Module 4. Adult congenital heart disease and sports

BARÇA INNOVATION HUB
Universitas

The aim of this chapter is to summarize the main congenital heart diseases that can be found in athletes. Our purpose is to give a general recommendation and guideline on the approach to follow regarding cardiological eligibility for sports practice.

Adult congenital heart disease and sports

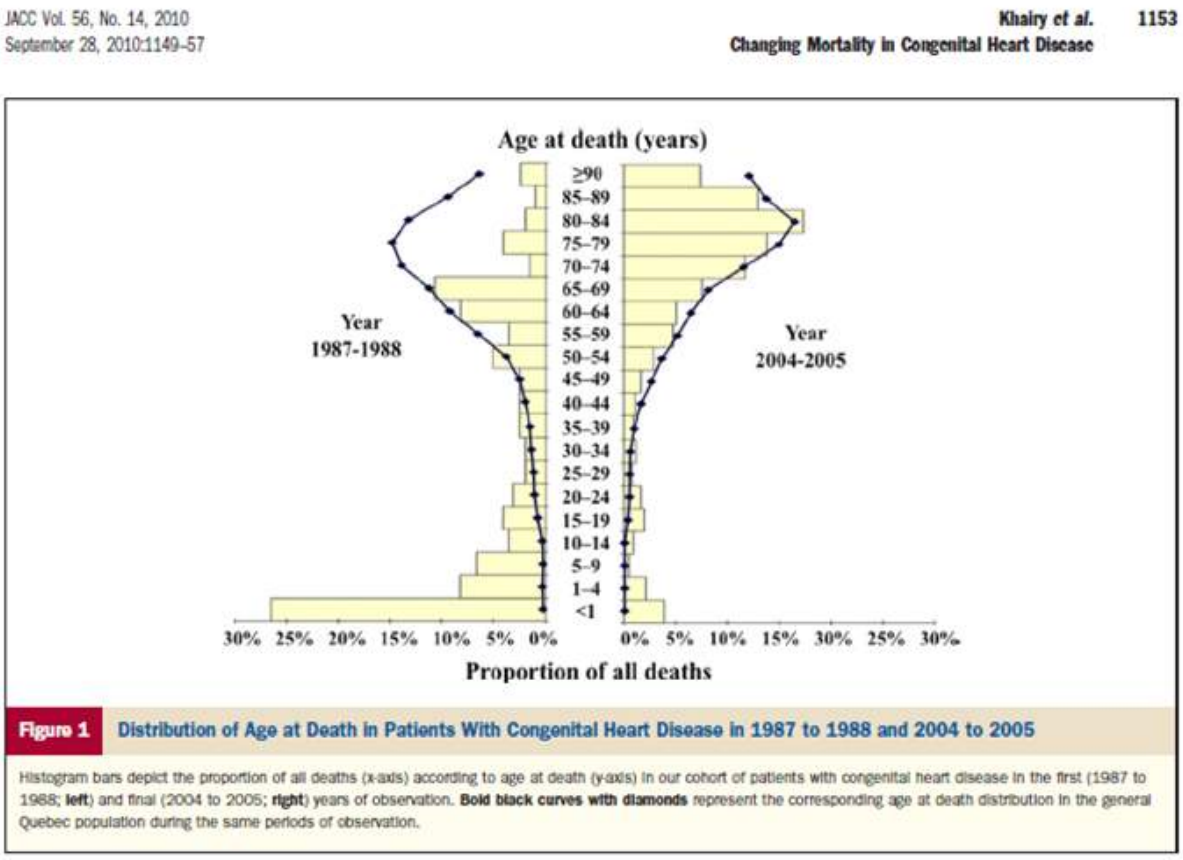
Congenital heart disease

Congenital heart disease (CHD) is the most common malformation at birth: 8-10 out of 1,000 children are born with a malformation of the heart or great arteries, and at least 6 out of 1,000 have moderate or severe heart disease. Until recently, only 50% of these children reached adulthood. According to the British Cardiac Society Working Party, 85% of children operated on for congenital heart disease reach adulthood, thanks to the development of cardiac surgery and percutaneous techniques.

Currently, it is estimated that the probability of survival to adulthood for mild CHD is 98%; 90% for moderately complex heart disease, and 56% for complex heart disease (Khairy et al., 2010). A prevalence of 11% of the population is estimated for 2030. The incidence of congenital heart disease in Spain is 13.6%. The most frequent were atrial septal defect (6.31%), ventricular septal defect (3.48%), patent ductus arteriosus (2.71%), coarctation of the aorta (0.55%), pulmonary stenosis (0.50%), transposition of the great arteries (0.49%),

atrioventricular canal (0.45%) and tetralogy of Fallot (0.41%) (Pérez-Lescure Picarzo et al., 2018).

Figure 1. Distribution by age at death of patients with CHD in 1987-88 and 2004-2005



Source: Khairy et al., 2010, https://goo.su/f5R1vrH.

Transmission. Risk factors/prevention

Most congenital heart diseases are sporadic, and some are hereditary. Hereditary transmission is low (atrial septal defect [ASD]: 1.5-6%, ventricular septal defect [VSD]: 2-10%, patent ductus arteriosus [PDA]:

2-4%, tetralogy of Fallot: 1.5-2.5%, transposition of the great arteries 2%, coarctation of the aorta 2-6.5%) (Baumgartner et al., 2021).

Overprotection in the form of physical activity restriction in children with CHD by parents, caregivers, and physicians leads to sedentary lifestyles, with increased likelihood of cardiovascular risk factors such as type2 diabetes, obesity and dyslipidemia, and increased risk of ischemic heart disease in adulthood. Promoting physical activity over sedentary behaviors, as well as providing appropriate sports prescriptions, should be a priority in every evaluation of these patients (Asociación Española de Pediatría de Atención Primaria, 2015).

Clinical manifestations

CHDs are a very heterogeneous group, so people present with a wide variety of symptoms and signs: Dyspnea, palpitations, loss of consciousness, chest pain, arterial hypertension, edema, diarrhea, and cyanosis. The vast majority of people are asymptomatic.

Diagnosis

Adult congenital heart disease is a very heterogeneous group and is classified as simple, moderate, and complex. Among the simple ones, we find atrial septal defect, ventricular septal defect, patent ductus arteriosus, anomalous venous drainage and congenital aortic, mitral and pulmonary valvulopathy. Moderate congenital heart diseases

include aortic and pulmonary supravalvular or subvalvular valvulopaties, tetralogy of Fallot, Ebstein's anomaly, sinus venosus or coronary sinus atrial septal defects, partial or complete atrioventricular canal defects, aortic coarctations, coronary anomalies, and coronary fistulas. The most complex are complete or congenitally corrected transposition of great arteries, single ventricles, double outlet right ventricles, pulmonary atresias, and Eisenmenguer syndromes.

In the assessment of patients with congenital heart disease, clinical examination, physical examination, ECG, and echocardiogram are essential (Asociación Española de Pediatría de Atención Primaria, 2015).

With the ECG, we will assess rhythm, PR duration, axis, QRS duration, QRS fragmentation, QT dispersion, and repolarization. Additionally, we will rule out ischemia, blocks, delta wave, Brugada syndrome, and epsilon wave.

In some patients, the study is expanded by performing additional diagnostic tests, such as the following.

Holter monitor: To record the heart rhythm for 24 hours.

Stress test: To assess functional capacity, ischemia, risk of arrhythmias, and blood pressure behavior. Currently, with associated

oxygen consumption.

Cardiac MRI: To assesses the same as an echocardiogram and a CT. It is the gold standard technique for assessing right ventricular volumes and function and ventricular fibrosis, without irradiation. It should be avoided in patients with claustrophobia.

CT: The gold standard technique to assess coronary malformations, pulmonary veins, the aorta and arteriovenous fistulas, collaterals, and extracardiac conduits.

Transesophageal echocardiogram: Performed under sedation. A probe is inserted through the mouth to visualize the heart from the esophagus. It is very useful for planning surgery or hemodynamics.

Stress echocardiography: The patient runs on a treadmill or rides a stationary bike. It produces images of the heart after exercise.

3D echocardiogram: It allows us to obtain an excellent assessment of left ventricular and mitral or systemic tricuspid valve volumes and function (Gallego and Montserrat, 2021). In addition, myocardial strain techniques help us to detect ventricular dysfunction early (Bijnens et al., 2021).

Cardiac catheterization: It requires puncture and catheters and is used to assess pressures and confirm the anatomy.

Electrophysiological study: It also requires puncture and catheters. It allows us to diagnose types of arrhythmias and, if necessary, treat them by ablation.

Treatment

Treatment depends on the type of injury, its severity, and the rest of the patient's characteristics. The goal is to prevent or control the symptoms and many of the complications of the disease. Pharmacological treatment may be necessary. Most patients in this advanced situation are treated surgically by repairing the defects or correcting cardiac function. In general, we always prefer to perform interventions that repair the patient's own structures; only in those cases where this is not possible, prostheses are implanted. Occasionally, some of these interventions can be performed in a minimally invasive, even robotic, manner.

Moreover, there has also been great progress in the percutaneous treatment of congenital heart diseases, which allows many defects to be corrected in a less invasive manner. Some examples of these techniques are closure of atrial septal defects (ASD), ventricular septal defect (VSD) or patent ductus arteriosus (PDA), and stent implantation in coarctation of the aorta.

In general, congenital heart disease requires close follow-up by the cardiologist throughout the patient's life, as lesions or dysfunctions

may remain and may evolve over time. New lesions or complications may also appear, and the corrections made previously may require reinterventions in the future.

Classification

Congenital heart diseases, as we said, are a very heterogeneous group, so we will focus on the most frequent ones.

- Acyanotic (left-rightshunt): ASD and VSD, less frequent, DAP, and anomalous venous drainage (AVD).
- Cyanotic: Tetralogy of Fallot (TF), transposition of great arteries (TGA) and single ventricles.
- Without shunt or cyanosis: Coarctation of the aorta, congenitally corrected TGA (ccTGA), and Ebstein's disease.

A) Acyanotic: Left-right shunt

The main congenital heart diseases with left-right shunt are atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), and anomalous venous drainage (AVD).

