into consideration when planning pregnancies in these scenarios. Women with congenital heart lesions that tend to worsen with age, such as systemic right ventricles, should consider having children earlier in adulthood when cardiac function is optimal [15]. For those with severe CHD (mWHO class IV or ACHD physiologic class D), such as failing systemic ventricles, Eisenmenger's syndrome, or select other high-risk lesions, pregnancy comes with a high risk for serious complications and is generally not advisable [15, 19]. Alternative means of having children should be explored with women who desire children but are unable to become pregnant, whether due to infertility or known risks associated with pregnancy. In vitro fertilization (IVF), surrogacy, or adoption may be alternative options for these patients.

All patients considering pregnancy, regardless of their risk level, should be counseled to establish care with a primary obstetrician/gynecologist (OBGYN). The primary OBGYN oversees important aspects of maternal health such as screening for diseases that may impact pregnancy, diagnosis and treatment of sexually transmitted infections, and management of routine aspects of pregnancy care. Referral to high-risk maternity is appropriate for patients with moderate or greater risk of complications with pregnancy (mWHO class II-III, III, IV). These patients should be managed at a tertiary care center by a team of ACHD cardiologists, anesthesiologists, obstetricians, and other specialists with experience in the care of pregnant women with congenital heart disease [2, 18]. As the time of delivery approaches, women at high risk for pregnancy complications who do not live near a tertiary care center may need to temporarily relocate in order to be nearer to their pregnancy heart team. Social workers may be able to assist with locating resources or temporary housing. Typically, patients in the lower-risk groups (mWHO class I, II) may deliver at a community hospital without the supervision of a specialized heart team [18].

13.5.1 Fetal Risk

Maternal functional class is a major determinant of fetal mortality with an incremental risk ranging from zero in the uncomplicated gravitas to a 20 to 30% fetal mortality rate in women who fall into class III and IV [26]. Maternal cyanosis threatens the growth, development, and viability of the fetus. Infants born to cyanotic mothers are typically premature and small for gestational age. The rate of spontaneous abortion is high, and the rate increases roughly in parallel to maternal hypoxemia [27].

13.6 Genetic Counseling

The biological children of ACHD patients are at increased risk for congenital heart disease, with incidence estimated at 3% to 15%, compared to a 1% risk of CHD in the general population. For parents with autosomal dominant defects, single gene defects, and chromosomal abnormalities, the fetal risk for CHD is higher, with an occurrence rate up to 50% [18, 28, 29]. Genetic counseling should be offered in particular to those patients with heritable syndromes such as Noonan and Holt–Oram Syndrome or with a family history of multiple cases of congenital heart lesions. Preconception genetic counseling and fetal echocardiography should be offered beginning at 14–16 weeks gestation.

13.7 The Clinical Assessment

13.7.1 Medical and Cardiac History

The APRN begins the assessment component of preconception counseling by taking a complete medical and cardiac history. This process may be cursory if the patient is already well known to the ACHD clinic. Some patients establish care with the ACHD program during pregnancy, either due to late diagnosis of their condition or due to having fallen out of care. For these patients, the APRN should attempt to obtain records from past cardiac surgeries and cardiac catheterizations, images, and reports from previous diagnostic imaging tests, stress tests (including cardiopulmonary exercise testing), and cardiac rhythm monitoring, as well as the most recent notes from other cardiology and primary care visits. While records are being collected, the nurse specialist should proceed with the assessment. Important medical problems to assess for include diabetes, hypertension, history of coronary artery disease or stroke, and history of blood clots [28]. Family history, particularly history of congenital heart disease, should be documented. Socioeconomic and psychosocial assessments should include information about support systems and housing stability, as well as a history of any past or present intimate partner violence. Substance use and smoking history should be assessed, and referrals to social work or treatment programs for substance abuse or smoking cessation should be entered expediently [18].

13.7.2 Physical Assessment

The physical assessment is cardiac-focused and can provide clues to a patient's congenital defect, prior surgeries, and current clinical status. Physical exam points include:

- Vital signs, including room air oxygen saturation.
- Jugular venous pulsations.

- Chest wall inspection, including scars from previous cardiac surgeries.
- Heart sounds, rate and rhythm, murmurs, rubs, or gallops.
- Extremities: peripheral pulses, clubbing, cyanosis, and edema.

In conjunction with the physical assessment, a review of systems should be completed with a focus on cardiac symptoms that could indicate underlying problems, such as dyspnea on exertion, edema, chest discomfort, orthopnea, syncope, dizziness, or palpitations [28]. The presence of any of these symptoms will guide the prepregnancy diagnostic workup to follow.

The APRN should review and update the patient's medication list and allergies and note whether any current medications are contraindicated in pregnancy. There are several medications commonly prescribed to the ACHD population that are not considered safe in pregnancy, and when possible, these should be discontinued or switched to an alternative prior to conception. Medications that are generally avoided during pregnancy include ace inhibitors, angiotensin receptor blockers, mineralocorticoid receptor antagonists, vitamin K antagonists, direct oral anticoagulants, amiodarone, and atenolol [6, 18]. Over-the-counter and naturopathic remedies should be avoided during pregnancy unless approved by a medical provider [28]. For women who take anticoagulants, the alternatives to warfarin and DOACs are subcutaneous unfractionated heparin or low molecular weight heparin. For those who will be switching to unfractionated heparin, create a plan for monitoring anti-Xa levels at a local laboratory. More information about cardiac medications in pregnancy can be found in the Congenital Heart Disease During Pregnancy section of this chapter.

13.7.3 Normal Signs and Symptoms in Pregnancy

The normal hemodynamic and structural changes that occur during pregnancy produce a series of signs and symptoms that may closely resemble a cardiopulmonary disease state. For example, hyperventilation is a common occurrence in pregnancy and may be described as breathlessness, which may be misinterpreted by the clinician as dyspnea. Fatigue, exercise intolerance, and orthopnea are other common complaints associated with an advancing pregnancy. Dependent lower extremity edema results from an enlarging uterus and high venous pressure in the legs caused by an increase in total body sodium, but may be interpreted as heart failure. Palpitations, or the sensation of the heart beating "faster or stronger," are not uncommon and are frequently associated with normal sinus tachycardia [30].

13.8 Diagnostic Testing

Women interested in becoming pregnant should undergo a full cardiovascular examination prior to conception, or as early as possible in the case of unplanned pregnancy. Once pregnant, diagnostic testing must be individualized with special consideration of the risk to both patient and fetus.

Diagnostic evaluations that should be obtained prior to conception and/or at first pregnancy visit include:

- *Laboratory testing*. Routine laboratory tests include complete blood count, serum electrolytes, urinalysis, calcium and magnesium levels, and renal and liver function tests. These levels should be measured at baseline, particularly for patients suspected of depressed ventricular function. Thyroid function tests should also be obtained as hypothyroidism as well as hyperthyroidism can cause or contribute further to systemic ventricular dysfunction [31]. In addition, the B-type natriuretic peptide (BNP) should be measured prior to and at onset of pregnancy. BNP is known to rise during pregnancy to twice the level of the nonpregnant person [32]. In patients with heart disease, BNP has been shown to be predictive of adverse maternal cardiovascular events [32–34].
- *12-Lead electrocardiogram*. Normal electrocardiographic (ECG) changes associated with pregnancy include a leftward shift of the QRS axis and shortening of the PR, QRS, and QT intervals at a resting heart rate. There may also be some nonspecific ST abnormalities including flattened and inverted T waves in lead III [30]. Rhythm disturbances are not uncommon during pregnancy, particularly during labor in the early puerperium. The ECG may show occasional premature atrial and ventricular beats, short paroxysms of supraventricular tachycardia, and sinus arrhythmia.
- *Echocardiogram*. Transthoracic echocardiogram (TTE) can be performed safely throughout pregnancy and should be repeated at regular intervals, such as at the beginning of each trimester, or as indicated should patient begin to show signs of ventricular dysfunction. Physiologic findings on a TTE may reveal chamber enlargement and physiologic aortic, mitral, or tricuspid regurgitation, as well as valvular dilatation. Ventricular size, shape, volume, and ejection fraction (EF), as well as signs of volume overload in the case of a shunt or valvular regurgitation, are detected by a good quality TTE.
- *Cardiopulmonary stress testing with echocardiographic imaging*. Cardiopulmonary stress testing (CPET) or exercise stress testing (EST) prior to conception helps to determine systemic ventricular adaptation to the hemodynamic changes of pregnancy and to the stress of labor and delivery. Unless strongly indicated, EST should be avoided in pregnant patients.

Additional investigations are only carried out in the setting of specific symptoms or pathologies. These may include magnetic resonance imaging or computed tomography (CT) as indicated for specific conditions such as aortic pathology, and Holter or event monitoring for complaints of palpitations. Because of the risk of radiation exposure to the fetus, cardiac catheterization is limited to rare cases where enough information cannot be obtained through noninvasive measures or hemodynamic compromise requires an emergent intervention [35].

13.9 Pregnancy Management: The Nurse Specialist Role

As the population of adults with congenital heart disease has grown, so has the cardiovascular disease (CVD) risk associated with pregnancy among females of reproductive age [36, 37]. As a result, the management of pregnancy and cardiovascular disease has been addressed internationally in cardiac and obstetrical guidelines as well as practice bulletins [19, 38–40]. The care of this population requires a multidisciplinary approach, starting with preconception counseling and categorization of maternal risk. Once pregnancy is confirmed, multidisciplinary care should continue throughout the antepartum, peripartum, and postpartum periods. Because of the wide spectrum of congenital heart defects, the 2019 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease is a helpful reference for categorizing simple, moderate, and high complexity defects by anatomical and physiologic criteria [19].

Patients with simple congenital heart defects who are clinically stable may be managed by a general obstetrician during pregnancy and delivered safely at a community hospital. For patients with moderate to complex CHD, existing pregnancy guidelines recommend they be managed and delivered at a regional or tertiary care center where a multidisciplinary team of caregivers is available [18, 38–40]. The multidisciplinary team may include maternal fetal medicine specialists, obstetrical anesthesiologists, neonatologists, cardiologists, geneticists, social workers, and advanced practice registered nurses (APRNs) who are experienced in the care of patients with CHD [7, 41–43]. Early coordination and ongoing communication between members of the multidisciplinary team is crucial to optimizing maternal outcomes. It is recommended that pregnancy heart teams meet regularly to review all pregnancies, with special attention to those which are moderate to high risk or present with special issues [43].

As previously outlined, APRNs play a pivotal role in initiating preconception and contraceptive counseling. Once pregnant, the APRN works collaboratively with the pregnancy heart team to individualize prenatal care based on maternal risk, the complexity and residual effects of the maternal cardiac disorder, and current functional capacity. The presence of any existing comorbidities such as diabetes, thyroid disease, or genetic syndromes should be integrated into the management plan. Psychosocial factors that may impact pregnancy management should be identified, including family support and resources, insurance coverage, employment, access to care, and proximity to the ACHD clinic and delivery hospital. For those living in geographically distant areas, a shared model of care should be developed with local providers in case the patient develops problems such as preterm labor and is not able to travel to the delivery hospital.

13.9.1 Antepartum Care

The initial meeting should include a review of the patient's cardiac history and preconception diagnostic evaluations and a comprehensive cardiovascular examination. Particular attention should be given to a history of arrhythmias or clinical

evidence of heart failure for which treatment was prescribed. The current medication regimen should also be reviewed for potential risks with pregnancy, as well as the need for dose adjustments or discontinuation. Lifestyle issues to be discussed include physical activity, diet and sodium intake, hydration practices, employment, and sexual activity.

It is important for patients to be familiar with normal signs and symptoms of pregnancy which can be mistaken for cardiac symptoms. For example, fatigue is normal finding in the first trimester, but peripheral edema is not; yet in third trimester, peripheral edema may be normally observed. Patients must also be advised of the importance of adjusting their schedules to allow sufficient intervals of rest. Excessive heat exposure can cause peripheral vasodilation with consequent decreased cardiac output; thus, attention to hydration and avoiding extremes in temperature are important.

The frequency of cardiology visits shall be largely individualized by the estimated maternal cardiovascular risk and the development of symptoms or complications as pregnancy progresses. Patients who remain clinically asymptomatic may be seen at the beginning of the second trimester and again at the beginning of the third trimester [7, 18, 39]. Those deemed at moderate to high risk (mWHO class II-III, III, IV, and physiologic stage C or D) or those with symptoms require more frequent visits, ranging from bimonthly to weekly. For women at the highest risk, such as those in physiologic stage D, a multidisciplinary meeting with the pregnancy heart team should be convened to discuss the management plan for both pregnancy and delivery once fetal viability has been reached (23–24 weeks gestation).

Delivery in a tertiary care center is recommended for mothers at potential risk for development of cardiac symptoms or decompensation during the peripartum period (mWHO class 3–4; ACHD physiologic stage C-D). Apart from those who present as physiologic class D, the preferred method of delivery in most women with congenital heart disease is vaginal delivery, with cesarean delivery reserved for obstetrical necessity.

13.9.2 Delivery Plan

As the pregnancy enters the third trimester, a delivery plan (DP) is developed outlining the management plan for labor and delivery and the postpartum period. The delivery plan is an organized, written means of communication which provides essential background information about the patient and outlines an individualized plan of care from time of admission to discharge [44]. While intended primarily for the labor and delivery teams, the DP must be accessible and reviewed by all medical and nursing staff members caring for the patient. Typically, labor and delivery nurses have little experience with cardiac issues such as the identification and treatment of arrhythmias, heart failure, or pulmonary hypertension in the patient with complex CHD. Therefore, the DP serves as a guide for proceeding with delivery, identifying potential cardiac complications, and determining who to call should symptoms or other problems arise. For patients at high risk, the DP further outlines

what cardiac support is required including monitoring, specific diagnostic tests to be ordered, and intravenous fluid allowances/restrictions [37].

A delivery plan begins with a brief history of the patient's cardiac disorder and any residual effects such as baffle leaks, arrhythmias, or signs of ventricular failure. This should also include the surgical history, a brief summary of the patient's past pregnancy history and outcomes, and any obstetrical or cardiac problems that may have occurred (such as preterm deliveries, arrhythmias, or ventricular dysfunction) as these issues may reoccur.

The delivery plan for the intrapartum period begins with risk category score assigned by cardiology and an outline of the potential complications that may occur. There should also be a summary of key diagnostic test findings, such as episodes of supraventricular or ventricular tachycardia, that may have occurred throughout the pregnancy as these may reappear during active labor or the postpartum period. The presence of a pacemaker and/or implantable cardiac defibrillator (ICD) should be listed. An ICD may be left on during delivery; however, if electrocautery is used for cesarean section, a magnet should be on standby to deactivate the ICD, with a portable defibrillator on standby [45, 46].

The body of the delivery plan includes the projected date for admission or induction of labor (IOL) and a brief antepartum summary to help the pregnancy heart team determine what level of care the patient might require, such as admission to the intensive care unit (ICU) versus the labor and delivery (L&D) unit. Timing and mode of delivery should be discussed in advance with the multidisciplinary team, and the patient's preference should be considered. The patient should be thoroughly counseled about the delivery plan and potential complications [16, 47]. A standardized checklist template may be used to track and edit details regarding planned mode of delivery, whether labor should be controlled (e.g., shortened second stage), anesthesia considerations, and specific diagnostic tests to be obtained upon admission such as BNP, echocardiogram, or EKG [44].

Details regarding peripartum cardiac monitoring, including when to initiate it, needs to be clearly delineated. The DP should provide specific guidelines as to what monitoring may be required (such as ECG or oxygen saturation) to ensure sufficient time to arrange the necessary staffing and unit adjustments and to guide the decision about whether the patient will be admitted to L&D vs ICU, where advanced hemodynamic monitoring such as arterial or pulmonary artery catheters may be used. L&D monitoring is generally limited to cardiac telemetry and oxygen saturation. Finally, because of the potential risk for thromboembolic events and infective endocarditis, the use of in-line IV filters, prophylactic antibiotics, and compression stockings during lengthy IOL should be included.

Because of the dramatic hemodynamic shifts that occur after delivery [7], the postpartum and discharge portion of the delivery plan should include plans for postpartum surveillance and the location where the patient should recover, as well as any diagnostic testing and monitoring that may be required before to discharge. Prior to delivery, discussions should be held with the patient regarding plans for breastfeeding as well as a postdelivery contraceptive plan. If the patient chooses to breastfeed, the reinstatement of cardiac medications (such as ACE inhibitors) may

be contraindicated. The mother's postdelivery contraceptive choice should also be documented in the DP, as some (like implants) can be placed before discharge. In the United States, a tubal ligation (TL) may be done prior to discharge if a signed written consent has been obtained prior to admission; if it has not, tubal ligation is deferred to a later date. To ensure compliance with follow-up care, discharge planning should include postdelivery dates for obstetrical and cardiac follow-up.

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Part VI

Physical Activity and Sport

Rehabilitation's Point Break: Changing for Improving

14

Barbara Baroni and Davide Girelli

14.1 Introduction

The World Health Organization (WHO) has defined Rehabilitation Cardiology (CR) as a multifactorial, active, and dynamic process, which aims to promote clinical stability, reduce disabilities resulting from the disease, and support the maintenance and the resumption of an active role in society, with the aim of reducing the risk of subsequent cardiovascular events, improving the quality of life and having an overall positive impact on survival. The role of Rehabilitation Cardiology has evolved a lot in the last 20 years. The first programs were developed in the 1960s–1970s following confirmation of the benefits of early mobilization during prolonged hospitalization after a coronary event.

Exercise was the main component of these programs which were essentially offered to patients who survived an uncomplicated myocardial infarction and started physical activity generally very late after the acute episode. In the years, the indications for cardiological rehabilitation have undergone important changes. The programs have also been successfully proposed to post-heart attack patients with complications or after coronary artery bypass grafting or coronary angioplasty. Finally, also as a consequence of the progressive aging of the population, the number of elderly people undergoing cardiological rehabilitation has increased considerably, many of them suffering from severe coronary artery disease, serious comorbidities, and dysautonomias. In the meantime, even the cardiological scenario of reference of our specific one has strongly changed: the increase in cardiac surgery centers and the refinement of medical-surgical techniques have made it possible to carry out a greater number of interventions with a progressive increase in

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children reaching adulthood. About 85% of newborns with congenital heart disease reach adulthood. These patients now constitute an estimated group in Italy of more than 100,000 subjects with at least 2000 patients entering adulthood every year.

It is therefore a completely new population of subjects, towards whom it is necessary to structure a dedicated recovery path starting from the already existing cardiological rehabilitation guidelines, adapting and perfecting them to the patient suffering from congenital heart disease. Undergoing cardiac surgery in childhood leads to a reduction in expectations, which naturally translates into a lower willingness of the patients themselves to assume an active lifestyle. The result is that the low physical activity of GUCH patients increases their predisposition to cardiovascular disease proportionally more than the general population [1].

Although the scientific literature is now full of confirmations about the benefits of physical exercise, not only in the healthy subject but also and above all in the heart disease, the recommendations issued to ACHD patients by specialized health structures do not appear adequate due to excessive fear and protection by healthcare professionals [2]. We must not forget that in people with congenital heart disease, the cardiorespiratory capacity is lower and deteriorates faster than in healthy people [3]. This has significant implications as cardiorespiratory capacity has been associated with increased mortality and morbidity [4].

In support of the safety and goodness of physical activity in ACHDs, the recent Cochrane review is cited, which quantitatively analyzes the effects of physical activity in people with congenital heart disease [5]. This review collects randomized controlled trials that compare the following:

- Any type of rehabilitation intervention and general physical activity, intended as the promotion of physical activity, physical training, and training of the respiratory system, with a control group "without physical activity".
- Which include all individuals with a diagnosis of congenital heart disease, regardless of age or previous medical interventions.
- With a median duration of surgery of 12 weeks.

The primary outcomes of the review were as follows:

- Cardiorespiratory capacity assessed on the basis of the oxygen peak with cardiopulmonary test on a cycle ergometer.
- The health-related quality of life determined by the HRQoL validated questionnaire.
- Objective measures of physical activity carried out by the patient during the day measured with a device supported by an app for continuous monitoring of movement.

Although the review highlights only small improvements in cardiorespiratory capacity, self-employed physical activity, and HRQoL of patients compared to the control group, there were no serious negative effects related to rehabilitation

interventions and set physical activity. These observations support the proposition that physical activity and exercise are safe and the benefits outweigh the potential risk.

Physical exercise is therefore proposed as an ideal preventive and therapeutic means, as it is physiological, effective, safe, and at low cost.

It is fundamental to define the rational principles and the ways in which to prescribe physical activity in individual heart diseases and therefore what is the right "dose" and the correct "modality" of exercise to be recommended.

To achieve a 20% reduction in mortality, an exercise intensity is required that leads to an energy consumption of approximately 4200 kJ per week (equal to 30 mins of physical exercise per day for at least 4–5 days per week). The greatest risk reduction is achieved with moderate intensity exercise, equivalent to 3–5 h of brisk walking, 2–3 h of jogging, or 1–2 h of running per week [6].

In analogy to what happens for a drug, in order to correctly prescribe physical activity to a sedentary, healthy, or cardiopathic subject, one must know the physiology and pathophysiology of the different types of physical exercise and sport, with particular regard to acute cardiovascular effects and chronic that they involve. For this reason, the various physical and sports activities, in relation to the responses of the cardiovascular system, have been classified by the experts of the COCIS (Cardiological Committee for Fitness for Competitive Sport) starting from the following essential physiological and pathophysiological concepts:

1. The cardiovascular effort can be constant over time, as in prolonged aerobic activities, or intermittent.
2. The cardio-circulatory effort primarily depends on the intensity of the effort, which in turn is proportional to the metabolic demands of the muscles involved. A simple measure of metabolic intensity is the MET or Metabolic Equivalent, which expresses the amount of oxygen consumed at rest (sitting silently on a chair), equal to approximately 3.5 ml of O2/kg/min corresponding to 1.2 kcal/min for a person of 70 kg.

   Compared to the calculation of the METs, an effort involving an expenditure of around 3 METS is considered of **light intensity**, of **moderate intensity** when the metabolic expenditure is between 3 and 6 METS, and of **medium to high intensity** when the effort involves an energy expenditure exceeding 6 METS.
3. The hemodynamic response to exertion is significantly influenced by the type of exercise. In activities where the technical gesture is cyclic, for example, in walking or pedaling, the muscle strength used is generally not high. These are aerobic activities in which the muscles, when the intensity of the effort is mild to moderate (<50–60% of the maximum), mainly use the energy released by the lipids, while for higher-intensity levels, the preferential substrate is represented by carbohydrates (glycogen). From a cardiovascular point of view, they are characterized by an increase in heart rate proportional to the intensity of exertion and a prevalent peripheral vasodilation with modest or no increase in mean arterial pressure. In power activities, on the other hand, the muscles use phosphocreatine as an energy source and only part of the carbohydrates through anaerobic gly-

colysis, with the production of lactic acid. In this type of exercise, the cardiocirculatory response is characterized by an important elevation of the mean arterial pressure due to the increase in peripheral vascular resistance, which, although of short duration, can be harmful in hypertensive patients and patients with pathologies of the aorta.

4. In terms of cardiovascular risk, the intensity of the exercise is a key factor. In fact, up to an intensity not exceeding 70–75% of the maximum, the regular practice of physical exercise is able to induce beneficial effects on the body and the cardiovascular system, without a significant increase in risk.

The incentive to a regular practice of physical exercise intended as a promotion and maintenance of health in subjects affected by congenital heart disease finds a specific application in childhood and adolescence not only for the important psychological and social aspects but also for the irreplaceable educational and training role.

What is required of the patient is to perform a certain amount of physical work in order to obtain with the lowest possible risk, an improvement in quality of life. To this end, a correct prescription of physical activity should combine two fundamental points:

- Meet the expectations of reintegration into active life and sports.
- Choose a physical activity and sport capable of providing benefits on the psychological and physical level with a risk of complications in the short and long term reasonably negligible or at least equal to the expected benefits.

The 2013 COCIS protocols [7, 8] devoted an entire chapter to competitive fitness criteria in the various types of congenital heart disease. Summarizing the general characters to prescribe any physical activity or sport in a child, adolescent, or young adult with congenital heart disease is necessary:

- A precise diagnostic framing of the pathology of the definition of its severity.
- A reasonable prediction of the possible evolution of the heart disease and of the possible impact on it of the chosen physical and sports activity.
- The evaluation of the functional capacity of the subject, through stress tests, or better still through a cardiopulmonary test.

There is no formula for adapting the program to each individual patient. Each subject differs in physical condition and consequences of the disease, with possible sequelae (pump dysfunction ischemia arrhythmias) that make it even more necessary and even more challenging to customize the work protocol. For this purpose, it is essential, when planning the program, to consider the following elements.

- Age.
- Sex.
- Risk class and associated pathologies.

