Recommendations for adult sport athletes with congenital heart diseases

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Received: 15.12.2015
Accepted: 09.03.2016

Summary

Safety of sport and physical activity in adult patients with congenital heart disease is still not well established. Recommendations on exercise in this subgroup of patients are usually pretty restrictive without having clear evidence for this, even though sport has shown significant cardiovascular benefits in both the general population and in patients with cardiovascular problems. Prevalence and survival of this population has increased considerably in recent decades due to early diagnosis, a substantial improvement in therapeutic measures, both surgical techniques and percutaneous, as well as aftercare and strict long term clinical monitoring. This is why it is increasingly common to find asymptomatic patients with good performance status and many doubts about their chances in sports. Doubts among professionals in establishing the best recommendations in relation to sport also arise. In the last years, it has become particularly relevant the study of the benefits and safety of these types of activities in this subgroup of patients. Researches focuses mainly on the safety of physical activity in patients with congenital heart disease, and fear that the practice of physical activity on a competitive level can significantly increase the risk of adverse events, especially arrhythmic events and sudden death. In this review, we analyzed numerous studies and current clinical practice guidelines, in order to establish recommendations for physical activity and its restrictions in terms of the different types of congenital heart disease.

Key words:
Exercise.
Congenital heart disease.
Sporting activity.
Competition.

Recomendaciones para la actividad deportiva en atletas con cardiopatías congénitas en el adulto

Resumen

La seguridad de la actividad física y deportiva en pacientes adultos con cardiopatías congénitas aún no está bien establecida. Las recomendaciones sobre el ejercicio físico en estos pacientes suele ser bastante restrictiva sin que haya clara evidencia para ello, a pesar de que el deporte haya demostrado importantes beneficios cardiovasculares tanto en la población general como en estos pacientes. La prevalencia y la supervivencia de esta población ha aumentado considerablemente en las últimas décadas debido a un diagnóstico precoz, una sustancial mejora de las medidas terapéuticas, tanto en técnicas quirúrgicas como percutáneas, así como en los cuidados posteriores y un seguimiento clínico estricto a largo plazo. Es por ello que cada vez es más frecuente encontrarnos con pacientes asintomáticos con buen grado funcional y con numerosas dudas sobre sus posibilidades en la práctica deportiva. Así mismo, surgen dudas entre los profesionales a la hora de establecer las mejores recomendaciones en relación a la actividad deportiva. En los últimos años ha cobrado especial relevancia el estudio de los beneficios y la seguridad de este tipo de actividades en este subgrupo de pacientes. Las investigaciones se centran fundamentalmente en la seguridad de la actividad física en pacientes con cardiopatías congénitas, y el temor a que la práctica de actividad física a nivel competitivo pueda aumentar significativamente el riesgo de eventos adversos, especialmente de eventos arrítmicos y muerte súbita. En esta revisión, analizamos numerosos estudios y las guías de práctica clínica actuales, con el fin de establecer las recomendaciones de actividad física, así como sus restricciones en función de los diferentes tipos de cardiopatías congénitas.

Palabras clave:
Ejercicio físico.
Cardiopatías congénitas.
Actividad deportiva.
Competición.

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Introduction

Today, clinical care for children with congenital heart diseases, as well as advanced surgical techniques, have given way to a considerable increase in the survival rate of patients who reach adulthood. Despite this, only a minority of patients with congenital heart disease (approximately 19%)[1] receive advise on recommended physical activity and they often follow a sedentary lifestyle, as a result of overprotection and uncertainty regarding the type and intensity of physical activity they should carry out. This is of particular importance when considering the trend within this demographic to become overweight, in comparison to the general public, fundamentally due to a lack of physical activity[2].

The participation in sports by adults with congenital heart diseases is considered a relatively new field, and many doctors may face difficulties when advising their patients. The main concern is based on patient safety and the fear that carrying out sports to a competitive level may increase the risk of adverse events, especially arrhythmias and sudden death. However, within this field there is a significant lack of prospective data, and there are still many disputed opinions. Moreover, it should be remembered that participation in sporting events might have beneficial effects on life quality, ischemic heart disease and heart failure[3]. As a result, we should not limit, above indicated levels, physical or recreational activity. Studies carried out on patients with congenital heart disease indicate that the majority of people participating in training programmes and receiving appropriate recommendations reveal a significant improvement in their exercise capacity and psychological state. The current challenge is to ensure safe participation in regular physical activity in order to avoid the detrimental effects associated with a sedentary lifestyle. This article describes recommendations for physical activity depending on the different type of congenital heart disease. Below is a classification of the static and dynamic components of the currently most practised sports (Table 1)[4].

