

Editorial

Percutaneous Intervention in Adult Congenital Heart Disease

Intervencionismo en las cardiopatías congénitas del adulto

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The diagnosis and successful management of congenital heart disease is one of the major triumphs of medicine and cardiovascular surgery in the 20th century. The publication of the book entitled *"Congenital Malformations of the Heart"*, by Dr. H. Taussig,¹ opened the door to an understanding of congenital heart disease. The same period witnessed the first milestones in the treatment of these conditions: John H. Gibbon performed the first open heart surgery in a patient with an atrial septal defect (ASD); Robert Gross described the ligation of patent ductus arteriosus for the first time; Clarence Crafoord, the resection of aortic coarctation; and Taussig herself, together with Blalock, published their experience with the palliative shunt to treat cyanotic heart disease. The development and incorporation of new surgical techniques, together with improvements in the medical care of these patients, have significantly reduced the mortality rate, from 20% in the 1970s to 5% at the present time. Consequently, the number of adults with congenital heart disease has increased rapidly, nearing 1 million in North America^{2,3} and estimated to be roughly 100 000 in Spain.⁴ Approximately 60% of cases of congenital heart disease are diagnosed in infants aged <1 year, 30% in children and 10% in adults. However, given the increase in survival, there are more adults than children with congenital lesions and most require lifelong follow-up and care. Moreover, many of these patients will need to undergo additional interventions during follow-up and, given that this population has required multiple previous surgical procedures, interest in the field of interventional cardiology is becoming especially intense.

Over the last 2 decades, the approach to the diagnosis and treatment of adults with congenital heart disease has been totally transformed. For many years, the place for diagnosis was the catheterization laboratory, whereas the site of treatment was the operating room. Now, the place for diagnosis is the imaging laboratory (echocardiography, magnetic resonance image [MRI], computed tomography [CT], etc.), and the first invasive treatment is increasingly performed in the cardiac catheterization and electrophysiology laboratory. The group of interventions carried out in the cardiac catheterization laboratory can be summarized as: a) valve disease; b) septal defects; c) vascular obstructions; d) fistulas and collateral vessels, and e) residual lesions and surgical wound dehiscence. In general,

the approach could be reduced to 2 groups of interventions: a) those requiring the correction of obstruction of a vascular conduit or a valve, which are treated with dilatation procedures using balloons, stents, or valved stents, and b) those requiring the occlusion of an abnormal communication between 2 cardiac chambers or vascular conduits, which are treated using closure devices or coils (Fig. 1).

ORGANIZATION OF THE LABORATORY AND THE CATHETERIZATION TEAM

Pediatric interventional cardiology has gradually incorporated increasingly complex procedures that require a multidisciplinary approach. On the one hand, this calls for the support of a group of cardiologists and pediatric surgeons to adequately evaluate each individual and discuss patients before referring them to the catheterization laboratory. On the other hand, it is essential to include a group of imaging specialists with extensive experience of congenital heart disease and training in the performance of CT, MRI, and 3-dimensional echocardiography. An adequate understanding of the clinical indication and a high-quality imaging study will facilitate the performance of the intervention. Finally, there must be an interventional cardiology team capable of performing hybrid procedures jointly, which ideally should include a pediatric interventional cardiologist, an adult interventional cardiologist, and a cardiac surgeon who is an expert in congenital heart disease.

To be able to carry out all types of procedures safely, the recommended setting is a hybrid operating room of approximately 80 m² that enables the simultaneous presence of anesthesia equipment (respirator, monitors, etc.), imaging equipment (echocardiograph), perfusionists (heart-lung machine), electrophysiology system (ablation and 3-dimensional mapping systems), and tables for surgical and catheterization equipment. An advantage of the biplane fluoroscopy system is that it allows the simultaneous acquisition of perpendicular images that facilitate anatomical examination, catheter placement, and subsequent 3-dimensional reconstruction of the images. Moreover, the new systems enable a rotation of –135° to 135°, while maintaining the head region accessible to the anesthetists and echocardiographers. The operating table can be tilted, placing the patient in the Trendelenburg position, and rotated laterally 20° to 30°. This facilitates a possible surgical approach. Integration of all the available information is aided by the presence of 6 flat panel