- Musculoskeletal situation.
- Current drug therapy, particularly for drugs that may interfere with exercise response.
- Exercise testing if available.
- Previous exercise habits and exercise enjoyment.
- Understanding of how the program is executed.

Prescribing an exercise program should take these four factors into account:

1. **FREQUENCY (F)** of sessions per week
2. **INTENSITY (I)** the amount of absolute energy expenditure during the sessions
3. **TYPE (T)** the modality of execution
4. **TIME (T)** the duration of the single session

1. **Frequency**: As has already been amply reiterated, there is no one-size-fits-all. In general, the frequency of execution of the exercise program is daily, especially during the first phases of rehabilitation, and tri-weekly at home.
2. **Intensity**: The intensity of the activity must be such as to produce an increase in physical training in proportion to the degree of tolerance to the effort and the period of time in which the immobilization determined by the acute event has lasted. The intensity of the exercise must therefore be personalized and put in relation with the duration: in fact, similar results, in terms of increase of functional capacity, can be obtained with prolonged periods at low intensity and vice versa.

   Since it is not always possible to measure the patient's VO2max, the maximum heart rate reached in the cardiopulmonary test or through an exercise test is used as a reference parameter. For the evaluation of the training intensity, the heart rate range within which to carry out the program under safe conditions (TargetHeart Rate or THR) is calculated according to:
   - The percentage of the maximum heart rate reached; if the recommendations of the American Heart Association are followed, the training is performed keeping the heart rate between 50 and 80% of the maximum rate reached; in the practice of our centers, this percentage is instead 70–85%.
   - The Karvonen formula. This method of calculation leads to a heavier program than the previous one and therefore should be reserved for uncomplicated subjects with good adaptation to the effort.

     It is important to educate the patient so that he recognizes his own response to the effort by means of simple methods of self-evaluation. The most commonly used method is the measurement of pulse beats both at maximum effort and in the first minutes of recovery immediately after the interruption of exercise. Widely used is the fatigue perception scale codified by Borg (or similar like OMNI) and validated by numerous studies. In association can also be used the Borg scale of dyspnea.
3. **Type**: The mode of carrying out the rehabilitation and training program can be interval training or endurance training. Intermittent exercise (interval), which can be performed both free-body and with equipment, alternates periods of work

at a predetermined intensity with phases of recovery with no or very light work. The application of the workload for short periods of time determines an adaptation to the effort that is useful in the most deconditioned patients or in the elderly who do not tolerate the application of a continuous workload in the first training sessions.

The general principle of all free-body programs is to mobilize the major muscle groups, with various repetitions for a duration of 1–3 mins per exercise and with increasing commitment. It should be emphasized that there is no such thing as exercise "for the heart patient," but calisthenics, free body, or small apparatus can be used, provided that the criteria set out in the quantification of intensity are followed.

The resistance training or continuous (endurance) is the most widely used because it allows the maximum increase in aerobic capacity, are traditionally preferred activities with a dynamic component carried out by cycle ergometers, ergometers or treadmills. Even in these cases, it is possible to provide exercises with weights and/or equipment, designed to determine an increase in muscle power. The rationale for integrating aerobic exercise with strength training derives from the observation that most human activities are characterized by both static and dynamic muscle work, in addition to contributing to work and social life activities. Strength work in the cardiopathic patient is not addressed to the development of hypertrophy and fast strength but must be aimed at the search of resistant strength with exercises, as we will see later, of low intensity, numerous repetitions and longer recovery times between series.

The mode of execution is related to the availability of equipment and risk stratification of patients: tendentially, patients at low risk and with a good tolerance to effort, after the first 2 weeks of aerobic training, can gradually begin a strengthening work with weights.

4. **Time**: During the first phases of rehabilitation, the single training session is generally set at around 30 mins, while in the later phases and especially in home maintenance programs, the session can last up to an hour. The single session is generally organized in 10′ of warm-up, 40′ of actual training, and 10′ of fatigue or recovery. Longer periods of exercise are not warranted. The overall duration of the training program must be planned according to the objectives set for each patient: the standard protocols of the main cardiology centers in Italy and abroad provide for a minimum of 12 sessions to a maximum of 40 sessions, in periods between 2 and 8 weeks.

   The progressive increase of the workload is a function of numerous variables that make it difficult to standardize a protocol. The simplest and most direct reference to regulate the progression of the program is the heart rate: the improvement of the response to the effort corresponds to a smaller increase in HR per equivalent load. This allows you to increase the workload until the target heart rate is within the set range.

   Placing in a graph on the y-axis the trend of the heart rate during the rehabilitation or training session and on the x-axis the progressive increase in intensity or time of the single work session, it is possible to monitor the improvements of

the cardiovascular system obtained and then customize the physical activity program.

Basically, the more the slope of the resulting line is reduced, the more the subject is trained; moreover, the greater is the cardiovascular adaptation to the effort, the lower will be the HR at the same load intensity.

The safety of physical training and the timing of initiating the rehabilitation program in the ACHD patient are a controversial topic.

Although the ability to perform prognostic stratification is now far more thorough, allowing the degree of risk of the patient to be established with sufficient sensitivity to minimize the risks potentially induced by exertion, the diversity and complexity of ACHD patients make the conclusions of the literature on the incidence of major complications during rehabilitation scarcely indicative.

On the topic of safety, attention is drawn to the following observations:

- It is necessary to consider multiple parameters of the response to exertion: linearity of cardiac frequency progression, body pressure (BP) behavior, ECG recovery phase, and patient's perception of fatigue.
- It is advisable to follow carefully the adaptation to the effort in the initial sessions of the program, in which the physical deconditioning or the difficulty in learning the pattern of execution of the exercise can determine an abnormal increase in heart rate (HR) and BP.
- It is recommended to adapt the modalities and the type of exercise to the physical capacities and motor aptitudes of the subject, up to a total customization of the program.
- It is necessary to be able to count on a staff (rehabilitation therapists, physiatrists, cardiologists, cardiac surgeons) specifically trained and updated and able to recognize those situations that require the immediate suspension of exercise (e.g., appearance of angina or complex arrhythmias), while in the case of subjective sensation of fatigue or exceeding the CF of training is sufficient to reduce the workload or lengthen the recovery time between exercises.

What has been stated above outlines the basis for the design and execution of a standard protocol of physical activity for cardiac patients and for heart patients.

The rehabilitation program is part of a therapeutic intervention whose main objective is to improve the quality of life, the subject's perception of his own state of health, independently of the actual clinical conditions. The resumption of motor activity and the consolidation of a better tolerance to effort obtained through training play a decisive role in maintaining a condition as close as possible to the "state of health," and this is all, the more important, the more precarious the physical, psychological, or social situation of the patient. It is therefore understandable that rehabilitation, and physical exercise as the central expression of the program, has a privileged position for the categories of patients most compromised, and therefore at greater risk of poor quality of life, which until a few years ago were excluded from rehabilitation itself because they were considered at high risk.

In practice, Cardiac Rehabilitation should take place in stages.

Phase 1 applies to hospitalized patients, often with a combination of low- to medium-intensity exercise, stress management techniques, and risk factor education programs. The program in this phase aims to ensure that the patient is discharged with the best possible physical and psychological conditions, with all possible information on a healthy lifestyle.

Phase 2, in which the exercise program is further individualized in terms of intensity, duration, frequency, type, and progression, as well as constant monitoring, is aimed at returning to social and professional activities. In some situations, cardiovascular rehabilitation can be performed on an outpatient basis.

Advantages of outpatient rehabilitation:

- Recovery in the usual environment.
- Involvement of the family in lifestyle adjustments.
- Assumption of greater self-responsibility without the fixed rules of a hospital.
- Ability to perform regular physical activity in a family environment.
- Disadvantages of outpatient rehabilitation:
- Danger of reexposing oneself too soon to professional and social stress.
- Less rigorous surveillance, so this type of rehabilitation is not for everyone.

In a rehabilitation center, the patient is assisted individually by the interdisciplinary team. The most important element is the physical activity program appropriate to the patient's efficiency.

Advantages of rehabilitation with permanence:

- No professional or domestic stressful situations.
- Global assistance.
- Thanks to continuous surveillance, more demanding physical activities are possible.

Disadvantages of rehabilitation with permanence:

- Separation from family and the usual environment for a few weeks.
- Less involvement of the family in lifestyle adjustments.
- The rules of the rehabilitation center with permanence reduce self-responsibility.

Phase 3 may or may not follow the previous one. Improving the physical condition is the main goal as well as improving the quality of life.

Phase 4, following long-term programs, has the main objective of increasing and maintaining physical fitness. Activities are not necessarily controlled, and sufficient time should be available for maintaining the exercise program and preference for recreational sports activities.

REHABILITATION does not mean "letting oneself be assisted," but actively dealing with the experience with the disease, hospitalization, and learning what is good for the body.

14.2 Phases 1 and 2

Cardiac rehabilitation programs aim at the physical, social, and psychological improvement of individuals, and studies show the reduction of anxiety and depression [9]. During immediate postoperative hospitalization (Phase 1), physiotherapy aims to avoid the negative effects of prolonged bed restraint, stimulates a quick return to daily activities, maintains functional capacity, develops patient confidence, reduces psychological impact, maximizes the chances of early discharge, and provides the foundation for a home program [10].

In Italy, the majority of congenital patients, after surgery, do not have access to cardio-specialist rehabilitation courses. This is mainly due to an excess of protection and uncertainty about how to carry out such rehabilitation, which in turn translates into an encouragement to a sedentary lifestyle, and to the further uncertainty about what physical activity and at what intensity it should be recommended.

The cardiopulmonary stress test (CPT) represents the gold standard for a correct drafting of the Individual Rehabilitation Project with specific target intensity.

But in the immediate postoperative period, the limitations given by sternotomy and restrictive respiratory dysfunction make it impossible to use a CPT because it would in any case be interrupted prematurely.

Added to this was the impossibility of being able to perform it during the COVID emergency, a circumstance that however allowed us to reconsider some evaluation methods [11].

To evaluate the functional capacity of each individual patient, stratify it in a correct training profile and create a targeted intervention in the absence of a CPT, some physical performance tests (field tests) can be used.

Among the most used we mention:

- *Six-minute walking test* (6MWT) [12], a test that is not strictly standardized as it is influenced by the patient's cooperation and by the level of encouragement (which takes place every 60 seconds) and requires a lot of space (corridor length). It has a moderate correlation with the VO2 peak. It can underestimate the performance in patients with a moderate level of cardiovascular fitness. It can be a valid alternative to CPT in more compromised patients.
- *Incremental shuttle walk test* (ISWT) [12]. The patient is asked to walk along a 10-m path marked by cones at the pace defined by an acoustic signal that increases the pace every minute. The patient, unlike the 6MWT, is not encouraged and ends when the cone is not reached or when symptoms occur. Being an incremental test, it correlates better to peak VO2 than 6MWT. Producing a cardiovascular response more similar to CPT, it is a valid alternative for assessing exercise tolerance.
- *Step test* [13–15]. One needs only a step or another solid surface as well as a method of counting time and tracking steps.
- *Gait speed test* (GST) [16]. It accesses an individual's functional mobility. It is simple to perform, and it requests minimal space, equipment, and time.

- *Sit to stand* (STS) [16]. It is a way to access in individual's legs strength and endurance by having them stand up from a sitting position repeatedly over the course of the seconds of the test.
- *Short physical performance battery* (SPPB) [17], a short battery of tests to evaluate the functional capabilities essential for the autonomy and performance of the lower limbs.
- *Time up and go* (TUG) [18]. It is the time that an individual uses to stand up from a sitting position, walking for 3 meters, turning of 180°, come back, and sit down.

Some test batteries were developed and validated as correlations of the underlying fitness domains, while others were developed and validated as predictors of disability and hospitalization. Making them requires little space, equipment, and costs; they can be administered by nonmedical personnel with minimal training. They are considered extremely safe in patients with clinical conditions to compensate.

The most used are excellent screenings of functional limits associated with a worse state of health.

They help in assessing the functional status of patients, especially at certain points in their medical history.

14.3 Exercise Prescription

The general principles of exercise prescription apply to adults of all ages. The relative adaptations to exercise and the rate of improvement of the components of physical fitness are important for maintaining health, functional capacity, and mitigating many of the physiological changes associated with congenital heart disease.

Low aerobic capacity, muscle weakness, and deconditioning are more common in this type of patient, and therefore the prescription should include aerobic, muscle-strength-endurance, and flexibility exercises. They may also benefit from specific neuromotor exercises to improve balance, agility, and proprioceptive training (Tai Chi Chuan), in addition to the other components of health-related fitness. Age should never be a barrier because positive improvements are possible at any age.

Physical exercise DOSE is defined as the total sum of energy expended, determined by the intensity, frequency, and duration of the activity performed. The MET is the most used unit of measurement which, multiplied by the duration and frequency of the exercise, provides the total sum of the energy expended. This is one of the most popular ways to identify the "dose." But in reality, it does not take into account individual variability, such as the different subjective effort between a trained patient and a deconditioned one. It is therefore difficult to identify a volume of physical activity that can be standardized for each individual person.

For performing adults, moderate-intensity and vigorous-intensity exercises are defined as such compared to METs, with moderate-intensity activities defined as 3–5 and vigorous-intensity activities such as ≥6 METs. Conversely, for more deconditioned adults, activities should be defined in relation to physical fitness in the

context of a perception scale of physical exertion (Borg or OMNI). Moderate intensity exercise should produce a minimal increase in heart rate (HR) and breathing, while one of vigorous intensity should produce a large increase in HR and breathing.

Considerations that should be made to maximize the effective development of an exercise program in deconditioned patients are:

- Increase muscle strength before you are physiologically able to engage in aerobic training.
- Intensity and duration of exercise should be mild at the beginning.
- The progression of the exercise should be personalized and adapted to tolerance and preferences; a conservative approach may be necessary for patients who are severely deconditioned or physically limited.
- If the clinical conditions prevent activity at the minimum recommended amount, the exercise must be performed "within tolerance."
- The initial load should be light (<3 MET), and the workload increases in the order of 0.5–1 MET.

It is desirable, after a correct stratification of patients, to apply the principle of "accelerated rehabilitation" and to start physical conditioning as soon as possible. Patients already deconditioned in the preoperative period or those who have undergone longer surgical times will hardly be able to tolerate training times longer than 10 mins in the immediate postoperative period: in these cases, it is useful to divide the treatment into at least two daily sessions. Once hemodynamic stability has been reached, the patient must be guided to use a pedal mini bike to be performed in the room even without supervision in order to create a "training volume effect" during the day.

Subsequently, the stepper (10–20 cm high step), a low-cost and easily transportable tool, usable in supervision, allows an infinite range of training and combinations of exercises. Initially performed slowly and for a few repetitions, it retrains strength, balance capacity, and, over the course of the days, it can be combined with numerous other exercises before and after the climb. The patient must be able at discharge to climb the stairs to return home. It is a tool that can also be used at home.

During training, in the absence of CPT performed, the rate of ascent of the heart rate must always be monitored, and with what progression, it is increased. Peripheral saturation control oxygen, blood pressure, Borg scale dyspnea (0–10), and perceived exertion (6–20) or alternatively Omni scale [19] are always monitored.

14.4 Phases 3 and 4

The reduced physical performance resulting from a sedentary lifestyle represents one of the most important negative predictors for the state of health: in fact, poor tolerance to effort is associated with a significant increase in mortality.

In the first two phases, in the absence of contraindications, the objectives include "preparing" the patient to tolerate an aerobic training [20, 21] with a training phase

lasting at least 20–30 mins. Considered the minimum level recommended by the guidelines, each program should be customized so that each patient does not only perform the type of physical activity preferred but that the modalities are those that are best suited to his degree of physical performance. It is useful to create structured protocols calibrated on the patient's level of availability, designed to help practice physical activity in daily life. Goals should be attainable and quantifiable by the patient himself, reinforcing successes and preventing dropouts. The message that must be conveyed is that physical activity must be promoted and must have the same importance as any other therapeutic treatment, and, to be effective, it must be done regularly throughout life.

The physical activity of aerobic resistance (endurance training), of light or moderate intensity (150 mins per week), can be divided into five weekly sessions of 30 mins and can be added to the activities of daily life if lasting more than 10 mins.

As for muscle strength training, when indicated, it can be trained two times a week, associated with aerobic activity, with 8–10 exercises for the main muscle groups in the number of 8–12 repetitions per exercise. These exercises should be performed without tearing and without holding breath to avoid pressure spikes.

Create groups of physical activity, associations between patients, forums on the Internet, applications, or anything that the construction of a "network" can give as an aid to the execution and continuation of the physical conditioning program.

In the third millennium, it would be desirable to create digital media, for the optimization of the prescription, the control of the activities carried out, and the timely resolution of any critical issues. The development of applications that can be easily downloaded on a telephone or computer would allow the patient not only to have indications on the type of training to be performed, but interacting with it could obtain instant variations on the workloads and the type of exercises to be performed, as well as see the objectives achieved and above all, by creating a feedback with the interdisciplinary pool that follows him, perform "remotely" (perhaps in supervision with video call) the functional tests used during the hospital stay. It should be remembered that various hospitals during 2020–2021 were forced to use this method to follow the changes in the clinical and fitness conditions of COVID patients previously undergoing physiotherapy during hospitalization after weeks [22, 23].

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Promotion of Daily Physical Activity and Exercise for Adults with Congenital Heart Disease

15

Hajar Habibi

15.1 Background

Congenital heart disease is a structural abnormality of the heart and/or great vessels that is present at birth and the most frequent congenital malformation diagnosed in children. The incidence is 1.1% of all newborns with heart defects resulting on over 90% of infants surviving into adulthood in the developed world [1, 2]. As this patient population gets older, their cardiac health can additionally be affected by acquired cardiovascular disease highlighting the need for awareness of risk factors to lead a healthy lifestyle.

The importance of physical activity, formal and unintentional exercise, should be communicated at a young age [2]. Physical activity is defined as 'any bodily movement produced by skeletal muscles that requires energy expenditure' [3].

As patients with CHD live longer, the focus has moved towards morbidity and functional statues rather than mortality alone. Exercise capacity is a predictor of long-term morbidity in these patients as a result of risk factors such as obesity and hypertension. Physical activity and exercise should be a regular part of everybody's daily life including patients with CHD [4]. Therefore, the preventive role of physical activity is the same in ACHD as in the general population [5]. ACHD patients are also at risk of developing obesity. Obesity and lack of exercise, among ACHD patients is multi-factorial and includes factors such as parental or environmental overprotection, health fears and misconceptions, poor role modelling and increased psychological stress. All of these can affect health adversely and trigger incidence of metabolic syndrome, diabetes mellitus and cardiovascular disease [6, 7]. Understanding why some ACHD patients are inactive can help plan physical activity programmes. There are multifactorial aspects to be considered such as biological

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(sex, ethnicity), psychological (behaviour, motivation), sociocultural (parental/family, peers influence) and environmental (access to facilities). In addition to these factors, there may be restrictions imposed by clinician due to fears about possible proarrhythmic potential of exercise [8].

The more physically active a person is, the greater the cardiovascular benefit would be. The general population is advised to do at least 150 minutes of moderate intensity aerobic activity per week. This also could be a target for CHD patients. The best and safest types of exercise are aerobic activities where the muscles use oxygen to extract energy from glucose and fatty acids. These increase the heart rate and result in heavy breathing [7]. Therefore, low−/moderate-intensity aerobic exercise is largely safe and effective for most ACHD patients. Examples include brisk walking, swimming, cycling, jogging, rowing, cross-country skiing, hiking or stair climbing. Team or court sports such as basketball, football, tennis, squash and volleyball are also aerobic activities [9].

15.2 Physical Aspect

Regular physical activity can delay progression of systolic and diastolic heart failure and coronary artery disease [8]. Over the last few decades, guidelines have focused on structured exercise to promote more moderate intensity. Such exercise regime can be incorporated into everyday life and include cycling, walking, gardening, taking stairs or cleaning [10]. Nurses should be aware of the importance of support and encouragement from the patient's family as that could have a great impact on ACHD patients being physically active [11].

In the past, ACHD patients were advised to restrict physical activity due to concerns that increased activity may be dangerous. But the sedentary lifestyle associated with this belief of 'self-protection' causes a decline in physical activity and puts the patients at risk of early cardiovascular disease [1]. The benefits of exercise for ACHD patients, within reason, outweigh a vast majority of the perceived risks. Physical activity improves health, exercise tolerance, quality of life, as well as enhancing emotional, social and cognitive skills. Exercise is a high-benefit/low-risk intervention for a majority of ACHD patients and therefore should be introduced to ACHD patients and their family early on during the transition process [7, 12, 13].

The effects of exercise are decrease in triglyceride levels, increase in HDL cholesterol level, decrease in blood pressure, reduction in insulin resistance and improvement in glucose control. Exercise also minimises the incidence of depression and social isolation. Combined, all these beneficial effects play an important role in the status of an individual's health [13, 14].

Consequently, there is an inverse relationship between physical activity and the rate of mortality and cardiovascular death [15, 16]. Some of the benefits of physical activity:

- Decreases risk of developing hypertension.
- Decreases cholesterol levels.
- Achieving and maintaining a healthy body weight.

- Improves mental, emotional and cognitive function and quality of life.
- Decreases morbidity and mortality associated with cardiovascular disease.
- Maintains functional fitness and improves self-confidence and body image.
- Increases the efficiency of muscles so they require less blood flow from the heart.
- Reduces the risk of coronary heart disease.
- Strengthens the heart muscle.
- Improves skeletal and diaphragmatic muscle strength (important in Fontan circulation).
- Improves sleep.

Nurses can discuss and raise awareness in the primary prevention methods, such as keeping active, to reduce the risk of being diagnosed with further heart disease. Therefore, promoting physical activity should be discussed at each clinic visit [17]. Even in patients in whom isometric activities may be restricted (left ventricular outflow tract obstruction, hypertrophic cardiomyopathy and aortopathy), dynamic activities of low-to-moderate intensity can have important health benefits [18]. The best and safest type of exercise is aerobic activities which increase the heart rate and may enhance myocardial reserve and improve enhance cardiac output during exercise [19]. Improving exercise capacity through aerobic training is probably worthwhile in almost all ACHD patients [20].

Nurses can educate and support patients that moderate-intensity activity noticeably increases the depth and rate of breathing. They can start in shorter bouts, such as three 10-mins sessions, then gradually increasing both intensity and duration [15], advising patients against doing too much physical activity too soon and to start slow and building up and starting with approximately 10 mins and gradually building by about 10% a day. This can be achieved by working collaboratively with physiotherapists, nutritionist, psychologist and other key persons in the patient's life [21].

15.3 Social Aspect

The aim of physical activity should be to motivate patients, provide pleasure and promote social cohesion while considering patient preferences. In most cases, having fun is one of the main reasons patients remain physically active, and this is the area that ACHD nurses can influence patients' behaviours [22]. Therefore, exercise should have a fun element and address patients unanswered questions and fears such as level of breathlessness [8].

This is the area that nurses can have an important role in creating a safe environment, being available, approachable and providing appropriate advice to enable patients to initiate and increase their physical activity [11]. We should also consider the social network and their support that could further facilitate behavioural change within the community such as relevant local, national and international charities for ACHD patients [23].

15.4 Psychological Aspect/Quality of Life (QoL)

Quality of life (QoL) is a complex entity; it is reported that a high level of physical activity such as training for over 3 hours per week is associated with a higher level of QoL. Many factors associated with low confidence in ACHD patients are modifiable and can translate into improved quality of life [24]. The attitude of both physicians and patients can influence inactivity more than the severity of the heart condition or level of physical fitness [8]. The other aspect to consider from a psychosocial perspective is the experience of impaired capacity compared to peers that could affect ACHD patient's self-confidence. Psychosocial barriers such as lack of self-confidence could affect patients taking part in physical activity; this is important as an improvement in the exercise capacity has been shown to be correlated with an improvement in quality of life [25].

Nurses need to consider nutritionist support as chronic stress due to psychosocial challenges in ACHD patients may alter food intake and in turn can cause stress-induced sympathetic and neuroendocrine activation contributing to increased fat storage and consequently increase cardiovascular risk factors [6]. Therefore, ACHD patients can benefit from nutritionist support to help them to maintain their body weight through physical activity and consequently decrease mortality [26].

Current research shows that there are a considerable number of ACHD patients experiencing psychological distress, such as anxiety, depression and low self-esteem, all of which have been identified as predictors for an increased risk of obesity. This has highlighted the importance of screening for early detection and timely referral for psychological input to enable patients to make better informed decisions [6].

Any type of physical activity can have associated improvements in self-esteem, behaviour and emotion. Even simple physical activity like regular walking is feasible and safe [2]. Current guidelines indicate that most ACHD patients should have no restrictions on participation in physical activity [27–29].

