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Review Article

Acyanotic Congenital Heart Disease in Adolescents and Adults

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Abstract

Congenital heart disease affects 1% of live births. These diseases are are divides in two groups : acyanotic and cyanotic and the those of the first group more frequent. Nowadays we have more facilities in diagnostic methods, surgical techniques, interventional cardiology, and perioperative care have led to a substantial increase in survival of patients with congenital heart disease (CHD), resulting in a growing population of adults with congenital heart disease (ACHD). Currently, more than 90% of children born with CHD reach adulthood, creating new clinical challenges related to long-term complications, reinterventions, arrhythmias, heart failure, pulmonary hypertension, and sudden cardiac death. This review aims to describe the most prevalent forms of acyanotic congenital heart disease in adult patients and describe their clinical characteristics, management strategies, complications, and long-term outcomes. The manuscript addresses major acyanotic defects, including bicuspid aortic valve,aoortic stenoosis, atrial and ventricular septal defects, atrioventricular septal defect., coarctation of the aorta. Particular attention is given to the pathophysiology of late complications, indications for surgical or transcatheter interventions, and the role of advanced imaging and lifelong follow-up in specialized centers. The increasing complexity of care in this population underscores the need for multidisciplinary management and structured surveillance programs to reduce morbidity and mortality and to improve quality of life in adults living with congenital heart disease.

Keywords

Acyanotic congenital Heart Disease, Adult Congenital Heart Disease, Long-Term Outcomes, Cardiac Surgery.

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Introduction

The congenital heart diseases (CHD) in adults are more and more prevalent with the advances in diagnostic methods, medical treatment and interventional techniques, surgeries and the perioperative care. Nowadays, more than 90% of children with repaired CHD survive into adulthood grown-up congenital heart disease (GUCH) patients. The majority of them survive into adulthood, leading to a significant shift in the demographic profile of patients with CHD. This improved survival has highlighted the importance of long-term management in grown-up congenital heart disease (GUCH) patients. As these individuals get older, they often face complications that differ from those seen in childhood, necessitating ongoing multidisciplinary care and a lifelong specialised follow-up [1].

Arrhythmias are the most common cause of unscheduled hospital visits for grown-up CHD (GUCH) patients, accounting for one third of emergency admissions. They often requiring advanced electrophysiological management and individualized therapy.

Some GUCH patients are also at increased risk for sudden cardiac death. Decompensated heart failure is the other cause of death.

Conventional medical heart-failure therapy for left ventricular dysfunction is not effective in GUCH because of right ventricular failure.

Careful haemodynamic assessment and structural interventions are the first step to consider in GUCH patients presenting with heart failure symptoms or pulmonary artery hypertension. Adults with moderate or complex CHD need regular follow-up in specialized team [2].

In a Brazilian Institution with a large experience in surgery of congenital heart disease the great majority of them had simple CHD. Among the 1168 adults registered at the hospital the more frequent defects were: atrial septal defect (ASD), ventricular septal defect (VSD), Aortic stenosis (Ao S with bicuspid aortic valve), coarctation of aorta (Co Ao), atrioventricular septal defect (AVSD) [2].

Bicuspid Aortic Valve(Bav) And Aortic Stenosis

The congenital bicuspid valve is the most common congenital malformation (2% of the population) and is the most common cause of isolated aortic stenosis with a male to female ratio 3:1. It can be functionally normally with no significant pressure gradient across the valve and no important regurgitation. However thickening and calcification can be detected in adults. Patients with bicuspid aortic valve should be monitorated for progressive valve dysfunction. The most common symptoms in cases of severe aortic stenosis are exercice intolerance, dyspnea on exercice, syncope, angina. In the examination is detected low amplitude parvus and tardus carotid pulse, systolic ejection murmur (cessation of murmur before A^2), and ejection clic [3,4].

There is no significant stenosis when the gradient less than 25 mm Hg, but the patient should be followed every two years, because later-on, frequently it will progress to fibrocalcic stenosis. When the gradient is more than 50 mm HG, can occur arrhythmias, angina, sudden deaths, syncope, endocarditis [3].

In adults patients mre than 30 years of age, severe aortic stenosis is defined as: peak velocity i ≥ 4m/s, mean gradient more than 40 mmHg,valve área less than 1.0 cm^2; they need Doppler echocardiography every year [3].

Dilatation of ascending aorta or aortic root is commonly seen in association with bicuspid valve with or without significant stenosis.

First degree relatives of patient with bicuspid aórtic valve should be evaluated for BAV and thoracic aortic disease [3].