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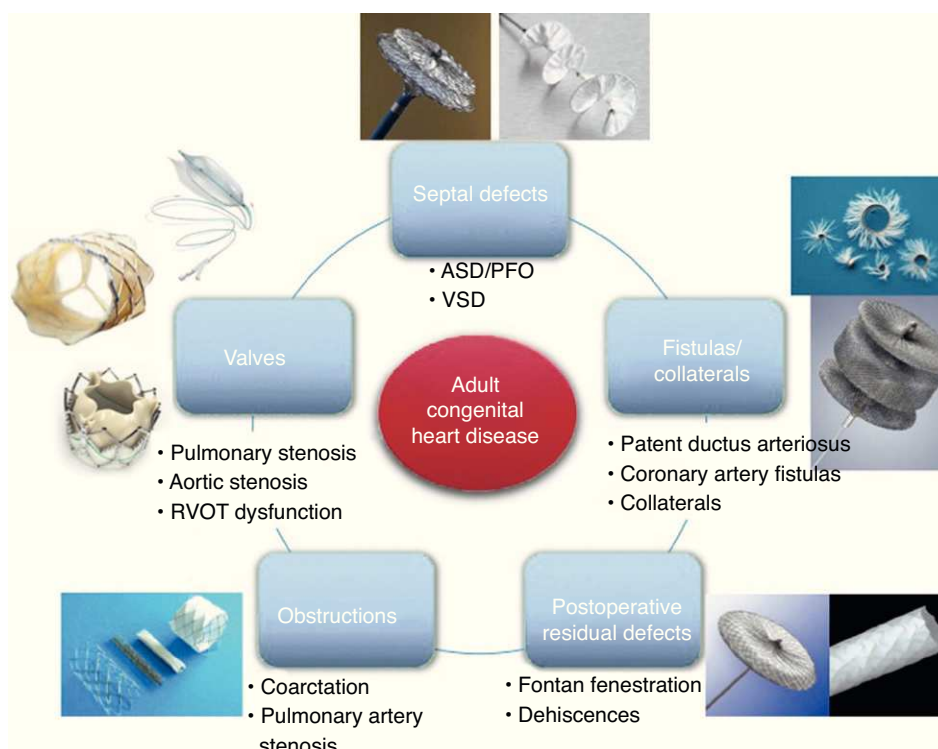


Figure 1. Illustrations of the different types of interventional procedures used in adult congenital heart disease, grouped according to septal defects, valve diseases, obstructions, postoperative residual defects, and fistulas/collaterals. Each group is accompanied by images of the device usually employed to treat the condition. ASD, atrial septal defect; PFO, patent foramen ovale; RVOT, right ventricular outflow tract; VSD, ventricular septal defect. Adapted with permission from Palacios et al.⁵

displays attached to swivel arms, which allows the integration of the polygraph, images from anteroposterior and lateral fluoroscopy, ultrasound, intravascular ultrasound, and CT/MRI. Software is available that enables the integration of the CT image over the fluoroscopic image, which greatly helps to identify each structure. Finally, the suite should be amply stocked with all the necessary material, including devices, balloons, stents (coated), catheters, and guide wires of every type and size. Rescue devices should always be available in case complications arise: snares of different sizes and types, coated stents of the proper size, electrode catheters, pericardial puncture kits, and even ventricular assistance devices, if deemed necessary.

VALVE DISEASE

Pulmonary Valvuloplasty

Pulmonic stenosis constitutes 7% to 12% of all congenital heart disease. There are 3 valve morphologies: a dome-shaped structure

(the most common type, which appears in isolated form); the dysplastic type (20% of cases, associated with Noonan syndrome); and the unicuspid or bicuspid valve, which is present in tetralogy of Fallot (Fig. 2).