Integrating physical activity programmes within ACHD services can improve access, compliance, tracking, frequency of counselling and patient confidence which ultimately will improve QoL and consequently long-term outcomes for ACHD patients [30]. Another aspect to consider is that exercise capacity may also guide and help in counselling women with CHD contemplating pregnancy [31].

15.5 Musculoskeletal Benefit

Congenital heart disease (CHD) encompasses a heterogeneous group of conditions. Reduced muscle mass and function is common in ACHD patients and can contribute to decreased tissue oxygen uptake [32]. They may also have reduced aerobic exercise capacity and reduction in skeletal muscle mass and muscle strength [32].

The complexity of the heart disease, associated with the extent of muscle dysfunction due to skeletal muscle mass, is correlated with peak VO2 and could be due to a variety of causes including neurohormonal activation, low cardiac output,

cyanosis, endothelial dysfunction, hypoactivity and high prevalence of scoliosis and/or kyphosis [33].

Patients with complex congenital heart disease may have impaired isotonic limb muscle function compared with patients with a simple lesion. Muscle dysfunction is associated with impaired exercise capacity. This could be the result of previous interventions, impaired respiratory function, ongoing medications, inappropriate advice relating to physical activity and parental/family overprotection [34].

Kyphosis is prevalent in ACHD patients due to musculoskeletal or orthopaedic problems, particularly deformities of the spine after sternotomy or thoracotomy such as scoliosis [35]. Aerobic exercise increases musculoskeletal strength and fitness [8]. Physical activities should target in improving muscle function and patient confidence such as Pilates or yoga which also are associated with improvement in self-esteem, confidence, quality of life and social skills [8, 34].

15.6 Respiratory Function Benefit

Abnormalities in respiratory function such as a tracheobronchial tree, lung parenchyma, pulmonary vasculature, chest wall and reduced maximum inspiratory and expiratory pressure are sometimes prevalent in ACHD patients and can compromise respiratory muscle strength. This ventilatory limitation has been also linked to surgical scarring, thoracic deformation, phrenic nerve injury and deconditioning, all of which could consequently be contributing to an overall impairment of physical capacity [33, 36].

ACHD patients have been reported to have restrictive lung disease and lung function abnormalities. Respiratory function is particularly important in patients with single ventricle physiology and Fontan type circulation, in which systemic venous return depends, in part, on negative intrathoracic pressure during respiration. The challenge for clinicians is to ensure physical activity is tailored to the individual's clinical status by assessing and then negotiating regular physical activity [37, 38].

15.7 The Fontan Circulation

In the Fontan circulation, cardiac autonomic nervous activity is reduced even without signs of heart failure (26). The autonomic nervous system in CHD patients has shown that they have a blunted heart rate response during exercise and delayed early heart rate recovery which are both associated with impaired sympathetic and parasympathetic cardiac autonomic nervous activity [8, 39].

Following the Fontan procedure, patients may benefit from inspiratory muscle training. Objectives should be individualised taking account of arrhythmias, significant ventricular dysfunction, elevated pulmonary pressure, decompensated heart failure, severe outflow tract obstruction, aortic dilatation and chronic pains [8, 33]. Since most patients with Fontan physiology have some degree of cyanosis, an increased haemoglobin concentration is necessary for adequate oxygen delivery to tissues; therefore, it is essential for nurses to check the iron level to prevent iron deficiency which has an impact on the exercise capacity [25].

15.8 Impact of COVID-19

During the COVID-19 pandemic, there has been a report of a decrease in the level of physical activity. This could be due to an increase in sedentary time such as sleeping and screen time [40]. Physical deconditioning and sarcopenia from bed rest and physical inactivity during COVID-19 might strongly impair the metabolism. This together with an increased consumption of unhealthy food could increase the prevalence of obesity [41, 42].

The complexity of physical, emotional, social and financial challenges that individuals faced during the pandemic could also influence food purchasing and food consumption habits. This could be a positive experience in terms of family's food behaviours (e.g. home cooking and increased attention to diet) but, in contrast among vulnerable groups, could have caused issues with affordability, leading to purchasing and consumption of cheap energy-dense foods. The concern would be that due to the bidirectional relationship of anxiety and depression with obesity, chronic fatigue and post-traumatic stress, symptoms may induce weight gain and thereby increase the risk of cardiovascular disease [42].

15.9 Impact of Technology

Current technology presents exciting opportunities utilising commercial wearable trackers, such as Fitbit and Apple watch, which can be useful in clinical monitoring of the patient's condition as well as for research purposes [43]. Promoting self-monitoring through fitness wearables and data sharing with their healthcare centre may motivate ACHD patients to adjust their levels of training [7]. Patients can start at a low level and steadily increase exercise intensity, training frequency and workout duration. Nurses can be a port of contact for patients on providing education and continuous support on having a healthy lifestyle [22, 44].

15.10 Nursing Assessment

Nursing assessment is the foundation of effective nursing care and integral to patient care which requires good cooperation between the healthcare team. The assessment includes systematic and continuous data collection, sorting, analysing and organising the relevant data to obtain information to provide an individualised care plan [45].

The role of the ACHD CNS includes education on preventive care, health coaching and chronic disease management where subjective and objective data collection are an integral part of this process [4]. ACHD CNSs need to be involved in the assessment process to provide a concise exercise regime that is individualised to each patient to encourage a physically active lifestyle [29].

One of the ways nurses can find out more about individual patients' physical activity is using a baseline questionnaire for better understanding of patients' daily

routine that can aid discussion and motivational interviewing where nurses help to improve short- and long-term patient engagement with active lifestyles and thus strengthening an individual's motivation for change [46].

This could be The General Practice Physical Activity Questionnaire (GPPAQ) which is a short self-reported brief measure of physical activity in patients that provide information in categories of active, moderately active, moderately inactive or inactive [47]. The discussion can be carried out with ACHD patients during clinic appointments in a variety of ways including waiting room questionnaires, promotional posters and videos [21].

The effect of ageing in ACHD patients needs careful attention. Physical activity includes basic daily tasks such as playing, working, house chores and recreational activities, all of which should be recognised as part of cardiovascular health. Activities of lighter intensity, such as walking, provides significant cardiovascular health benefits even if there are no changes in objective measures of exercise capacity [8, 11].

Nurses can contribute in developing a comprehensive assessment for a personalised exercise regime for each ACHD patient using a specific algorithm and gathering relevant information [28].

This could include:

Taking the patient's medical history (including medication):

- Carrying out a physical examination (including observation).
- Performing an ECG.
- Holter monitor if history of or risk of arrhythmias.
- An assessment of **five** baseline parameters that can be obtained from their investigations (such as echocardiogram, MRI, Holter) to assess ventricular function, pulmonary artery pressure, aortic size, arrhythmia, saturation at rest/during exercise.
- The type and relative intensity of each exercise as well as cardiopulmonary testing.
- Six-minute walk test (6MWT).
- Follow-up and review to discuss the progress.

Assessment includes a history of chest pain, dyspnoea, palpitations, arrhythmias, device implantation and the presence of syncope [48]. Exercise intolerance such as dyspnoea and fatigue on exertion is the most common complaint made by ACHD patients. That can be assessed either subjectively by the New York Heart Association (NYHA) classification or objectively by using a cardiopulmonary exercise test (CPET) or 6-minute walk test [7].

Patients with reduced ventricular function due to their cardiac condition are recommended to perform lower levels of exercise. The same low-level exercise recommendation applies to patients with pulmonary hypertension and dilated aortic dimensions as intensive exercise may be associated with syncope and in turn could be life-threatening [49].

The exercise test objectively quantifies the maximal oxygen uptake (peakVO2) measurement and documents any adverse responses to exercise in a closely supervised environment. If CPET is not accessible, then a 6-minute walk test (6MWT) is a simple, economical and a safe test that is moderately correlated with peak VO2 in ACHD patients which can be done by the ACHD CNSs [22, 33].

Nurses are in a key position to counsel the ACHD patients on the importance of engaging in healthy lifestyle behaviours such as eating a healthy diet, participating in regular physical activity, getting adequate sleep, reducing stress and avoiding tobacco and excessive alcohol intake [50]. Nurses can initiate discussion and assessments about physical activity including topics like sex, age, exercise experiences, preferences, skills, motivation and training goals. Then an individualised training schedule should be designed by the ACHD consultant with input from other teams.

Members include physiotherapists. The tailored programme should include recommendation on frequency, intensity, workout time as well as the type of exercise (FITT principle) [22].

Finally, a thorough assessment in an ACHD specialist centre is needed to ensure that all relevant areas are taken into consideration prior to exercise prescription and the dynamic and static components tailored to the individual patient assessment with regular review [7]. When all the relevant investigations are available, then the ACHD CNS can use the Performa (Table 15.1) in order to facilitate the prescription of the physical activity plan.

Table 15.1 (Assessment (form) carried out by the ACHD CNS)

Exercise/physical activity assessment form completed by the adult congenital heart disease (ACHD) clinical nurse specialist
Date of assessment
Patient's name
Date of birth and hospital number
Congenital heart disease (CHD) lesion and relevant medical history
Baseline assessment (ECG, SpO2, BP, NYHA class)
Baseline blood test: HB, iron level, BNP, INR
Level and duration of current activity, interest area
Relevant medication: e.g. warfarin
Latest cardiopulmonary exercise test (CPET) (date and summary)
Latest echocardiogram (date and summary)
Latest 6-minute walk test:6MWT (date and summary)
Devices: Permanent pacemakers, ICD yes/no
Lung function needed yes/no
Further investigation needed yes/no
Referral needed YES/who to refer/no
To the named cardiologist
To physiotherapist
To rehabilitation team
To psychology/counselling team
To nutritionist

Table 15.1 (continued)

Exercise/physical activity assessment form completed by the adult congenital heart disease (ACHD) clinical nurse specialist
Clinical nurse specialist (CNS) and consultant discussed and recommended on types of exercise: Low/moderate/vigorous
Types of exercise to avoid:
Information leaflet/website address/charity support
Review in 3/6/9/12 months
Additional comments
ACHD CNS details

15.11 Role of the ACHD CNSs

The increasing prevalence of obesity has led to global concern about physical inactivity and how to promote active lifestyles involving multidisciplinary team members. CNSs are in an optimal position to be influential in increasing levels of physical activity among ACHD patients across their lifespan [44]. ACHD CNSs have frequent contact with patients and the ability to develop trusting therapeutic relationships, listening skills to encourage and engage with patients during their clinic visit by talking to them to find out how physically active they are in their daily lives therefore triggering awareness about health behaviour and inspire and motivate patients to make informed choices about their health [51]. This can be achieved by identifying and addressing barriers, encouraging patients to set achievable goals and monitoring their achievements [24]. The goal of physical activity should be realistic considering each individual's culture and personal factors to help the individual build self-efficacy, and the ACHD CNS can facilitate a team-based care [24].

Even though exercise is considered safe and beneficial for almost all patients, still hesitancy remains among physicians regarding the safety and efficacy of regular exercise in ACHD patients with complex lesions. Some of the reasons given are they do not have the knowledge, skills, resources or time needed to implement, promote or counsel regarding physical activities [8]. ACHD CNS can have a positive impact on the future health of ACHD patients by discussing with them on the impact of their lifestyles on their health outcomes [52]. This can be done by explaining the rationale for the physical activity recommendation and informing them of the warning signs and symptoms such as exertional dizziness, persistent palpitation or chest pain [7]. Nurses can advise patients about **talk test** which is a simple way to measure the intensity of their physical activity and reassure them that if by increasing their physical activity they can still carry on a conversation that they are likely to benefit from a safe level of activity [53, 54].

One area that nurses can be involved in is the transition stage when young adults transfer from a paediatric setting into the adult care system. This is a critical time to introduce and reinforce the habit of a healthy lifestyle [7]. Any contact with an ACHD patient is an opportunity for a discussion about physical activity which can

help and motivate a patient to become more physically active. This can be done by asking simple questions like, 'What exercise do you enjoy and undertake regularly?' The question can then be followed by an assessment of the patient's ideas about becoming more active, followed by goal setting and planning [21]. One way of working jointly with ACHD patients and incorporating their personal preferences is to use strategies to help make the physical activity goal realistic and reasonable by using the SMART method (**S**pecific, **M**easurable, **A**chievable, **R**ealistic, **T**ime-specific). In this way, both nurses and patients can create achievable plans that permit consistent evaluation of progress and reviewing the progress [55]. This goal setting can improve motivation and adherence to an exercise programme [56].

Physical activity counselling combined with supporting written material can encourage patients to increase their activity. Nurses working with the multidisciplinary team such as the physiotherapist can develop a programme and template for mix upper limb, lower limb and trunk/core exercise programme [11]. This can be sustained by episodic follow-up, reinforcing positive behavioural change and referral to community programmes [15]. Other programmes could be ACHD CNS's planning an informal physical activity day. Patients and their family/carer can attend the day to talk to the multi-military team about the benefit, available resources such as relevant health department websites, charities and online physical activity resources and toolkits. This will help to create a safe forum for ACHD patients to talk about their fear or any concern they might have about physical activity [57]. This will enable the ACHD patients to meet with people in similar circumstances from the ACHD community to engage in physical activity and to build relationships and social network [46].

15.12 Executive Summary

There is widespread recognition of the pivotal role that nurses play in promotion of physical activity and exercise for prevention and management of cardiovascular disease. The range of benefits associated with regular physical activity is irrefutable.

The emerging population of adults with CHD is ageing and living longer. Nurses as part of the wider team need to address preventive cardiac care to empower patients to make informed decisions with respect to cardiovascular health and health care, particularly when it comes to nutrition and exercise.

A sedentary lifestyle is one of the most important modifiable risk factors for morbidity and mortality. Physical activity is a key element in the prevention of cardiovascular disease. Exercise training not only improves functional capacity and mental health but also promotes and maintains positive self-esteem.

Nurses, as part of key team members, are ideally placed to play a greater role in patient education and empowerment where it is related to physical activity and the harmful effects of a sedentary lifestyle. It is paramount to tailor advice regarding the intensity of the exercise programme for each individual ACHD patient. The importance of the recommendations and healthy physical activity should be highlighted

and formalised in the form of exercise prescriptions. Patients may require baseline assessment to guide the team to prescribe an individual plan for them.

In addition to nutritional guidance, psychological counselling might be necessary to reduce psychological distress. It is important for early detection and management of psychological issues to ensure patients remain empowered and are able to make wiser decisions regarding their physical health, in turn improving their long-term outcomes. Commercial activity trackers can be individualised to provide a unique way of monitoring physical activity levels, with the possibility to integrate physical activity and counselling and optimising cardiovascular health in the ACHD population.

There is much uncertainty about the optimal strategy, timing and format (e.g. home-based working, new technology) of exercise interventions; therefore, more research is needed in this field, and ACHD nurses should be part of that to confirm the survival benefit with respect to physical activity in ACHD patients.

Exercise is safe in the vast majority of ACHD patients and deliverable where patients are given guidance, structure and support. The intensity of the exercise recommendation should be adjusted according to individual assessment and regular review to adjust the exercise prescription. This is an area that needs further investment and exploration with the wider national and international collaboration which may ultimately assist in improving the quality of life and long-term outcomes for ACHD patients.

Physical activity assessment and counselling should be an important aspect of holistic care for all ACHD patients. Integrating physical activity programmes within ACHD services may improve access, compliance, tracking and confidence; therefore, continuous support and follow-ups should be planned.

Nurses should enhance patient knowledge and help improve their understanding of the multiple benefits of regular physical activity and strongly advocate the importance of an active lifestyle with a focus on abilities. This is also key to creating a feeling of safety for patients of every ability and at all ages from transition into the old age.

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Part VII

Patient Self-Care and Nursing Role for Patients' Education

Connecting Dots for Framing Health: The Self-Care Process

16

Federica Dellafiore, Cristina Arrigoni, and Barbara Riegel

16.1 Introduction

Self-care refers to a process of maintaining health through health-promoting practices and managing illness when it occurs [1]. This seemingly simple idea is more complex than initially thought, especially for individuals with congenital heart disease (CHD). As described in this chapter, the self-care process itself is multidimensional, and the switch of self-care from family to the individual as the patient ages makes self-care even more complex for those with CHD [2].

This definition of self-care reflects the Middle Range Theory of Self-Care of Chronic Illness [1]. The theory evolved from the Situation-Specific Theory of Heart Failure Self-Care because the theorist was repeatedly asked permission to use the situation-specific theory with varied patient populations [3]. Joining with other experts in self-care, Professor Tiny Jaarsma and Professor Anna Stromberg, Riegel and colleagues developed a theory designed to address the self-care needs of the wide variety of individuals with chronic illness. That theory, originally published in 2012, was updated in 2019 [4]. The update was prompted by further consideration of the importance of symptoms in the self-care management process described further below.

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16.2 What Is Self-Care?

Regardless of the chronic illness under consideration, self-care is characterized as a process with three sets of behaviors: self-care maintenance, self-care monitoring, and self-care management [1]. Self-care maintenance is defined by those behaviors used by the patient with a chronic illness to maintain physical and emotional stability. Examples of self-care maintenance include taking medications as prescribed and routine exercise. Self-care monitoring involves self-observation for changes in signs and symptoms. Symptoms can be defined as subjective physical or mental experiences, appraised and defined by the patient, and reflecting an altered health state or change [4]. "Body listening" is a core process in self-care monitoring [5]. Self-care management is the response to signs and symptoms when they occur. That response may involve changing one's behavior in some fashion or consulting with others for advice. Self-care management is particularly challenging because people often have difficulty interpreting their symptoms; this interpretation is required before management can take place. These three behaviors are thought to be mastered in sequence because self-care maintenance is often easier to master than self-care monitoring and self-care management. The performance of adequate or sufficient self-care encompasses the constellation of all three behaviors.

A term commonly used in place of self-care is self-management [6]. The National Institute of Nursing Research, within the National Institutes of Health in the United States, defines self-management as strategies to help individuals with chronic conditions and their caregivers better understand and manage their illness and improve their health behaviors. This definition is very similar to the definition of self-care in the Middle Range Theory of Self-Care of Chronic Illness, so for the purposes of this chapter and searching the literature, the two terms are used synonymously.

Patients make decisions about whether or not to engage in self-care behaviors. Some of the self-care maintenance decisions are influenced by habit formation (e.g., dietary choices). But many of the self-care monitoring and management decisions reflect naturalistic decision-making processes. This type of a descriptive model of decision-making emphasizes that decisions change as the context changes [7]. Naturalistic decision-making is one descriptive model of decision-making that emphasizes that real-world decisions have ill-defined, shifting, or competing goals [8]. In such real-world decisions, the context may be uncertain, ambiguous, and evolving dynamically. The decision-maker is often missing information, the stakes are high, and the decision needs to be made quickly.

Since the original publication in 2012, the Middle Range Theory has been tested 26 times and referenced 361 times. The enthusiasm of the research community is probably due to the variety of self-report theoretically based instruments that are available free of cost to users through this website: www.self-care-measures.com. The instruments designed to reflect the Middle Range Theory of Self-Care of Chronic Illness address patients with hypertension [9], coronary heart disease [10], diabetes [11], chronic obstructive pulmonary disease [12], and general chronic illness or multimorbidity [13]. Because of the availability of these instruments, most

of the self-care research conducted to date has been done in these populations. Notably, many of these valid and reliable research instruments have been translated into other languages and made freely available to potential users.

16.3 Why Self-Care Is Important?

To date, the international research on self-care is surrounded by a lively and heated debate, illustrating the high interest of the scientific community in the beneficial effects of self-care on patients' and caregivers' health outcomes. The need for people with chronic conditions to take care of themselves and to assume responsibility for their own health is now recognized by researchers and clinicians as an imperative to stay healthy [14]. The first self-care publication was in 1946, and a considerable number of different definitions and theories have been published since then, in order to implement and support the birth of this new paradigm of care for chronic illness. These publications have centered on supporting and empowering persons with chronic conditions and their care partners to assume responsibility for their own health and to be actively involved in self-care.

Despite acceptance of the need for self-care behaviors to stay healthy, much of the empirical evidence has revealed the complexity of self-care, discovering several aspects that positively or negatively influence the decisions of chronically ill patients regarding taking care of themselves [14]. Emotions or mood states (e.g., depression [15], anxiety [16], and other forms of distress), personality traits [17], social [18], or interpersonal factors and clinical factors have been recognized as factors influencing the level of self-care performed. Evidence shows that financial worries, financial strain, and economic hardships are also sources of stress that may influence self-care [19]. When chronically ill patients don't implement self-care behaviors, the consequences can be dramatic: emergency admissions, recurrent hospitalizations, and complications that could have been prevented. Poor adherence to treatment and to the management of disease by chronically ill patients is the main cause of illness exacerbation.

Adequate self-care behaviors are the cornerstone of clinical pathways and care processes for persons with a chronic illness who are determined to stay healthy. Multiple benefits have been highlighted in chronically ill patients who engage in an adequate level of self-care. Particularly beneficial behaviors include limiting the intake of recreational drugs, monitoring of symptoms, adherence to diet, restriction of alcohol, physical exercise, and smoking cessation, which have been shown to limit hospitalization, control healthcare costs, and improve quality of life [20]. Higher levels of self-care involve adherence to taking prescribed medications, accomplished with support for the many common reasons for nonadherence to drug therapy described in the literature such as cost, attitudes about drugs, and the effect of some drugs on sexual function [21]. Higher self-care levels also involve greater adherence to dietary recommendations, and the restriction of sodium intake, especially in patients with severe symptoms of heart failure [22]. Often, chronically ill patients have problems adhering to a prescribed diet or poor eating habits, which

can further worsen their pathology [22]. Substance abuse, such as alcohol intake, drug use, and tobacco addiction may interfere with self-care behaviors [23]. Another self-care behavior of crucial importance is daily physical activity, which improves oxygen supply and reduces inflammation, even in people with severe heart dysfunction [24]. Furthermore, comorbid conditions pose an important risk factor for rehospitalization and readmission, especially for patients with chronic illness. For this reason, prevention and health promotion are essential in chronically ill patients, and self-care could be useful to prevent or help manage some lifestyle-related comorbidities.

16.4 What Is Known About Self-Care in Adults with Congenital Heart Defects?

In spite of the generic nature of self-care as described in the theory and the availability of research instruments measuring the construct, very few researchers have applied the concepts of self-care in the field of CHD. Only six published studies [25–30] and one unpublished doctoral dissertation were located [31].

Specifically, three articles focused on self-care status or self-care behaviors in children with congenital heart disease [25, 27, 28]. Nematollahi and colleagues [25] conducted a mixed-method study using a sequential explanatory design, assessing self-care status in 124 CHD children in Iran. They discovered that the self-care of these children was optimal, thanks to educational and supportive nursing interventions that enhanced self-care behaviors and reduced the complications associated with the disease [25]. Two studies by Christensen and colleagues [27, 28] centered attention on self-care management and the consequences of oral anticoagulation therapy, for example, rapid fluctuations in international normalized ratio (INR) values, interruption in daily life due to frequent hospital/doctor visits, and difficulties and pain due to the performance of venipuncture. The results of their studies demonstrated that self-management of oral anticoagulation therapy is safe and provides a good quality of treatment for selected children with CHD [27, 28].

Synthesizing the results of these few studies, the science of self-care is poorly developed in adult with CHD. McCabe and colleagues [26], Hays and colleagues [29], and Lee and colleagues [30] illustrate the challenges faced by these patients, but only Fleck [31] identified a theme related specifically to self-care: inconsistency in performing self-care. Specifically, Hays et al. [29] found that adults with CHD needed a plan for the future education about health and life expectations. But when Lee et al. [30] provided such a program, knowledge improved but self-care performance and health-related quality of life did not improve. Modifiable determinants of self-care were identified: self-efficacy, depressive symptoms, and perceived family support [26].

Additionally, only two articles of those mentioned above were guided by the Middle Range Theory of Self-Care of Chronic Illness [4]. The quasi-experimental study by Lee and colleagues [30] recognized that self-management must be practiced by ACHD patients and developed a self-management efficacy promotion

program testing the effects on disease-related knowledge, self-management implementation, and health-related quality of life [30]. Articles by McCabe and colleagues (2015) [26] were unique in that they provided an overall description of self-care maintenance, self-care monitoring, and self-care management of adults with CHD. Using an instrument based on the Middle Range Theory [4], they described the practice of self-care and identified possible antecedents of self-care in a cross-sectional, descriptive study with a sample of 132 adults with moderate or severe CHD. The investigators discovered that ACHD patients performed low levels of self-care; self-efficacy, depressive symptoms, and perceived family support were recognized as factors associated with self-care. Adequate self-care maintenance, monitoring, and management were found in only 44.7%, 27.3%, and 23.3% of participants, respectively. Moreover, self-efficacy, education, gender, perceived family support, and comorbidities influenced self-care maintenance; age, depressive symptoms, self-efficacy, and NYHA class influenced self-care monitoring, and self-efficacy and NYHA class influenced self-care management. Self-care self-efficacy influenced self-care maintenance, monitoring, and management [26].