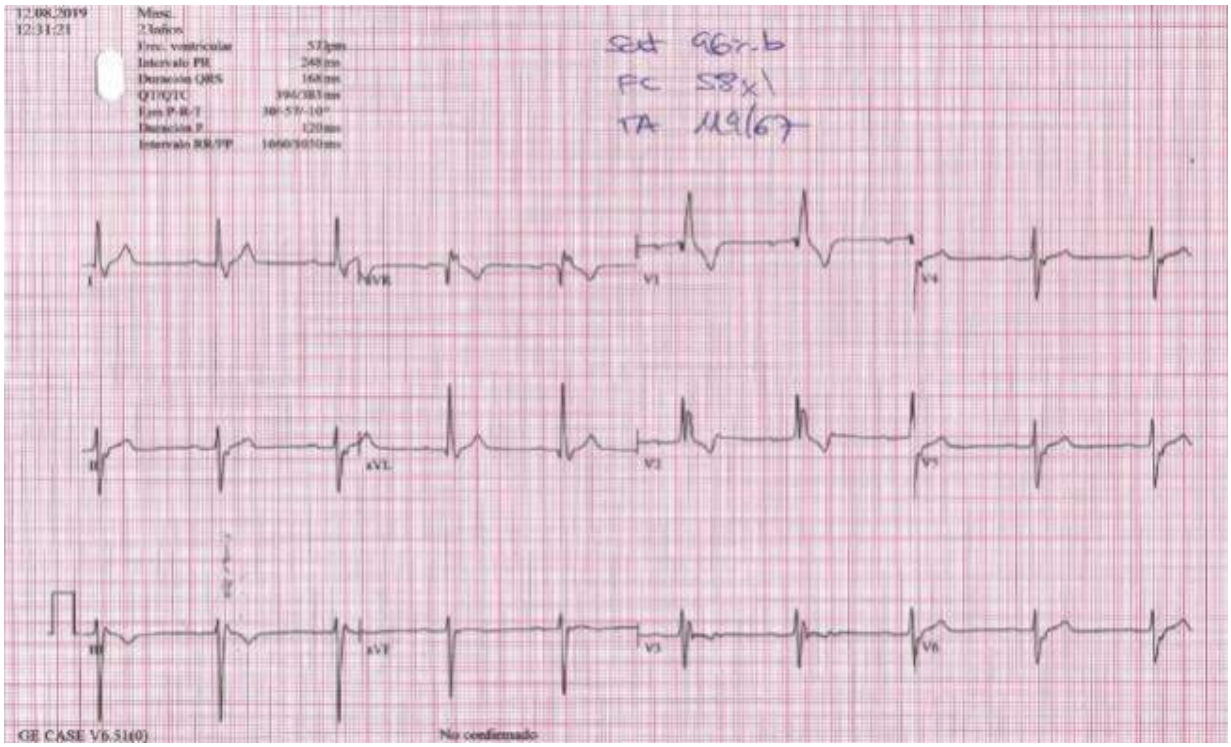
1 **Atrial septal defect**

It is characterized by a defect (hole) at the level of the interatrial septum. Most people are asymptomatic until the age of 30-40years. In the long term, some

present atrial arrhythmias (atrial flutter or atrial fibrillation), stroke, right heart failure, or pulmonary hypertension.

On a physical examination, patients show a fixed splitting of the second heart sound. The typical ECG of ostium secundum (OS) ASD is right bundle branch block (RBBB). The typical ECG of ostium primum (OP) ASD associated with mitral cleft is anterior hemiblock (AHB) + RBBB (Figure 2).

Figure 2. ECG with anterior hemiblock and right bundle branch block



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The diagnosis of ASD is echocardiographic. There are 4 types of ASD, as shown in Figure 3: Ostium secundum ASD (OS ASD), which is the most frequent (80%); ostium primum ASD (OP ASD); sinus-venous ASD, and sinus-coronary ASD at the level of the superior vena cava, more frequent than that of the inferior vena cava.

Figure 3. Transthoracic echocardiogram (first three images) and transesophageal echocardiogram (last image)

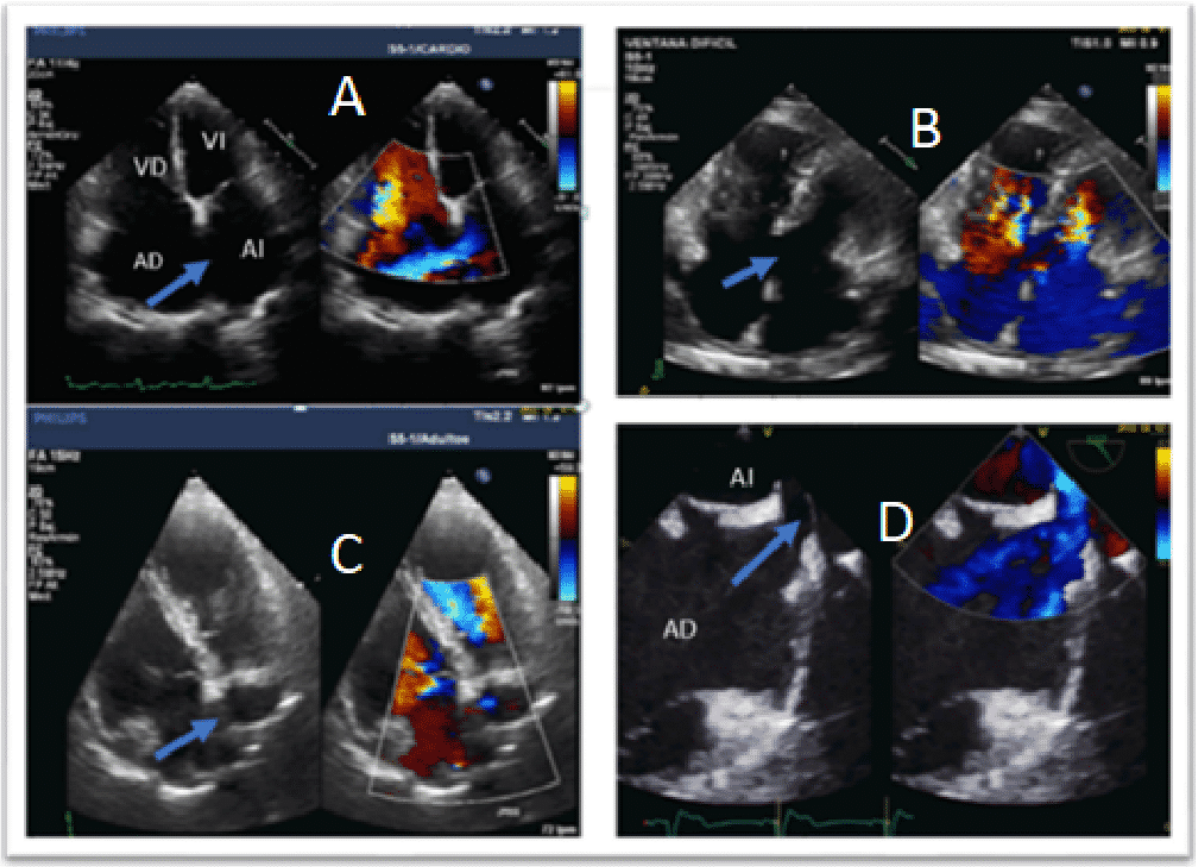

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Figure references

AD: Spanish acronym for right atrium (RA); VD: Spanish acronym for right ventricle (RV); LA: Spanish acronym for left atrium (LA); LV: Spanish acronym for left ventricle (LV). Figure A: OS ASD at the level of the fossa ovalis; Figure B: OP ASD at the level of the crux cordix; Figure C: Coronary sinus ASD at this level; Figure D: Sinus venosus ASD at the level of the superior vena cava.

In these cases, a cardiac MRI is performed to assess the right ventricle, Qp/Qs, assessment of coronary sinus ASD, CT angiography, in case of suspicion of anomalous venous drainage and before the surgery to study the anatomy of the coronary arteries.

The transesophageal echocardiogram is used to assess whether percutaneous closure can be performed. Treatment will be percutaneous (Figure 4) or surgical (Figure 5), when there is right cavity overload or Qp/Qs >1.5.

Figure 4. Percutaneous closure. A: Interatrial septum with 3D transesophageal echocardiography (TEE) and image of atrial septal defect; B: 3D TEE with catheter passage through the ASD; C: 2D TEE with device implanted as an open umbrella that closes the ASD

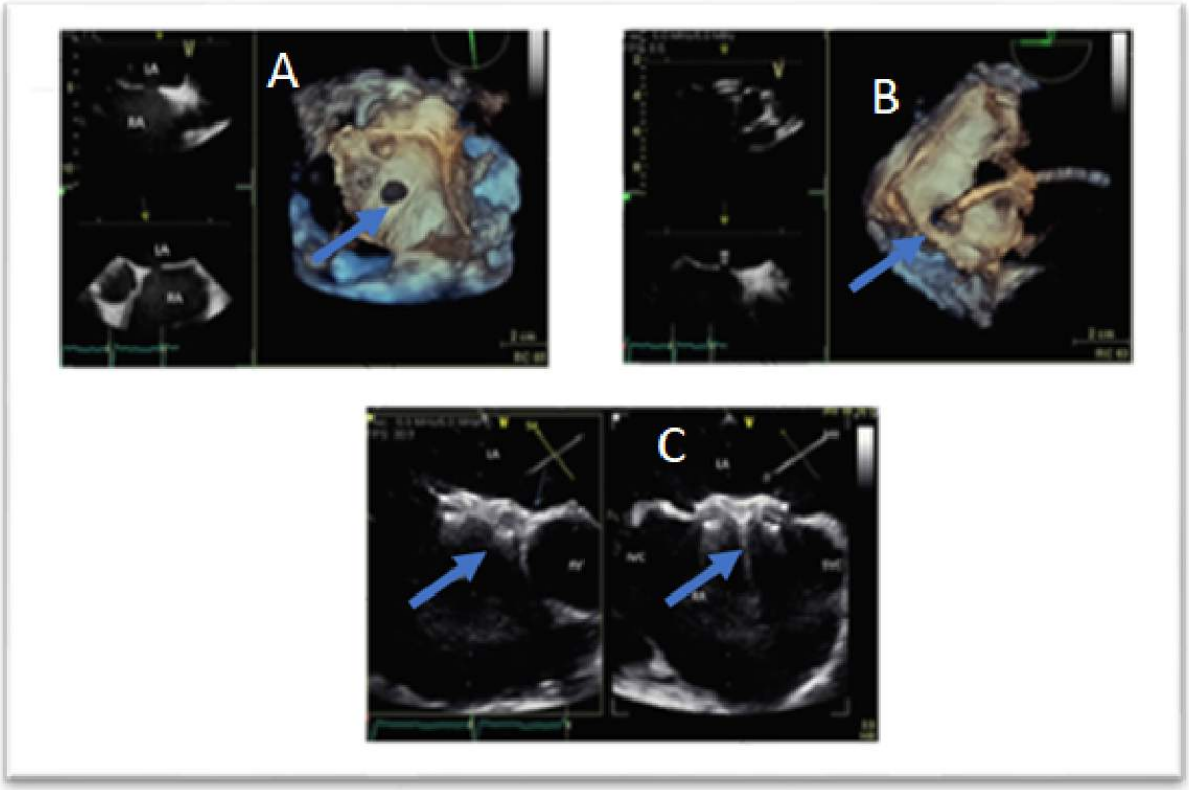

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Figure 5. Mini-invasive surgical closure or robotic surgery

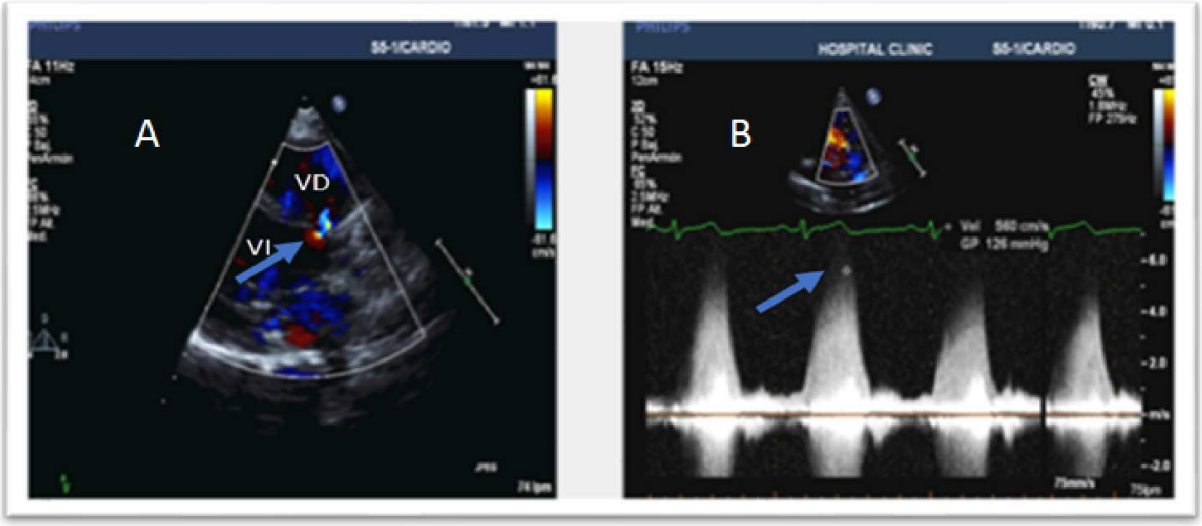
Source: Barnaclínic Barcelona, n. d., https://goo.su/TH2oN.