The history of pulmonary valvuloplasty has been well documented, as it is one of the first percutaneous interventions to be successfully performed. This procedure is indicated in cases of dome-shaped valves with a pressure gradient >30 mmHg in symptomatic patients and 50 mmHg in asymptomatic patients⁷ (Table 1). The Valvuloplasty and Angioplasty Congenital Anomalies Registry included 533 patients treated with pulmonary valvuloplasty. Over a mean follow-up of 3 years, the investigators observed that 84% of the patients did not require reintervention. Predictors of a suboptimal result were a small annulus size (typical in dysplastic valves), a high residual pressure gradient (>30 mmHg), and a balloon diameter-to-annulus diameter ratio of <1.2.⁸ The complications rate was low, although a certain degree of pulmonary regurgitation was common. Mild and moderate regurgitation are usually well tolerated, but the long-term clinical impact of

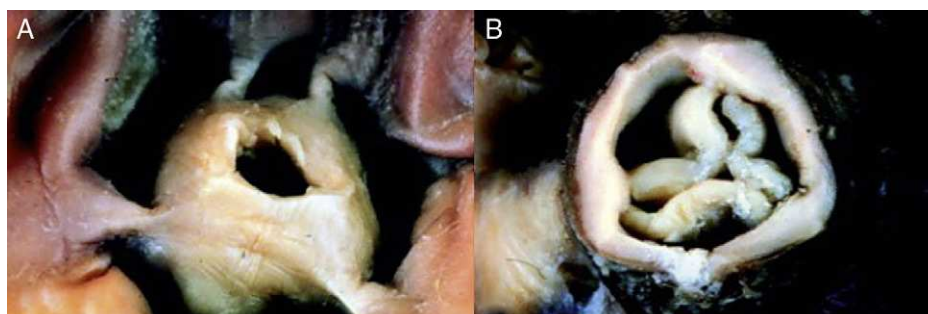


Figure 2. Macroscopic views of a stenotic pulmonary valve. A: dome-shaped valve. B: dysplastic valve. Adapted from Bruce et al.⁶ with the permission of the editor.

Table 1

Indications for Pulmonary Valvuloplasty According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for pulmonary valvuloplasty	
Class I	Dome-shaped valve with peak/mean Doppler pressure gradient of 50 mmHg/30 mmHg in symptomatic patients Dome-shaped valve with peak/mean Doppler pressure gradient of 60 mmHg/40 mmHg in asymptomatic patients
Class IIb	Dysplastic valve with peak/mean Doppler pressure gradient of 50 mmHg/30 mmHg in symptomatic patients Dysplastic valve with peak/mean Doppler pressure gradient of 60 mmHg/40 mmHg in asymptomatic patients
Contraindications for percutaneous closure	
Surgery is recommended in cases of severe pulmonary regurgitation, hypoplastic annulus, or supra-annular or sub-annular stenosis, and in patients with severe valve dysplasia	

severe regurgitation is unknown because of the lack of follow-up data in the published series.

When this disease is first detected in an adult, the symptoms are more marked than in younger patients; there is usually subvalvular obstruction due to the hypertrophy produced by chronic valvular obstruction, the valves show a greater degree of calcification, and considerable poststenotic dilation of the pulmonary artery is very common (since the valve is usually dome-shaped). For valvuloplasty, the balloon's diameter should be 1.25-fold larger than that of the pulmonary annulus; thus, in adults, the simultaneous use of 2 balloons is often required⁹ (Fig. 3). In dysplastic valves, the use of a balloon with a larger diameter, which can be up to 1.5-fold greater than the annulus, is recommended. In pediatric patients, there is a certain amount of positive experience with cutting balloons, but the diameters of those currently available are too small for adults. However, the development of balloons for use in valvuloplasty, such as the AngioSculpt[®] scoring balloon (AngioScore Inc.; Fremont, California, United States), could prove to be the ideal alternative for dysplastic valves.