Evidence from the few articles described above highlight the heterogeneous self-care behaviors performed by ACHD patients. Accordingly, it was necessary to deepen the literature search by including articles describing specific self-care behaviors advocated for adult patients with CHD, in accordance with the guidelines and the middle-range theory of self-care in chronic illness [4, 32]. Specifically, we searched for articles describing physical activity, dental hygiene, adherence to drug therapy and medical checks, and substance abuse. These are all behaviors considered essential for the treatment and health outcomes of CHD patients [32].

In this secondary search, we found mainly articles focused on some aspects of self-care maintenance. Although contraindicated [32], more than half of young adults (54%) with CHD reported smoking cigarettes, using marijuana or other illicit drugs at least once, and consuming alcohol [33]. Specifically, alcohol consumption was habitual in 61% of ACHD patients [23], and cigarette smoking addiction ranged from 10% [34] to 25% [23]; 13% of ACHD patients reported occasional drug abuse [34]. Excellent dental health (daily brushing and flossing, as well as an annual dental visit) was performed by 15% of ACDH patients [33], and approximately 40% of the ACDH population was underweight or overweight-obese [34]. The WHO-recommended optimal level of physical activity of 2.5 h per week was implemented by 30% of ACHD patients [35, 36], with variations among nations ranging from 10% (India) to 53% (Norway) [35]. Only one study [33] analyzed the willingness of ACHD patients to undergo to seasonal flu vaccination, describing that one in every three ACHD patients did not receive the flu vaccine, most likely because they believed it was unnecessary. Finally, approximately one in five ACHD patients (18.1%) reported poor adherence to medical treatment and follow-up visits [34], despite most of the ACHD patients enrolled in the study having received advanced education about their medical condition [37].

Self-care monitoring has gained very little attention in the field of ACHD patients; it was only studied by McCabe and colleagues [26]. Self-care management in ACHD patients was analyzed further by other investigators in addition to the

McCabe's study and Lee's study described above [26, 30]. By assessing self-care management associations, Hays and colleagues [29] identified four main themes: (a) desire for connectivity, psychological support; (b) plan for the future, health education and life expectancy; (c) coping with needs, ability to handle stress; and (d) access to care, navigation of the health system [29].

Finally, two studies analyzed self-care self-efficacy in ACHD patients in addition to the study by McCabe [26]. Bay and colleagues [38] investigated self-care self-efficacy in the context of ACHD patients' willingness to engage in physical activity, demonstrating that ACHD patients had low levels of self-care self-efficacy [38]. In addition, age was considered to be a predictor of self-care self-efficacy [38]. Another study by Thomet's [39] showed that a lower level of general self-care self-efficacy was associated with female gender, unemployment, and a lower functional class. Additionally, lower general self-care self-efficacy positively predicted health status and quality of life while negatively predicting anxiety and depressive symptoms [39].

Although few investigators have studied self-care in persons with CHD, various determinants of self-care are specified in the latest update of the theory [4]. One factor we have found repeatedly to influence self-care decisions is self-care self-efficacy [18, 40–42], as noted by other CHD investigators [26, 31]. Self-care self-efficacy reflects confidence in the ability to perform self-care [43]. This type of confidence is extremely important in predicting someone's ability to perform self-care. Other factors that probably influence self-care in individuals with CHD include symptoms, access to care, experience, and culture [4]. But support from others may be the most important factor influencing self-care in this population because the family of children with CHD initially performs self-care for the patients, but responsibility shifts to the patient as they age [2].

16.5 Conclusion

Even if adequate self-care behaviors are recognized as essential to staying healthy for persons with a chronic illness, many ACHD patients don't perform them. Even if the evidence regarding ACHD self-care behavior is poor, studies underline an alarming scenario, which makes necessary further research and implementation of tailored health support to the peculiarity of this population. At this regard, the role and competencies of advanced clinical nurses are fundamental because they act as intermediaries between ACHD healthcare need and the specific contribution from each healthcare provider, working in a multidisciplinary team.

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Patient Education to Self-Management

17

Arianna Magon, Deena Barber, and Theresa Faulkner

Abbreviations

ACHD	Adult congenital heart disease
ACHD-NC	ACHD-Nurse Clinician/nurse coordinator
AHA	American Heart Association
BMI	Body mass index
CDRIE	Cardiac devices-related endocarditis
CHD	Congenital heart disease
ECS	European Society of Cardiology
HF	Heart failure
IE	Infective endocarditis
INR	International normalized ratio
ISACHD	International Society for Adult Congenital Heart Disease
NOACs	Novel oral anticoagulants
NYHA	New York Heart Association
OAC	Oral anticoagulation therapy
POCT	Point-of-care coagulometer
PSM	Patient self-management

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PST	Patient self-testing
TEE	Transesophageal echocardiography
VKAs	Vitamin k antagonists

17.1 Patient Education to Self-Management: Education to Pharmacological Therapy

Arianna Magon

Adults with congenital heart disease (ACHD), particularly those with moderate or complex congenital heart malformations, are at increased risk of thromboembolic complications due to several potential acquired or genetic risk factors such as the complexity of the congenital heart disease (CHD), the presence of cyanosis, supraventricular arrhythmias, and surgical sequelae [1, 2]. Overall, it has been estimated that ACHD patients have a ≈ 1- to 100-fold elevated thromboembolic risk compared with non-congenital cohorts [3]. In this scenario, oral anticoagulation therapy (OAC) becomes one of the leading therapeutic choices for primary and secondary thromboembolic prevention in ACHD; currently, roughly 10% of ACHD patients require OAC with a rising tendency [4]. Therefore, based on the most recent guidelines (i.e., ESC GUCH [5]; Pediatric and Congenital Electrophysiology Society and Heart Rhythm Society (PACES/HRS) [6]; ESC AF [7]; ESC [4]), the OAC is a long-term treatment in patients with recurrent or sustained intra-atrial reentrant tachycardia (IART) or non-valvular atrial fibrillation in the presence of moderate or complex CHD with CHA_2DS_2-VASc score ≥1, otherwise, in those with previous intracardiac repair, cyanosis, Fontan palliation or a systemic right ventricle with CHA_2DS_2-VASc score = 0 [5–7]. Nonetheless, the current management approach of OAC in these patients is mainly based on expert opinion. A recent study conducted by Yang [8] has provided insights into implementing the current OAC guidelines in ACHD with atrial arrhythmias (AA). Accordingly, the results of this study showed that only 37% of the patients had a strict indication for OAC considering all three guidelines (i.e., ESC GUCH [5]; PACES/HRS [6]; ESC AF [7]), and its implementation ranged from 54 to 80% depending on which guidelines were used. In fact, a higher heterogeneity was highlighted, and only two recommendation criteria were consistent in all three guidelines: Fontan circulation and cyanosis [8]. Thus, future research should be orientated to evaluate the possibility of enhancing and unifying the guidelines for more effective prevention of thromboembolism in ACHD patients.

Oral vitamin k antagonists (VKAs) are still the standard of care for most ACHD patients and the primary choice for those with mechanical heart valve replacement, even if the novel oral anticoagulants (NOACs) have increasingly replaced VKAs in the last decade. This trend is not unexpected considering the most easier management of NOACs (i.e., it is not required regular laboratory monitoring of coagulation, lower drug or food interactions) and their profiles of efficacy and safety in acquired

cardiovascular conditions [3]. Nonetheless, there is limited clinical experience and a lack of robust evidence about the risk factors for thromboembolism and bleeding complications associated with NOACs treatment in the ACHD subgroup [3, 9]. Therefore, the literature suggests that the initiation of NOACs should be reserved for experienced congenital cardiologists/ACHD centers to ensure patients' adherence and reduce complication rates [3, 10]. However, beyond the pros and cons of each oral anticoagulant, the OAC therapy should be considered carefully on a case-by-case basis. Moreover, OAC is commonly associated with critical treatment adherence and low health-related quality of life in the ACHD population [11, 12]. Accordingly, the rate of treatment nonadherence ranges between 27% and 60% [13]. In addition, in a recent Italian study, the prescription of OAC has been associated with lower physical health status. This last result can find an explanation considering the potential burden related to the complex management of OAC and its indication that is often placed in cyanotic defects or more complex CHD. Thus, identifying the main predictors of good quality of anticoagulation control is pivotal to designing adequate and standardized ACHD educational interventions/programs.

The more recent literature has described several modifiable and non-modifiable predictors of anticoagulation control quality in the ACHD population. Regarding non-modifiable predictors, some clinical and individual variables were identified as significant predictors of poor quality anticoagulation control, such as younger age, female gender, lower treatment time, the presence of comorbidities, and psychiatric disorders [14]. On the other hand, an adequate educational background, patient disease-specific knowledge, and family support were identified as crucial modifiable predictors in sustaining self-management of care in the ACHD population [15]; likewise, higher levels of patients' self-efficacy, engagement, and psychological well-being play a pivotal role in promoting good healthcare behaviors [11]. Among these modifiable predictors, patients' knowledge about the disease and its treatment is often insufficient [12, 16]. The lack of knowledge is mainly concerned with managing abnormal heart rhythms or heart infection symptoms (e.g., sometimes patients do not recognize the symptoms relevant to endocarditis or over-anticoagulation) [15–17]. Moreover, low levels of knowledge concerned the effects of destructive health behaviors as risk factors for poorer anticoagulation control, such as alcohol consumption, smoking, and the interaction between food or vitamins products and OAC. In this scenario, many endeavors are still necessary to address these educational gaps and identify new potential educational needs considering that the population of adults with CHD is newly evolved and the lack of longitudinal research in these perspectives. Thus, educational support should be a lifelong process considering the cardiac care challenges that the ACHD population needs to face during their growth and the risk of their loss to follow-up visits [13].

Although patient education is an essential component for the entire clinical pathway, from childhood through adulthood, the summary of empirical evidence about the efficacy of educational strategies/interventions adopted in the ACHD is still limited. Consequently, there is high heterogeneity in the strategies/approach and educational tools adopted in this population, not enabling the definition of a standardized ACHD educational intervention. Nonetheless, most empirical studies aimed to

promote adequate self-management behaviors in ACHD patients as the primary goal of the educational intervention. In other words, in the adulthood phase, the educational interventions should be focused on enabling patients to function independently in managing their own care with other life responsibilities such as work, family, and school [18]. Therefore, self-care management is recognized as a vital component of chronic illness management, and identifying the self-management needs of this population is a fundamental step to caring for the ACHD. In this regard, Leslie et al. [19] showed that congenital heart disease survivors who fell more competent in managing their health were less likely to experience future anxiety and depressive symptoms and reach better health status and treatment adherence [13, 19]. Furthermore, the literature suggests that transitioning into adulthood is the most opportune time to encourage adolescents to take responsibility for their care. Overall, four main categories of educational needs were described in the ACHD population to perform self-care management: (a) desire for connectivity and psychological support; (b) a plan for the future with educational support about health and life expectations; (c) coping skills for mental stress; (d) and support to navigate in healthcare systems and access to care [20]. Thus, these broad categories should be considered during the planning and monitoring of educational interventions aimed to promote and maximize self-care management.

More specifically, the self-care management of OAC could be performed according to two approaches of care: patient self-management (PSM) and patient self-testing (PST) [21]. The first approach (PSM) requires more advanced patients' abilities to detect and analyze a drop of blood using a point-of-care coagulometer (POCT) and doses the drug based on the coagulation parameters. Conversely, patients autonomously perform blood sampling and analysis in PST, while healthcare providers decide on dosage adjustment [22]. Overall, there is an international consensus about the efficacy and safety of PST and PSM in real-life practice to achieve a better quality of oral anticoagulation control in adult patients with acquired heart disease [22, 23]. However, the implementation of PSM/PST has been primarily tested in children and adolescents with CHD [21, 24, 25]. Accordingly, PSM and PST should be considered valid options than conventional management (i.e., physician-managed approach) in highly selected patients [21, 24–26]; for example, one of the essential criteria is the interest and motivation of the patients to perform PSM/PST. In this scenario, the lack of evidence of PST/PSM in ACHD could undermine the possibilities to identify the most suitable patients (upon clinical, functional, and psychological criteria) by healthcare providers. In addition, the strict refund policy of POCT and the lack of standardized educational interventions for self-care management can further limit its spread in routine clinical settings and discourage patients from being responsible for their medications management. In fact, as reported in the study of Van Damme et al., only a few patients were under self-management approach, due to at the time of the study (2007–2008), the refund system policy did not reimburse patients for buying point-of-care equipment for checking international normalized ratio (INR) [12]. Currently, only in some European countries (e.g., Denmark, Germany, the UK, Spain, Austria, and Holland),

the POCT is refunded by the public health service, enabling patients to effectively adhere to self-management approaches [23].

This last paragraph aims to synthesize the role and the key functions of nurses in providing educational interventions for ACHD under OAC treatment. ACHD nurse specialists play a pivotal in providing comprehensive patients' education and adherence-promoting strategies to manage complex pharmacological treatment regimens [18, 27]. More specifically, the nurse coordinator (ACHD-NC) role is crucial to coordinate OAC management within a multidisciplinary team approach and across the network of ACHD services. ACHD-NC could perform several functions such as scheduling the follow-up visits, detecting INR values, providing guidance and educational interventions, and assessing the potential barriers for treatment adherence (e.g., financial constrain, limited access to pharmacy, fear of side effects, lack of knowledge, or cognitive deficiencies that preclude self-management) [18]. However, Moons et al. showed that in many ACHD specialist centers, the insurability and nonadherence with prescribed regimen or lifestyle were the educational issues less addressed by the nurse specialists [27]. Thus, it is crucial to reach standardized educational interventions to address the needs of ACHD comprehensively. Accordingly, the position statement of the international society for adult congenital heart disease (ISACHD) recommends how an educational intervention on anticoagulation therapy should be considering the following main topics: dietary guidance (e.g., the impact of vitamin K-containing foods on INR), adherence, and interactions with other medications; the importance of timely INR testing; and guidance around activities that may involve impact, falls, or other injuries that may place patients at risk of bleeding complications; furthermore, the patients' capabilities and their level of engagement should be continuously reinforced at each visit or contact by ACHD-NC to identify any potential challenges or limitations [18]. In conclusion, considering the pivotal role of ACHD-NC and the rising indication of OAC therapy in CHD survivors, there is room for improvement of structured patient education programs to support self-management practice.

17.2 Patient Education to Self-Management: Education to Lifestyle Habits

Deena Barber

Nursing care and education of adults with congenital heart disease (ACHD) is an ongoing process occurring over a patient's lifetime [7] and is a top priority for nurses [27]. Education must always emphasize the importance of lifelong specialized congenital cardiac care [28], with the focus on enabling the patient to function independently in making educated, informed decisions on lifestyle choices across the lifespan [18]. All ACHD team members should educate patients [7], but expert, supportive nurses, providing individualized, patient-specific education are the hallmark of ACHD nursing care [18, 27].

All nursing interactions, including in-person, Telemedicine encounters, via electronic message, or on the telephone with an ACHD patient will include some form of education. Often, that education will focus on healthy lifestyle choices related to the patient's congenital heart disease [18].

According to Collins English dictionary, 1994, "the lifestyle of a particular person or group of people is the living conditions, behavior, and habits that are typical of them or are chosen by them"; and "there is a choice of things, there are several of them and you can choose the one you want." ACHD patients are faced with daily lifestyle choices affecting their unique congenital heart disease. Patients not only need, but more importantly, deserve individualized and thorough information to make thoughtful, educated lifestyle choices [29]. Each patient's age, education level, emotional maturity, and individual history must be taken into consideration when providing education at the level of the patient's understanding [30]. Nurses are a highly trusted member of the healthcare team and in a unique position to best understand the patient's expectations and needs [18].

The following modifiable areas should be discussed early and often:

- Heart-healthy nutrition
- Physical activity
- Smoking and tobacco use
- Substance use, including alcohol, cannabis, and illegal drugs
- Body art (tattooing and piercing)

17.2.1 Heart-Healthy Nutrition

All people, including adults with congenital heart disease, are advised to follow a heart-healthy diet. This should include a balanced, nutrient-rich diet with particular attention paid to sodium content [31]. The American Heart Association (AHA) recommends no more than 2300 milligrams of sodium per day for any adult and notes most adults would benefit from reducing sodium intake [31]. ACHD patients should be aware of the role sodium plays in their diet, including identifying high-sodium foods and the importance of reducing processed foods and restaurant meals. ACHD patients may be on a sodium restriction; it is important the nurse knows and understands the patient's sodium limit while educating about diet and nutrition.

Learning to understand nutrition labels and food ingredients is a helpful skill for patients throughout their life. This is helpful when advising patients to avoid excess sodium, saturated fat and trans-fatty acids, and heavily processed foods. This essential skill will prove useful as ACHD patients are also advised to limit sugar intake and caffeine intake [31].

Caffeine ingestion may be high risk for some ACHD patients and should be limited [18]. Caffeine increases the activity of the central nervous system, and some patients may experience negative side effects such as palpitations, increased blood pressure and heart rate, feelings of anxiety, and insomnia. The AHA recommends moderation in caffeine consumption, which is described as one to two cups of

coffee per day [31]. A review of foods, beverages, and products high in caffeine, including coffee, tea, caffeinated sodas and energy drinks, products with chocolate and cocoa, medications (especially over-the-counter), and any product with added caffeine may be surprising to patients and will help when making low or caffeine-free choices.

Some ACHD patients may have a daily fluid restriction, typically ranging from 1 to 2 L per day [32]. This restriction will be decided by the ACHD provider and is patient specific. Not all patients are aware of the importance of fluid management and will benefit from education on the reasons for fluid restriction, restriction amount (in a patient-preferred measurement, i.e., ounces/cups versus milliliters/liters), measuring fluids, and tracking fluid intake. Engaging patients on the daily difficulties and frustrations of fluid restriction and acknowledging their feelings will encourage problem-solving and increase compliance [30, 33].

Following a heart-healthy lifestyle is important to maintain a healthy body weight. Although the rate of obesity for all US adults is 39.8% [34], Jackson and colleagues found half of all CHD survivors were overweight or obese [33]. Addressing a patient's weight and body mass index should be done at each visit, in a supportive and nonthreatening manner. Considering weight and BMI as an additional vital sign to be reviewed has met with considerable success in my practice. Most patient's goal BMI is between 18.5 and 24.9 kg/m^2, and a BMI over 29 is considered obese [34]. Encouraging patients to make a conscious decision to maintain a healthy weight and BMI provides the opportunity to discuss heart-healthy diet choices as well as the importance of physical activity [30, 33].

17.2.2 Physical Activity

The benefits of regular exercise on overall health and quality of life in ACHD patients have been documented in many reports [35, 36]. Many ACHD patients, however, were restricted by medical providers or parents from physical activity as children, and the shift from physical activity restrictions to recommendation may be unusual for some patients [37, 38]. Investing time to investigate past experiences and practices will help the nurse and patient appreciate the potential struggle to incorporate safe physical activity into daily life.

Dua et al. concluded a gentle graded exercise training program, like walking, is safe in all ACHD patients and improves quality of life. They further state ACHD clinicians should promote physical activity via an exercise prescription [39]. This prescription reinforces the value of physical activity and will include recommendations on type, amount, and frequency of exercise and physical activity. Nurses facilitate the conversation from abstract to concrete. Calling an activity "exercise" may lead to resistance and discourage engagement. Discussing how to integrate physical activity into everyday life will empower patients to include daily physical activity in their lives. Reminders of the importance and safety of daily walking help [40] patients move from perceived difficulties with exercise to adopting walking and physical activity in their lifestyle choices [39].

Clinicians may place limitations on some ACHD patient's activities based on specific congenital heart disease, heart failure, pulmonary hypertension, NYHA class, or arrhythmia history [38]. Counseling patients with limited functional capacity is a priority. Referring to the exercise prescription while providing physical activity counseling will encourage the patient to explore alternate approaches with the guidance and support of the nurse [39]. Patients may need additional cardiac testing and referral to cardiac rehabilitation or another medically appropriate exercise program [18]. All patients, even those with reduced functional capacity, benefit from physical activity tailored to their unique needs [39].

Education of ACHD patients on heart-healthy lifestyle choices for diet and physical activity is often well-accepted. Patients are familiar with many of the heart-healthy dietary and physical activity recommendations, common for most adults.

The next topics to be addressed may elicit more stress and resistance, in my experience. These high-risk lifestyle choices include smoking and tobacco use, substance use, including alcohol, cannabis and illegal drug use, and body art. Nurses caring for ACHD patients must be respectful and sensitive to the difficulties discussing behavior typically labeled as negative. Kovacs et al. recommend discussing the challenges faced by ACHD patients and normalizing these challenges with education [41].

17.2.3 Smoking and Tobacco Use

Tobacco use is detrimental to all people and globally is one of the leading causes of preventable death, as well as a major modifiable risk factor for cardiovascular disease and stroke [42, 43]. Unfortunately, congenital heart disease patients also smoke and use tobacco, albeit at rates less than the general population. Fox and colleagues note congenital heart disease survivors have increased risks from smoking, the overall rate for smoking in the US general population is 15.3%, and a lower smoking prevalence for CHD patients of 9.3%, although 23.3% of CHD patients with limitations consistent with NYHA Class IV reported smoking [44]. In addition, HOLBEIN et al. found 12% of CHD patients reported smoking from an international sample of 4028 ACHD patients [45].

Patients may be reluctant to share their actual smoking and tobacco use. ACHD nurses are in a unique position to establish trust between themselves and their patients, by utilizing open, nonthreatening dialogue, and supporting patients' rights to make their own lifestyle choices, including smoking and tobacco use [18, 41]. Nurses must ascertain not only if a patient smokes but what form is used for tobacco ingestion. Virani et al. note the dramatic rise in the use of electronic nicotine delivery systems (commonly known as vaping), cigars, cigarillos, filtered cigars, hookah, and smokeless tobacco [42].

The negative side effects of smoking and tobacco use are well known to nurses. Patients need to hear there is no safe amount or form of tobacco [42]. Remaining nonjudgmental and supportive while reviewing tobacco use at each visit provides a safe environment to discuss each patient's unique experiences, everyday patterns of

life, previous education, and concerns about tobacco use [43]. For nonsmokers, the ACHD nurse should reinforce continued abstinence from smoking and tobacco use. If the patient currently smokes or uses tobacco, the ACHD nurse should acknowledge smoking cessation as a challenging and difficult process while reinforcing reduction in tobacco use. Smoking cessation is promoted and encouraged, and support is provided to the patient. In our practice, we celebrate every victory, no matter how small [43]. The use of smoking cessations aids will be determined by the ACHD physician, and patients may be referred to their PCP for ongoing therapies. The ACHD nurse is in an ideal position to promote consistent education and long-term support while encouraging this positive lifestyle change [43].

17.2.4 Substance Use, Including Alcohol, Cannabis, and Illegal Drugs

Few studies are available for substance use (cannabis, alcohol, illegal drugs) in congenital heart disease patients. These studies did find overall lower rates of substance use in congenital heart patients than in the general population; however, studies are rare, and substance use remains an important topic to discuss with ACHD patients [46].

Medicinal use of cannabis ("medical marijuana") is legal in many countries across the world, 36 US states and 4 US territories [47]. Recreational use of cannabis is also increasing, and patients may ask about the safety of both medicinal and recreational use of cannabis. According to Page et al., "cannabis use has been associated with tachycardia, premature ventricular contractions, atrial fibrillation and ventricular arrhythmia," as well as cardiac clinical presentations including arrhythmia, pulmonary edema, and sudden death [48]. CHD patients may have compromised cardiac status, and ACHD nurses are essential to educate patients on the possible consequences of cannabis use. Few studies were found reporting cannabis use in CHD patients. Recently, Moons et al., as part of APPROACH-IS Consortium and the International Society for Adult Congenital Heart Disease (ISACHD), found 8% of men with CHD and 4% of women with CHD used cannabis containing products in a study including 4028 patients with CHD from 15 countries [49]. As there are few, if any, cardiovascular benefits to cannabis use, ACHD patients should be advised to refrain from all use of cannabis. In addition, drug interactions with cannabis are mostly unknown and difficult to predict. The AHA has noted potential drug-drug interactions with common cardiac medications, including but not limited to antiarrhythmics (amiodarone, dronedarone, flecainide, propafenone), PDE5 inhibitors (sildenafil), statins, beta blockers (carvedilol, metoprolol), losartan, diltiazem, and warfarin [48].