2 Ventricular septal defect (VSD)

It is the most frequent CHD at birth (30-40% of CHD), but the vast majority of defects close spontaneously. It is characterized by a defect (hole) at the level of the interventricular septum. The clinical presenation depends on the size and hemodynamic repercussion (Qp/Qs >1.5). Large VSDs cause left heart failure, whereas small VSDs are asymptomatic and present a very intense systolic murmur. In the case of restrictive (small) ones, in the echocardiography we detect a

high gradient (126 mm Hg), so the murmur is intense (the more murmur, the smaller and the more benign) (Figure 6).

Figure 6. Transthoracic echocardiogram. VD: Spanish acronym for right ventricle (RV); VI: Spanish acronym for left ventricle (LV) Figure A: Small restrictive VSD; Figure B: Elevated gradient of 126 mm Hg



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The diagnosis is also echocardiographic and the treatment of large VSDs is mainly surgery with patch closure or, in some cases, closure with a percutaneous device.

3 Patent ductus arteriosus (PDA)

Patent ductus arteriosus is the persistent patency of a fetal structure, usually located between the pulmonary artery (usually the left pulmonary artery) and the descending aorta, and accounts for 5-10% of CHD, excluding premature newborns. The clinical presenation depends on the size and hemodynamic repercussion (Qp/Qs >1.5). Large PDAs cause left heart failure, whereas small PDAs are asymptomatic and present a continuous machinery-type murmur. The diagnosis is also done by echocardiography and CT, and treatment may be percutaneous or surgical.

4 Anomalous venous drainage (AVD)

They can be total or partial, depending on whether all or some of the pulmonary veins drain to the right atrium and not to the left. Partial venous drainage can be of 1 vein, almost always asymptomatic and without hemodynamic repercussions, or of 2 or 3 veins, which usually cause overload of the right cavities and the same symptoms as ASD, occasionally associated with ASD.

By echocardiography, whenever we detect a more than slight dilation of the right ventricle, we must rule out anomalous venous drainage

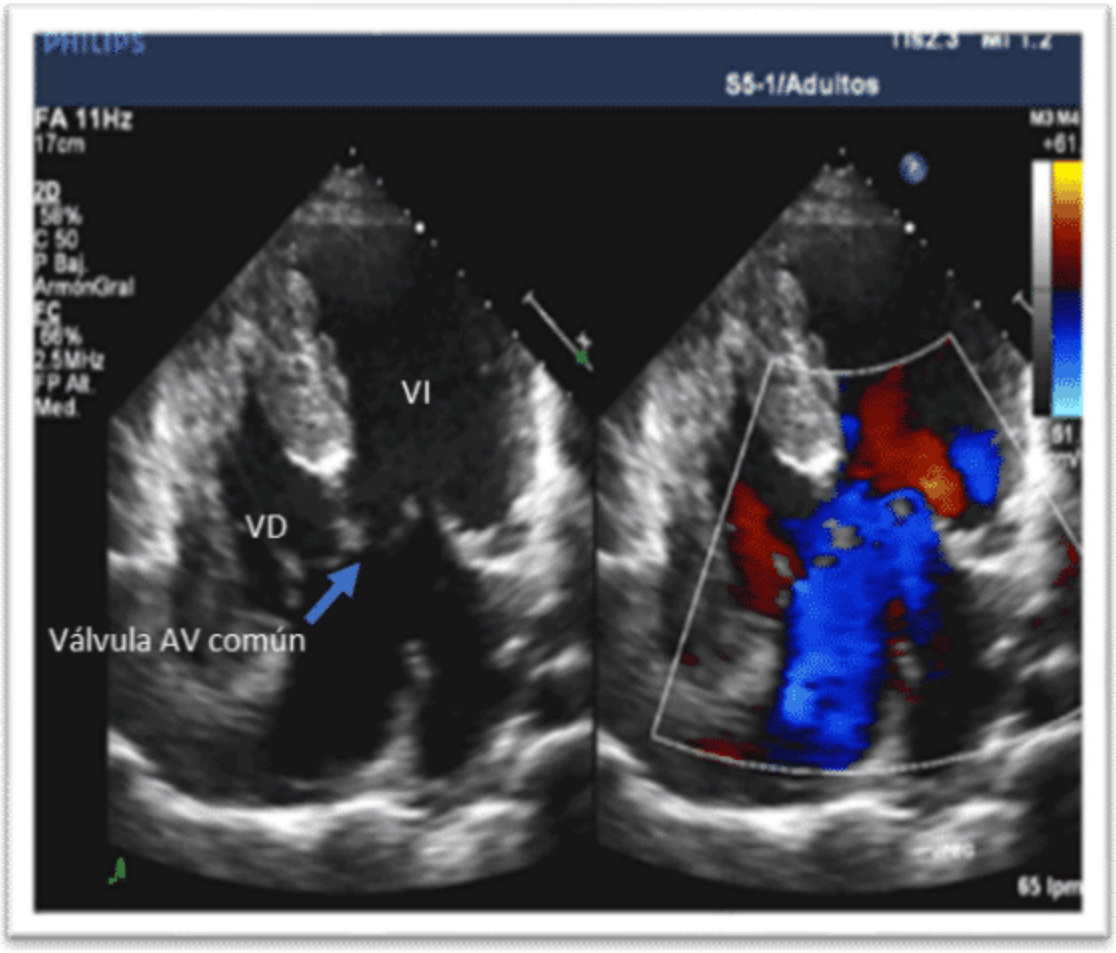
and arrhythmogenic right ventricular cardiomyopathy, although most of the time it is simply exercise adaptation.

The diagnosis is done by pulmonary vein CT angiography or cardiac MRI. Treatment is almost always surgical.

5 Atrioventricular (AV) canal

It occurs when, in the fetal stage, the fusion of the endocardial cushions to form the crux cordis fails. The atrioventricular (AV) canal can be totally cyanotic (Figure7) or partial (OP ASD with cleft or mitral valve cleft) non-cyanotic. Treatment is surgical. Once repaired, follow-up and complications are similar to those of ASD, except that, in this case, there is more risk of common AV block.

Figure7. Full AV canal. Wide ASD and VSD, with common AV valve



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B) Cyanotic

Cyanotic CHDs are tetralogy of Fallot, transposition of the great arteries, and single ventricle.

The transposition of the great arteries is always cyanotic. There is ventricle-arterial discordance, i.e., the atria and ventricles are normally related. The right atrium connects to the morphologically-

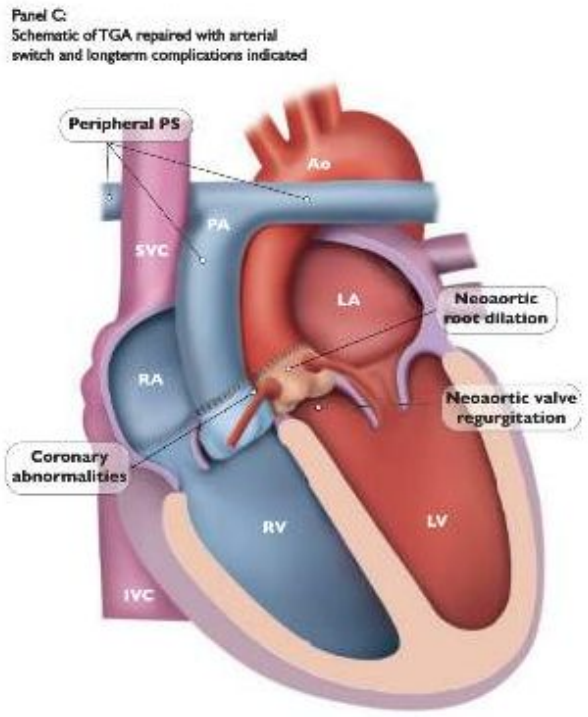
right ventricle and the left atrium connects to the morphologically-left ventricle, but the right ventricle connects to the aorta, and the left ventricle connects to the pulmonary artery. The systemic and pulmonary circuits are parallel (Figure 9), and the existence of a shunt (ASD or patent foramen ovale, VSD or PDA) that allows oxygenated blood to reach the systemic territory is mandatory for the survival of these patients.

Figure 8. Fetus with transposition of the great arteries. The aorta communicates with the anterior right ventricle, and the vessels arise and run in parallel

Source: 14th European Echocardiography course on congenital heart disease Barcelona 9-12 October 2019.

Surgical repair currently consists of arterial swich (Jatene surgery), vessel exchange, connecting the pulmonary artery to the right ventricle, the aorta to the left ventricle, and reimplantation of the coronary arteries (Figure 9).

Figure 9. Surgical repair



Source: Baumgartner et al., 2021, https://goo.su/bg2cdF.

The most frequent complications in adults are the following:

1. Suprapulmonary stenosis due to suture; it is usually not very symptomatic.

2 Dilation of the aortic root; it is asymptomatic until complications occur.

3 Stenosis or kinks at the origin of reimplanted coronary arteries, which can cause acute or chronic myocardial ischemia.

The **Tetralogy of Fallot** may be cyanotic or pink. It consists of a large VSD, overriding of the aorta (misalignment of the septum with the aorta), right ventricular outflow tract obstruction (RVOT and pulmonary), and right ventricular hypertrophy. Patients with more RVOT and pulmonary obstruction are more cyanotic. Surgical repair is performed in infancy and consists of VSD closure and RVOT enlargement (sometimes with patching and sometimes a conduit is required). At times, palliative surgeries such as Blalock-Taussig are performed (Figure10).

In 1946, Dr. Blalock (surgeon) and Dr. Taussig (cardiologist) gave life to the "blue babies" with tetralogy of Fallot.

Figure10. Dr. Blalock (surgeon) and Dr. Taussig (cardiologist)

NEW YORK HERALD TRIBUNE, FRIDAY, FEBRUARY 15, 1946

'Blue Babies' Who Were Restored to Health and Doctors Who Did It

Herald Tribune—Bliss
Michael Rose, five and a half, of 220 Boscobel Place, the Bronx

Herald Tribune—Rice
Alan Beck, three and a half, of 1700 Sterling Place, Brooklyn

Herald Tribune—Acme
Marilyn Firsenbaum, nine and a half, 80 Chester Street, Brooklyn

Herald Tribune—Kavalliner
Harry Goldweig, seven, of 935 Fifty-seventh Street, Brooklyn

How 2 Doctors Give New Lives To Blue Babies

Blalock-Taussig Operation, First Tested on Dogs, Reroutes Flow of Blood

By Lester Grant

BALTIMORE, Feb. 14.—This is the story of the work of two doctors—a man from Georgia and a woman from Massachusetts—who met in Baltimore and combined their talents to save the lives of "blue babies."