Bicuspid aortic valve

The bicuspid aortic valve (BAV) is the most common congenital heart disease within the general population, with an incidence rate of around 0.5-2.4%, with higher prevalence in males. This may often be associated with certain anomalies, such as narrowing of the aorta, ventricular septal defects or obstruction of the left ventricular outflow tract. The prevalence among athletes is still little known, though literature describes how there may be a similar incidence rate as that of the general population[5].

Despite the majority of cases being sporadic, an important family burden has been identified, with a BAV incidence rate between 10 and 17% in first-degree family members[6]. This is why it is important to carry out a good anamnesis and a more thorough study on athletes with BAV family antecedents.

The presence of bicuspid aortic valve does not usually present an obstacle when it comes to initiating sporting activity; however, the impact of high-intensity training on the heart of an athlete is unknown. In theory, the physiological stress of intense and on-going exercise on an abnormal aortic valve may cause its early deterioration and dilatation of the ascending aorta[7]. This is why early identification in these athletes may help in the follow-up and prevention of adverse consequences that may arise from intense training by these carriers.

This is particularly important if we consider that the majority of these athletes have absolutely no symptoms whatsoever, with the first symptoms emerging after 40 years of age. In most countries, the pre-participation screening protocol carried out includes an appropriate anamnesis and complete physical examination, including cardiac auscultation.

However, if the BAV is not associated to stenosis or heart failure, its diagnosis via auscultation is highly unlikely. This is why performing a transthoracic echocardiogram (or transesophageal echocardiogram, which is more sensitive) is a fundamental tool in detecting the presence of BAV, however, it is not carried out routinely.

Table 1. Classification of sports depending on cardiovascular needs (based on combined static and dynamic components) adapted to sports carried out in Spain.

Sports	Low dynamic	Moderately dynamic	Highly dynamic
Low static	Billiards, Bowling, Golf, Throwing	Baseball, Softball, Table tennis, Tennis (doubles), Volleyball	Badminton, Cross-country skiing (classic), Grass Hockey[a], Orienteering, Running, Athletics (long-distance), Football[a], Squash Tennis
Moderately static	Archery, Motor racing[a,b] Diving[a,b], Horse riding[a,b], Motorcycling[a,b]	Fencing, Athletics (jumping), Figure skating[a], American Football[a] Rugby[a], Athletics (speed) Synchronised swimming[b], Surfing[a,b]	Basketballa, Ice Hockey[a], Cross-country Skiing (skating), Athletics (middle-distance) Swimming, Handball
Highly static	Athletics (throwing) Gymnastics[a,b], Karate/Judo[a] Sailing, Rock climbing[a,b] Water skiing[a,b], Weightlifting[a,b] Windsurfing[a,b]	Bodybuilding[a,b] Wrestling[a]	Boxing[a] Canoeing, Cycling[a,b], Athletics (Decathlon), Speed skating, Rowing

[a]: danger of corporal collision; [b]: increased risk in the event of a syncope.

The most frequent complications associated with BAV are: aortic stenosis (15-51%), heart failure (7%), endocarditis (5%) and aortic dissection (4-5%)[8].

The bicuspid aortic valve is also considered to be an independent cause of aortic dilatation. Given the high prevalence of BAV among the general population, this represents the most common aetiology of aortic dilatation, and eventually dissection. In cases of BAV with coexisting ascending aorta dilatation, intense physical exercise may lead to a significant risk of aortic dissection or rupture, especially during isometric exercises. In fact, during phases of high-intensity competition, it may even induce major arterial hypertension, which stimulates the appearance of these complications. The presence of BAV significantly increases the risk of dissection, especially among young and previously asymptomatic patients.

This risk increases considerably when the aortic diameter is greater than 50mm and in the presence of accompanying arterial hypertension (AHT).

According to the Clinical Practice Guides of the Spanish Cardiology Society regarding physical activity performed by the heart disease patient, in the evolution of this disease an ECG must be performed along with an echocardiogram to establish the severity of the valvular heart disease. In cases of aortic stenosis, a distinction is made between the mild forms (< 20 mmHg), moderate forms (21-49 mmHg) and severe forms (>50 mmHg), based on the peak systolic gradient measured with an echocardiographic Doppler[9].

Likewise, anyone with BAV wishing to practise competitive sports is recommended to carry out a stress test before starting the physical activity, and a Holter monitor to improve the risk stratification and to suitably establish the intensity of the exercise.

Prescribing physical and sporting activity in aortic stenosis[10]:

- Mild: no limitations on physical activity or competitive sports.
- Moderate:
  - Physical activity: mild intensity: allowed. Intense: prohibited.
  - Competitive sports: Mild intensity: allowed with low static charge and low or moderate dynamic charge and with moderate static charge and low dynamic charge. Intense: prohibited.
- Severe:
  - Physical activity: mild intensity: allowed. Intense: prohibited.
  - Competitive sports: prohibited.