Holbein et al. note a substantial portion of the CHD population engages in substance use although at a lower rate than the general population. Substance use includes alcohol consumption in the form of binge drinking (six or more drinks consumed at one occasion), and illicit drug use in the form of cannabis, amphetamines ("speed"), 3,4-methylenedioxymethamphetamine (MDMA; "ecstasy"),

cocaine, and psychedelic mushrooms. Even with lower rates than the general population, the percentages noted in this international study of 4028 CHD patients included 10% of patients who reported binge drinking and 6% reporting at least monthly use of recreational drugs [45]. Janssens et al. also reported substance use significantly lower than in matched controls, but again, higher rates than would be hoped for in this medically fragile population [45, 46, 49]. Particularly concerning for adults with single ventricle physiology (SVP) is the reported rates of alcohol use and binge drinking. Overgaard et al., in a report of single ventricle patients, note this population is at particular risk for negative health outcomes from unfavorable lifestyle choices [50]. Although reported rates of alcohol use and binge drinking are lower than the control group, 85% of SVP patients report drinking alcohol from time to time, with 26% reporting binge drinking once a month or greater. Education for SVP patients must focus on the concerns for alcohol use in SVP, especially the potential for arrhythmias, and the importance of protecting the liver when making lifestyle choices for alcohol use [51].

There is no current recommendation for a "safe" amount of alcohol use for SVP or for any ACHD patients. Expecting no engagement in substance use may be unreasonable and may present more challenges to this population [46]. Remaining open-minded and nonjudgmental while reviewing substance use with our ACHD patients provides an opportunity for an honest assessment of the risks of these behaviors and pertinent counseling for all ACHD patients.

17.2.5 Body Art (Tattooing and Piercing)

Body art, including tattooing and piercing, is increasing in popularity, especially in adolescents and young adults. Body art is considered low risk for infection in the general population, but data on the safety of body art in ACHD patients is sparse [40]. Discussing the clinician's concern of endocarditis from an infected tattoo or piercing may help patients understand the recommendation to avoid body art, even though endocarditis after body art is rare in CHD patients [40]. Both the American Heart Association (AHA) and the European Society of Cardiology (ESC) emphasize education on the potential hazards and recommend discouraging patients from obtaining body art, particularly piercing of the tongue or mucous membranes for all patients, and especially those in the highest-risk group [52, 53].

According to the AHA and ESC, this high-risk group includes unrepaired cyanotic CHD, including palliative shunts and conduits, completely repaired CHD with prosthetic material or device for the first 6 months after the procedure, and repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch. Also included is any patient with prior endocarditis, and patients with any prosthetic valve, including a transcatheter valve or when prosthetic material was used for the valve repair [52, 53]. Despite these straightforward recommendations, antibiotic prophylaxis prescribed for patients prior to obtaining body art remains at the discretion of the ACHD provider. The ACHD nurse must remember the choice to obtain body art remains with the patient. Patients choosing to obtain body art must be

educated to have the tattooing or piercing performed under sterile conditions, care for the area as instructed by the body artist, and promptly report any signs of infection [40, 53].

In conclusion, ACHD patients are faced with the same lifestyle choices as the general population. It is paramount the ACHD nurse educates patients on the consequences of these lifestyle choices, in a supportive and nonjudgmental manner. We remain the first line of care and are in a unique position to support and encourage our patients to make healthy lifestyle choices. For adults with congenital heart disease, this education should start with our first patient encounter and remain a focus of our ACHD nursing care across the life span.

17.3 Patient Education to Self-Management: Infective Endocarditis

Theresa Faulkner

The number of adults with congenital heart disease (ACHD) has increased markedly over the past few decades as a result of astounding successes in pediatric cardiac care. Nevertheless, it is now well understood that CHD is not cured but palliated, such that lifelong expert care is required to optimize outcomes [54]. Infective endocarditis (IE) is a severe complication in patients with congenital heart disease (CHD). Infective endocarditis (IE) is a life-threatening, multisystemic illness initiated by microbial infection, mostly bacterial, targeting the endothelial lining of the heart chambers and valves. It could also affect the mural endothelium, chordae tendineae, interventricular septum, and other intracardiac tissues or devices [55]. A substrate of prosthetic material and residual lesions, constantly evolving as surgical techniques change over time, predisposes these patients to the potentially devastating complication of infective endocarditis (IE) [55]. Epidemiology, etiology, and outcome in this group are different to those of patients with acquired heart disease [56]. The reported incidence of IE in CHD is 15–140 times higher than that in the general population. Infective endocarditis (IE) is a leading cause of morbidity and mortality in patients with congenital heart disease (CHD) [57]. IE in CHD carries a mortality rate of 4–10%. This prognosis is lower compared with acquired heart disease and may reflect the higher proportion of right-heart IE or the better care in CHD centers [58, 59].

CHD is linked to the development of IE through endothelial shear stress, the presence of intracardiac prosthetic material (valves and conduits) and implanted cardiac electronic devices, and recurrent healthcare procedures [60, 61]. IE occurs in prosthetic valves but can also affect the adjacent structure of the valve, mural thrombus, or cardiovascular devices [62]. Some research has shown valve-containing prosthetics as an important independent risk factor for IE in ACHD patients, short and long term after implantation, whereas non-valve-containing prosthetics are associated with greater risk only in the short term after implantation [63]. Over the

last 30 years, there have been significant advances in CHD care, and the number of patients with repaired or palliated defects has expanded significantly [60, 61].

In the presence of classical features such as fever, cardiac murmur, bacteremia, and peripheral stigmata, the diagnosis of IE may be established easily. Unfortunately, in everyday clinical practice, this presentation is rarely seen, and atypical presentation occurs more frequently. The clinical diagnosis of IE relies on integration of clinical, microbiological, echocardiography, and laboratory findings [62]. Therefore, the skills needed by the ACHD nurse clinician (NC) as a member of the Endocarditis Team is to be able to recognize, organize, coordinate, and facilitate the care of the ACHD patients with IE. This includes assessment, patient education to recognize IE, and support for self-care management and self-advocacy [18].

IE occurs if bacteria, fungi, or other microorganisms invade your bloodstream and attach to abnormal areas of your heart. The infection can damage the heart and cause serious and sometimes fatal complications. IE can develop quickly or slowly depending on what type of microorganism causing it and whether you have an underlying heart problem.

There are two forms of infective endocarditis, also known as IE:

1. Acute IE develops suddenly and may become life threatening within days.
2. Subacute or chronic IE (or subacute bacterial endocarditis) develops slowly over a period of weeks to several months.

The types of microorganism that cause IE do not differ from the types found in acquired heart disease, with *Streptococci* and *Staphylococci* being the most common.

Microorganisms found to cause IE in ACHD patients [64]:

- Penicillin-susceptible (MIC ≤0.12 mcg per ml) viridans *Streptococci* and *S. bovis*
- Penicillin-intermediate (MIC >0.12 and ≤ 0.5 mcg per ml) viridans *Streptococci*
- *Enterococci* and penicillin-resistant (MIC >0.5 mcg per ml) viridans *Streptococci*
- *Staphylococci*
- HACEK strains
- Enterobacteriaceae
- *Pseudomonas aeruginosa*
- Fungi

Changing trends in etiology of IE with emerging infections from *Staphylococci*, bacteria of the HACEK group and fungi, have resulted in an increased frequency of culture-negative IE. Sepsis or persistent fever despite appropriate antimicrobial therapy, recurrent emboli, heart failure, or new pathologic murmurs suggests hemodynamic impairment and/or infection extending beyond the valve leaflet or prosthetic valvular annulus [65].

There is a correlation between the risks of IE in ACHD patients that appears to be lesion specific. The greatest is in the context of a patient having recent medical interventions. There is an increased 1-year mortality following IE diagnosis,

irrespective of CHD lesion classification [28]. Lesion-specific risk factors related to the development of IE and the CHD lesions were grouped as follows:

Highest- to lowest-risk factor lesions:

1. Cyanosis at birth (tetralogy of Fallot, univentricular heart, complete transposition, truncus arteriosus, hypoplastic left heart)
2. Endocardial cushion defects
3. Ventricular septal defect
4. Left-sided lesions (aortic coarctation, stenosis, and insufficiency, mitral stenosis, and insufficiency)
5. Right-sided lesions (Ebstein anomaly, anomalies of the pulmonary artery or valve, congenital tricuspid valve disease)
6. Atrial septal defect, patent ductus arteriosus, and other CHD (other congenital anomalies)

Clinical risk factors for IE include the following:

- Artificial heart valves. Bacteria are more likely to attach to an artificial (prosthetic) heart valve than to a normal heart valve.
- A history of endocarditis. Endocarditis can damage heart tissue and valves, increasing the risk of a future heart infection.
- Damaged heart valves. Certain medical conditions, such as rheumatic fever or infection, can damage or scar one or more of your heart valves. This can make them more prone to endocarditis.
- A history of intravenous (IV) illegal drug use. People who use illegal drugs by injecting them are at a greater risk of endocarditis. The needles used to inject drugs can be contaminated with the bacteria that can cause endocarditis.
- Heart valve disease—either stenosis (a narrowed valve) or regurgitation (a leaking valve)
- Heart devices (such as a pacemaker or implantable cardioverter defibrillator)
- Diabetes mellitus
- Recent cardiac surgery (<6 months)
- Recent medical interventions (≤6 weeks)
- Genitourinary
- Gastrointestinal
- Respiratory
- Advanced age (above 58 years)
- Male gender
- Patients who have prolonged vascular access

Several comorbidities have been correlated with increased risk of IE development, including:

- Chronic kidney disease (CKD)
- Cancer

- Human immunodeficiency virus (HIV)
- Previous dental procedures

Healthcare-associated IE represents up to 30% cases of IE, justifying aseptic measures during venous catheters manipulation and during any invasive procedures. Bacteria in other parts of the body, e.g., mouth, throat, and the gut, cause endocarditis when they migrate into the bloodstream then to the endocardium. In developing countries, rheumatic heart disease remains the most common predisposing cardiac condition for IE [66].

CHD lesion subsets, male sex, diabetes mellitus, and recent medical interventions were associated with an elevated risk of developing IE. Notably, right-sided CHD, atrial septal defects, and patent ductus arteriosus were at comparatively lowest IE risk [28]. The most affected valve in IE patients is the mitral valve, followed by the aortic, tricuspid valve, and, less frequently, the pulmonary [67].

Infective endocarditis (IE) can cause a range of signs and symptoms that can vary from person to person. Signs and symptoms also can vary over time in the same person. Signs and symptoms differ depending on whether you have an underlying heart problem, the type of organism causing the infection, and whether you have acute or subacute IE. Signs and symptoms to educate the ACHD patient to recognize related to IE include:

- Flu-like symptoms, such as fever, chills, fatigue (tiredness), aching muscles and joints, night sweats, and headache
- Shortness of breath or a cough that won't go away
- A new heart murmur or a change in an existing heart murmur

Skin changes such as:

- Overall paleness.
- Osler's nodes—small, painful, red, or purplish papulopustules type bumps under the skin, on distal pads of digits on the fingers or toes.
- Janeway's lesions—small, dark, painless, flat spots erythematous macules on palms of hands and soles of feet.
- Petechiae common—found on extremities, mucosa, and palate. Tiny spots under the fingernails, on the whites of the eyes, on the roof of the mouth and inside of the cheeks, or on the chest. These spots are from broken blood vessels.
- Splinter hemorrhages—dark, linear lesions in nail beds Roth's spots—retinal hemorrhages with small, clear centers.
- Conjunctival hemorrhages—bright red/dark red patch on the sclera.
- Nausea (feeling sick to your stomach), vomiting, a decrease in appetite, a sense of fullness with discomfort on the upper left side of the abdomen, or weight loss with or without a change in appetite.
- Hematuria—blood in the urine.
- Swelling in the feet, legs, or abdomen.

IE also can affect other organs in the body, such as the lungs, kidneys, and spleen.

- Lungs—The lungs are especially at risk when IE affects the right side of the heart. This is called right-sided infective endocarditis. A vegetation or blood clot going to the lungs can cause a pulmonary embolism and lung damage. Other lung complications include pneumonia and a buildup of fluid or pus around the lungs.
- Central nervous system complications—These complications occur in as many as 20 to 40 percent of people who have IE. Central nervous system complications most often occur when bits of the vegetation, emboli, break away and lodge in the brain. These emboli can cause brain abscesses and meningitis. Emboli also can cause a stroke or seizures. These complications can cause long-lasting damage to the brain and may even be fatal.
- Kidneys—IE can cause kidney abscesses and kidney damage. IE also can cause inflammation of the internal filtering structures of the kidneys. Signs and symptoms of kidney complications include back or side pain, blood in the urine, or a change in the color or amount of urine. In a small number of people, IE can cause kidney failure.
- Spleen—In as many as 25 to 60 percent of people who have IE, the spleen enlarges (especially in people who have long-term IE). Sometimes, emboli also can damage the spleen. Signs and symptoms of spleen problems include pain or discomfort in the upper left abdomen and/or left shoulder, a feeling of fullness or the inability to eat large meals, and hiccups [58].

There is a lack of scientific evidence for the efficacy of infective endocarditis prophylaxis. Strict oral hygiene and regular dental checkups are more important than antibiotic prophylaxis to decrease the risk of IE. Antibiotic prophylaxis is recommended only for patients with the highest risk of IE undergoing the highest-risk dental procedures. Strict sterile technique is required during venous catheter manipulation and all invasive procedures to decrease the rate of healthcare-associated IE. Skin hygiene needs to be emphasized, and tattoos and skin piercing are discouraged specifically tongue piercings [53].

According to the American Heart Association (AHA) and European Society of Cardiology (ESC), high-risk group includes unrepaired cyanotic CHD, including palliative shunts and conduits, completely repaired CHD with prosthetic material or device for the first 6 months after the procedure, and repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch. Also included is any patient with prior endocarditis, and patients with any prosthetic valve, including a transcatheter valve or when prosthetic material was used for the valve repair [52, 53]. The distribution of lesion severity in the ACHD population depends, in part, on the age, category, and available healthcare services. For example, in developed countries, most adults with CHD have had surgical palliation. In contrast, in an outpatient population of 1157 adults in New Delhi, India, only 14% had had cardiac surgery [54].

The ACHD patients with complicated IE should be referred early and managed in an ACHD center with surgical facilities and preferably with an Endocarditis Team. This multidisciplinary approach has been shown to significantly reduce the 1-year mortality in infective endocarditis [18]. A multidisciplinary approach is mandatory, and the team involves ACHD cardiology, ACHD cardiothoracic, infectious diseases, ACHD radiology, nuclear medicine, research fellows, nursing, and other subspecialties (e.g., neurologist, anesthesiologists, etc.) on a case-by-case basis with close follow-up. ACHD patients with complicated IE, i.e., endocarditis with HF, abscess, embolic or neurological complication, should be referred early and managed in an ACHD program [18].

Diagnosis of IE is frequently difficult, particularly in some subgroups (prosthetic valve IE [PVE], intracardiac device and blood culture-negative IE [BCNIE]). The Modified Duke criteria are useful for the classification of IE, but they are of limited value in those subgroups [53].

Topan et al. clarify below the terms in modified Duke criteria for the diagnosis of infective endocarditis including both major and minor criteria [62].

17.3.1 Major Criteria

1. *Blood culture positive for IE*
   (a) Typical microorganisms consistent with IE from two separate blood cultures.
      - Viridans *Streptococci*, *Streptococcus bovis*, HACEK group, *Staphylococcus aureus*
      - Community-acquired *Enterococci*, in the absence of a primary focus

   (b) Microorganism consistent with IE from persistently positive blood cultures, defined as follows:
      - At least two positive cultures of blood samples drawn >12 h apart
      - All of three or a majority of ≥4 separate cultures of blood (with first and last sample drawn at least 1 h apart)

   (c) Single positive blood culture for *Coxiella burnetii* or antiphase I Ig G antibody titer > 1 > 800
2. *Evidence of endocardial involvement*
   (a) Echocardiogram positive for IE (transesophageal echocardiography [TEE] recommended in patient with prosthetic valves)

   IE by clinical criteria or complicated IE (paravalvular abscess) TTE as first test in other patients) is defined as follows:
   - Oscillating intracardiac mass on valve or supporting structures, in the path of regurgitant jets, or on implanted material in the absence of an alternative anatomic explanation
   - Abscess
   - New partial dehiscence of prosthetic valve

   (b) New valvular regurgitation (worsening or changing of preexisting murmur not sufficient)

17.3.2 Minor Criteria

- Predisposition, predisposing heart condition, or injection drug use
- Fever, temperature >38 °C
- Vascular phenomena, major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, and Janeway's lesions
- Immunologic phenomena: glomerulonephritis, Osler's nodes, Roth's spots, and rheumatoid factor
- Microbiological evidence: positive blood culture but does not meet a major criterion as noted above[1] or serological evidence of active infection with organism consistent with IE

17.3.3 Definite Infective Endocarditis

Pathological Criteria

- Microorganism: demonstrated by culture or histology in a vegetation or in a vegetation that has embolized, or in a intracardiac abscess
- Pathologic lesions: vegetation or intracardiac abscess present confirmed by histology showing active endocarditis

Clinical criteria using specific definitions for these terms as listed below.

- Two major criteria
- One major and three minor criteria
- Five minor criteria

Possible infective endocarditis, at least:

- One major and one minor criteria
- Three minor criteria

Rejected

- Firm alternate diagnosis explaining evidence of infective endocarditis
- Resolution of infective endocarditis syndrome, with antibiotic therapy for 4 days or less
- No pathologic evidence of infective endocarditis at surgery or autopsy, with antibiotic therapy for 4 days or less

[1] Excludes single positive cultures for coagulase-negative *Staphylococci* and organisms that do not cause endocarditis Modified Duke criteria.

Twenty years after their launch, the Duke criteria for the diagnosis of IE continue to be important tools. Nevertheless, the final diagnosis should not rely only on these criteria; clinical judgment and integration of whole clinical and laboratory findings should be performed. Low index of suspicion of IE and inappropriate use of antibiotics may have a great negative impact on the diagnosis of IE. Nowadays, the scarcity of classical Osler manifestations—bacteremia, fever, and peripheral stigmata—makes the diagnosis of IE a challenge [62].

Echocardiography and blood cultures are the cornerstone of diagnosis of IE. TTE must be performed first, but both TTE and TEE should ultimately be performed in the majority of cases of suspected or definite IE. If initial diagnosis cannot be confirmed or even rejected but with a persisting high-level suspicion, echocardiography and blood culture should be repeated, and other imaging techniques should be used [53].

Pharmaceutical treatment is largely unchanged and based on classical antibiotics in monotherapy or as combination therapy, but for staphylococcal endocarditis, gentamycin is no longer required. As cardiac surgery is needed in 50% of the cases during the course of the disease, the urgency for surgery depends on the extent of cardiac insufficiency, the persistence of the pathogen despite antibiotic treatment, and on neurological complications [68]. The treatment of IE relies on the combination of prolonged antimicrobial therapy and—in about half patients—surgical eradication of the infected tissues [53].

Early consultation with a cardiac surgeon is recommended to determine the best therapeutic approach. The three main indications for early surgery in IE are its three main complications, i.e., HF, uncontrolled, infection, and prevention of embolic events. HF is the most frequent and severe complication of IE. Unless severe comorbidity exists, the presence of HF indicates early surgery [53].

Embolism is very frequent in IE, complicating 20–50% of cases of IE, falling to 6–21% after initiation of antibiotic therapy. The risk of embolism is associated with the size and mobility of the vegetation and is the highest during the first 2 weeks of antibiotic therapy [53]. Embolic events are a frequent and life-threatening complication of IE related to the migration of cardiac vegetations. The brain and spleen are the most frequent sites of embolism in left-sided IE, while pulmonary embolism is frequent in native right-sided and pacemaker lead IE. Stroke is a severe complication and is associated with increased morbidity and mortality [53].

Symptomatic neurological events develop in 15–30% of all patients with IE and are associated with excess mortality. After a first neurological event, if cerebral hemorrhage has been excluded by cranial CT and neurological damage mild, surgery is recommended for HF, uncontrolled infection, abscess, or persistent high embolic risk [53].

Cardiac device-related IE (CDRIE) is one of the most difficult forms of IE to diagnose and must be treated by prolonged antibiotic therapy and device removal. Percutaneous extraction is recommended in most patients with CDRIE. Surgical extraction should be considered if percutaneous extraction is incomplete or impossible or when there is associated severe destructive tricuspid IE [53].

Other organ systems that can be affected by IE include splenic infarcts. They are common and very frequently asymptomatic. Persistent or recurrent fever, abdominal pain, and bacteremia suggest the presence of complications (splenic abscess or rupture) [53]. Furthermore, cardiac failure may be due to myocarditis, which is frequently associated with abscess formation or immune reaction. Ventricular arrhythmias may indicate myocardial involvement and imply a poor prognosis. Conduction disorders are uncommon complications of IE. According to data from patient registries, their frequency is between 1% and 15% of cases, and their presence is associated with worse prognosis and higher mortality. Conduction abnormalities (mainly first-, second-, and third-degree atrioventricular blocks, rarely bundle branch blocks) are due to spread of the infection beyond the endocardium, from valves to the conduction pathways, and are generally associated with perivalvular complications. Complete atrioventricular block is most often associated with involvement of the left-sided valves (aortic, 36%; mitral, 33%) [69]. Musculoskeletal symptoms (arthralgia, myalgia, back pain) are frequent during IE [70]. Acute renal failure is a common complication of IE and may worsen the prognosis of IE. Acute renal dysfunction occurs in about 6–30% of patients [68].

In conclusion, infective endocarditis in patients with CHD is an ongoing clinical challenge. The incidence of IE is increasing in adults with CHD. Morbidity caused by a broad clinical spectrum of cardiac and extracardiac episode-related complications is high. The risk of IE in ACHD patients is lesion-specific and is greatest in the context of recent medical interventions. Infective endocarditis is a complex disease, and despite new techniques for diagnosis and microbiological therapy, mortality rate is still up to 10–20%. Reducing the burden of IE in patients with ACHD is complex and requires a multifaceted approach. The skills needed by the ACHD nurse clinician (NC) as a member of the Endocarditis Team are vital as a frontline provider to recognize, organize, coordinate, and facilitate the care of the ACHD patients with IE. An integral part of the nurse's role in IE management is to educate the patient to be able to recognize IE causes and symptoms. This is key to early treatment and self-care management. *Streptococci* remain the most common causative organism, and antecedent dental or medical procedures. The presence of an atrioventricular septal defect or staphylococcal infection is associated with significantly increased risk of early mortality [71].

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The Health Engagement of Adult Congenital Heart Disease Patients

18

Rosario Caruso

18.1 Introduction

In the last decades, the health engagement of adults with chronic conditions has been considered strategic to meet citizens' demands and expectations toward healthcare services [1]. Focusing on engaging patients is not merely an approach to meet expectations, as it also implies several positive influences on patient- and system-level outcomes [2]. In this context, the disciplines involved in research and practical initiatives to enhance the health engagement of patients are growing continuously, from psychology, nursing, clinical disciplines to public health. Adults with congenital heart disease (ACHD), especially people with moderate or severe conditions, require to be engaged in health-promoting practices (salutogenic approach), treatment schemes, and follow-ups to optimize their health status over time. In other words, the failure of health professionals to engage ACHD as skilled self-managers of their condition and health might lead to poor clinical outcomes and worse health-related quality of life (HRQoL). From a salutogenic perspective, ACHD have to perform health-promoting behaviors (e.g., social and physically active life), specific illness-related behaviors (e.g., adherence to treatments and follow-ups), proficient symptom recognition, and self-management behaviors to increase their physical and mental health within their chronic condition. This chapter highlights the worth of focusing on health engagement in ACHD and some practical hints to actively engage patients toward their health.

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18.2 What Is Health Engagement?

The term engagement has been used with several nuances, such as patient engagement, health engagement, or as a related concept to patient activation or empowerment. Recently, a consensus conference tried to clarify the meaning of engagement by involving several clinicians, institutions, and patient organizations [3], and then, a theoretical framework was proposed [4]. The framework is given by considering the patient's point of view within a dynamic psychological process (patient health engagement model), patient-related factors, health professional-related factors, organization-related factors, and community-related factors.

The "patient health engagement" model defines the dynamic psychological process, which is driven by emotional components underpinning patients' ability to adjust and adapt to the change of self role identity involved in the engagement experience [4]. Therefore, the dynamic psychological process is given by individuals' personal choices to change their attitude toward the healthcare services, from being passive patients to being partners and co-developers of their specific paths (patients' journeys) with healthcare providers. These changes in the personal role identities reflect an evolving adjustment to their health condition; for this reason, the process of engagement is dynamic. The phases of health engagement have then been described: blackout, arousal, adhesion, and eudaimonic project. The blackout phase is given when patients feel overwhelmed, shocked, fragile, and are passive and prefer to rely on others (e.g., family caregivers). The arousal phase is given by anxiety, which is an over-reactive status when patients acquire knowledge about their condition or newly changed condition. In this phase, patients start to copy with the changed condition and are very focused and hypervigilant over their body and body signs and symptoms. When patients succeed in emotional regulation and coping with illness, they experience the third phase of the PHE model: adhesion. In this phase, patients show a good acceptance of their condition, being aware of their health status and its impact on their lives and life habits. When patients become completely aware of their condition and its implications in terms of changed life habits and therapeutic requirements, they can be skilled and effective co-developer of their care project, working with healthcare providers (eudaimonic project). Overall, the trajectory of engagement from the blackout phase to the eudaimonic project is not necessarily linear.