On electrocardiogram: the symptomatic patients are detected ST or T wave abnormality in left precordials leads.

On transthoracic echocardiography should evaluate the size of aortic root and ascending aorta measuring the maximum diameter perpendicular to aortic flow and aortic annulus at the hinge point of the aortic valve leaflets using the inner edge to inner ledge. If the aortic root and ascending aorta above sinotubular junction If cannot be adequately visualized magnetic resonance imaging(MRI) or contrast cardiac tomography CT
is recommended [4].

Treatment: is percutaneous aortic valvuloplasty is recommended in selected population with significant stenosis defined as a peak gradient is ≥ 60mmHg or ≥ 50 in symptomatic patients in young adults without significant regurgitation or calcification [4].

In symptomatic adolescents or young adults, or in cases that the mean gradient is over 50mmHG, the medical therapy is limited. The percutaneous aortic valvuloplasty recommended in aortic stenosis most commonly in bicuspid commissural fusion without significant valve calcification or regurgitation. The surgical procedure can be done with valve replacement in adults with significant aortic disease. Ross procedure is pulmonary autograft aortic replacement is the surgical procedure of choice [3]. Surgical repair should be done or replace in ascending aorta when aortic root is more than 5,5 cm.

Patients with BAV should continue to undergo surveillance [4].

Mid term results of this procedure shows excellent results. In long follow-up the patient may develop neoaortic regurgitation and dilatation of neoaortic root.

Ventricular Septal Defect

VSD is the most common congenital heart disease in infants but many of them close spontaneously and accounts for only 10% in adults. It is known that spontaneous closure really takes place,particularly the perimembranous defect. Where septal aneurysmal tissue can partially occlude the defect.in approximately 48% of cases, within the first nineteen months of life. From that date, the closure rate falls drastically, reaching near zero at 7 years of age [5,6]. Functionally small (restrictive), which were imagined having benign evolution, may show some severe complications along adult life, especially infectious endocarditis (10%), aortic regurgitation (19%) and need for surgery (12%) [6]. False security might exist when subpulmonic defect, seems to be getting smaller, the aortic cusp can prolapse and partially or completely occlude the defect. In these cases operation is necessary to protect the integrity of aortic valve coaptation [3].

Indications for closure of VSD in adults are:

1. Pulmonary-to-systemic flow ratio(Qp/Qs) ≥ 2 and clinical evidence of left ventricular volume overload.
2. History of infective endocarditis.
3. Intervention may be considered for mild shunts (Qp/Qs ≥1.5) if there is evidence of left ventricular systolic or diastolic dysfunction.
4. Ventricular septal defect should be considered for closure when pulmonic vascular resistence (PVR) is less than 3 WU and the shunts significant, as long as no Eisenmenger

physiology or desaturation on exercise [5].

To the evaluation of benefit the VSD closure is important clinical, echocardiographic and invasive hemodynamic assessment to determine the most appropriate timing of surgery or percutaneous repair surgery or percutaneous.

Surgical repair was done for several years with low mortality rate. Amaral et al. related 73 cases of VSD among 468 cases of interventions in congenital heart diseases in adults (15%) with very low mortality [2].

Elafifi et al. conducted a prospective analysis from 151 patients who underwent percutaneous closure of muscular and perimembranous VSD using a Lifetech Konar-multifunctional occluder, procedure anterograde and retrograde approach with very good result [7].

In the series of cases Chamie et al. is described interventricular VSD surgical closure using Amplatzer occluder (Musc or Member occluder) and no complications in the cases of muscular type of VSD and few compliocations in perimembranous type [8].

Retrograde transcatheter VSD occlusion with double disk devices proved to be effective in different types of VSD. The authors suggest this technique as an alternative to percutaneous occlusion of all types of VSD [9,10].

Complications of transcatheter closure: there are risks of complications that include atrioventricular block, other early or late arrhythmias, post-pericardiotomy, pulmonary, mediastinal infections, and even death, not mentioning maintenance of residual VSD, which is much more frequent in our milieu [7]. In the study of Yan Yang from 105 cases utilizing eccentric occlusion of transthoracic subarterial VSD 13 cases (16%) had as complication aortic regurgitation (coronary cuspid prolapse) [9].

Surgical closure by direct suture or with patch was done for many years with low perioperative mortality and high closure rate. Patch leaks might occur. Another complication is late sinus node disease [10].

Atrial Septal Defect (Asd)

After bicuspid aortic valve ASD are the most common congenital heart disease found in adults.