Prescribing physical and sporting activity in aortic insufficiency[10]:

- Mild: no limitations on physical activity or competitive sports.
- Mild-moderate: with normal ECG without Left Ventricular (LV) dilatation:
  - Physical activity: no limitations.
  - Competitive sports: mild intensity: allowed. Intense: allowed if the stress test is normal.
- Moderate-serious, with alterations in the ECG and LV dilatation:
  - Physical activity: mild intensity: allowed. Intense: prohibited.
  - Competitive sports: prohibited.

The cardiac stress test is recommended to asymptomatic patients that wish to practise competitive sports. These athletes, despite having a very low sudden death incidence rate, may suffer from this especially in very serious cases in relation to exercise-induced ischemia; in these cases performing the stress test is clearly contraindicated. Patients that have undergone effective valvular repair, with no residual gradient or with a minimum residual gradient, should wait for at least 3 months after the percutaneous valvuloplasty or 6 months after surgery before returning to sporting activities.

The presence of at least one of the following criteria may be considered to be associated with a greater cardiovascular risk in patients with bicuspid aortic valve[11]:

- Aortic systolic gradient > 20mmHg.
- Aortic root dilatation (diameter > 40mm).
- Moderate-severe heart failure.
- LV dilatation (LVD 60 mm) connected to symptoms (syncopes, prolonged palpitations) or arrhythmias, such as supraventricular or ventricular tachycardia documented on the ECG or electrocardiographic monitoring.

In a study performed by the National Institute of Sports Medicine in Rome in 2006[12], 8000 patients were assessed, among which 81 had BAV. In this study, athletes with none of the previously described criteria were classified in a low-risk group, and were considered apt for competition. However, the patients that presented at least one risk factor were discouraged from participating in training and competition. In the 13-year follow-up of the patients at low risk (the high-risk patients were excluded), 12% presented some kind of complication (aortic root dilatation, increase in gradient, arrhythmic events, etc.) and were moved into the high-risk group. In conclusion, this study reveals the importance of undergoing strict follow-up of these patients, as well as their high probability of presenting an unfavourable clinical evolution.

The clinical and prognostic course of athletes with bicuspid aortic valve still remains uncertain. However, numerous studies suggest that with competing athletes, the BAV should not simply be considered to be a variation of normal, rather a possible association with a worsening of the valve morphology and functioning, as well as clinical deterioration.

Given the high prevalence of aortic valvular pathology and other potential risks that this entails in athletes subject to high levels of sporting activity, it is recommended to expand the pre-participation cardiovascular study to identify and adjust the stratification in athletes with BAV. It can be confirmed that practising high-level sport is not in itself the cause behind the deterioration of this disease; however, some results have revealed that long-term training may be associated in some individuals with a progressive worsening of valvulopathy, aortic root dilatation and the appearance of clinical symptoms.

Left-right shunts

Atrial Septal Defect (ASD) and Ventricular septal defect (VSD)

The most common defects with left-right shunts are ASD and VSD, and patent ductus arteriosus. In these diseases, oxygenated arterial

blood passes to the venous side, causing an overload in volume of the cavities through which the excess blood passes, including the lungs, also potentially causing major pulmonary hypertension (PHT).

Its detection requires clinical diagnosis, an x-ray of the thorax, electrocardiogram and echocardiogram. It may be considered to be both lung pressure as well as a shunt (pulmonary/systemic flow relation (Qp/Qs)) via the echocardiogram; however, these measures are best established via catheterisation, if considered necessary.

With regards to atrial septal defect (ASD), the majority of patients are asymptomatic till adulthood. However, a large proportion of them develop symptoms from the age of forty: reduced functional capacity, dyspnoea related to efforts, palpitations, and right cardiac failure… Occasionally, cases can be seen of secondary pulmonary hypertension with overloading in right cavities, especially in older patients, or in shunts that have gone unnoticed for a long time.

In terms of performing exercise, no clear restrictions have been established for asymptomatic patients before or after intervention, always in the event that they do not have pulmonary hypertension, major arrhythmias or right ventricular dysfunction. In patients with severe pulmonary hypertension or Eisenmenger Syndrome (inversion of the shunt), physical activity or recreational sports should be limited to those of medium-low intensity[13].

In ventricular septal defects, there are similar restrictions to those previously mentioned for atrium defects. There are no limitations for patients following the closure of this shunt with restricting VSD of small size, without PHT, arrhythmias or right ventricular dysfunction. Patients with PHT may limit themselves to low-moderate intensity activities or sports for non-competitive purposes.