Patient-related factors to the patient health engagement process are given by diverse aspects, such as age, ethnicity, education level, income level, beliefs, disease-specific characteristics, and cultural-social elements [5, 6]. In other words, some patient-level characteristics might influence patients from gaining the skills necessary to play an active role in their care path.

Considering the organization-related factors, organizational models oriented to patient-centered care and infrastructural characteristics are pivotal to enabling patients to co-develop their care path: factors regarding healthcare providers might influence the engagement process. In fact, the role identity of the health professionals, their set of communicative and relational skills, and their attitude toward shared decisions about their care process might potentially foster or mitigate the process of

patients' engagement [7]. From a broader perspective, some social and community-related factors might also influence the process of health engagement, such as the family, the media, and the community of peers [8].

18.3 Health Engagement of Adults with Congenital Heart Disease

ACHD are at risk for multiple possible health concerns, such as endocarditis, pregnancy complications, cardiac-related complications, stroke, coronary artery disease, hypertension, and congestive heart failure [9]. Some possible complications might be mitigated by healthy lifestyles, such as eating a diet low in saturated fat and being physically active; therefore, the health engagement of ACHD is strategic to prevent or slow the development of amenable complications.

Considering that the clinical journeys of ACHD are clinically and socially dynamic, understanding their health engagement is strategic to adopt a tailored approach to educational and motivational initiatives. The first step to engage ACHD in their care path and toward healthy behaviors might be represented by the needed alignment between patients and healthcare providers. Failure to support the patients' engagement and align with patients' expectations might result in dissatisfaction with care, and poor adherence to treatments, undermining the care path. For this reason, the adoption of specific and validated self-report assessments might be strategic to assess in which phase of the health engagement process a patient is.

The first useful tool for an overview of the engagement process might be the Patient Health Engagement (PHE) scale [10]. The PHE scale was developed in accordance with the PHE model. The PHE scale comprises five ordinal items and identifies four characteristics along a continuum of engagement from the blackout phase to the eudaimonic project. The PHE scale showed evidence of validity and reliability in several clinical contexts [10].

ACHD characterized by a worsened clinical condition or changes in their lives might show the blackout phase characteristics (and scores). In this stage of the engagement process, overcoming the blackout is important, and it is relevant to understand whether ACHD recognize healthcare providers as gatekeepers to catalyze the engagement process to achieve health results. Therefore, ACHD in the blackout phase require to be listened to, and the involvement in the process of the family caregivers might be important to support ACHD in coping with the challenges of unfavorable changes. In this phase, healthcare providers have to create a sense of trust and supportive actions.

In the phase of "arousal," ACHD might be scared and hyperreactive over their symptoms, showing anxiety. Healthcare providers might be perceived as the protectors of patients' health and with a vicarious role regarding many simple decisions that ACHD have to undertake, such as the possibility of performing some physical activities or similar. In other words, ACHD in this phase perceive themselves to be behaviorally unequipped to effectively manage their condition. To cope effectively with their condition, ACHD in this phase require specific knowledge about what

they can do and what they should do. Therefore, health education initiatives in this phase might be effective in boosting the needed awareness to move forward in the emotional and psychological maturation of the engagement. In this step, to facilitate ACHD engagement to their care path and toward healthy behaviors, an alignment between healthcare professionals and patients regarding the patients' susceptibility to a particular negative outcome if a set of specific behaviors are not properly performed is required. Some individuals might underestimate specific risks, while others overestimate the same risks; for this reason, the alignment regarding what and why it is important to stay in health is challenging. In other words, risk knowledge and perceived risk are the first elements that should be addressed in the engagement process of ACHD. In the specific field of ACHD, the impact of health engagement of specific outcomes remains understudied, despite evidence suggesting ACHD are at greater risk for developing amenable cardiac-related complications [11]. In this regard, disease knowledge should be assessed, and some valid and reliable self-report measures might be helpful. For instance, the CHD Assessment of Information Measure (CHD-AIM) is a 24-item measure to assess three domains: recall knowledge, risk knowledge, and general knowledge. Recall knowledge encompasses three free-response items asking participants to recall the specific information regarding their conditions (i.e., diagnosis, current medications, and cardiac surgical history), and each answer has to be scored 0, 1, or 2 depending on the level of accuracy when compared to medical chart. Risk knowledge encompassed items to establish the accuracy of patients' ability to identify cardiac-related conditions for which they are at risk due to their CHD, including arrhythmia, heart failure, stroke, aortic aneurysm, coronary artery disease, and hypertension. Considering that there is no consensus about which possible complications are specific to each possible congenital lesion, the authors of the CHD-AIM developed an a priori scoring procedure based on the literature. General knowledge encompasses 11 items exploring a wide range of CHD self-care behaviors, including diet (i.e., low sodium and saturated fat), exercise, duration of follow-up care, endocarditis, and whether ACHD are at an increased risk of having offspring with CHD. In this domain, men were given two additional items about the presence of erectile dysfunction and sexual performance difficulty, while women received two additional items about pregnancy. Items have to be scored incorrect or correct.

In the phase of adhesion, ACHD have a sense of trust toward healthcare initiatives and might consider healthcare providers as trusted experts. ACHD in this phase are able to learn behavioral skills to effectively adhere to treatments and healthy behaviors and feel sufficiently confident to cope with their health condition. Generally, in this phase, ACHD understand and manage symptoms based on the counseling with their healthcare providers and the increasing ability to perform "body listening" (i.e., recognizing signs and symptoms, or changes).

The last phase of the health engagement process is the eudaimonic project. In this phase, ACHD fully accept their condition and seek advice and support to build a "normality" and develop sustainable life plans. In this phase, healthcare providers are perceived as trusty experts and as "trusted allies" to undertake personal development and growth [2]. In this phase, ACHD are experienced masters and testimonials

of proficient self-care behaviors and might be able to support other people with CHD as peer-counselors. ACHD are co-developers of each aspect of their life and focused on improving their HRQoL, recognizing the (internal and external) resources they require to meet their goals.

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The Essential Impact of Counseling in Improvement of Quality of Life in ACHD Patients

19

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It is easier to build strong children than to repair broken men.
Frederick Douglass (1817–1895)

19.1 Introduction

The population of adults with congenital heart disease (ACHD) is constantly growing. It is estimated that more than 90% of patients born with a heart defect reaches adulthood in the current era [1], thanks to the extraordinary advances in pre- and postnatal diagnosis, cardiac surgery, and interventional cardiology. Considering that the incidence of congenital heart disease (CHD) is around 0.8% to 1.2% of live births [2], it has been estimated that about 13 million adults worldwide are affected by a congenital heart anomaly, accounting for 60% of the total CHD patients, thus surpassing the prevalence in pediatric population [1]. Moreover, complex cardiac lesions as tetralogy of Fallot (TOF), truncus arteriosus (TA), transposition complex (TGA), endocardial cushion defects, univentricular heart, and hypoplastic left heart syndrome have dramatically improved survival rate, and it has been reported an overall prevalence of 55% in the adult cohort over a 10-year observation period [1].

Although most of the CHD can be corrected or palliated, it may be perceived as "never going away" [3], and ACHD have to deal with specific medical, psychosocial, and behavioral problems throughout their life span.

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CHD comprises a large variety of cardiac conditions, and even if some patients may never experience symptoms or require treatment, a significant proportion of patients suffer from some physical limitations, undergo one or multiple cardiac procedures, and need frequent medical attention. Thus, the majority of ACHD patients need a lifelong medical follow-up to assess functional state, necessity of reintervention, specific clinical problems as arrhythmias, pacemakers or ICD implantation, endocarditis, and evaluation of medical therapy. The personal cardiac history and the unique way each individual relates to its heart condition growing up deeply impact the individual neuropsychological and behavioral sphere and are strongly related to the risk of mental health problems. If the initial clinical efforts in managing ACHD were all directed toward the goal of increasing survival and reduce hospitalizations, patients and healthcare professionals are now recognizing the importance of quality of life (QoL) as part of the individual well-being, inseparable from the physiological health [4, 5].

The uncertainty in view of disease course and prognosis, the signs and symptoms related to CHD, and restrictions in activity level have a profound impact of patient's QoL. However, QoL is determined not only by the patient's physical condition but—above all—by the emotional responses to problems raised by the disease [6].

19.2 Health-Related Quality of Life

The World Health Organization (WHO) defines health as "a state of complete physical, mental and social well-being, and not merely the absence of disease and infirmity" [7]. The mental and social aspect of health has gained more and more relevance in a world where medical advances have extended life expectancy, and individual realization and satisfaction go way further than the freedom from a physical condition [8]. This assumption has led to the notion of "quality of life" as "a conscious cognitive judgment of satisfaction with one's life" [9] and "health-related quality of life (HrQoL)" as "those aspects of self-perceived well-being that are related to or affected by the presence of disease or treatment" [10]. Mental health status, social and economic issues, physical limitations, and functional class can all influence QoL in patients with chronic disease. Accordingly, ACHD specialists have recognized the critical importance to explore these issues in order to improve their HrQol and medical care. Evaluation of HrQoL is not always straightforward, since it involves mostly subjective perception. For this reason, different questionnaires have been created to quantify it, and the most popular are the Short Form 36 (SF-36) [11] and EQ-5D [12]. The questionnaires have been translated in multiple languages worldwide and provide a simple and fast measurement of HrQoL, applicable to most medical specialties, and can be self-administered. These health surveys are widely used in ACHD medical practice to help monitor patients' well-being and subjective perception of their health status, with standardized and exhaustive questions that assess the main aspect of the patients HrQoL:

- SF-36 includes one multi-item scale that investigates eight aspects of health: physical activity, social activities, usual role activities in relation to physical health and emotional problems, pain, mental status, vitality, and general health perception.
- EQ-5D created by the international group EuroQol for both clinical and economical research on perceived individual health status; it is a generic questionnaire that explores five dimensions of health: mobility, self-care, usual activity, pain/discomfort, and anxiety/depression.

Patients' perception of their own health status has gained more relevance over the years and should be considered part of a comprehensive clinical evaluation, since it also seems to influence prognosis [13].

19.3 CHD Complexity and Functional Status

Analyzing ACHD quality of life, most of the studies focused on objective variables most likely to be relevant: the complexity of the heart defect [14, 15], the type and number of interventions [16], NYHA class, and reduced functional capacity [17]. Most researchers suggest that more complex CHD, such as univentricular heart, Eisenmenger, and cyanotic heart defects, are associated with the worst QoL, while aorthopaties (aortic coarctation, aortic valve disease) have the best outcome [18–20]. However, results from studies on this topic vary greatly, sometimes with unexpected results: more complex ACHD had sometimes reported QoL comparable to healthy peers and simple CHD [2, 21]. It has been postulated that functional status may be more relevant than anatomic complexity in influencing QoL, since it directly impacts daily performance and activities [19, 22].

19.4 Psychological Distress

Mental health is an emerging problem in ACHD. It has been estimated that one-third of this population experiences anxiety and/or depression [23, 24]. Of note, children with CHD have more difficulties in behavioral, emotional, and neuropsychological aspects in comparison with their healthy peers [25], and thus, as they grow into adulthood, they remain at an increased risk of psychological and emotional distress, neurocognitive deficits, and social challenges [26]. If left untreated, chronic emotional distress can have deleterious consequences on the cardiovascular (CV) system and QoL [27].

In most cases, the clinical diagnosis is difficult, as patients are not adequately screened and interviewed by a mental health specialist and most researchers rely on self-administered surveys; however, symptoms consistent with mood disorders are reported by up to 50% of patients in some studies [28]. The undoubted evidence gathered from exploring ACHD emotional status is that psychological disorders are

more common than in general population and are underdiagnosed and undertreated [29].

Anxiety in ACHD often manifests as "hart-focused anxiety," described as "the fear of cardiac-related stimuli and sensations based upon their perceived negative consequences" [30]. Affected patients express frequent worry for cardiac symptoms or cardiac issues; they avoid situations and activities that they fear could cause heart problems; and they tend to monitor cardiac parameters frequently (i.e., checking their pulse) and implement behaviors aimed at preventing a negative heart event (i.e., they feel safer only when in a medical environment) [30, 31].

Depression, on the other end, is suspected when the patient has generally low mood, has small interest in leisure activities, experiences a change in eating and sleeping habit, fells fatigued most of the time or has a frequent sensation of agitation and uneasiness, finds difficult to concentrate, and has negative thoughts of guilt, worthlessness, and suicidal ideation [32].

Studies have showed that health-related mental stress increases with age in ACHD, as the concern for future invasive procedures and worsening of their heart condition arises [33].

Depressed and anxious patients are more likely to engage in unhealthy behaviors such as smoking, physical inactivity, poor eating habits, and reduced adherence to medical regimens and to have impaired social relationships, low QoL, and life expectancy [32, 34, 35]. However, more research is needed to clarify the impact of clinical anxiety and depression on the risk of adverse CV events.

Mood disorders appear also be associated with the higher prevalence of neurocognitive deficit among ACHD and might further impair processing speed, attention, and executive functioning that, at least in some extent, are already generally compromised in severe CHD [36]. In adult age, adverse clinical events such as arrhythmias, heart failure, and reinterventions may negatively impact a preexisting neurocognitive deficit and be the cause of more evident psychological disorders [37].

Unfortunately, mood disorders often do not receive adequate medical attention in ACHD treatment. One study shows that only 3.3% of ADHD patients with diagnosed depression were in fact taking appropriate medication, and they had a worse outcome and were less adherent to scheduled outpatient visits [38]. Behavioral therapy protocols are being implemented in these patients with promising results in preliminary studies [33, 39]. In everyday practice, it is often difficult to address mental disorders in ACHD patient for deficiencies in resources, time, and education. The best strategy is to increase the awareness toward these topics and institute screening programs for patients at risk. The cardiologists and specialized nurses play a central role in helping the patients in this journey, but other healthcare professionals such as general practitioners, psychologists, and psychiatrist should intervene when appropriate in order to better address the patients' psychological needs. The approach needs to be individualized and tailored on the thorough knowledge of patients' clinical history and psychosocial background [26].

Current guidelines recognize the importance of psychosocial support for ACHD and the necessity of adequate screening and early identification of neuropsychological and emotional problems and concerns, e.g., fears, depressive symptoms,

self-esteem, family functioning, and social integration, approaching the clinical problems in holistic view including physical, mental, and social profile of the patients [26]. Several screening tools have been proposed as the Hospital Anxiety and Depression Scale (HADS) for assessment of depressive and anxiety symptoms that can be easily administered by expert nurses or doctors during each follow-up clinic visit [32, 40].

Notably, it has been reported that early interventions can help to avoid the persistence of psychological problems later in life [41, 42]. Anxiety and depression are indeed often associated with posttraumatic stress disorder (PTSD), and early childhood trauma associated with CHD may present as chronic stress in the ACHD patient [27, 43]. It has been theorized that many events surrounding living with CHD (i.e., numerous hospitalizations, continuous medical emergencies, and developmental milestone constraints) in conjunction with an individual's maladaptive internalized response to those events may result in long-term trauma. The hospital experience is often painful or even traumatic for young patients and memories of grief related to procedures as injections and surgeries can provoke persistent anxiety and affect later pain sensitivities and trust and adherence to medical care in adulthood [44].

By virtue of its complex nature, psychological trauma often remains mostly unconscious making it more challenging to recognize and treat. It manifests by ruminating thoughts and fears related to heart health stored in the body, developing over time as chronic, traumatic stress, and anxiety [27, 45]. Adult clinicians should inquire into a patient's childhood medical disorders and treatments as these early experiences surely influence illness identity as they develop into adults. This is a very important issue, because patient's beliefs about their illness and the results of its management may strongly influence their acceptance of the disease and QoL.

Illness identity can be defined as the degree to which a chronic health condition, like in CHD, is internalized and integrated into one's sense of self including four states: engulfment (feeling of being consumed by the disease), rejection, acceptance, and enrichment. The first two states are linked to an increase in emotional distress and, thus, increased symptoms of anxiety and depression [27, 46, 47].

Recurrent themes found in a narrative analysis of ACHD, looking back on their bad experience as children and adolescents, are feelings of embarrassment from others and denial, fears and pain, insecurity, physical limitations, isolation, and feeling different from others. These dolorous memories and feelings act as building blocks of ACHD's adverse illness identity [27, 46, 47].

19.5 Social Challenges

Stable social relationships and self-realization in the education and work fields greatly contribute to a good QoL. Socioeconomic outcomes have been explored in ACHD patients with contrasting results.

Nieminen et al. examined the social aspect of ACHD using data from national registries rather than relying on questionnaires. They found lower rates of

education, employment, and marriage and higher retirement rate than the general population [48]. Educational achievements could be influenced by the impairment in neurodevelopment more frequent among CHD patients [49]. Unsurprisingly, patients with cyanotic defects and complex CHD had the worst results in terms of education and career when compared with simple CHD and general population [50].

The social-economic background should be also taken into consideration when interpreting these data. Low familiar socioeconomic status, in fact, is associated with worst prognosis in CHD and decreases QoL [51, 52].

Adolescents and adults with CHD might experience physical limitations and school absences that prevented full interaction with siblings and peers, feeling different from them, causing isolation and social awkwardness. The feeling of "being different" seems to be a key theme in the lived experiences of patients with CHD. They not only perceive themselves as being different but also have the perception that their environment (parents, school, friends, etc.) does so [53–55].

Visible signs, such as cyanosis, digital clubbing, or scars, due to the heart defect or operations may trigger the perception that their body deviates from the norm [53, 54].

Moreover, depending on the severity of the lesion, patients may also be confronted with physical limitations and restrictions, possibly preventing them from achieving social independence equivalent to that of their peers. This feeling of being different seems to emerge predominantly in adolescence, a life period during which physical performance and bodily appearance are particularly important [53–55].

The feeling of being different is strengthened when parents create a protected world by hiding the heart defect from their child. This creates the risk that patients are confronted with their limitations later in life, without enough preparation, only when becoming adult [55, 56].

Moreover, patients with CHD might not have achieved typical educational milestones or developed employment skills because of restrictions (real or perceived) and/or fear of limited life span, often conditioned by parent overprotection, with consequence of denying them the ability to support themselves independently [53–57].

Accordingly, family, social, and healthcare environment have a deep impact on emotional growing and identity of these patients and delayed progression into full adulthood, due to insecurity and parental overprotection strongly contributing to frustrations and stress.

19.6 Sense of Coherence

Surprisingly, several studies have reported that patients with CHD, although all these adversities, felt that they had a good quality of life, even better than that of healthy counterparts [58, 59]. It has been speculated that it is related to increase sense of coherence (SoC) in these patients [60]. Antonovsky developed the concept of SoC to explain why some people become sick when stressed, whereas others remain healthy [61]. SoC is the central construct of Antonovsky's salutogenic

model, which posits that—to create healthy well-being—it is more important for people to focus on their resources and capacities rather than to focus on their disease (as in the classic pathogenic perspective).

Patients with a strong SoC are generally more resilient to stress and are better able to seek a solution to cope with the problem. Hence, SoC is a general expression of an individual's view of the world and is a mixture of optimism and control. SoC expresses the processes through which people manage to stay healthy despite being exposed to ubiquitous stressors, based on the extent to which the individual perceives: (1) stimuli as structured and predictable (*comprehensibility*), (2) resources available to meet the demands posed by these stimuli (*manageability*), and (3) the ability of the individual to believe that the own life has a scope, finding motivation and assuming the control over the own life (*meaningfulness*). It has been theorized that SoC exerts its positive influence on health through adaptive health behavior [60, 62].

Since individuals with CHD are born with this condition, they develop mechanisms to cope with their condition at an early age, and they continue to develop and refine these mechanisms well into adulthood. Accordingly, many patients, without getting caught up in despair, might acquire a greater sense of appreciation for life and expectations consistent with their capabilities and limitations, which positively influence their satisfaction and QoL [63, 64]. Clinical staff involved in ACDH care have a crucial role in helping patients to accept and cope with disease, improving SoC.

19.7 Counseling in ACHD

By performing strategic counseling, through openly and compassionate active listening and by discussion about their concerns and possible solutions, healthcare professionals can significantly help the ACHD patients to deal with disease, to find resistance resources, and to gain the ability to use them, contributing to the development of a strong SoC [62, 65]. The goal of counseling should empower patients and enhance their feelings of comprehensibility, manageability, and meaningfulness by providing information, making patients aware of the own strengths, and supporting patients to take up an active role in their process of care and control on disease and life.

It has been proposed that the *4 A's strategy* detects and manages psychosocial issues: ask, advise, assist, and arrange referral [66].

1. The first step is to ask the patient about specific challenges, trying to bring out problems and current experience of distress, anxiety, and depression. This step is very important since it allows the patients to recognize the problems, improving awareness. Through the sapient use of strategic questions, the counselor guides patients to question limiting beliefs and to struggle with a maladaptive narrative, downsizing perceived problems and eventually to change the perspectives, toward a new realized sense of self to include acceptance and enrichment.

2. Later, the healthcare provider can advise the CHD patient on the common challenges that lie ahead and how to plan for their management. Research shows that many adult patients know very little about their heart defect, the treatment, prevention, and prophylaxis. Educational efforts should be strengthened throughout the lifelong follow-up by expert nurse and doctors. Efforts should be made to allow the patients to have the right and complete knowledge of situation, reducing the sense of insecurity and confusion and accordingly increase sense of control, responsibility, and adherence to therapy, improving health behavior.
3. Moreover, the provider can assist the patient by addressing the immediate concerns of the patient, normalizing the most common challenges, and providing brief problem-solving. Initiating proactive discussions allows patients to recognize the normalcy of their concerns and the openness of healthcare providers to address them. Healthcare workers can make their patients aware of their resistance resources by informing and encouraging them in decision-making. When patients feel they are competent to deal with the problems associated with their disease, they develop an increased sense of manageability and increased confidence in the own power and self-esteem. Thus, counseling might help patients to feel that they have control over their fate and to find a scope of their own life. Patients often divide their lives into two periods: before and after their operation, building up from a painful experience their sense of meaningfulness.
4. Finally, the healthcare provider should arrange a consultation for those patients who eventually would benefit from speaking with a mental health specialist.

There is robust evidence of the favorable impact of psychological interventions for anxiety and depression among cardiopathic patients often involving a combination of education, relaxation training, cognitive restructuring (i.e., examining and challenging dysfunctional thoughts), and/or improving stress management skills [67, 68].

Kovacs and colleagues demonstrated the efficacy of a cognitive-behavioral protocol for the treatment of depression and anxiety among adults with CHD. The protocol emphasized psychoeducation on living with CHD, cognitive-behavioral techniques such as relaxation training, cognitive restructuring, and strategies to improve social interaction/communication [69].

There is growing data proving how a positive health psychology may impact QoL for someone living with a chronic illness [70]. Promoting positive protective factors, particularly self-efficacy, self-esteem, humor, optimism, and positive affect, is important throughout the life span of patients with CHD [71]. Opportunities to cultivate resilience can be explored through engaging patients in direct, structured interventions [72], or during interactions with members of an ACHD Center team, ideally formed by interdisciplinary specialists (including the primary healthcare providers, cardiologists, cardiac surgeons, expert nurse, psychiatrists, psychologists, social workers) that cooperate to improve psychosocial functioning of ACHD patients [73].

It is crucial for ACDH healthcare providers to consider the psychosocial dynamics in the family of the patients and to make efforts to enhance stability and serenity

of the relationship, considering the patient and the family as a single unit [74]. Family environment has the most important rule for children's and adolescents' healthy development. High level of parenting stress is common in parents of children with severe heart diseases as they continue to experience repeated hospitalizations and accommodate the uncertainties and challenges that arise from the disease. Parental distress, adverse family relationship, poor parental control skills, single parent status, maternal anxiety and fears, controlling parental style, low income, and problems related to unemployment have been all proven to be associated with behavioral and psychosocial difficulties and low QoL in children with CHD [75].

Counseling of patients and their family is really important to early tackle these problems and obstacles since early identification, explanation, and easing potentially traumatic and stressful situations have a significant impact on the comfort of the child and the whole family and help to strengthen support relationship and accordingly improving QoL [76]. It has been proven that parents would like to receive more counseling than cardiologists believe is desired or warranted [77]. The aim of parent education programs is to assist families through enhancing the parent's knowledge, behavior, and cognition, improving parent-child interactions and family relationships [78]. The common core intervention approaches are the use of narrative therapy enabling families to tell their own stories, thus facilitating emotional processing and co-construction of meaning, enhancing parents' skills in communication, and stress-behavioral management and provision of psycho-education to deepen parents' understanding of their child's condition and associated developmental challenges [78, 79].