Transcatheter Prosthetic Pulmonary Valve Replacement

Given the large number of adults with congenital heart disease due to right ventricular outflow tract (RVOT) dysfunction, the availability of catheter-based procedures for valve replacement represents a drastic change in the treatment of these patients. Thus, one of the most important advances in the treatment of heart disease in adults is the development of the Melody valve (Medtronic Inc.; Minneapolis, Minnesota, United States) and its approval for compassionate use by the US Food and Drug Administration (Fig. 4). This device is made of a bovine jugular vein sutured to a Cheatham-Platinum stent. For delivery, it is mounted on a balloon-in-balloon catheter and its implantation requires a 22-Fr catheter. Implantation of the valve does not differ much from placement of a stent in the RVOT, but the absence of compression of the neighboring structures (coronary arteries) must be confirmed and the size, morphology, and distensibility of the conduit (most of which are highly calcified) must be taken into account. In the United States, its use has been approved in circumferential conduits that connect the RVOT to the pulmonary artery and measure 16 mm to 22 mm in diameter, with more than moderate regurgitation or stenosis. However, most of the indications for pulmonary valve replacement apply to patients in which the RVOT has been dilated due to repair of tetralogy of Fallot with a transannular patch. To extend the use of the valve to this group of patients, some authors have proposed placing a stent in the RVOT prior to implanting the valve.^{10–12}

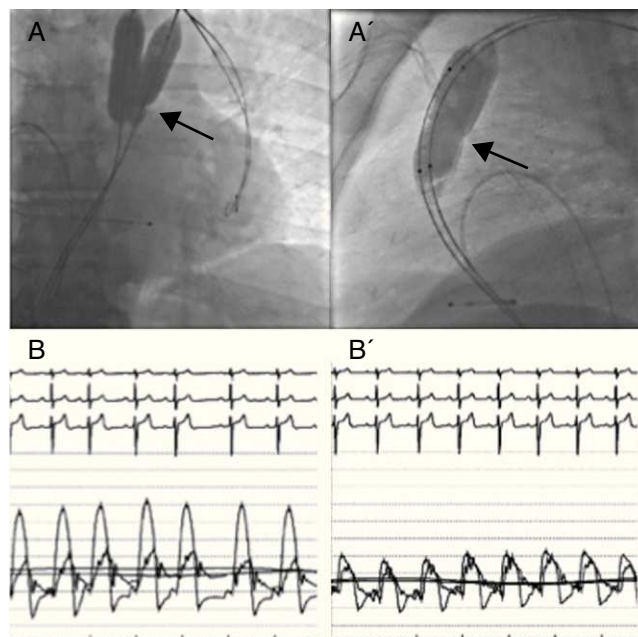


Figure 3. Pulmonary valvuloplasty in an adult, using the double-balloon technique. A: posteroanterior fluoroscopic view of the simultaneous inflation of the 2 balloons at the level of the pulmonary valve. A': lateral view of the simultaneous inflation of the 2 balloons; the arrow indicates the indentation produced in the balloon at the level of the stenosis. B: simultaneous pressure curves in the pulmonary artery and right ventricle prior to valvuloplasty; a peak-to-peak pressure gradient of 60 mmHg is observed. B': simultaneous pressure curves in the pulmonary artery and right ventricle following valvuloplasty; a significant reduction of 10 mmHg is observed in the peak-to-peak pressure gradient.

The medium-term results are good, with adequate valve competence and with no need for reintervention at 2 years in 80% to 90% of the patients^{13–17} (Table 2). Most of the reinterventions are due to obstruction of the tract, often produced by stent fracture. Systemic placement of stents prior to valve implantation seems to have reduced this complication, and the long-term results are eagerly awaited.

The development of Edwards valves of smaller and larger diameters has allowed these valves to be used in the pulmonary position, and the initial results are positive. Moreover, because many of these adult patients have aneurysmal dilation of the RVOT and thus require devices with a larger diameter, the development of the 29-mm valve has facilitated their treatment.

Aortic Valvuloplasty

Congenital malformations of the aortic valve are relatively common, either alone or accompanying more complex cardiac malformations. The spectrum of morphological abnormalities ranges from unicuspid dome-shaped valves or the more common bicuspid valves (present in 1% to 2% of the general population) to dysplastic valves. In adults, there is disease progression, with thickening and calcification of the valves, which is superimposed on the underlying congenital disease. This results in valve stenosis and, less frequently, valve regurgitation.