Small ASD close spontaneously in infants but is unusual in older children and in adults. Often asymptomatic until adulthood might occur complications including atrial arrhythmias, right ventricular failure, paradoxical embolization leading to cerebrovascular accident or transient ischemic attacks, cerebral abscess, pulmonary hypertension that can become irreversible and lead to right-to left shunt [11,12].

Atrial septal defects are classified into secundum, primum, sinus venosus, and coronary sinus defects. Secundum ASD, the most common type of all ASDs (80%), is located within the fossa ovalis. Small secundum ASD needs to be distinguished from patent foramen ovale as the latter is not a true deficiency of atrial septal tissue, but rather a tunnel-like communication between the septum primum and septum secundum located in the anterosuperior portion of the atrial septum [13].

Amaral related a series of cases of adult CHD and among 468 cases submitted to surgical treatment and 37% were ASD [2].

Atrial septal defect results in a left-to-right shunt and the magnitude of blood flow depends on atrial pressures related to the compliance of the both ventricles. Most small defects (<10mm) are associated with a small shunt and minimum or no enlargement of the right heart structures. Larger, long-standing shunts result in right atrial and right ventricular dilatation, myocardial cellular stretch. Increased pulmonary blood flow may trigger a pathological mechanism due to shear stress causing pulmonary endothelial cell activation and activation of growth factors, vasoconstrictors and smooth muscle hypertrophy, thus contributing to the development of pulmonary arterial hypertension (PAH). Rise in pulmonary artery pressure (PAP) with and the development of pulmonary vascular disease.

The symptoms of ASD are: abnormal intolerance to exercice, fatigue, dyspnea or others complications as mentioned above. The physician suspects on basis if cardiac murmur or abnormal EKG (tall P wave indicative of right atrial enlargement, incomplete right bundle branch block pattern, and right axis deviation. The rhythm is typically sinus, but in adult patients can be atrial flutter or fibrillation. Right ventricular hypertrophy is evident in patients with pulmonary hypertension. Left axis deviation with a superior axis is suggestive of ostium primum defect [13].

Transthoracic echocardiogram is the Imaging of choice for the diagnosis and the trans-esophageal may aid sizing of defects, definitive for diagnosis of ostium secundum or ostium primum defect, or sinus venous defect of superior vena cava or inferior vena cava. Cardiac magnetic resonance (CMR) or cardiac tomography (cardiac CT) can be used for localization, estimate shunt flow and detect anomalous pulmonary venous connection. A 3D reconstruction requires data from CMR or cardiac CT; the 3D model built, for instance, the sinus venosus defect and anomalous pulmonary veins, may be printed allowing for direct inspection or viewed virtually and analysed with dedicated software [13].

ASD closure It is necessary in the symptomatic patients. Percutaneous device closure is in alternative for surgical closures in patients with ostium secundum with Amplatzer septal occluder. Atrial septal defect closure has been shown to be safe and associated with a decrease in PAP and improvement of symptoms when pulmonary vascular resistance (PVR) < 5 WU. The good results of occlusion procedures suggest that the use of devices may be considered the treatment of choice for ostium secundum atrial septal defects ≤ 6 mm [12].

The recent development of new material biodegradable occlusion

devices has attracted increasing interest with the potential of providing a temporary scaffold for tissue endothelialization following controllable degradation over time leaving only 'native'tissue behind [13].

A new and promising technique of catheter ASD closure pertains to sinus venosus defects.Robotic assisted ASD closure is a type of minimally invasive procedure with an endoscopy [1].

Chamie related 141 cases of transcatheter closure of large defects. From the most complex cases, 58 had stretched catheter, six had multiple ASD, 9 presented atrial septum aneurysm. Procedure was unsuccessful in 4 cases (5.47%) and in one, with double orifice. No major difficulties were posed in closing multiple defects or in aneurysm closure performed. However, large ASD with deficient or absent rims were most difficult to close. In those cases, the possibility of residual shunt should be considered. Although device occlusion is feasible, surgery should always be considered as an option [12].

The hemodynamic response after atrial defect.closure is right and left cardiac remodelling, including electrical remodelling immediately after closure and appear to continue for one year. Persistent right heart dilatation and residual functional tricuspid regurgitation are more prevalent in older patients with late ASD closure.

Coarctation of the Aorta

Aortic coarctation is a narrowing in the thoracic aorta. Most frequently it is located at the insertion of the ductus arteriosus just after the left subclavian artery (postductal or adult type). It comprises 6-8% of all congenital lesions.