The doctors are Alfred Blalock, forty-six, surgeon in chief at Johns Hopkins Hospital here and professor of surgery at the Hopkins Medical School, and Helen B. Taussig, forty-seven, physician in charge of the cardiac clinic of the Harriet Lane Home for Invalid Children. The Harriet Lane Home constitutes the pediatrics division of Johns Hopkins Hospital.

The surgery, known as the Blalock-Taussig operation, first was used on an infant on Nov. 29, 1944. Its development since then is one of the most exciting stories in

Herald Tribune—Acme
Dr. Alfred Blalock and Dr. Helen B. Taussig at Johns Hopkins Hospital in Baltimore

Source: Terumo, n. d., https://goo.su/OHPEcC.

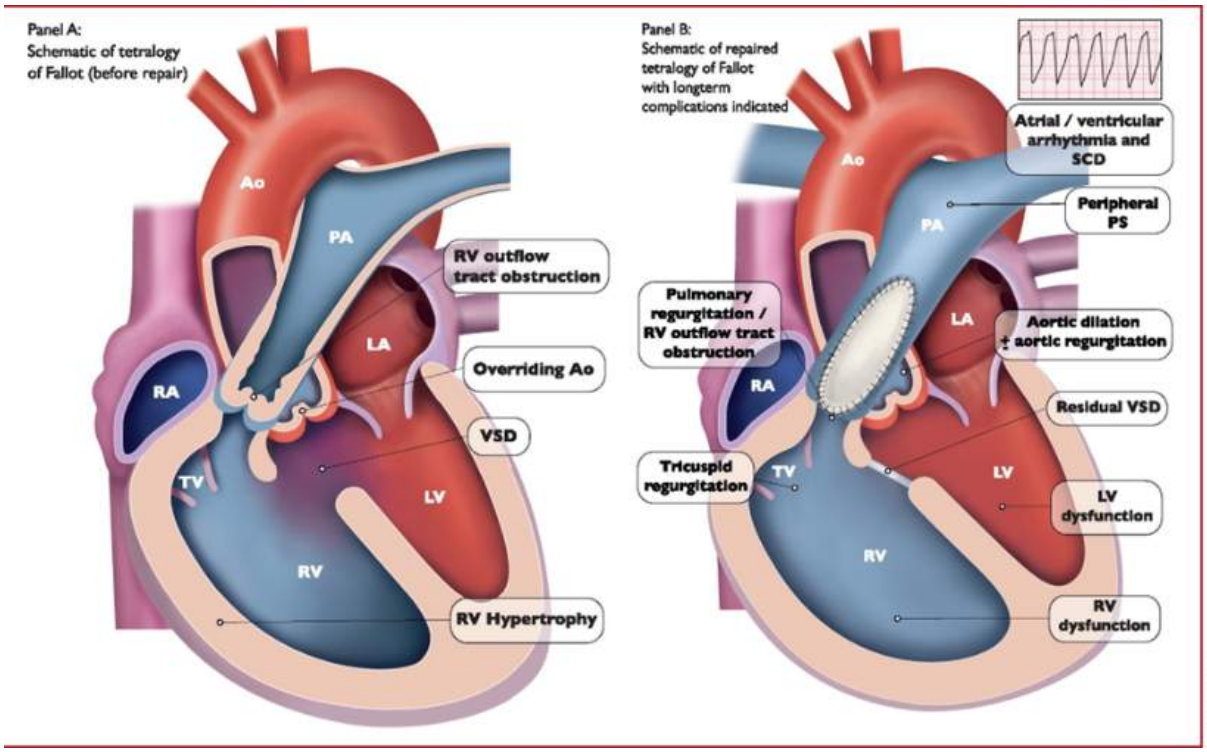
The most frequent complications (Figure11) in adulthood are the following:

1. Pulmonary insufficiency in 30% of patients at 20years old, after transannular patch. 10-15% require pulmonary valve replacement. This complication is not very symptomatic until very advanced stages.

2. Aortic root dilation, which is asymptomatic until complications occur.

3. Sudden death in 2.6- 6% of cases, so the following risk criteria are defined: RV or LV dysfunction with ejection fraction (EF) <40%; severe pulmonary insufficiency; uncontrolled atrial or ventricular arrhythmia; QRS →180ms; fragmented QRS; QT dispersion; extensive fibrosis on MRI; non-sustained ventricular tachycardia (NSVT) on Holter monitor or ventricular tachycardia (VT) induction on electrophysiological study (EPS); long-lasting palliative shunts; age of repair; LV end-diastolic pressure >12mmHg, and coronary abnormalities.

Figure 11. A: Tetralogy of Fallot pre-repair and post-repair. Long-term complications



Source: Baumgartner et al., 2021, https://goo.su/bg2cdF.

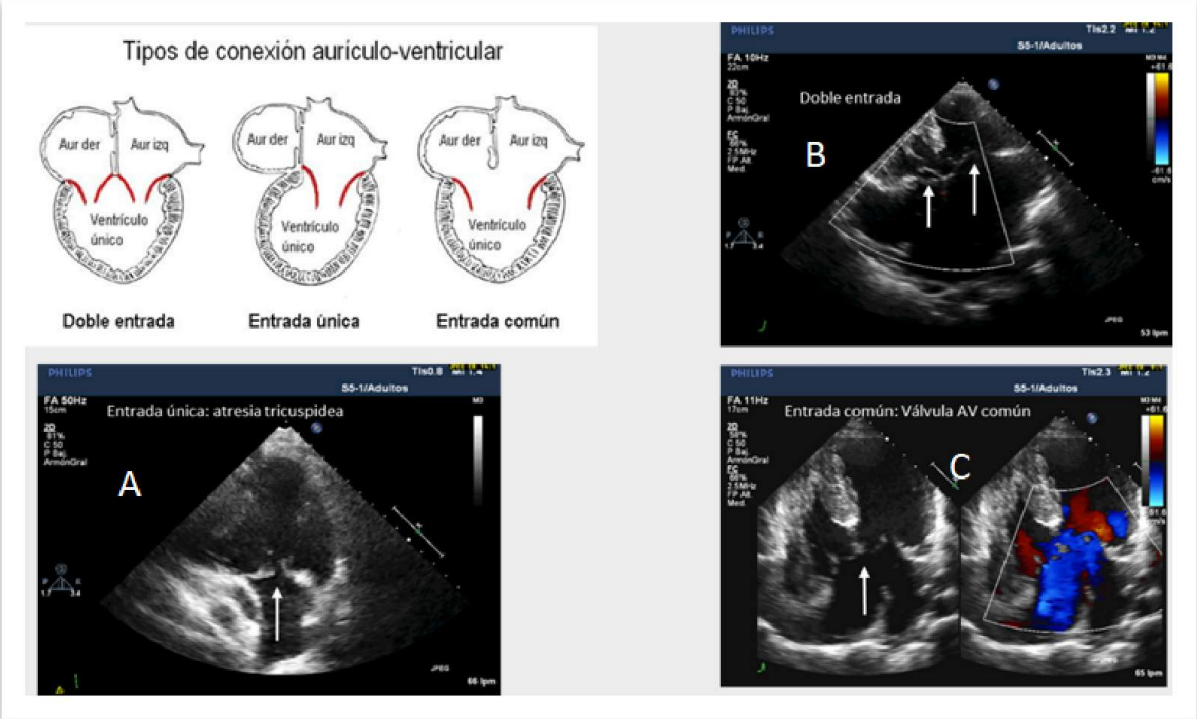
In both transposition of the great arteries and adult tetralogy of Fallot, systematic follow-up is essential, since complications are not very symptomatic, but serious if not correctly assessed.

Single ventricle. Glenn and Fontan

A single ventricle is defined as an anatomical entity characterized by a single functional cavity (with complete and well-developed

trabeculated, inlet, complete, and well-developed portions). There are different types of single ventricles. We will mainly define the following: Normal atria, right or left isomerism or mirror image; atrioventricular connection (Figure12), and morphology of the right, indeterminate, or left ventricles.

Figure12. Transthoracic echocardiography of the three types of single ventricles, according to the atrioventricular connection. A: Single inlet (tricuspid atresia); B: Double inlet; C: Common inlet (common VA valve)



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The most commonly used surgical treatment at present, if bi-ventricular correction cannot be performed, is total cavo-pulmonary connection. This is a connection of the superior vena cava to the pulmonary artery (Glenn surgery) and of the inferior vena cava to the pulmonary artery (extracardiac Fontan surgery).

c) **Without shunt or cyanosis**

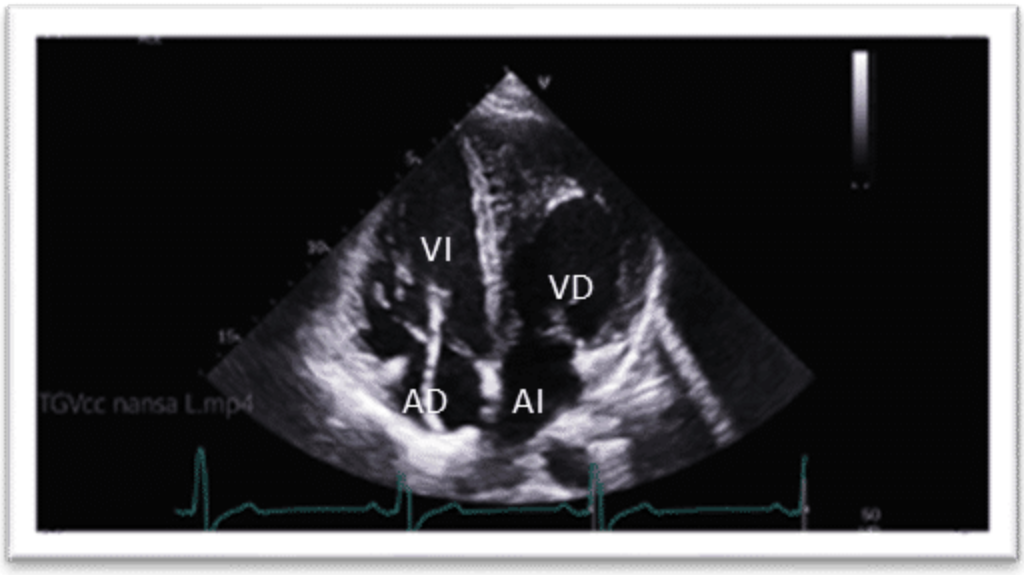
CHDs without shunt or cyanosis are congenitally-corrected transposition of the great arteries, coarctation of the aorta, and Ebstein's malformation.

Congenitally-corrected transposition of the great arteries (ccTGA)

There is a double discordance, atrial-ventricular and ventricular-arterial, i.e., the atria and ventricles are abnormally related. The right atrium (RA) connects to the morphologically-left ventricle (LV) and the left atrium (LA) connects to the morphologically-right ventricle, but the right ventricle (RV) connects to the aorta, and the left ventricle connects to the pulmonary artery (Figure13). Newborns do not present cyanosis, but are at increased risk of complete VA block and systemic right ventricular dysfunction. They may be asymptomatic, even into adulthood.