In the clinical guidelines of the American College of Cardiology/American Heart Association for adult congenital heart disease, a class I indication for aortic valvuloplasty is defined as the presence of a noncalcified valve and a transvalvular pressure gradient of 50 mmHg in symptomatic patients and of 60 mmHg in asymptomatic patients⁷ (Table 3).

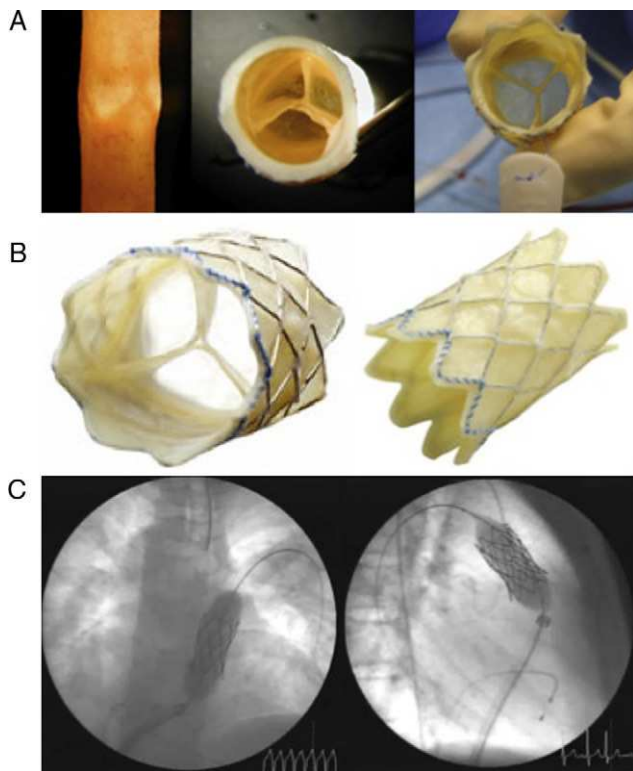


Figure 4. The Melody valve. A: view of the jugular vein from which the valve is obtained and short-axis view of the valve used to construct the valved stent. B: valve sutured over the Cheatham-Platinum stent. C: inflation and expansion of the valve carried out with rapid ventricular stimulation in a patient with tetralogy of Fallot and pulmonary transannular resection.

A balloon-to-annulus ratio of 0.9-1 is recommended. The procedure usually involves retrograde access and crossing the valve from the aorta. There are different types of balloons, but a length of >40 mm is recommended and the procedure should be performed with rapid ventricular pacing using an electrode catheter placed in the right ventricle. Nevertheless, many congenital heart disease patients have already undergone catheterization a number of times and, in these patients, arterial access may not be suitable. Consequently, antegrade venous access and transeptal puncture should be considered. Once the guide wire has crossed the aortic valve, it is snared with a loop in the descending aorta and an arteriovenous loop is created. Valvuloplasty can be performed with routinely used balloons, but the Inoue balloons (Toray Inc.; Texas, United States) that are employed for mitral dilatation have a suitable profile for these procedures.

In terms of the results, the rate of restenosis at 5 years is greater than 50%.¹⁸ Thus, many of these patients require reintervention. It seems that proper patient selection is essential to obtain good results and that the most unfavorable outcomes are obtained in older individuals, those with more calcified valves, and those with previous valve regurgitation. Nevertheless, the development of balloons with a better profile and percutaneous closure techniques for arterial access have reduced the complications associated with this procedure, and several centers are retrieving this technique as a bridge to definitive treatment, whether aortic valve replacement or transcatheter valve implantation.

Transcatheter Aortic Valve Implantation

To date, most of the studies carried out with percutaneous valves, both the Edwards-SAPIEN (Edwards Lifesciences Inc., Irvine, California) and CoreValve (Medtronic, Minneapolis, Minnesota), have included inoperable or generally elderly patients, individuals who are at high surgical risk.^{19,20} Despite the promising initial results, data on the long-term behavior of these valves will not be available for a few more years. In adult congenital heart disease, the experience is very limited and usually involves valves in the pulmonary position.¹⁷ However, positive outcomes have been reported in implantation over biological valves and, thus, this procedure can be considered an alternative in patients who have undergone multiple interventions and are at high surgical risk.