The clinical manifestations it depends of the severity of narrowing. It may remain unnoticed throughout the childhood or, in contrast, progress to epistaxis, hypertension, heart failure and aortic dissection [15]. Unfortunately despite of adequate childhood surgery, patients are in risk for several concerning long-term complications include systemic hypertension, recoarctation aortic aneurysm and dissection. All patients with coarctation of aorta should be monitored with lifelong follow-up. The Mayo Clinic series describing long-term outcome included more than 800 patients the survival rates fere 93% in 10 years and 74% in 30 years. The most common cause of death was coronary artery disease [16].

The image methods are the key for diagnoses and the severity of defect. The first method is echocardiogram with doppler. The MRI is more accurate and can demonstrate other complications as aorta aneurysm and recoarctation.

The treatment is usually transcatheter stent therapy to relieve aortic obstruction [17,18].

Recoarctation refers to restenosis after an initially successful intervention. The patient can be asymptomatic or develop severe hypertension. The rate of recoarctation is 5-14% after surgery [3]. It is seen primarily usually due to inadequate aortic wall growth at the site of repair before the aorta has reached adult size. Following balloon angioplasty in children are in great risk for recoarctation compared with the adults.

Systemic hypertension is one of the major long term problems following repair of coarctation. Although typically falls after successful repair, persistent or recurrent hypertension and disproportionate systolic hypertension with exercise. Patients who had delayed initial repair often have residual hypertension despite surgical or transcatheter intervention3. The adult with hypertension should be controlled by beta-blockers, angiotensin-converting-enzyme (ACE) inhibitors or angiotensin-receptor blockers.

Aortic aneurysm may develop at site of prior coarctation following surgery(especially after patch angioplasty), balloon dilatation or stent implantation of native coarctation [17].

For the majority of patients aneurysm repair requires surgical intervention with resection of aneurysm and graft placement. Alternativelly endovascular stent graft have been used [3].

Atrioventricular Septal Defect (Avsd)

AVSD are anatomic defects that arise from faulty development of embryonic endocardial cushions. This spectrum ranges from a primum atrial septal defect. And cleft mitral valve, partial AVSD), to defects of both the primum atrial defect and inlet ventricular septum and the presence of a common atrioventricular valve (complete AVSD) [3].

Surgical correction for a complete AVDS is generally perfprmed because of the significant morbidity and mortality. The management approach is based on the type of AVSD determining whether thereare other associated anatomic and hemodynamic abnormalities (as Patent ductus arteriosus-PDA). Also it is importante find out the size of ventricle,AV valve regurgitation, right and left outflow obstructin. Patients with significant left AV valve regurgitationare at risk of atrial fibrillation.

Patients with partial defect. are generally assymotoimatic during childhood but can develop symptoms of right heart overload.

The mortality rate for surgical repairis about 3% and 10 years survive is around 90% [19]. Before yhe surgical management of complete AVSD is important to treat the heart failure, and an incomplete repair or palliative intervention [3].

There is a significant morbidity and mortalty associated with uncorrected lesion. An autopsy results in young individuals with congenital heart disease in Scandinavia showed that structural malformations, including ventricular hypertrophy and dilatation,as well as fibrosis and acute inflation or infaction , were recorded [19,20].

Pulmonary Hypertension In Adults With Congenital Heart Disease

Pulmonary hypertension is defined as a pulmonary artery pressure is ≥ 25 mm Hg at rest in the presence of normal capillary edge pressure (≤ 15 mm Hg). It'is common in patients with congenital heart disease with left to right intracardiac shunts. When they are large and non restrictive due to an increased pulmonary blood volume or pressure overload [13].

The most severe stage of pulmonary arterial hypertension related to shunt is Eisenmenger syndrome. Significants right to left shunts unrepaired or those repaired a later age may develop pulmonary hypertension: Septal defects (ASD, VSD, atrial ventricular septal defects, patent ductus arteriosus), Single ventricle complexes (double outlet right ventricle, double inlet left ventricle), transposition of great arteries, truncus arteriosus.

Eisenmenger syndrome: patients with left to right shunts leading to severe pulmonary arterial hypertension, later may have reversal shunt and chronic cyanosis, secondary erythrocytosis and multiple organ involvement. Patients with small or coincidental defects who develop pulmonary arterial hypertension that cannot be attributed to the congenital defect.

Conclusion

We tried to describe the main aspects of the most common congenital heart diseases in growing-up patients and it is evident that during the last decades the prognosis for congenital heart disease has gone through an enormous development due to improvement in diagnostic methods, and treatment. Despite this, further developments are needed to decrease the morbidity and mortality,still very high in many of these congenital heart diseases.

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