Living with CHD can be extremely stressful for the family, because both the complexity and uncertainty of the condition can initially overwhelm both child and parents. By educating the child about his disease, parents and healthcare professionals can make the condition more predictable for the child and allow him/her to accept and cope with it [56, 60]. By contrast, parents who conceal the heart defect from their child (to protect their child from adverse experiences) increase the risk of delusion and insecurity when, later in life, adults with CHD have to face with the impact and limitations of the disease without enough preparation [80]. Structured education programs are indispensable to arise awareness.

It is important also to deal with anxiety of parents that have been used to manage the disease of their sick child and have now, when grown up, concerns about the maturity and the child's ability to engage in self-care monitoring and care [81]. Diagnosis of CHD is experienced by parents as a trauma that deeply influences the relationship with the kids. Parents have often experienced extreme sufferance and exhaustion for all the stress related to fighting to survive of the kids [82, 83]. Because of the uncertainty of survival during adulthood for some children with CHD, parents often viewed their children as vulnerable. Frequently, they don't feel the kids are prepared to be an adult, they still need to protect their kids, and they are reluctant to favor independence [75, 84]. These feelings of insecurity affect parents' ability to set long-term goals for their children's future and can have an impact on educational and employment and social achievement of their child, preventing his/her independence and realization. Of note, overprotection of the parents determines a vicious

circle that prevents self-esteems, experience, and maturity of the child that feels insecure to avoid involvement on his care and is scared to break the chains [75, 81, 84, 85].

Counseling of the family should allow to recognize and deal these fears, and efforts should be made to reassure and promote independence and responsibility of the adolescent [74, 86].

19.8 Counseling in Transitional Age

The adolescents' knowledge and understanding of their cardiac condition need to be addressed by health professionals who must develop specific age- and gender-appropriate education programs to meet the specific psychological needs of the affected adolescents [87, 88].

Adolescence is a complex period of transition from childhood to adulthood. Specific issues emerged during puberty, such as research of autonomy, personal identity, sexuality, education, and vocational choices, become more difficult for an adolescent who is also coping with chronic illness [76, 89]. Upon entering adolescence, patients with CHD face the challenge of autonomy struggling with parental overprotection [84, 85]. Furthermore, adolescence is a crucial consolidation phase of health healthy behavior, such as, for instance, the adoption of good food practices and exercises, but also a period in which risk behavior is manifested for the first time, such as the use of drugs and alcohol and risky sexual practices [90, 91].

The transition program has been developed to facilitate the journey from pediatric to adult cardiology, to identify specific patient's needs, to emphasize the peculiar challenges associated with adolescence, to educate and counsel patients and their families, to screen for psychosocial problems, and appropriately to refer patients to social worker, psychologist, or other professionals [88, 92]. It may be difficult for chronically ill adolescent patients to break ties with their pediatrician, and the transition program helps introducing to new adult healthcare staff during a bridge period, where both pediatric and adult doctors and nurses ideally work together to guarantee the correct clinical continuity by building a relationship of trust. Two important goals are to prevent loss of follow-up care and to foster and encourage patients' self-care behaviors [88, 92].

Counseling helps educate patients about the heart defect and its implications. The purpose of counseling in the transition program is to prepare the patient for the transfer to adult medicine by gaining an understanding of the clinical picture of their disease and of the treatment goals and possibilities and developing a personal responsibility for medications and lifestyle [74, 87]. A carefully planned counseling in the transition to adult health care should improve self-reliance, enhance autonomy and independence, and support young people in attaining their maximum potential [55, 60].

There are specific issues that need to be addressed during counseling in adolescent CHD patents:

Body image—A distorted and not accepted body image related to surgical scars or cardiac lesions that resulted in more severe bodily disfigurement (smaller stature, deformity/dysmorphism) is an important issue, especially felt during adolescence that can cause negative impact on self-esteem, symptoms of depression, and impaired psychosocial interactions [45, 76, 89]. The onset of sexual maturity and the difficulty accepting the physical aspects of might exacerbate the discomfort [93].

Employment—Patients may experience difficulties in finding a satisfying job, eventually because of a low educational level and also because of potential physical disabilities. This problem is very important since it affects economic status, independence, and security of patients [94, 95]. Counseling helps to achieve personal life projects, stressing their abilities rather than disabilities [70, 76].

Pregnancy—Discussions about future pregnancies, family planning, and contraception should begin in adolescence to prevent accidental and potentially dangerous pregnancies in women with CHD [96]. For many women with mild cardiac defects, pregnancy is generally well tolerated with no long-term sequelae. However, for women with more complex defects, pregnancy, labor, and delivery should be monitored closely or may even be contraindicated in some patients [97].

The risk for pregnant women with CHD of having adverse cardiovascular events—such as symptomatic arrhythmia, stroke, pulmonary edema, overt heart failure, or death—is determined by the ability of their cardiovascular system to adapt to the physiological changes of pregnancy. An individualized approach and expert opinions remain paramount in counseling women with CHD with a pregnancy wish [96, 98]. Different congenital conditions carry specific risks based on their morphological features, previous operations, and current hemodynamic status. Patients with Eisenmenger syndrome (or other severe forms of pulmonary arterial hypertension), severe/progressive aortic dilatation, or severe left-sided obstructive lesions carry a very high risk associated with pregnancy and should be made aware of such risks and eventually explicit advice not to embark pregnancy but to prevent devastating situations. If an unplanned pregnancy occurs, early termination should be considered. If the patient chooses to proceed with pregnancy, however, there is essential need for planning care in a tertiary, multidisciplinary unit providing all kinds of support. In this regard, early consultation on contraceptive issues is very important. The impact of heart disease on childbearing potential needs to be explained clearly and sympathetically [96, 98].

Overall, there is a higher incidence of fetal and neonatal adverse events—including intrauterine growth restriction, premature birth, and fetal loss—in women with CHD compared with the general population [99–102]. This risk is highest in women with poor functional class, cyanosis, and left heart obstruction to flow (which restricts cardiac output and thus flow to the placenta) and is amplified by any other obstetric risk factors [102].

Counseling has to address how pregnancy may affect not just them/others but also the fetus and the rest of the family. This allows women to make an informed choice. Considering the risk that the cardiac defect will occur in their offspring

(estimated for pregnant women with CHD between about 3% and 12%), compared with the background risk for the general population (0.8%), genetic counseling is recommended and also early fetal echocardiography and prenatal counseling [97, 98].

Specific counseling is also necessary regarding medication during pregnancy. Several drugs require withdrawal to avoid any teratogenic effects and interference with normal embryonic development (e.g., ACE inhibitors, angiotensin receptor blockers, warfarin). Pregnancy increases the risk of thromboembolic event in patients with metallic prosthetic valve; it is necessary to balance the risk of embolic event switching warfarin with heparin that doesn't cross the placenta but is less effective for thromboprophylaxis and the risk of teratogenic effect to the fetus related to maintaining warfarin treatment [96–99]. On the other hand, when suggesting contraception, it's also necessary to take into account that estrogen pills used for contraception might be contraindicated for women with cyanotic CHD or residual pulmonary hypertension because of risks of fluid retention or thrombosis, and intrauterine devices increase risk of endocarditis [96–98].

Acknowledging the limits of our knowledge or our predictive capabilities provides a credible backdrop to establish a constructive discussion in order to provide risk stratification and tailored care [96, 98]. The prognosis is not a simple informative event, but a *communication*, i.e., accompanying the patient in difficult choices, one of the main tasks of the medical profession [79].

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Part VIII

End-of-Life Care: Nursing Care in the Final Stage of Life

Advance Care Planning and Advance Directives

20

Caroline Scribner and Kristina Fontecha

20.1 Part 1: Developing a Comprehensive Advance Care Planning Program

20.1.1 Introduction

The number of adults with congenital heart disease (CHD) outnumbers children born with congenital heart defects. Advances in surgical and procedural techniques have allowed patients born with congenital heart disease to live well into adulthood, though frequently with symptoms and at risk of premature death [1, 2]. As medical care for adults with CHD continues throughout their life span, it should be recognized that death and dying are a part of this continuum and that preparing patients for this part of their illness is an integral part of the medical care they receive. Although advance care planning (ACP) discussions are generally considered an important part of adult congenital heart disease (ACHD) care by both patients and providers, there is no standardized approach to these conversations. However, we are seeing a shift toward ACP program implementation, and the incorporation of palliative care discussions into the 2018 American College of Cardiology/American Heart Association ACHD Guidelines emphasizes the importance of this movement [3].

Advance care planning is often misunderstood by patients and families, and is often challenging for healthcare providers, who struggle with difficult prognostication and concerns regarding their patients' readiness to have these discussions [4, 5]. It is important to recognize that advance care planning should not be reserved for conversations at the end of life. These discussions should occur early in the disease process as they provide valuable information for the clinician about the patients'

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wishes, goals for the future, and how to optimize quality of life. They also provide valuable information for many patients who lack understanding of their disease process. In one North American study evaluating patients' preferences for ACP discussions [4], it was noted that, for those with simple complexity of disease who underestimated their life expectancy, ACP discussions were valuable in relieving anxiety regarding their perceived life expectancy. The awareness of ACP prepares the patient for what can be expected, not always with the objective of highlighting the seriousness of their illness, but often as a way to reduce anxiety and stress [4]. For many, these conversations focus on their future care and emphasize patients' values and beliefs, which can be important to guide clinicians in caring for CHD patients throughout their lives. For some, having these conversations when patients are facing death is too late. It deprives patients of psychological support during their illness while coping with debilitating symptoms [1, 4].

20.1.2 When to Have these Conversations with Patients

Knowing when to have these conversations with patients is often a challenge for providers. Although timelines for initiation of advance care planning conversations have been proposed by researchers in this field [2, 6], there is no specified timeframe to initiate these conversations—it is dependent on patients' comfort level with these discussions, and each situation is unique. Despite some providers' hesitation to broach the topic of advance care planning with patients in general [7], patients have shown interest in having these discussions with their providers. In a survey of 200 adult outpatients with CHD, only two patients reported having end-of-life discussions with their providers, although 54% reported having discussed their wishes with at least one other person and 78% agreed that a medical team member should bring up these discussions [8]. In a small qualitative study evaluating patients' views on palliative care and ACP [7], most patients expressed interest in learning about palliative care and ACP and stressed the importance of their providers normalizing these conversations with them. The majority of the participants had thought about or initiated discussions with their family members about ACP. At one point in this study, 72% of participants described having negative feelings about living with CHD and wanted to know more about their condition and prognosis as the knowledge provided them with some control over their illness. Despite the majority belief that these conversations would be standardized practice, timing of conversation varied greatly, from onset of diagnosis to older age [7], and in other studies, early adulthood has been suggested as the optimal timeframe for initiation of these discussions and most preferred discussions while healthy, before being diagnosed or hospitalized for a life-threatening illness [2, 4].

Another study examined the transition of care from repair to palliation in adults with congenital heart disease [2]. By incorporating advance care planning discussions into the management of ACHD patients, the study proposed that clinicians should preempt these conversations on ACP. These discussions are warranted much sooner in the disease process as they can help reduce stress and anxiety, improve

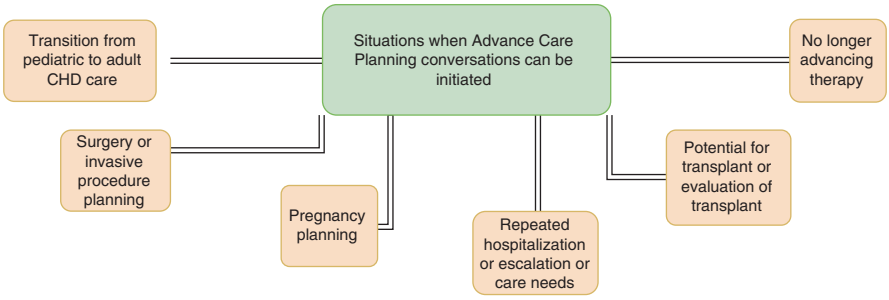

Fig. 20.1 Diagram of opportunities over the life span to initiate ACP conversations

quality of life, and reduce symptoms. The research team in this study developed a timeline for advance care planning discussions with suggested key moments in providing information about outcome and integration of palliative care and end-of-life care. Notably, the identification of patients at risk of heart failure, arrhythmias, those who require re-intervention and possible transplantation were most likely to benefit from these conversations [2]. In another study from an American northeastern tertiary ACHD center, 152 adults with CHD were observed and found that overall, 18-year olds were considered the most appropriate age to begin having these conversations, taking into consideration a patient's maturity level [4]. Depending on the maturity of the patient, the transition from pediatric care to adult care is often ideal to begin conversations about expectations of disease process, allowing for the ability to tailor lifelong advance care. As the disease process itself is dynamic, these conversations are as well. Figure 20.1 provides a diagram of opportunities to initiate the advanced care planning conversations. Maintaining an open dialogue with the purpose of free-flowing communication between provider and patient is beneficial.

20.1.3 What Is the APP/RN Role

There is a lack of research within the ACHD community that highlights the advanced practice provider (APP) or nurse coordinator/registered nurse (RN) role in having ACP conversations. Although nurses are ideally situated to facilitate ACP, there is often discrepancy in their roles [9]. In the outpatient setting, the APP or RN typically has a strong relationship with these patients and often provides a holistic approach in their patient care. In the inpatient setting, it is appropriate for nurses to teach patients and their families about why planning is important and to help initiate a conversation while they are in the hospital [9]. In both the inpatient and outpatient setting, nurses are frequently the strongest patient advocates and can ensure these conversations are had with their patients and that their wishes are carried out. Nurses may also play the role of educators, communicating to providers why conversations about ACP are important and addressing barriers the providers may face in having these conversations with their patients. Despite their potential role as advocates for the ACHD patient, nurses face barriers to participating in these conversations. In

Table 20.1 Patient/provider goals of ACP

Goals of advance care planning discussions	
Patient	Provider
• Reduce anxiety related to outcomes and life expectancy • Verbalize goals related to health and quality of life • Communicate to family and friends patient's desires regarding care • Build trust with provider as wishes and goals are clarified and documented	• Provide information to patients regarding expectations of future care • Understand patient perspective to tailor appropriate care plan • Reduce potential future conflict among care team members including family, friends, and loved ones • Develop relationship with patient that allows patients needs to be met

one Quality Improvement project performed at an ACHD center on the East Coast of the United States, Shigeko Izumi, PhD, RN, found that nearly half of the nurses surveyed reported never to rarely ever being involved in ACP conversations [9]. This project presented several perceived barriers including the lack of sufficient time, lack of physician support, as well as the belief that ACP was not a nursing responsibility. Additionally, some physicians in this particular QI project commented that ACP conversations should not be done by nurses given the prognostication required. This emphasizes the misunderstanding that ACP is the same as end-of-life planning. Due to the nature of the nursing role as first-line caregivers, it reasons that nurses have the potential to play a vital role in these conversations to support and empower patients to take an active role in advance care planning. Table 20.1 describes the goals of ACP for both the patient and the provider.

20.1.4 Model of Delivery of Discussions in Outpatient Clinic

Incorporating the difficult conversations about ACP into practice requires a structured approach utilizing several team members who understand their involvement. Patients with congenital heart disease require complex care and coordination throughout different points in their life. The transition from pediatric care to adult care can be challenging. In some cases, providers' initial meeting with complex patients occurs in adulthood after the patient had already been through several operations, hospitalizations, and faced critical health milestones in their lives. Because of the complex needs of the patients, having a structured model of delivery of these discussions is beneficial to minimize stress on both patient and provider [6]. ACP discussions are not end-of-life conversations but rather discussions on goals with the objective of understanding a patient's wishes about their care.

20.1.5 The Serious Illness Conversation Guide

The Serious Illness Conversation Guide [10] is a framework for standardizing serious illness conversations and was developed by the palliative care team at Ariadne Labs, with the goal, "for every seriously ill patient to have more, better, and earlier

conversations with their clinicians about their goals, values, and priorities that will inform their future care." This guide has become a useful tool for palliative care programs worldwide, and its usage is being expanded into the routine clinical practice of patients with chronic illness. For non-palliative care providers, training on how to have these conversations is imperative for effective advance care planning discussions. Specifically, incorporating nurses and advanced practice nurses into this framework is key in this process. Nurses frequently develop long-lasting relationships with patients, and the roots of a holistic approach in nursing care makes them well suited to support patients in achieving their goals regarding their care.

An approach to ACP discussions may be a shared-team approach, incorporating APPs, RNs, and social workers, into the conversations to ensure all needs are addressed. APPs and RNs may assist with a holistic patient assessment, whereas social workers may provide significant emotional support [1].

20.1.6 Patient Selection

Two-thirds of adults with congenital heart disease will die a CHD-related death with death from heart failure peaking around the fifth decade of life [2]. With such a shortened life expectancy, patients' focus on quality of years becomes paramount. For ACHD centers who seek to implement advance care planning into their care of patients, a stepwise approach to program development can be useful.

Step 1: Identify patient group(s)—Focus on complex lesions for early discussion before worsening cardiac status.

Step 2: Introduce the concept of advance care planning during visit—Introduction can be initiated in-person or through telecommunication. Additional visits may be required.

Step 3: Tracking and follow-up—Add to database for tracking, and document changes to ACP as needed.

It might be best to engage a specific subset of ACHD patients and then expand the pool once providers are more comfortable in having these conversations. When selecting patients to engage in these conversations, consider beginning with patients whose natural trajectory of disease likely involves premature death, to be able to facilitate a smoother transition through the process. For example, select only patients with Eisenmenger physiology and Fontan circulation as initial patients and identify these patients during each weekly clinic visit. For clinics utilizing APPs, the APP introduces the concept of advance care planning during the visit. Additional information may be sent to patients via mail or online health provider portal following the visit with the intent to set up a dedicated ACP visit in the future [2]. During these future visits, nurses, APPs, social workers, or physician providers may continue the conversation ensuring patients' requests are addressed, their expectations are met, and their questions are answered. Figure 20.2 provides a template for ACHD programs to initiate serious illness conversations with their patients.

The conversations may continue as needed during future visits. In cases where a stable patient's clinical status changes, past conversations regarding ACP may be

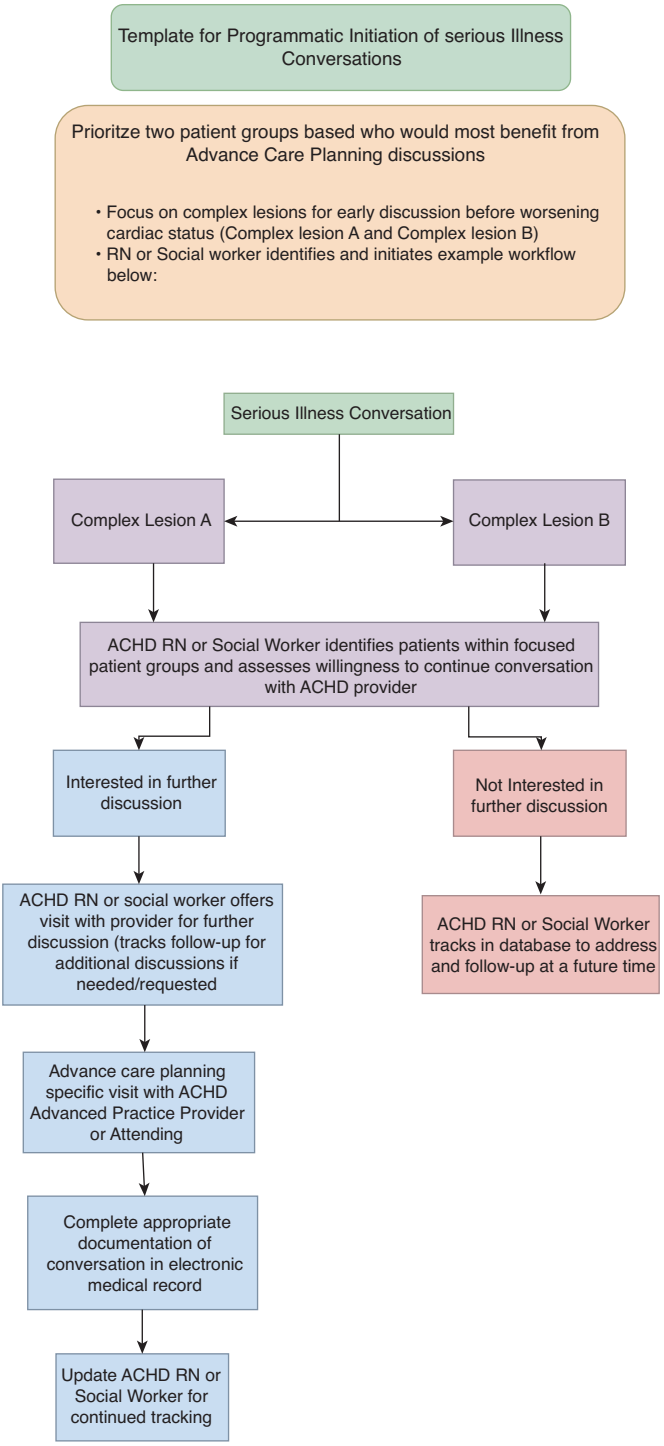

Fig. 20.2 Template for programmatic initiation of Serious Illness Conversations

used as a reference to guide treatment and aid in making difficult decisions in approach to care. For instance, if a patient is clear that their ability to verbally communicate with family members is critical to their quality of life, then providers could remind them about this aspect of the ACP when they face the possibility of a permanent tracheostomy. Having ACP conversations when the patient is in good health also eases the pressure of the conversation on the patient, allowing the patient to think about their wishes and the qualities in their life they could not live without [5], rather than worrying about the possibility of losing those capabilities.

20.1.7 Having the Conversations

Introduction to these conversations can be at any type of visit, but to allow for adequate time and understanding, having a dedicated ACP discussion visit is beneficial. Following the Serious Illness Conversation Guide, providers can use the Ask-Tell-Ask framework allowing a patient to share their understanding of their disease process and expectations for the future by asking them about their worries and concerns, what information they would like to have, and what they understand about their disease [1, 8]. This is followed by the provider sharing information about disease prognosis and concludes with ensuring the provider understands the patient's wishes clearly and that all the patient's questions are answered. This method ensures clarity and understanding by both parties.

20.1.8 Post Conversation Nursing Assessment and Follow-Up

The patient's initial conversation with their provider should include the patient's comprehension of their disease process, their goals and wishes for future care, as well as any questions related to the future and receiving information about their prognosis. The nurse or social worker should then follow up as needed on any outstanding discussion items, setting up communications as needed to continue the conversation. Future visits could be arranged to continue the conversation, answer additional questions, or involve other family members as requested by the patient.

20.1.9 Tracking

Following the initial advance care planning conversation, the patient is entered into a tracking database. Keeping track of the record of the patient's ACP status ensures the patient receives the attention needed. These recorded previous conversations provide valuable resources in any event when an acute illness exacerbates and a patient faces new choices in care and serious illness or death becomes a possibility. Tracking conversations in the medical record is imperative for those who subsequently care for the patient or when a patient is hospitalized and their primary ACHD provider is not part of their inpatient care. Additionally, as a program

development tool, it's important to keep track of which patients have had ACP conversations, who needs a follow-up conversation visit, and as a method to keep track of any outstanding items.

20.1.10 Referral to Palliative Care and Hospice

The ACHD RN and APP work in tandem with the patient and medical team to approach end-of-life care in a holistic and supportive environment to prioritize the needs of the patient. Similar to Advance Care Planning, palliative care consultation can be initiated at any time as an added facet to patients' ACHD care. Often, it is initiated too late, and patients are frequently deprived of the psychosocial care they need at the end of life. In a retrospective study in 2011 of 48 patients with congenital heart disease who died from non-perioperative congenital heart disease-related deaths, only 10% of patients were reported to have documented end-of-life discussions prior to their final hospital admission, and those who had documented advance care planning wishes were significantly less likely to undergo attempted resuscitation than those without documented conversations [11].

The PAL-HF trial was a pivotal trial in the discussion of integrating advance care planning with standardized management of heart failure patients, and its importance is being recognized in the care of adult congenital heart patients as well. The trial integrated advance care planning into standard care of chronic heart failure using interdisciplinary palliative care interventions, and it showed that incorporating palliative care with usual care resulted in improved depression and spirituality scores with similar anxiety scores and no effect on hospitalization or mortality [12]. lt further accentuates the value of integrating palliative medicine into patient care and emphasizes a more holistic approach.

The primary focus of palliative care is to empower patients' decision-making to prioritize their comfort whenever possible. Colloquially known as "comfort care," palliative care supports relief of distressing symptoms whether it be in the form of pain or psychosocial factors like anxiety and depression [1, 13]. As the ACHD RN and APP can be in closer day-to-day communication and follow-up with patients, assessment of patients' needs should be discussed not only between patient and provider but also provider to provider as another avenue of supporting ACHD patients. It is important to understand what resources are available to the patient within an ACHD program, healthcare facility, and health insurance carrier.