Physical and sporting activity in left-right shunt defects

- Small or moderate (Qp/Qs<2) with normal pulmonary pressure:
  - Physical activity: no limitations.
  - Competitive sports: no limitations.
- Large (Qp/Qs >2): with normal or slightly elevated pulmonary pressure:
  - Physical activity: no limitations.
  - Competitive sports: Mild intensity: low-charge static and dynamic sports allowed. Intense: prohibited.
- With pulmonary hypertension:
  - Physical activity: Mild intensity: allowed. Intense: prohibited.
  - Competitive sports: Mild intensity: allowed if the stress test is normal. Intense: prohibited.
  - Both prohibited if it is Eisenmenger Syndrome.

Following the correction of inter-atrial and inter-ventricular septal defects (this may be performed using conventional surgery or percutaneously), specially targeted recommendations should be followed if there is residual defect, as well as pulmonary hypertension. Furthermore, it should be remembered there are other possible limiting factors to exercise, such as the appearance of arrhythmias and ventricular dysfunction. For these patients the recommendation is to avoid sporting activity for at least 6 months following the intervention[14].

In general, for mild residual defects, no limitations are set for physical exercise or competitive sport. However, with moderate-serious defects, intensive sport is contraindicated, though low-intensity sport can be performed. Moreover, it is recommendable to perform an ergometer on patients that still display signs of pulmonary arterial hypertension, symptomatic arrhythmias or ventricular dysfunction after 6 months following intervention.

Patent ductus arteriosus

Patent ductus arteriosus (PDA) is defined as the persistent communication between the pulmonary artery (predominantly the left pulmonary artery proximally) and the descending aorta (just distal to the outflow left subclavian artery). On numerous occasions, it may be associated with other congenital heart diseases; however, in adulthood the presence of PDA is usually an isolated cardiac finding.

The majority of patients with this pathology are usually completely asymptomatic, with small-sized ductus that does not affect the cavities, or significant pulmonary hypertension. However, on some occasions there may be secondary impacts to this shunt: overloading of the left cavities, with dilatation or accompanying ventricular dysfunction, pulmonary hypertension, Eisenmenger physiology... There is also a potential risk of endarteritis, though this seems to be rare. The formation of aneurisms in the ductus level is a very rare complication, and may compress the left coronary tree. The suspected diagnosis is usually performed with an echocardiograph, though a CAT scan or MR is recommended to establish a confirmed diagnosis and to quantify and more precisely define the anatomy of the ductus.

In terms of performing sporting activity, there are no restrictions in place for asymptomatic patients either before or after intervention; the only limitation is to practise low-intensity sports when pulmonary hypertension is detected[15]..

Permeable ovale foramen

The persistence of a permeable ovale foramen (POF) in adults is a common condition, with a prevalence of approximately 25% of the general population. In the majority of cases, the presence of POF is a casual finding with no clinical repercussions; however, the link between POF and clinical conditions has been frequently described in POF association literature, such as embolic cerebrovascular accidents, platypnea-orthodeoxia syndrome, gas embolism, or migraines.

In terms of athletes, a special mention should be given to divers, among whom major complications may arise such as gas embolism. Decompression syndrome is a term used to name the gas embolism produced, especially in divers and pilots, due to a rapid drop in atmospheric pressure, which leads to a reduction in the solubility of gases and the possible release of these gas bubbles into the blood stream[16].

Diagnosis of POF is made by performing a transthoracic echocardiogram with/without bubbles, and a transesophageal echocardiogram.

Regarding the treatment of choice for POF, today there is still no clear definition; there are no specific recommendations for athletes as opposed to the general public carrying POF. However, there are some specific clinical situations, such as recurring cryptogenic ictus in young patients (<55 years) with evidence of venous thrombosis or high-risk anatomies, in which the percutaneous closure may be justified[17]. Likewise, the percutaneous approach has become the treatment of choice among patients with platypnea-orthodeoxia and requires the closure of the atrial septal defect18. In these patients, the percutaneous shunt closure has proven to be safe and effective, with a success rate of around 86-100%[19].

Coarctation of the aorta

Coarctation of the aorta is one of the most common congenital heart diseases, making up between 5 and 8% of congenital heart defects. It can occasionally be treated as an isolated defect, though in over 60% of cases patients present some kind of accompanying pathology: bicuspid aortic valve (30-40%), sub-valvular or aortic supra-valvular stenosis, complex congenital heart diseases or Turner Syndrome.

Diagnosis of coarctation is defined as the narrowing of the intraluminal calibre that causes an obstruction to the aortic flow. In the majority of cases, the coarctation area is usually localised in the descending thoracic aorta distal to the outflow left subclavian artery with different degrees of extension, and in some cases is associated with hypoplasia of the aortic arch. Diagnosis of coarctation should be suspected in young patients with refractory hypertension and weakness or an absence of pulses to the lower limbs.