Figure13. Congenitally-corrected transposition of the great arteries. RA connected to the LV and the pulmonary artery outflow; LA

connected to the systemic RV (with the presence of a moderator band and the tricuspid valve inserted more apically). Pacemaker device with resynchronization for RV dysfunction and complete atrioventricular block (AVB)



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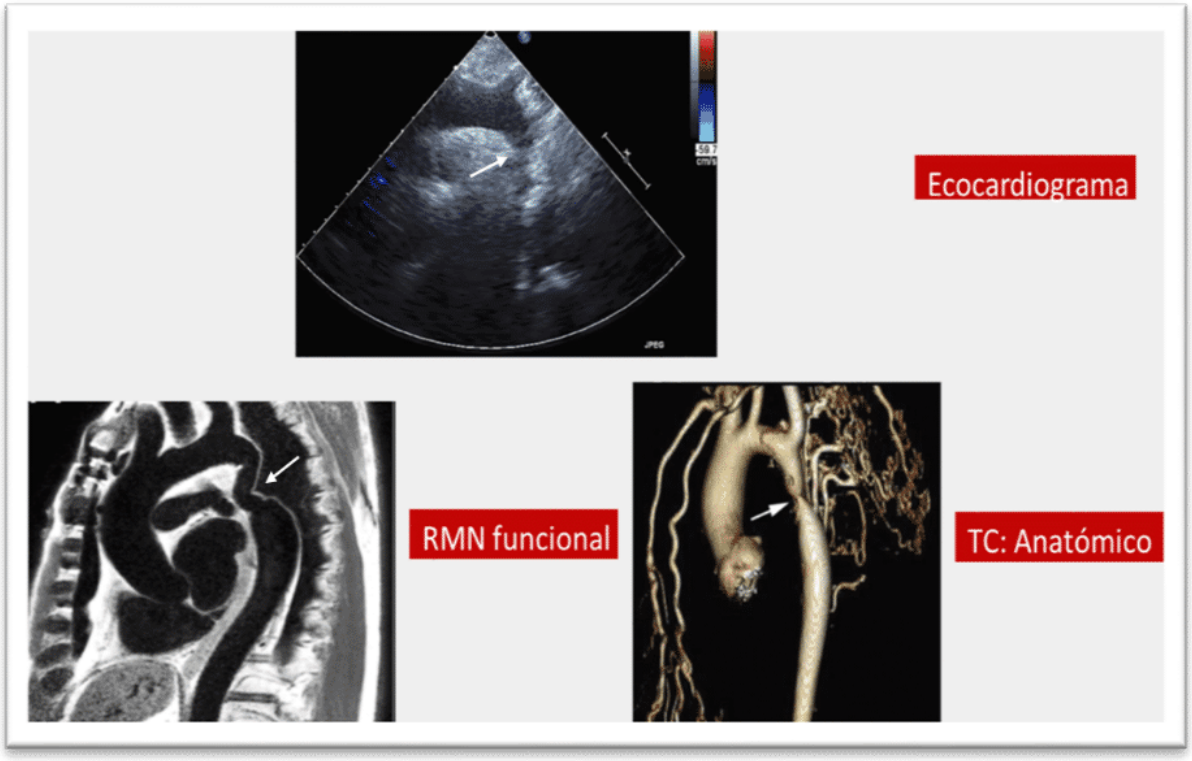
Coarctation of the aorta

Coarctation of the aorta is considered part of a generalized arteriopathy and not only as a circumscribed narrowing of the aorta at the level of the aortic arch. It accounts for 5-8% of CHDs. The most frequent associated lesion is the bicuspid aortic valve, in 85% of cases. The clinical presentation in newborns is left heart failure and, in children and adults, arterial hypertension (upper half of the body). Physical examination reveals a systolic murmur at the interscapular

level, with arterial hypertension and weak femoral pulses. Diagnosis is made by echocardiography, CT and MRI (Figure14).

Figure14. Coarctation of the aorta. Echocardiogram, MRI and CT (the arrow indicates site of coarctation, narrowing)



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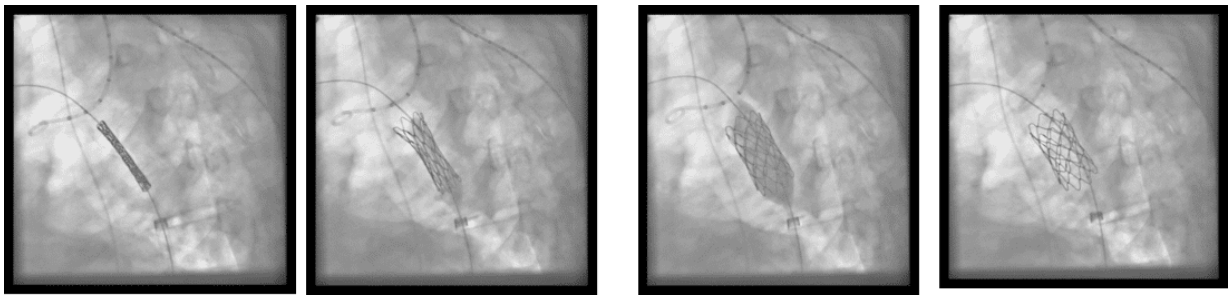
Treatment is performed percutaneously, with balloon angioplasty and stent implantation (Figure15), as well as surgically (aortoplasty). Blood pressure should be strictly controlled.

Figure 15. Angioplasty with stent in postductal coarctation of the aorta



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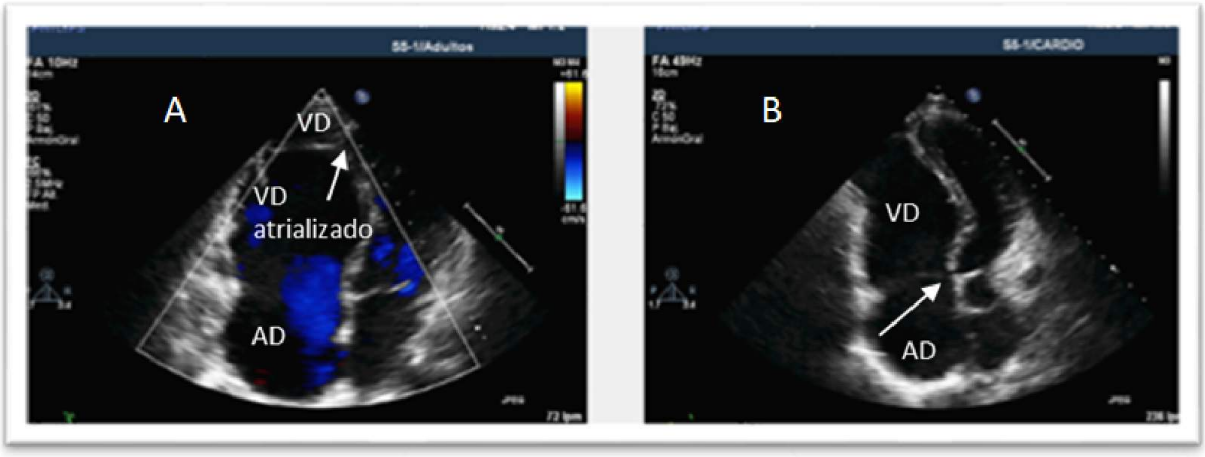
Ebstein's anomaly

Ebstein's anomaly accounts for <1% of congenital heart disease. It is characterized by an apical displacement of the insertion base of the septal and posterior leaflets of the tricuspid valve more than 15 mm or 8 mm/m2 with respect to the atrioventricular annulus, due to a delamination defect of the inner layer of the myocardium. About 25% of patients have multiple accessory pathways (Wolf-Parkinson-White syndrome). The clinical manifestations and prognosis depend, to a large extent, on the age at the onset, the degree of anatomical severity, the degree of tricuspid regurgitation, the ventricular function, the arrhythmias, and the presence of associated lesions. The right side of the heart is divided into three parts: the right atrium (RA), the atrialized portion of the RV, and the RV. The treatment of choice in

patients who do not require treatment is anatomical repair (cone reconstruction) by experts (Figure16).

Figure16. Ebstein's anomaly. A: Right heart is divided into 3parts — RA, atrialized portion of the RV, and RV— with apical insertion of the septal leaflet (arrow); B: Anatomical repair, with insertion of the tricuspid septal leaflet at the atrioventricular groove (arrow), and right heart divided into 2parts —RA and RV— with the atrialized portion of the RV integrated into the RV



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There are several guidelines aimed at adult patients with CHD with recommendations on competitive sport (Sandoval et al., 2023; Grazioli et al., 2017). Now, we will focus on the latest published guidelines on competitive sport (Budts et al., 2020). European guidelines aimed at pediatric-age (Takken et al., 2012), adolescent and adult patients

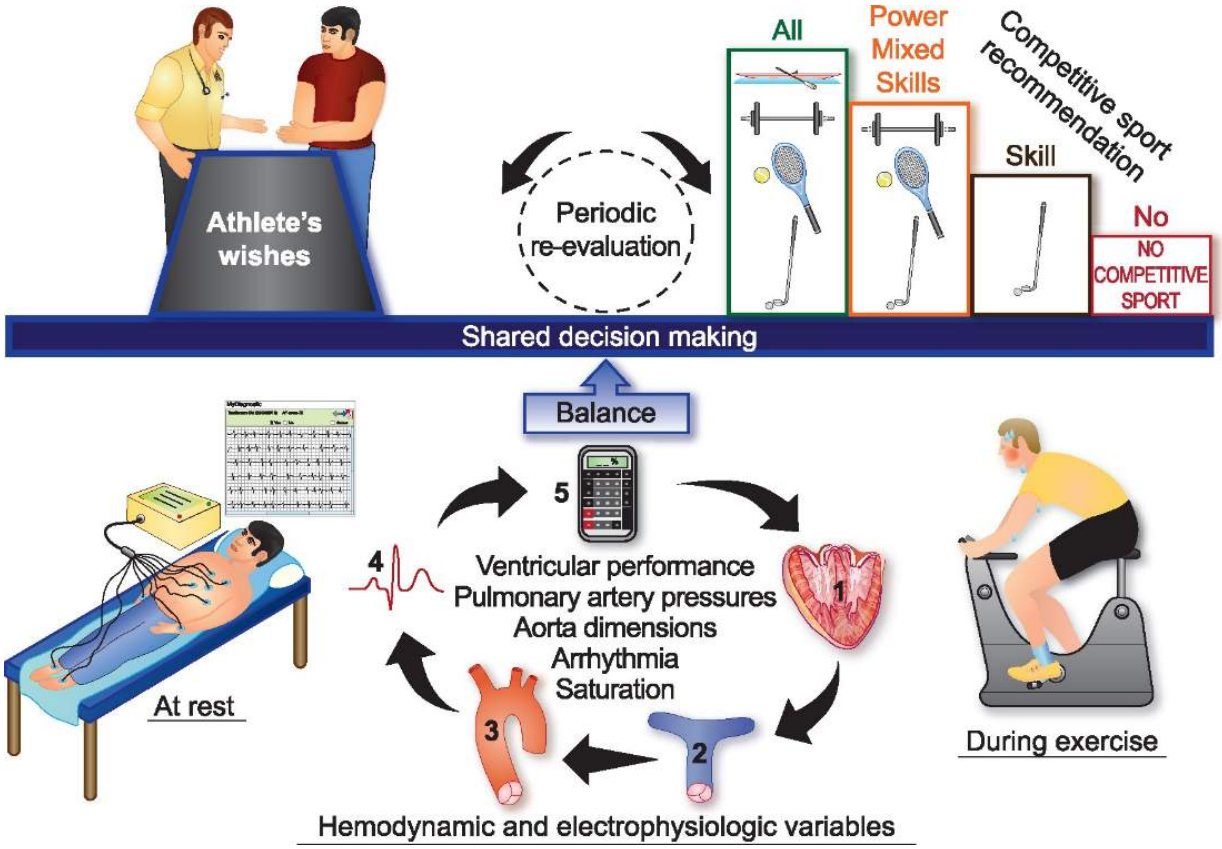
(Foster et al., 2001; Budts et al., 2013) have also been published, with reference to common and recreational physical exercise and training programs. In most of these patients, even when competitive sports are contraindicated, it is important to prescribe an active lifestyle and physical training programs appropriate to their situation.