Hospice is another facet of end-of-life care focused on providing comfort for patients who have been identified by their provider as approaching end of life. In the United States, this is often quantified as likely to die in 6 months or less [11]. Referrals for hospice can be met with reluctance from patients and loved ones since it is sometimes viewed as "giving up" or "exhausting all options," especially in a setting where prolonging life remains the primary patient goal [7]. Furthermore, many ACHD providers have reported similar hesitation with initiating a referral for hospice due to little training and knowledge regarding the referral process. It is especially important for the ACHD RN and APP to assess patient readiness when

referring to hospice [14]. This ensures that both the patient and the healthcare team are all in agreement and understanding of the expectations of care at end of life.

20.2 Part 2: Advance Directives

20.2.1 What Is an Advance Directive

Advance directives are legal documents that identify the patient's wishes about end-of-life care ahead of any significant clinical or critical events [15]. This document, created by patients, clearly outlines what treatment and care options the patient would be agreeable to in the event they are not able to fully communicate these wishes. There are two types of advance directives that patients can complete. Table 20.2 compares the two types of advance directives. Many preprepared forms can include both types as a way to further decrease confusion and burden of completion, but the patient has the prerogative to partially or fully complete both types of advance directives.

20.2.2 Who Should Complete an Advance Directive

All young adult and adult patients with congenital heart disease are encouraged to complete an advance directive prior to surgery, hospitalization, or significant progression of CHD. As an ACHD nurse or APP, it is important to acknowledge cultural considerations for patients when discussing the need for an advance directive [16]. In order to provide holistic care of the CHD patient, the clinician should be aware whether the patient would like to initiate this discussion with family, friends, or someone they trust prior to finalizing the document.

Table 20.2 Comparison of advance directives

Living will	Durable power of attorney
• Lists treatments or interventions a patient does or does not consent for in the event they are unable to verbalize themselves. Examples can include: – Resuscitation efforts – Mechanical ventilation – Tube feeding – Analgesia – Organ/tissue donation • Does not designate individual who can make healthcare decision on the patient's behalf if unable to do so • Can address more than one scenario or treatment option • Can be specific to the patient's diagnosis (i.e., heart failure requiring IV inotrope)	• Designates a specific individual who can make decisions for the patient in the event the patient is unable to • Chosen individual who can make decisions on patient's behalf can be called: – Healthcare proxy – Healthcare agent • Proxy can make both healthcare and financial decisions on behalf of the patient • Proxy cannot be doctor or part of the patient's medical care team • Does not list treatments or interventions a patient consents for in the event they are unable to verbalize

CHD progresses across the life span; therefore, developmental considerations should also be recognized by the nurse or APP [16]. This is especially true with young adult CHD patients as they take a more active role in healthcare decision-making compared to their previous pediatric care. A patient who has developmental or neurological delays would require clear identification of the patient's legal guardian and ensure the appropriate documentation is present in the patient's physical or electronic medical record. In this circumstance, an advance directive is not necessary since a court-appointed guardian has already been deemed appropriate and able to make decisions on behalf of the patient.

20.2.3 Why Complete an Advance Directive? Why Is It Important?

In the event a patient is unable to make healthcare decisions, an advance directive allows the healthcare team to provide care aligned with the patient's preferences and wishes. The advance directive is able to be a flexible document that can be updated regularly in the setting of the patient's life events like surgery, pregnancy, and transplantation [16]. The advance directive acts as a guide for the healthcare team, family, and friends for how to best prioritize the patient's comfort and care.

As a part of the clinician's role in patient care and care planning discussions, it is necessary to understand the importance of advance directives in maintaining a trusting and active care relationship with patients. A cross-sectional study of ACHD providers focusing on providers' perspectives has shown that palliative care discussions including advance care planning, end-of-life care, and advance directives can be difficult for providers to initiate [5]. By utilizing a specific framework and protocol for initiating the discussions, healthcare providers could become more confident and proficient in implementing these processes as a part of their standard practice.

20.2.4 When to Complete an Advance Directive

The American College of Cardiology and the American Heart Association recommend that an advance directive be completed when it is culturally and cognitively appropriate as well as before the patient is extremely ill or hospitalized [16]. Conversations regarding advance directives can be initiated at any time by either the clinician, social worker, or the patient.

The World Health Organization recommends early discussion of advance care options in the progression of a chronic illness [1]. A psychosocial evaluation of CHD patients who perceived themselves as "cured" emphasized to providers the importance of gauging patient preferences in advance and, further, that a patient will generally lean toward less aggressive management options/interventions when given adequate time and preparation [17].

In the situation a patient pursues aggressive management and/or treatment options, particularly associated with end-of-life or critical cardiac decompensation,

there may be an increased mental toll on the patient, family, and friends. Additionally, the stress of financial hardship further emphasizes the need for an advance directive [18]. A systematic literature review of heart failure costs between 2014 and 2020 found the average annual cost per hospitalization alone was roughly $24,383 per patient though this estimate was variable among subspecialties [19]. In the USA, the total cost of care for patients with heart failure is estimated at $43.6 billion of which 70% is attributed just to medical costs [19].

20.2.5 How Is an Advance Directive Completed/Initiated by the ACHD Provider

Topical conversations on congenital heart disease published in *Current Cardiology Reports* in 2020 reviewed the perspective that patients preferred their CHD care provider to initiate conversations related to advance care planning, including discussion about advance directives [1]. Serious illness conversation models can be used to introduce advance directives to patients and caregivers. Many ACHD centers introduce advance directives when possible surgical/transcatheter planning discussions are initiated. Figure 20.3 is a sample electronic message that may be sent to patients:

Many patients may undergo a wide range of procedures with varying complexity in both a children's hospital and adult hospital, no matter the age. An advance directive on file further helps to guide the patient's care team regardless of age or CHD complexity. Advance directives can be a universal tool for communicating among providers regarding patient wishes since many times the primary ACHD cardiologist is not the primary treating physician in an inpatient setting.

20.2.6 Nursing Assessment

When the ACHD RN or APP is first initiating the conversation regarding advance directives, the nurse or APP should take into consideration patient readiness while in clinic or discussing via telecommunication (secure patient portal messaging,

We request all patients preparing for a procedure to fill-out and bring an advance directive to their pre-op or pre-cath appointments. We encourage all individuals to have an advance directive on file to be clear about their medical wishes. It is best to complete these forms while you are well and have the opportunity to discuss with your family and loved ones. If you do not already have an advance directive, you can search online for an interactive electronic form that can be completed from your desktop, laptop or tablet. You can complete the whole letter or just part of it.

Fig. 20.3 Example of patient messaging discussing advance directives as a part of pre-procedure planning

telephone calls, or video visits) [7]. It is crucial to review the patient's current health status [14] and understand the effect on patient decision-making. Table 20.3 compares examples of patient's decision-making in an inpatient versus outpatient setting.

While completing the nursing assessment related to advance care planning, specifically advance directives, the RN or APP should evaluate the patient's level of understanding of clinical situations [14]. While a patient may not fully comprehend the specific nuances of their clinical care, discussing these topics in different communication mediums and more than once can help to reinforce the patient's understanding. For example, depending on a patient's learning style, they may prefer print materials to review instead of discussions over the phone. Additionally, for patients whose first language is not English, educating patients in their preferred language should be a practice standard for the RN or APP to provide high-quality culturally competent care [7]. Other alternatives may also include a visual sliding scale for patients to depict priorities within preferences visually.

When completing the advance directive, the ACHD RN or APP should allow the patient adequate time to consider preferences and complete discussions with loved ones should they so desire. This provides the patient the option to complete advance directive decisions at home and may be beneficial in the long term for advance care planning.

20.2.7 Future Nursing Research Opportunities

The breadth of data on advance care planning conversations in patients with serious illness, including those with congenital heart disease, is growing. Despite this, there is little data on the role of nurses and APPs in these conversations. Utilizing standardized approaches and inserting these conversations into general ACHD practice allows for the roles of ACHD providers to be clarified as well as presents the opportunity for more studies to be conducted to investigate the best ways to utilize these providers.

Table 20.3 Comparison of patient example decision-making scenarios

Inpatient	Outpatient
54-year-old Fontan with unexpected admission for worsening heart failure and arrhythmias requiring discussion from the care team for advance care planning Patient verbalized feeling ill-prepared and not ready to have discussions while in the hospital Patient emphasized strong preference for only identifying healthcare proxy at the time of discussion without identifying wishes for interventions and treatment options	40-year-old Fontan with worsening heart failure engaged in multiple conversations with the care team Patient utilizes regular video visits and online patient portal messaging to interact with care team and remain an active advocate for her care

20.3 Conclusion

Advance care planning, the use of advance directives, and referrals to palliative medicine and hospice are underutilized within care of the adult with congenital heart disease. As many of these patients face premature death and often endure debilitating symptoms at end of life, incorporating these components of care is integral. To encourage patient support and care across the life span, providers should strongly consider initiating advance care planning prior to any sudden changes in health status and when it is culturally and developmentally appropriate for the patient. As a part of advance care planning, patients should be given adequate time and information to formally complete an advance directive to guide the patient's care per their wishes. Finally, despite the interpretation that palliative care is viewed as "giving up," reinforce the specialty as a holistic approach to ACHD care with resources including hospice for care at end of life.

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Part IX

Coronavirus Disease 2019 (COVID-19) Pandemic Implications in ACHD

COVID-19 and Adult Congenital Heart Disease

21

Ivana Babić and Margarita Brida

21.1 Introduction

Infectious diseases pose a global threat to human health and life without knowing political or geographical boundaries. The consequences can be detrimental, and such is an example of the Spanish flu pandemic caused by the H1N1, A type virus in 1918. Spanish flu pandemic led to the deaths of more than 50 million people, which in comparison, accounts for more casualties than in both World Wars combined. Furthermore, this same virus caused repeated outbreaks through its mutations in the years to come, including the last H1N1 pandemic in 2009 informally called the swine flu [1].

In the more current era, coronaviruses have shown epidemic propensity similar to influenza viruses [2]. The name was derived from the crown-like spikes on the virus surface (lat. Corona, crown), and in 2002 and 2012, coronaviruses caused severe acute respiratory syndrome coronavirus (SARS-CoV) and the Middle East respiratory syndrome coronavirus (MERS-CoV) epidemics, respectively [3, 4]. The most recent severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) was first detected in humans in late 2019 in Wuhan, China, and has been a global threat with an ongoing pandemic of coronavirus disease 2019 (COVID-19) infecting more than 400 million and causing death of six million people to date [5, 6].

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21.2 Basic Characteristics of SARS-COV-2 Infection and Its Management

In general, SARS-CoV-2 infection results in mild flu-like symptoms in the majority of healthy and young patients affected [7]. The virus primarily attacks the respiratory system but can also affect multiple other organ systems. It uses glycoprotein spikes to bind to cellular ACE2 receptors with the help of a specific cellular protease that allows it to enter target cells [8]. Beyond airway epithelial cells, ACE2 receptors are expressed in multiple other organs, including vascular endothelial cells, immune cell tissue, the heart and the intestine.

The most common symptoms of SARS-CoV-2 infection are chills, fever, dry cough, runny nose, headache, fatigue, myalgia and/or diarrhoea. Depending on the strain, loss of smell and taste is also recorded. More severe presentation of the disease is clinically characterized by the onset of dyspnoea, occurring in approx. 14% of patients [7] and can progress to acute respiratory distress syndrome (ARDS). In general, patients at an increased risk of severe form of the disease are elderly patients with chronic illnesses such as cardiovascular patients, patients with chronic lung disease, severe chronic kidney and liver disease, patients with uncontrolled diabetes, patients with malignancy and individuals with morbid obesity (BMI > 40 kg/m^2) [9, 10].

The SARS-CoV-2 infection is confirmed by positive nasopharyngeal swab test [11]. The virus replicates in the upper respiratory tract, resulting in a possible spread through social contacts. A person is contagious a few days before the onset of symptoms (3–4 days), which poses an additional threat for individuals in the immediate environment. From the beginning of symptoms, the duration of the disease is, in vast majority of cases, 10–14 days.

Management is largely supportive and includes symptomatic therapy such as oral antipyretics and analgesics as per need. Social distancing is necessary in order to prevent spread of the disease. In case of dyspnoea hospitalizations with diagnostic workup and prompt treatment measures are indicated. Chest radiography at this stage usually shows bilateral atypical pneumonia, while laboratory testing commonly exhibits lymphopaenia, increased C reactive protein (CRP), and not uncommonly elevated levels of procalcitonin (PCT), which are regarded as adverse prognostic signs. Oxygen therapy is often required [11].

21.3 Potential Impact of SARS-COV-2 on the Heart

The cardiovascular involvement induced by COVID-19 disease has generated considerable concerns due to various degrees of myocardial injury and arrhythmias reported [12, 13]. A subset of afflicted patients may present with chest pain, an increase in cardio-selective enzymes, changes in the ECG, and/or an increase in NT-ProBNP levels. Viral myocarditis, hypoxia or myocardial ischaemia due to pre-existing coronary disease, mismatch of coronary flow and increased demand, stress cardiomyopathy or systemic hyperinflammation may play a role. The increasing occurrence of venous and arterial thrombosis reported is multifactorial and may be

related to immobilization, hypoxia, hyperinflammation and diffuse intravascular coagulation [11]. Myocardial injury in association with COVID-19 infection is associated with adverse outcome; however, the prognosis of patients with underlying cardiovascular disease but without myocardial injury in the setting of COVID-19 infection seems to be relatively favourable [14].

21.4 Resilience and Response of Adult Patients with Congenital Heart Disease During the COVID-19 Pandemic

Adult patients with congenital heart disease (ACHD) by definition have underlying cardiovascular disease. Many patients with ACHD are afflicted with residual haemodynamic lesions, such as valve dysfunction, diminished ventricular function, arrhythmias or cyanosis [15]. As cardiovascular diseases have been recognized as a risk factor for mortality in COVID-19 infection [16], it was feared this risk might have been further accentuated in ACHD, especially when associated with diverse comorbidities such as pulmonary hypertension, heart failure, or in the presence of complex congenital heart disease [2, 13]. However, ACHD patients still belong to relatively younger population and, hence, do not belong to the increased risk group according to age. Limited data showed that general risk factors, primarily age, obesity and multiple comorbidities, are associated with an increased risk of complicated COVID-19 course also in this population. Indeed, studies to date showed that type of ACHD, including the most complex defects, was not associated with worse clinical outcomes related to COVID-19 [17–20]. On the other hand, ACHD patients at advanced physiological stage seem to be at the highest risk for a moderate/severe course of COVID-19 disease; particularly high risks are cyanotic lesions and patients with pulmonary hypertension [19, 20].

During the COVID-19 pandemic, the goal is to protect ACHD patients from COVID-19 exposure. In the healthcare systems, this has been successfully accomplished by rapid conversion of outpatient clinics to digital clinics. Tele- or video-clinics are now standard, functional and have been well received by ACHD patients globally. They may substitute rather than replace conventional face-to-face clinics in a revised, hybrid model of care beyond the COVID-19 pandemic [21].

21.5 Nursing Role During the COVID-19 Pandemic

As during the time without pandemic, the role of the specialist nurse during pandemic is multifactorial and includes organizing, coordinating and facilitating care for ACHD patients [22]; it can broadly be divided into two main segments. The first being care and monitoring of all ACHD patients and the second segment is specificity of care and monitoring for ACHD patients afflicted with COVID-19 disease (Fig. 21.1). The latter includes the assessment of severity of patient condition and the need for hospitalization during and after the disease.

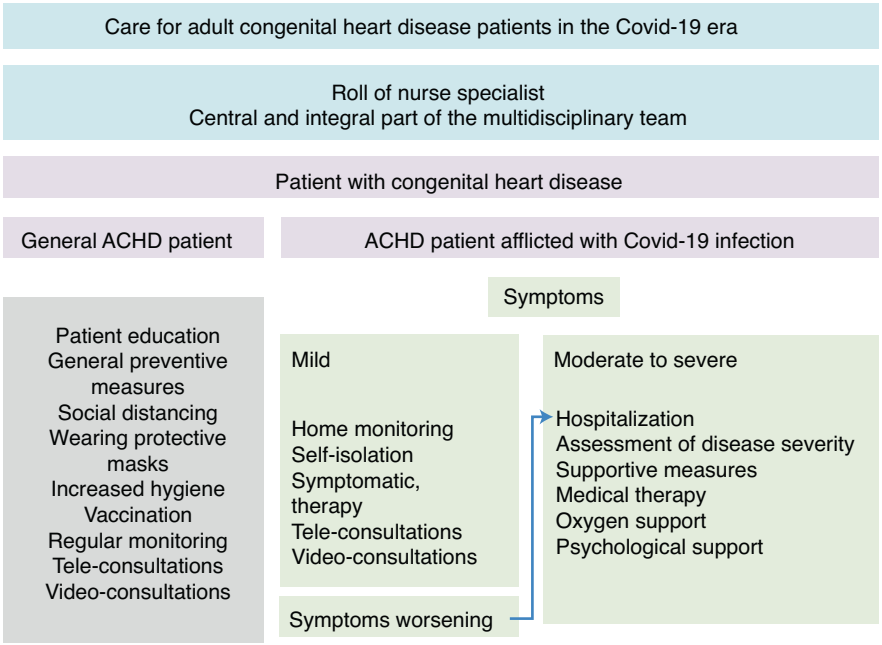

Fig. 21.1 Main illustration. Proposed algorithm in the time of COVID-19 pandemic for nurses in the care of adult patients with congenital heart disease

In order to ensure optimal care and treatment of ACHD patients with COVID-19 disease, nurses must be educated and familiar with the general guidelines in the treatment and care of ACHD patients [22, 23] as well as educated on general issues pertaining to COVID-19 infection [15] so that together with other members of the multidisciplinary team, they can provide optimal patient care.

Regarding all ACHD patients during pandemic, tele- or video-consultations and clinics have proven to be very good in providing outpatient care. The nurse specialist should provide advice regarding importance of social distancing, benefits of wearing protective masks and increased self-hygiene, importance of vaccination, self-monitoring and importance of healthy lifestyle including regular exercise and healthy diet, maintenance of regular doctor appointments as indicated, either via digital means or in-person, as per needed. Furthermore, nurse specialist can help decrease anxiety and provide reassurance for the patient in the time of need. A holistic and individual approach to each patient is needed [24].

In the nurses' work with patients, the following should be highlighted:

1. *Patient education and empowerment.* Patients should understand the basis of their congenital heart disease and the importance of maintaining healthy lifestyle choices, especially during the pandemic time to avoid unnecessary risk related to

both their underlying congenital heart disease and COVID-19 infection [25]. This includes adherence to the health advices related to regular medical therapy [22]. In line with professional advice, indications and dosages of drugs and knowledge such as in chronic heart failure patients, when to self-manage diuretic therapy and regulate fluid intake, should be explained. Patient should be educated regarding healthy diet with limited salt intake, reduced sugar and trans-fatty acid intake, as well as the advice regarding physical activity – encouraging the achievement of a healthy BMI (<25 kg/m^2) and engaging in activity adequate to the patient's functional capacity [22]. The importance of regular preventive, diagnostic, and therapeutic examinations should be emphasized. ACHD patients have an increased risk of infective endocarditis [23], hence, the importance of dental and skin health care, with the education regarding the need for antibiotic prophylaxis in certain ACHD cases, especially if dental procedures are indicated. The nurse specialist should strongly recommend appropriate regular vaccination against COVID-19, influenza, and pneumococcus, especially in older patients and patients with pulmonary hypertension. Finally, it is paramount to reassure the patient to seek medical help at the time of need. This can be via online consultation or in-person, and in the latter, the patient should be aware that the health system has taken all measures of precaution to reduce the risk of COVID-19 infection spread. Consequently, to all above-mentioned information, patients should take responsibility for their lives.

2. *Recommendations on how to reduce the risk of SARS-Cov-2 infection.* Preventive measures are crucial. This includes compliance with measures adopted by the government and public health institutions such as recommendations on wearing protective masks, self-isolation and adopting social distancing (maintaining physical distance >1.5 m), avoidance of unnecessary contacts outside the family environment, regular hand hygiene during the day especially after physical contact (washing hands for at least 20 seconds with soap and using alcohol disinfectant), timely detection of symptoms of COVID-19 infection and timely testing to reach diagnosis [25]. The main general measure is regular vaccination against SARS-Cov-2 infection. Smoking cessation should be encouraged. Evidence suggests that several mechanisms increase the risk of respiratory tract infections in smokers. Smoking impairs the immune system; smokers in general have increased risk of infection that may contribute to a more severe clinical presentation and poorer outcome of COVID-19 disease [26].
3. *Counselling on self-care during the COVID-19 pandemic.* The nurse specialist should strongly recommend appropriate regular vaccination against COVID-19, influenza, and pneumococcus, especially in older patients with comorbidities. As in general population, vaccination is considered safe and beneficial in ACHD patients. Furthermore, it is important to assess patients' and their families' awareness regarding signs of potential worsening of patient's underlying disease and to encourage them to timely seek for help. Nurse specialist should provide psychological and emotional support to patients in regards to fear of COVID-19 infection [27].

4. *Patients with Confirmed or Suspected COVID-19 Infection*

   For ACHD patients with confirmed or suspected COVID-19 infection, a rapid and concise triage is needed. Initial assessment should optimally include assessment of the degree of COVID-19 symptoms, followed by symptoms severity, and assessing for ACHD-specific high-risk features. For mildly symptomatic ACHD patients, usually with only one symptom, home management with appropriate telephone counselling within 48 h seems reasonable. Patients presenting with moderate symptoms, broadly referring to having two or more typical COVID-19 symptoms or severe symptoms, which include end-organ dysfunction, should be referred to hospital. After talking to the patient, the nurse learns the number of symptoms and severity of COVID-19 infection and based on these information and the degree of risk can refer the patient for outpatient nasopharyngeal swab testing or emergency service [28]. In case of moderate or severe symptoms, the nurse should refer the patient to hospitalization in which case, depending on the severity of the symptoms and the risk, they can be admitted to the general COVID-19 ward or to the COVID-19 intensive care unit. In these specific medical units, there is in general no specialized ACHD staff. Hence, appropriate and regular communication and consultation between doctors and nurses working in COVID units with ACHD cardiology team are highly advisable.

There is a recommendation to determine the degree of risk according to the complexity of the underlying congenital heart disease and its symptoms [29, 30]. In broad terms, low-risk patients could be considered patients with biventricular anatomy, normal function and heart size, without haemodynamically significant arrhythmias, without pulmonary arterial hypertension, without haemodynamically significant shunts and valvular lesions and with normal physical capacity. In contrast, moderate- and high-risk ACHD patients include patients with more complex congenital heart defect, chromosomal diseases, patients with univentricular anatomy, patients with complex or palliative heart anatomy (such as Fontan circulation), heart defects with cyanosis, with severe valvular stenosis or regurgitation, with severe ventricular dysfunction and with pulmonary arterial hypertension [11]. As a part of clinical assessment, it is important to take into account whether the patient is stable – compensated or unstable – decompensated; any change in the patient's condition should be timely addressed in order to prevent possible complications. In hospitalized patients, clinical symptoms, vital signs (blood pressure, pulse, respirations, temperature, SpO_2), fluid intake, diuresis, physical capacity and laboratory findings (inflammatory parameters – CRP, SE, Lkc, procalcitonin, coagulation, NT-pro-BNP) should be carefully monitored. In severe forms of COVID-19 infection, special attention should be paid to ECG recordings (the issue of timely recognition of arrhythmias that could rapidly lead to clinical worsening), basic blood oxygen saturation, the need for assisted ventilation and the occurrence of acute respiratory distress syndrome (ARDS) [31].

The nursing role in the patient care process is also to provide psychological and emotional support to the patient with regard to fear of isolation and separation from family and the fear of disease outcome. Nurses together with physicians should provide information about the patient's condition to family members, since visits during the pandemic are not allowed. Upon discharge from hospital, each ACHD patient should be scheduled for a follow-up visit at the appropriate ACHD centre to evaluate his condition after COVID-19 infection and continue with the most optimal treatment for his underlying congenital heart disease and eliminate possible post-COVID-19 consequences.

21.6 Conclusion

The COVID-19 pandemic has led to a change worldwide in the organization of patient care and, thus, also in the care of ACHD patients. Nurses, as an integral part of the multidisciplinary team, play a central role in the patient education and empowerment, counselling regarding preventive measures, necessary examinations and procedures during the pandemic, as well as provide an indispensable care and support to patients afflicted with COVID-19 disease. In the challenging time of COVID-19 pandemic, the multidisciplinary team should aim to ensure and provide the best possible care for the ACHD patients according to the contemporary recommendations, both in preventive measures and in treatment, in order to reduce the complications and consequences of COVID-19 diseases.

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