To diagnose and follow up coarctation of the aorta, a good anamnesis is needed, such as an electrocardiogram, an echocardiogram, and occasionally, an ergometer. However, performing a CAT or MR scan of the thorax is a key tool in establishing a definitive diagnosis for these types of diseases. When the coarctation is mild, all kinds of physical or sporting activities can be carried out. However, when the coarctation is major or the stress test reveals the presence of serious systematic arterial hypertension in connection with the exercise, only low static and dynamic component sports can be carried out[20].

Anatomical correction may be performed using conventional surgery or percutaneously with dilatation and posterior implant of a stent at the level of the coarctation. Following intervention, sporting activity will depend on the presence of residual gradient. It is recommendable to wait approximately 6 months following the intervention before starting sporting activity. It is recommendable to avoid sports with high static charge and contact sports for the first year after surgery. After this first year, any kind of sport is permitted except for weightlifting. However, if an aneurismatic area or aortic dilatation persists as a residual injury, exercise will be restricted to low static and dynamic component activity.

Physical and sporting activity in the coarctation of the aorta

- Base gradient between 10 and 20 mmHg:
  - Physical activity: no limitations.
  - Competitive sports: mild intensity: allowed. Intense: allowed if the ergometer is normal.
- Base gradient higher than 20 mmHg:
  - Physical activity: mild: allowed. Intense: allowed with low static and dynamic component.
  - Competitive sports: allowed with low static and dynamic component.
- Base gradient higher than 50 mmHg:
  - Physical activity: mild intensity: allowed. Intense: prohibited.
  - Competitive sports: prohibited.

To conclude, patients without residual obstruction that are normotensive both resting and during exercise, may carry out unrestricted sporting activity, except for sports with a high static component. On the other hand, patients with AHT, significant residual obstruction or other complications, should avoid high-intensity isometric exercises in proportion with the severity of their pathology[10].

Marfan Syndrome

Marfan syndrome is a dominant autosomal hereditary disease that generates an alteration of connective tissue, causing an impact on various levels: cardiovascular, ocular, muscular-skeletal or pulmonary. It has an estimated prevalence of around 1 in each 5,000 live new-borns[21].

Cardiovascular manifestations pose a particular interest because they entail a high risk of sudden death in individuals with this condition. The most frequent manifestations are dilatation of the aortic root, mitral valvular prolapse, coarctation of the aorta or ASD. On the other hand, the worst finding is aneurism or aortic dissection. Approximately 60% of patients with Marfan syndrome have aortic root dilatation, predominantly males. In general, medical treatment for these patients is based on avoiding aortic root dilatation and dissection by aiming to reduce arterial pressure and cardiac inotropes. Treatment with beta-blockers[22] has been widely recommended in numerous studies with the aim of avoiding the progressive dilatation of the aorta. Moreover, recent studies have further investigated the use of ARB II (angiotensin receptor blockers II) to inhibit TGF-B signals that are involved in the dilatation of the aortic root in Marfan syndrome.

Currently, the criteria reviewed by Ghent are used to diagnose Marfan syndrome (Table 2)[24]. The main tools used to assess the cardiovascular impact are performing transthoracic echocardiogram and the CAT/MR scan of the thorax. Stable patients require annual check-ups with an echocardiogram. The use of a CAT/MR scan is recommended every 5 years if there is no aortic dilatation; in the case of aneurism or aortic dilatation, these image tests should be repeated annually[25].

Given the higher level of vulnerability patients with this disease face, limits for intervention and surgery are different to those of the general population. With regards to surgical repair, according to 2014 European guidelines regarding the diagnosis and treatment

Table 2. Diagnostic criteria of the Gante nosology for diagnosing Marfan syndrome

Organ/System	Requisites for classifying the major criteria	Requisites for the impact on the organ/system
Skeletal	At least four of the following: 1. Pectus carinatum 2. Pectus excavatum requiring surgery 3. Ratio between segments reduced or ratio size and height elevated (<1.05) 4. Thumb and wrist signs positive 5. Scoliosis (20º) or spondylolisthesis 6. Reduced elbow extension (<170º) 7. Medial displacement of the internal malleoli causing flat foot. 8. Acetabular protrusion	At least two findings for major criteria, or one from this list and two from the following minor criteria: 1. Moderately severe pectus excavatum 2. Joint hypermobility 3. Palate with pronounced arch or dental crowding 4. Characterised facial appearance (dolichocephaly, hypoplasia malar, enophtalmos, retrognatia, low palpebral fissure)
Ocular	Ectopia *lentis*	At least two of the following minor criteria: 1. Abnormally flattened cornea 2 Increase of axial length of the eyeball 3. Hypoplasia of the iris or Ciliary muscle, causing reduced miosis
Cardiovascular	At least one of the following: 1. Dilatation of the ascending aorta with or without regurgitation, affecting the Valsava sinuses 2. Dissection of the ascending aorta	At least one of the following minor criteria: 1. Prolapse of the mitral valve, with or without regurgitation 2. Dilatation of the pulmonary artery, in absence of stenosis or another cause in individuals under 40 years 3. Dilatation or dissection of the descending or abdominal thoracic in individuals under 50 years
Pulmonary	None	At least one of the following minor criteria: 1. Spontaneous pneumothorax 2. Apical bullous
Teguments	None	At least one of the following minor criteria: 1. Marked stretch marks in the absence of important weight variations, pregnancy or repeated stress 2. Recurring or incisional hernia
Hard	Lumbosacral dural ectasia	None