In a study of 25,790 adults with congenital heart disease, sudden death with exercise was estimated to be 8% in adults (Zomer et al., 2012) and 0.06% in children (Jortveit et al., 2016). Hence, recent studies were conducted to detect predictors of sudden death risk in congenital heart disease (Koyak et al., 2012; Vehmeijer et al., 2019).

Adult congenital heart disease and competitive sports

The most recent guidelines (Budts et al., 2020) advise performing a complete assessment in individuals older than 16years old with CHD (Figure17).

Figure17. Comprehensive assessment of patients with congenital heart disease



Source: Budts et al., 2020, https://goo.su/wfPc1Gu.

In patients with congenital heart disease, in addition to a correct medical history and physical examination, an assessment at rest of cardiac structure (1), pulmonary hypertension (2), aortic diameters (3), arrhythmias (4), and oxygen saturation will be performed.

Besides, the cardiac response during exercise will be assessed with an oxygen consumption test or exercise echocardiogram. Subsequently, it will be decided together with the patient, according to his wishes and the complete assessment, which sport is advised (green: All; orange:

All, except high-intensity, high-volume endurance sport; brown: Skill sports only; and red: No competitive sport) (Budts et al., 2020).

Step 1: Medical history and physical examination

- Medical history: Family history (although hereditary transmission is low) and cardiological symptoms (syncope, dizziness, chest pain, dyspnea, palpitations); type of sport, intensity and level of competition. Finally, the environment where the sport is practiced.

- Physical examination: Blood pressure (if it is high, we will measure the difference between the right arm and leg), heart rate, oxygen saturation, murmurs, and pulses.

Step 2: Assessment of 5 parameters at rest (Table 1)

- Cardiac structure: Ventricular function, valvulopathies, shunts, obstructions, and aorta. The fundamental imaging techniques are transthoracic echocardiogram, cardiac MRI, and CT angiography.

- Pulmonary pressure with echocardiogram: Assess the velocity of tricuspid insufficiency (normal <2.8 m/s) and indirect signs of pulmonary hypertension. If necessary, perform right catheterization.

- Aortic dimension by echocardiogram. If it is borderline or pathological, by CT or cardiac MRI. Normal (−2 < z > 2).

- Arrhythmias: ECG, Holter monitoring, and, in patients at risk of sudden death, electrophysiological study and cardiac resonance may be performed to assess fibrosis.

- Oxygen saturation: At baseline and during exercise. Normal >95%.

Table 1. Assessment of 5 parameters at rest

Variables	Definitions
1. Cardiac structure (a)	
RV and LV dysfunction No Mild Moderate Severe	 • EF ≥55% • 45% ≤ EF <55% (normal systemic RV) • 30 ≤ EF <45% • EF <30% (reduced systemic RV)
LV hypertrophy (qualitative RV) No Mild	Septal thickness (cm) Mass (g/m^2) ♂ <1.1 ♀ <1.0 ♂ 50–102, ♀ 44–88
Moderate Severe	♂ 1.1–1.3 ♀ 1.0–1.2 ♂ 103–116 ♀ 89–100 ♂ 1.4–1.6 ♀ 1.3–1.5 ♂ 117–130 ♀101–112 ♂ ≥1.7 ♀ ≥1.6 ♂ ≥131 ♀ ≥113
RV and LV pressure overload No Mild Moderate Severe	PSV assessment in RVOT/LVOT/PV and arm-to-leg gradient of coarctation of the aorta • PSV <2.6 m/s • 2.6 m/s ≤ PSV <3 m/s Arm-leg gradient <20 mm Hg • 3 m/s ≤ PSV ≤4 m/s • PSV >4 m/s Arm-leg gradient ≥20 mm Hg
RV and LV volume overload (b)	
No Overload without ventricular remodeling Overload with mild ventricular remodeling Overload with severe ventricular remodeling	• Mild/moderate insufficiency or shunt without volume overload • Severe-grade insufficiency or shunt without dilation and ventricular dysfunction • + ventricular dilation • + ventricular dysfunction
Ventricular physiology	Single ventricle or bi-ventricular circulation Systemic LV or systemic RV
2. Pulmonary pressure No PH without RV dilation or dysfunction PH with ventricular dilation or dysfunction	• Maximum TR velocity ≤2.8 m/s and without indirect signs of PH. • PAPm ≥20 mm Hg (right heart catheterization) • PAPm ≥20 mm Hg + ventricular dilation or dysfunction
3. Aorta	Aortic diameter
No/mild Moderate Severe If there are risk criteria, surgery	• Normal (≤35 mm) or borderline (≥35 to <40 mm) *z*-score ≥2 to <3 • ≥40 to <45 mm, *z*-score ≥3 to <4 • ≥45 to <50 mm, *z*-score ≥4 • ≥50 mm
4. Arrhythmias No Not significant/not malignant Significant/potentially malignant	 • <500 PVC/24 hr on Holter monitor, which do not worsen with exercise • Frequent PVC, doublets, or controlled AF/A flutter, which do not worsen with exercise • AF/A flutter or PVC which worsen with exercise, NSVT or SVT
5. Cyanosis No Mild-moderate Severe	Oxygen saturation; at rest or during exercise 96%–100% 90%–95% <90%

Table references

RV: Right ventricle; LV: Left ventricle; A: Atrial; PVC: Premature ventricular contractions; EF: Ejection fraction; AF: Atrial fibrillation; TR: Tricuspid regurgitation; PAPm: Mean pulmonary arterial pressure; RVOT: RV outflow tract; LVOT: LV outflow tract; PV: Pulmonary valve; PSV: Peak systolic velocity; PH: Pulmonary hypertension; TVNS: Non-sustained ventricular tachycardia; SVT Sustained ventricular tachycardia.

The following should be considered:

1. Interpretation of ventricle measurements should contemplate the race and sport discipline of the athlete.

2. Serial follow-up should be performed, especially in cases where we do not know whether the ventricular dilation is secondary to the cardiac lesion or to the sport performed.

3. In individuals with systemic RV, the maximum velocity of mitral regurgitation will be assessed.

Step 3: 5 parameters during exercise

Cardiopulmonary stress tests with oxygen consumption provide very relevant information related to the physiological sequelae of anatomical lesions, the risk of morbidity and mortality, and the timing of the intervention.

- Peak oxygen consumption (VO2 peak) is one of the best predictors of morbidity and mortality. In addition, the following parameters can help quantify exercise capacity: Heart rate reserve; ventilatory efficiency slope as a useful parameter in the context of submaximal testing; O2 pulse to assess systolic volume; gas exchange threshold to detect alterations in aerobic and anaerobic metabolism.

- Detection of ischemia

- Desaturation during exercise, in case of shunts or pulmonary pathology.

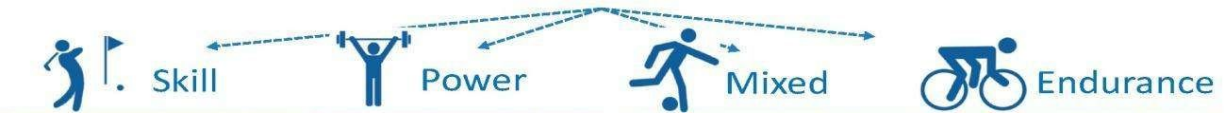
- The behavior of blood pressure. The normal value is a rise from >25 mm Hg with exercise up to a maximum of 220 mm Hg(♂) y 200 mm Hg (♀).

An exercise echocardiography also allows us to assess ventricular function, segmental alterations, gradients, mitral (or systemic tricuspid) regurgitation, and the degree of pulmonary hypertension during exercise.

Step 4: Exercise type recommendation

The following image shows a schematic representation of the 4 types of sports disciplines.

Figure 18. The 4 types of sports disciplines



Source: Pelliccia et al., 2020, https://goo.su/ih3p9nP.

Table 2. Recommendations according to the type of sport discipline

HR	+/++	HR	++	HR	++/+++	HR	+++
BP	+	BP	+++	BP	++	BP	++
CO	+	CO	++	CO	++/+++	CO	+++
Volume of training	-	Volume of training	+	Volume of training	++	Volume of training	+++
Remodeling	-	**Remodeling**	+	**Remodeling**	++	**Remodeling**	+++
Golf Motorcycle racing** Sailing Horseback riding* Ski jumping* Scuba diving* Archery Table tennis Shooting		Calisthenics Alpine skiing Snowboarding Javelin Climbing* Weightlifting Wrestling		Soccer** Basketball Handball** Rugby** Tennis Squash Hockey** Volleyball Waterpolo* Gymnastics		Biathlon Triathlon Pentathlon Canoeing Cycling Mid-long distance skating Running Swimming*	

Source: own source based on Budts et al., 2020, https://goo.su/wfPc1Gu.

Table references

HR: Heart rate; CO: Cardiac output; BP: Blood pressure;* indicates sports with risk of collision; ** indicates sports with risk of severity or death in case of syncope.

Based on the 5 parameters assessed, each patient will be recommended, according to his congenital heart disease and on an individual basis, which competitive sport he should practice (Table 3).

Table 3. Recommended sports according to CHD

1. Structure	No dysfunction No/mild hypertrophy No/mild pressure overload No volume overload	Mild dysfunction Volume overload without remodeling	Moderate dysfunction Moderate hypertrophy Moderate pressure overload Volume overload with mild remodeling Single ventricle Systemic right ventricle	Severe dysfunction Severe hypertrophy Severe pressure overload Volume overload with severe remodeling
2. Pulmonary hypertension	Low probability	PH without ventricular remodeling		PH with ventricular dilation or dysfunction
3. Aorta	No/mild dilation	Moderate dilation	Severe dilation	Dilation with criteria to assess surgery
4. Arrhythmias	No arrhythmias	Non-significant/ non-malignant arrhythmias		Significant or malignant arrhythmias
5. Oxygen saturation	No central cyanosis		Mild central cyanosis	Severe central cyanosis
	A	B	C	D
	All of A	≥1 parameter of B, but none of C or D	≥1 parameter of C, but none of D	≥1 parameter of D
Competitive sports	All sports	Skill, strength, or mixed-type sports	Skill sports	Non-competitive sports

Source: own source based on Budts et al., 2020, https://goo.su/wfPc1Gu.

For the sake of simplicity, cases in which competitive sports are contraindicated are shown in **red.** The cases in which competitive sports are allowed, but only skill sports, are shown in **orange.** The cases in which competitive sports are allowed, but only skill-, strength-, mixed-type sports are shown in **yellow. Green** color indicates patients fit for all sports. Nevertheless, it should be emphasized that the decision should always be individualized and personalized according to the severity of the heart disease, the subject's comorbidities, the sporting modality and sport discipline, and, especially, the subject's personal environment.