To diagnose Marfan syndrome in patients without family antecedents of the disease, two organs/systems should be involved that bring together the criteria and at least an impact on a third organ/system. In patients with a family history of Marfan syndrome, only one major criterion is required, with data that suggests an impact on a second system (De Paepe, *et al*).

of the pathology of the aorta[26], surgery is recommended to patients with Marfan syndrome and with a maximum aortic diameter that is greater or equal to 50 mm, or 45mm if there are risk factors, such as family antecedents of dissection, growth > 3mm/year (in various exams using the same technique and with confirmation in another), serious aortic regurgitation or the intention to become pregnant. Patients with Marfanoid manifestations through disease of the connective tissue, without complete Marfan criteria, must be treated as Marfan patients.

With regards to the participation of athletes with Marfan syndrome, the most important aspect to consider is the early detection of athletes with this condition. Athletes with a marfanoid phenotype or family antecedents should be examined immediately to rule out this pathology, prior to initiating sporting activity. More aggressive screening strategies are recommended for sports that typically involve athletes with this specific profile and with certain marfanoid habit, such as basketball and volleyball. The prevalence of Marfan syndrome is usually higher within this demographic, which is why screening these high-risk groups may improve the early detection of this pathology and avoid the progression of this disease among these athletes[27].

Today, significant restrictions are in place regarding physical activity performed by Marfan syndrome patients. For example, activities that involve collisions or intensive contact sports are considered to be particularly high risk for these individuals, given the cardiovascular and skeletal susceptibility that these individuals present. Likewise, athletes carrying this syndrome should not carry out high-risk activities, or at least they should aim to minimise their exposure to these kinds of activities.

In regard to current recommendations, isometric exercises should be prohibited for athletes with Marfan syndrome, given the damaging haemodynamic effects of straining the aortic wall, which produce a significant increase in the risk of aortic dissection or rupture. According to that established in the 36th Bethseda Conference[28], these athletes should only participate in low-intensity activities and with a low dynamic and static component (such as hiking, bowling, golf, etc.).

Congenital coronary anomalies

Congenital coronary anomalies present major implications for athletes. These anomalies have been described in around 1% of the general population[29]. This incidence rate is usually underestimated, given that the majority of cases proceed asymptomatically and the condition can be undetectable. Furthermore, it has not yet been clarified whether the rate of coronary anomalies in athletes is different to that of the general population; however, currently there are numerous studies underway based on screening and autopsies, which aim to confirm whether or not prevalence among athletes is greater than among the general population.

In a recent review of coronary anomalies, researchers classified them into four different groups:

- Anomalous source and course,
- Anomalous intrinsic coronary arterial anatomy,
- Anomalous coronary termination, and
- Anomalous anastomotic vessels[30].

Numerous image methods have been used to diagnose coronary anomalies, including conventional coronary angiography, transthoracic echocardiogram, transesophageal, multi-cut CAT scan and thorax MR. This group of diseases has frequently been diagnosed using a coronary catheterisation; however, despite the efficiency of this technique when it comes to detecting the source and final course of coronary arteries; these are limited as they do not give a view of the spatial orientation of the coronary arteries in relation to other intra-thoracic structures involved in coronary arterial anomalies.

The importance of diagnosing this pathology in athletes is particularly based on the significant increase in the risk of sudden death resulting from this condition. In the United Kingdom, this constitutes the second most frequent cause of sudden death in athletes[31]. Occasionally, sudden death can be the first manifestation of the process in athletes, with no previous episodes of angor ever appearing with strain or other symptomatology.

The anomalous source of the left coronary artery in the right Valsalva sinus and the anomalous source of the right coronary artery in the left coronary sinus are the most frequent coronary anomalies associated with the risk of sudden death in athletes. Despite the lower incidence rate, the presence of one coronary artery has also been associated with an increased risk of sudden death during exercise.

The general mechanism of death in these athletes is produced through coronary ischemia and ventricular arrhythmias. However, the precise mechanism of this pathology is still unknown and may be different depending on the structural anomaly.

As we have previously mentioned, the main concern in the diagnosis and follow-up of coronary anomalies is fundamentally the high risk of sudden death. In accordance with recommendations from the 26th Bethseda Conference, detection of these abnormalities should entail the exclusion from participation in any competitive sport, although surgical treatment (if possible) significantly reduces the risk of sudden death. Likewise, participation in sporting activity could be allowed from six months following surgery, always as long as a maximal stress test has been carried out beforehand with no evidence of induction of ischemia[32].

To conclude, the basic recommendation for athletes with coronary anomalies is non-participation in competitive sports, unless reparation or surgical intervention is performed. In the most common types of coronary anomalies described above, excision and reintroduction is usually performed, which involves repositioning the coronary arteries in the correct Valsalva sinuses. In cases where the coronary anomalies proceed with an intramural trajectory between the pulmonary artery and the aorta, an intracoronary stent implantation may be performed to treat this anomaly[33]. Moreover, a coronary artery bypass implant can be performed, though given the associated risks with these procedures, these are not routinely recommended for young active athletes.

Currently, preliminary studies reveal promising evidence that these techniques are effective. Literature on the subject describes an important series of cases involving post-operative patients demonstrating the absence of signs of ischemia during exercise based on symptoms, a basal electrocardiogram and a stress test, in a 2-year follow-up after surgical intervention. Likewise, today there is a drive to confirm the effectiveness of these procedures in protecting athletes from presenting a high risk of sudden death in a long-term follow-up.

Cyanotic congenital heart diseases

In general, patients with cyanotic congenital heart disease present diverse degrees of arterial desaturation and intolerance to exercise, making it highly unlikely they will be able to participate in any kind of sporting activity. However, in cases where there is capacity to carry out physical exercise, they will only be recommended sports with a low static and dynamic charge, taking into account that with strain the hypoxemia will increase even further in comparison to basal conditions. In alleviated cases, in which there is an on-going increased pulmonary flow, there is frequently a certain degree of arterial desaturation, meaning that only low static and dynamic component exercises can be performed, as long as the patient is asymptomatic and the hypoxemia is mild. In dubious cases, especially when it comes to light sporting activities, it would be recommendable to perform a stress test to analyse desaturation with the exercise. Below is a description of recommendations for sporting activity in some of the most frequent cyanotic congenital heart diseases.

Tetralogy of Fallot

Tetralogy of Fallot is the most common congenital heart disease after one year of life, with an approximate incidence rate at around 10% of these heart diseases. Diagnosis of this pathology consists in the association of these four elements: infundibula pulmonary stenosis (obstruction of the right ventricle outflow tract), ventricular septal defect, overriding ascending aorta (overriding aorta) and right ventricular hypertrophy. On occasions, these patients may also present associated atrial septal defect.

The majority of these patients require surgical treatment in their first years of life. This treatment has two tracks: one palliative and the other corrective. The first consists in performing a Blalock Taussig fistula (systemic-pulmonary), which connects the right subclavian artery with the right pulmonary artery and thus improves oxygenation. Corrective treatment is more used more nowadays and is principally based on unblocking the right ventricle outflow tract, closing the ventricular septal defect with Dacron patches and performing a correct alignment of the aorta.

In principle there are no restrictions on sporting activities in asymptomatic patients with good functional capacity. However, patients at high risk of arrhythmias or sudden death (numerous cases of sudden death have been described in patients with this pathology), or that present other complications such as bi-ventricular dysfunction, major residual pulmonary failure, or the advanced pathology of the ascending aorta, should limit their sporting activity to exercises with a low static and dynamic component, and should avoid performing isometric exercises34. Prior to this, the patient needs examining with an electrocardiogram, echocardiogram, Holter monitor and stress test.

Transposition of the great arteries

LTransposition of the great arteries comprises 5% of the cyanotic congenital heart diseases and is characterised by a ventricular-arterial discordance: the right ventricle connects to the aorta, whilst the left ventricle connects to the pulmonary artery. When there is also atrial-ventricular discordance, it becomes a transposition of the great arteries corrected congenitally. If there are no other associated cardiac malformations, it is a simple transposition; whereas complex transpositions often present different types of associated malformations: ventricular septal defect, obstruction of the outflow tract, coarctation of the aorta… The majority of adults are seen after undergoing surgical interventions in their childhood.

With regards to carrying out sporting activity, patients with an excellent haemodynamic capacity may perform regular activity, aiming to avoid extremely intense activities. However, patients that present worse functional capacity should limit their sporting activity and avoid contact and high-charge sports; they should be advised to perform regular low-mid intensity physical activity. Carriers of this pathology with symptoms or with a history of previous arrhythmias should be thoroughly assessed individually, given the high risk of exercise-induced arrhythmias. To do so, these patients are recommended to undergo a stress test, a Holter monitor and a transthoracic echocardiogram[35].