The following tables of contraindications are an adequate and reasonable guide for making decisions regarding the granting of eligibility for the practice of a sport, and they complement and guide good judgment and clinical criteria.

Step 5: Follow-up

Patients with congenital heart disease who practice competitive sports should be evaluated every 6-12 months, according to their heart disease, risk of arrhythmias, hemodynamic sequelae, and the sport they practice.

Special considerations

Patients taking blood-thinning medications should not practice contact sports. Patients with ICDs and pacemakers should follow the sport recommendations of these devices, and those with arrhythmias, coronary abnormalities, and pulmonary hypertension should also follow the corresponding guidelines.

Sports at high altitudes can increase pulmonary resistances, reduce cardiac output, oxygenation, hypercoagulability, and arrhythmias. Unrepaired or palliated cyanotic patients with complex congenital heart disease or pulmonary hypertension should not practice sports at moderate or high altitudes (>1,500 m). After closure of an ASD, VSD, PDA or stent in a coarctation of the aorta, a complete re-evaluation is performed at 3-6 months to assess eligibility for sports.

Finally, for each congenital heart disease, we summarize below which are the recommendations for competitive exercise. For each of the alterations, a recommendation of the recommended exercise intensity is given, according to the type of competitive sport.

In the case of ASD, AVD (anomalous venous drainage), VSD, and PDA and after closure of the defect, avoid diving before closing the ASD, and avoid high altitudes if there is pulmonary hypertension or cyanosis.

Table 4. Recommendations in congenital heart disease - ASD, AVD (anomalous venous drainage), VSD, and PDA

Syncope, chest pain, palpitations, dyspnea, or any of the following symptoms: 1. RV dysfunction (for ASD and AVD) or with LV dysfunction (for VSD or PDA) EF <45% 2. Tricuspid insufficiency >3.5 m/s suspected (right catheterization PAPm >20 mm Hg or pulmonary vascular resistance (PVR) >3 WU) 3. Aortic dilation 4. Atrial arrhythmias (uncontrolled atrial fibrillation or flutter, ventricular NSTV, PVC that increase with exercise) or 2nd or 3rd AVB 5. Desaturation at baseline or during exercise <95%	Assess treatment and after 3-6 months from closure
Only mild ventricular remodeling	Assess closure
Only tricuspid insufficiency 2.8-3.5 m/s and without RV dysfunction (for ASD and AVD) or LV dysfunction (for VSD or PDA) with right catheterization PAPm >20 mm Hg o PVR >3 WU Only mild RV or LV dysfunction EF 45-50%	At 6 months, assess closure
Only controlled atrial arrhythmias (atrial fibrillation or flutter) or only PVC >500 h/24 h, doublets that disappear with exercise	At 6 months, assess ablation
All normal, no symptoms or arrhythmias, tricuspid insufficiency <2.8 m/s and no RV (for ASD and AVD) or LV dysfunction (for VSD or PDA)	Control after 1 year

Source: own source.

It is recommended, as well as ASD, AVD, VSD, or PDA and according to residual valvular lesion (mitral insufficiency [MI] or mitral stenosis [MS], tricuspid insufficiency [TI] or tricuspid stenosis [TE]), see valvulopathies.

Transposition of the great arteries with intervention (anatomical correction - arterial switch Jatene) Coronary arteries CT angiography: Rule out coronary stenosis or angulation.

Table 5. Recommendations for congenital heart disease - repaired AV canal

Only one of the following: Myocardial ischemia during exercise Ventricular dysfunction LV EF <30% Severe neoaortic insufficiency with dilated LV and LV EF<55% Severe pulmonary stenosis	6 months
Only one of the following: Severe neo-aortic insufficiency with dilated LV and LV EF >55% Moderate pulmonary stenosis Ventricular dysfunction LV EF 30-45%	6 months
Only one of the following: Moderate-severe neoaortic insufficiency Mild LV dysfunction EF 45-50% with normal stress test	6 months
Asymptomatic. Mild neoaortic insufficiency, mild pulmonary stenosis 1. LV and RV EF >50% 2. No or mild RVOT obstruction 3. No aortopathy 4. No arrhythmia on Holter monitor, normal stress test 5. No residual shunt	1 year

Source: own source.

In the case of the tetralogy of Fallot, fibrosis should be ruled out by MRI and, if there are criteria for risk of sudden death, an EPS should be performed.

Table 6. Recommendations in congenital heart disease - Tetralogy of Fallot

Syncope, palpitations, or any of the following symptoms: 1. RV or LV dysfunction EF <45% or severe RV dilation (>160 ml/m²) with severe pulmonary insufficiency (PI) 2. Right ventricular hypertension (> 50% of systemic pressure), transpulmonary gradient >60 mm Hg or maximum velocity >4 m/s (severe) 3. Severe ascending aortopathy >50 mm 4. Uncontrolled atrial or ventricular arrhythmia, QRS ≥180ms, fragmented QRS, QT dispersion, extensive fibrosis on MRI, NSVT on Holter monitor or VT induction on EPS 5. Desaturation at baseline or during exercise <90% Other risk criteria: Long-lasting palliative shunts, older age of repair, LV end-diastolic pressure >12 mm Hg, coronary abnormalities	Assess treatment
Only one of the following: Severe pulmonary insufficiency with slightly dilated RV and RV EF >55% Moderate RVOT obstruction (transpulmonary gradient 40-60 mm Hg or maximum velocity 3-4 m/s) Aorta 45-50 mm Desaturation at baseline or during mild exercise 90-95%	6 months
Only one of the following: RV or LV EF 45-50% Moderate pulmonary insufficiency Aorta 40-45 mm Controlled atrial or ventricular arrhythmia	1 year
Asymptomatic and without risk criteria: 1. LV and RV EF >50% and normal or slightly increased RV size or mild PI 2. No or mild RVOT obstruction (transpulmonary gradient <40 mm Hg or maximum velocity <3 m/s) 3. No aortopathy. Aorta <40 mm 4. No arrhythmia on Holter monitor, normal stress test and MRI with no significant fibrosis 5. Desaturation at baseline or during exercise $SatO_2$ >95%	1 year

Source: own source.

In the case of total cavopulmonary connection and Fontan surgery, the recommendations are presented in the following table.

Table 7. Recommendations in congenital heart disease - Total cavopulmonary connection and Fontan surgery

Asymptomatic for heart failure and without risk criteria 1. LV and RV EF >50% 2. No or mild RVOT obstruction 3. No aortopathy 4. No arrhythmia on Holter monitor, normal stress test 5. No residual shunt $SatO_2$ >95% Normal stress test (no ischemia, arrhythmias, or arterial hypotension)	6 months
Symptoms of heart failure or any risk criteria	3-6 months

Source: own source.

Table 8. Recommendations in congenital heart disease - Congenitally-corrected transposition of the great arteries

No risk criteria and normal stress test (no ischemia, arrhythmias, or arterial hypotension) * RV EF 35–45%	6 months
Risk criteria: 1. Systemic RV EF <35% 2. Right ventricular hypertension (>50% of systemic pressure) 3. Severe ascending aortopathy >50 mm 4. Recurrent or uncontrolled atrial or ventricular arrhythmia, VT on Holter monitor or stress test or MRI with significant fibrosis 5. $SatO_2$ <90% Other risk criteria: Previous VSD, QRS >180 ms, fragmented QRS, heart failure, ischemia, coronary abnormalities	3–6 months

Source: own source.

In the case of untreated coarctation of the aorta, isometric exercises should be avoided.

Table 9. Recommendations in congenital heart disease - Untreated coarctation of the aorta

1. Aortic dilation score ≤3.0 2. Systolic blood pressure gradient between right upper extremity and right lower extremity <20 mm Hg 3. Peak systolic blood pressure <95th percentile predicted by age (stress test with BP: <220 in men and <200 mm Hg in women)	1 year
No, any of the above 3 items	At 6 months, assess treatment

Source: own source.

In the case of coarctation of the aorta treated with stent placement or surgical repair, isometric exercises should be avoided.

Table 10. Recommendations in congenital heart disease - Coarctation of the aorta treated with stent placement or surgical repair

After 3 months, if all these items apply: 1. Aortic dilation z-score ≤3.0 2. Systolic blood pressure gradient between upper and lower right extremities <20 mm Hg 3. Peak systolic blood pressure <95th percentile predicted by age 4. No aneurysm associated with coarctation 5. No contraindicating aortic valvulopathy Note: Evaluate the association with bicuspid valve	1 year
Aortic dilation z-score 3-4 Note: Evaluate the association with bicuspid valve	6 months

Source: own source.

Recommendations for patients with Ebstein's malformation are shown in the following table.

Table 11. Recommendations in congenital heart disease - Ebstein's malformation

Severe TR with symptoms or any of the following symptoms: 1. Moderate-severe RV and/or LV dysfunction EF <45% or moderately-severely dilated RV 2. Right ventricular hypertension (>50% of systemic pressure) 3. Aortic dilation >50 mm 4. Uncontrolled atrial arrhythmias or malignant ventricular arrhythmias 5. Desaturation at baseline or during exercise <90%	Assess treatment
Severe TR with only RV and/or mild LV dysfunction EF 45-55% and non-significant or non-malignant arrhythmias (rare isolated PVC) Severe tricuspid insufficiency with slightly dilated RV and RV EF >50-55%	Assess treatment (stress test, oxygen consumption, and NT pro-BNP)
Mild, moderate, severe TR with no symptoms 1. Without RV and/or LV dysfunction with non-dilated RV 2. No high blood pressure 3. No aortic dilation 4. No arrhythmias 5. No desaturation at baseline or during exercise	1 year

Source: own source.

Table references

A: Atrial; PVC: Premature ventricular contractions; EF: Ejection fraction; AF: Atrial fibrillation; TR: Tricuspid regurgitation; PAPm: Mean pulmonary arterial pressure; RV: Right ventricle; LV: Left ventricle.

Green: All competitive sports.

Yellow: Skill, strength, and mixed-type competitive sports.

Orange: Skill competitive sports.

Red: Competitive sports are contraindicated.

In most of these patients, even when competitive sports are contraindicated, it is important to prescribe an active lifestyle and appropriate physical training programs on an individual basis.

For recreational sports or cardiac rehabilitation, we must know the static and dynamic component of the sport and the degree of intensity, according to the stress test, ideally with oxygen consumption.

The Michel's classification divides sports according to their static and dynamic components. Regarding the static component, Michel classifies sports according to the estimated percentage of maximal oxygen consumption (A: <50%; B: 50-75%, C: >75%). As for the dynamic component, it consists of the estimated percentage of the maximum voluntary contraction achieved (I: <10%; II: 10-30%; III: >30%).

Table 13. Michel's classification

III. High static	Martial arts* Climbing*^ Artistic gymnastics*^ Weightlifting Ski jumping *# Sailing Windsurf	Bodybuilding*^ Alpine skiing*^ Snowboarding*^ Wrestling*	Boxing* Cycling*^ Speed skating*^ Rowing Triathlon*^ Water polo
II. Moderate static	Motorsports*^ Diving*^ Equestrian sports*^ Motorcycling*^ Archery Motor boating*^ Recreational fishing Polo	Sprint events Speed - Jumping Fencing American Football* Rhythmic gymnastics Synchronized swimming^ Skating* Rugby Surf Caving	Middle-distance athletics Basketball* Handball* Ice hockey* Roller hockey* Swimming Modern pentathlon Tennis Paddle
I. Low static	Pool Bowling Golf Pitch and putt *Pétanque* Olympic shooting Yoga	Baseball Softball Ball **Tennis (doubles)** **Table tennis** **Volleyball** Hunting	Distance running Badminton Cross-country skiing*^ Soccer* Field hockey Orienteering
	A. Low dynamic	**B. Moderate dynamic**	**C. High dynamic**

Source: own source based on Grazioli et al., 2017, https://goo.su/dSTYdW.

Table references

*Contact sports with risk of bodily collision.

^Sports with risk of death, in case of syncope.

In recreational sport, there is insufficient evidence. Despite this, it is advised that the prescribed exercise intensity be determined by the stress test with O2 consumption. The percentage of the maximum heart rate during the stress test and the Borg scale will be calculated. For the sake of simplicity, based on the tables presented above, individuals who are assigned the color red can perform low-intensity, low-static recreational sports (with an intensity of less than 60% of heart rate or Borg scale 11-12). Individuals who are assigned orange and yellow can perform moderate-static recreational sport with moderate intensity (less than 60-75% or Borg scale 13-14). Finally, those who are assigned the green color can perform all sports and at maximum intensity >75-90% or Borg scale 15-17. For patients who insist on practicing sports with a greater static component than the one previously prescribed, it is advisable to reduce its intensity.

The usual cardiological follow-up indicated in every patient with a CHD should be sufficient to reevaluate their sports eligibility. Patients should monitor their symptoms (Borg scale) and heart rate.

Table 13. Borg scale, intensity, sweat, VO2 max % and percentage of maximum heart rate (HR) of the stress test with ocygen consumption

Borg (6-20)	Intensity	Sweat	VO_2 max %	Maximum HR %
<11	Mild	No/Scarce sweating	<20	<54%
12-13	Moderate	Scarce sweat	20-39	55-69%
14-16	Heavy	Very sweaty	40-59	70-89%
17-19	Very heavy	Very tired	60-84	>90%
20	Maximum	Unable to breathe	>85	100%

Source: own source based on Budts et al., 2020, https://goo.su/wfPc1Gu.

In summary, individuals with congenital heart disease are a growing population with a good prognosis and may be suitable for competitive sports. In some cases, cardiological treatments may allow the patient to resume competitive sports. Similarly, recreational sports/appropriate exercise prescription should be advised in all patients with congenital heart disease to improve their prognosis.

Clinical case

34-year-old male patient, competitive basketball player. On medical examination, atrial fibrillation and right His bundle branch block are detected. A transthoracic echocardiography (TTE) study showed an atrial septal defect (ASD) of the ostium secundum type with left-right shunt and right ventricular volume overload.

Step1

Medical history: No family history of sudden cardiac or unexplained death, no history of cardiac pathology before the age of 50. He denies cardiological symptoms. He had no syncopal episodes, chest pain, dyspnea, or palpitations. He does not take regular medication and has not required any intervention.

Sports history: He has been playing basketball, a mixed-type sport, since childhood, and has been playing competitively since adolescence. He currently competes at national level, which is

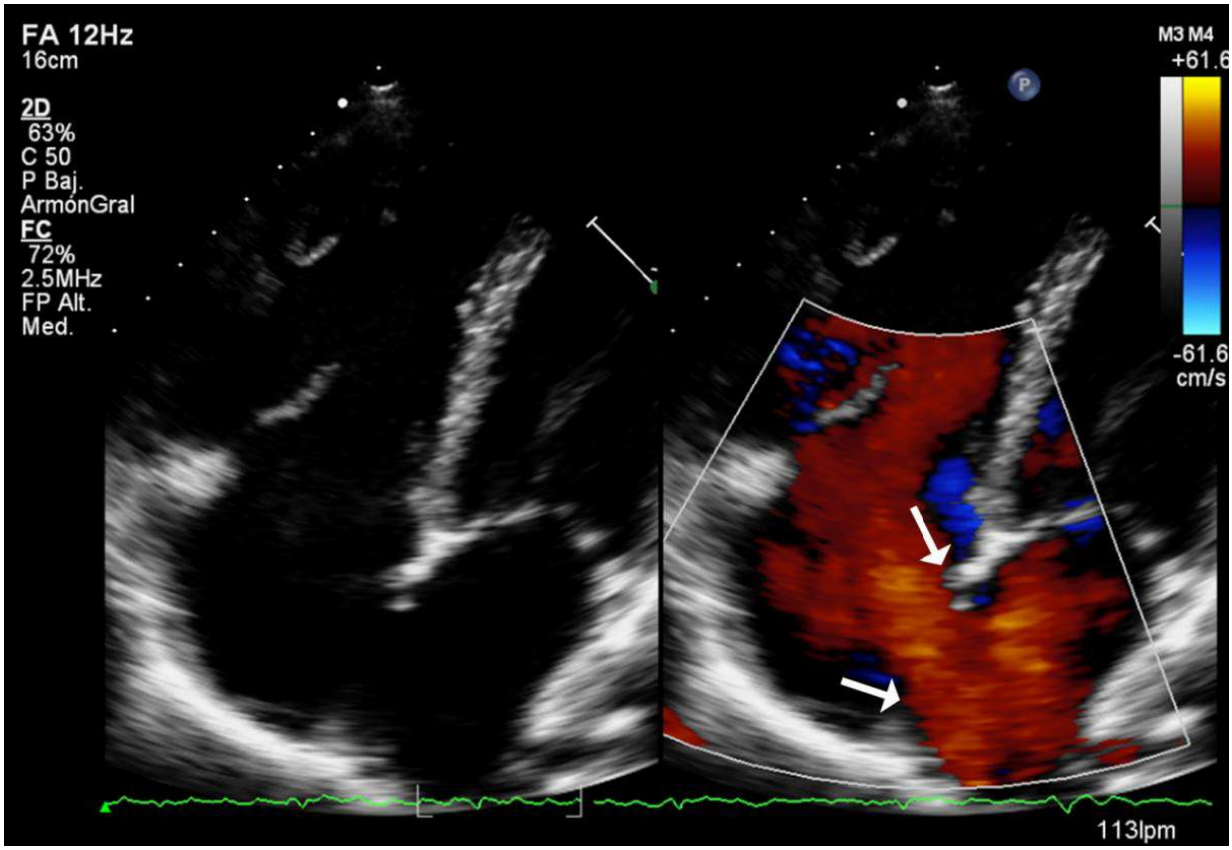
associated with moderate-high training loads of mixed type (8 hours/week).

Physical examination: Blood pressure: 100/60 mmHg; heart rate: 90 bpm; oxygen saturation: 100%; normal pulses. Cardiac auscultation shows irregular heart sounds with fixed splitting of the second heart sound.

Step 2

The transthoracic echocardiogram shows a severely-dilated right ventricle with normal ventricular function, low probability of pulmonary hypertension. The etiology of this volume overload with mild ventricular remodeling (ventricular dilation without ventricular dysfunction) is due to the presence of an atrial septal defect of the ostium secundum type (see Figure 18). The left ventricle is of normal size and function, systolic-diastolic left ventricular function is preserved. The size of the root and ascending aorta are normal, and the origin of the coronary arteries is normal. Volume overload with mild remodeling. The ECG shows baseline atrial fibrillation with a mean ventricular rate of 90 bpm and complete right bundle branch block.

Figure 18. Transthoracic echocardiogram. Apical view of 4 chambers. Comparison with and without Doppler echocardiogram



Source: own source.

The image shows the usefulness of a color Doppler echocardiogram to observe the passage of blood through the interatrial septum defect through the ostium-secundum-type ASD (arrows).

Step 3

A conventional maximal stress test is performed, achieving 16METs. The tension and adrenergic response are correct. No arrhythmias or ST segment changes were observed.

Step 4

Basketball is a mixed-type sport discipline, following the current European Association of Preventive Cardiology (EAPC) classification. In this context, volume overload with mild ventricular remodeling contraindicates the practice of this sport at a competitive level.

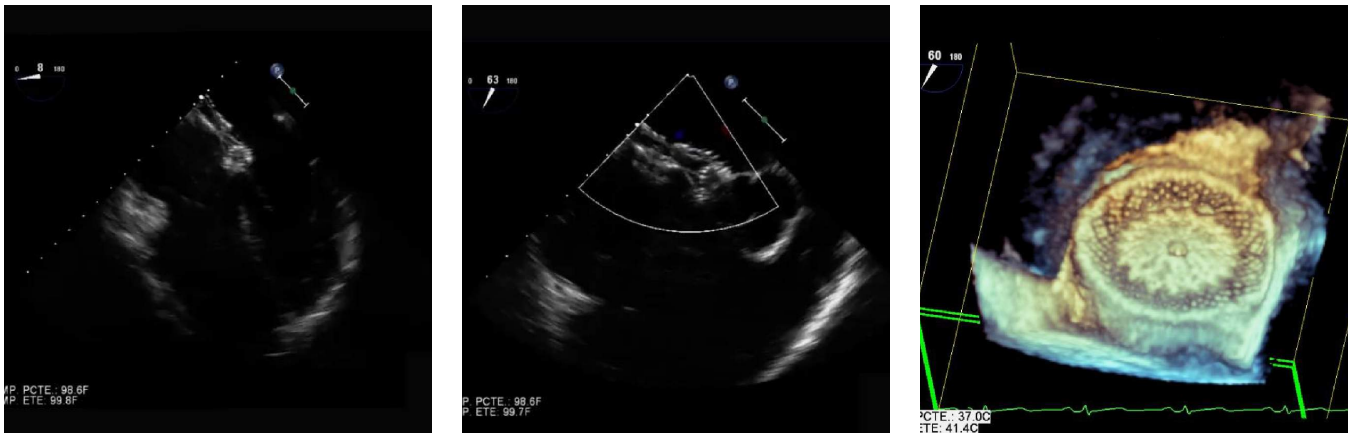
Step 5

It is therefore considered that it is the closure of the ASD what is causing such volume overload in the right cavities. Atrial fibrillation ablation is also considered.

A transesophageal ECG was performed to rule out atrial appendage thrombi and to evaluate the ASD in detail to confirm its suitability for percutaneous closure. The test confirms that the maximum dimensions from the insertion of the interatrial septum to the margins of the ASD are greater than 4-5mm at all edges (a requirement for percutaneous closure, with the exception of the aortic edge, where it is not mandatory). It is considered suitable for percutaneous closure of ASD.

Prior to closure, atrial fibrillation ablation should be performed, since access to the pulmonary veins is through the interatrial septum. Pulmonary vein ablation was successfully performed. Six months after the procedure, percutaneous closure of the ASD was performed.

Figure19. Percutaneous ASD closure with Amplatzer device guided by transesophageal echocardiogram



Source: own source.

Six months after closure, the right ventricle normalizes, without dilation, and the normal right ventricular function is maintained. The patient remains in sinus rhythm and asymptomatic. In agreement with the patient, it is considered that he can progressively resume his usual training routine and competition. At the age of 38 (4 years after diagnosis), he decided to stop competing. Currently, he is 44 years old, plays mixed-type recreational sports (basketball and paddle) and remains free of atrial fibrillation, with normal right cavities and no pulmonary hypertension.

CONTINUE

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