In the case of physiologically corrected transposition of the great arteries using Mustard or Senning surgical techniques, given that the morphologically right systemic ventricle develops long-term major ventricular dysfunction, low static and dynamic component sports are recommended. In any case, each case should be considered independently depending on the degree of systemic ventricle dysfunction.

The anatomical repair of the D-transposition of the great arteries (Jatene operation) has become the procedure of choice for surgical correction. Given that with this surgery normal anatomy is recovered, in principle, patients can perform all kinds of sports, as long as they present an optimal haemodynamic situation and that there are no residual injuries where the anastomosis took place[36]. Likewise, due to the diversity of the anatomy of the coronary arteries in the TGA and the need to reposition them in the Jatene operation, the appearance of a coronary ischemia is always assessed in these patients. However, it is recommendable to abstain from performing high static charge exercises, which may significantly increase arterial strain.

Patients following Fontan surgery

Patients with functionally uni-ventricular hearts undergo a series of interventions that aim to passively direct the flow of the systemic veins to the pulmonary circulation, leaving the one ventricle to drive systemic circulation, which is known as a Fontan situation or physiology.

Following a Fontan correction type for a uni-ventricular heart or for complex heart diseases, it is common for a certain degree of intolerance to exercise to persist, such as the appearance of early or late arrhythmias. Only patients with normal ventricular function, an absence of hypoxemia, absence of arrhythmias and a good tolerance of exercise proven using a stress test, may perform low static and dynamic charge exercises, with others not recommended. In general, these patients should only be recommended to perform recreational-purpose sports[37].

Physical and sporting activity in cyanotic congenital heart diseases

- Not corrected:
  - Competitive sports: prohibited.
  - Physical activity: Mild intensity: allowed with low static and dynamic component. Intensive: prohibited.
- Corrected:
  - Competitive sports: Mild intensity: allowed if the ergometer is normal and the patient is asymptomatic. Intensive: prohibited.
  - Physical activity: Mild intensity: allowed if the patient is asymptomatic and the hypoxemia mild. Intensive: prohibited.

Ebstein's anomaly

Ebstein's anomaly is a rare congenital heart disease, characterised by a malformation and apical displacement of the different tricuspid valve leaflets. The apical displacement of the tricuspid valve means that the right part of the heart is basically made up of the right atrium, an atrialised portion of the right ventricle and the residual region of the right ventricle, which generally functions normally. It is common to find that these patients also suffer from tricuspid insufficiency, which can occasionally be major.

The most common associations usually displayed in patients with this anomaly are: atrial septal defect (especially ostium secundum type ASD and permeable ovale foramen) and the presence of accessory pathways, given that many of these patients present Wolf Parkinson White syndrome[38].

In terms of practising sporting activity, patients carrying this malformation with no residual anomalies, may perform unrestricted physical activity, except for high static charge sports at a competitive level. However, patients experiencing heart complications, such as moderate-severe tricuspid insufficiency, shunt, right ventricular dysfunction or arrhythmias, should avoid intensive isometric exercises, in proportion to the severity of their pathologies.

Conclusion

It has been proven that practising exercise is beneficial in health adults as well as in patients with acquired cardiovascular diseases (coronary disease, heart failure, etc.). Advances in dealing with congenital heart diseases have allowed for an improvement in the survival rate and quality of life of these patients. This has meant the functional development of this demographic has become similar to that of the general population. From this, new issues arise, such as follow-up and recommendations regarding physical and sporting activity. Despite literature regarding sporting activity recommendations being limited, and with few studies performed on reduced groups of patients with short-term follow-up, the results are consistent in revealing that the benefits of exercise are applicable to people with congenital heart diseases.

At the start of the paediatric cardiology development, physical activity is, for the vast part of these patients, limited to performing recreational-type sporting activity. Follow-up of operated patients that also present a positive evolution has opened up new expectations regarding recommended activity. To this, we must add the fact that heart rehabilitation has acquired a very important role over recent years, and many studies have begun to include patients with congenital heart diseases to establish the benefits of physical exercise on this demographic.

It is important to highlight that the nature and intensity of physical training should be adapted to each individual in accordance with the type of congenital heart disease and the surgical correction, taking into account the clinical profile of each patient, including residual haemodynamic injuries, ventricular function, associated arrhythmias, etc. Additional research is required to identify the optimum exercise regimes for achieving the desired results in patients with congenital heart disease, and to establish strategies to effectively promote exercise-training recommendations as an integral component within a healthy lifestyle.

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