OBSTRUCTIONS

Aortic Coarctation

Adult coarctation of the aorta is usually located in the isthmus, but complex lesions are not uncommon and the morphology can vary widely (Fig. 5). When not treated, this defect is associated with a high rate of comorbidity due to hypertension, early coronary artery disease, heart failure, and stroke.²¹ The standard criteria for the indication for treatment in coarctation is a peak systolic pressure gradient >20 mmHg, although other factors such as the presence of collateral branches, ventricular hypertrophy, systemic hypertension, and other comorbidities should be considered⁷ (Table 4). Angioplasty, with or without stent insertion, is generally accepted as the treatment of choice for patients with recoarctation following surgery or previous stent placement. The percutaneous approach to native coarctation seems to be more controversial, although experienced centers advocate endovascular treatment. There are no direct comparisons with surgery, but a study by the Congenital Cardiovascular Interventional Study Consortium,²²

Table 2
Results of the Major Series of Transcatheter Valve Implantation in the Pulmonary Position

Group	Device	No.	Implant, %	Homografts, %	Preimplant gradient, mmHg	Postimplant gradient, mmHg	Complications, %	Reintervention-free, %
Lurz et al. ¹³	Melody	155	95	81	37±20	17±10	4.5	93
McElhinney et al. ¹⁴	Melody	136	91.1	76	37 (10-70)	12 (0-37)	6	93.5
Vezmar et al. ¹⁵	Melody	28	100	25	36±15	12±7	7	91
Boone et al. ¹⁶	Edwards	7	100	71.4	60.4±27.3	14.9±6.9	14.2	100
Kenny et al. ¹⁷	Edwards	36	97.1	80.5	26.8±18.4	11.7±8	19.4	97

Complications, device embolization, conduit rupture, coronary compression, compression or rupture of pulmonary branches, major vascular complications, and ventricular tachycardia; homografts, percentage of patients with implantation over a homograft; implant, percentage of catheterized patients who underwent implantation; postimplant gradient, right ventricle-pulmonary artery pressure gradient following implantation; preimplant gradient, right ventricle-pulmonary artery pressure gradient prior to implantation; reintervention-free, percentage of patients not requiring further interventions in the conduit during the first year of follow-up.

Table 3

Indications for Aortic Valvuloplasty According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for aortic valvuloplasty	
Class I	Young adults with noncalcified valves and without aortic regurgitation who: <ul style="list-style-type: none"> • Have symptoms and a peak-to-peak pressure gradient >50 mmHg • Are asymptomatic but show ST segment or T wave changes on electrocardiogram and a peak-to-peak pressure gradient >60 mmHg
Class IIa	Young asymptomatic adults with a pressure gradient >50 mmHg who want to participate in sports or women who plan to have children
Class IIb	Can be considered a bridge to surgery for hemodynamically unstable patients or those at high surgical risk or who are inoperable due to high comorbidity
Contraindications for valvuloplasty	
Aortic valvuloplasty should not be considered an alternative to surgery in operable adult patients and is not recommended in those who have more than moderate aortic regurgitation	

Table 4

Indications for Transcatheter Intervention for Coarctation According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for transcatheter intervention for coarctation	
Class I	Peak-to-peak systolic pressure gradient >20 mmHg Peak-to-peak pressure gradient <20 mmHg but with imaging study showing anatomical evidence of severe coarctation and the presence of collaterals In short-segment native coarctation, transcatheter intervention is recommended over surgery Transcatheter intervention is recommended in recoarctation with a pressure gradient >20 mmHg
Class IIb	Stent implantation in long segments is considered a possible therapeutic option, but there are no data on long-term efficacy and safety

which included 302 consecutive patients who underwent implantation of a stent in the aorta demonstrated an immediate success rate of 96% and a complications rate of only 1% with the endovascular approach (including 1 dissection and 3 aneurysms). These results appear to be better than those found in surgical series, in which the morbidity rate is higher (and includes bleeding, hemothorax, laryngeal and phrenic nerve paralysis, and aneurysm formation).

Studies directly comparing stenting with simple angioplasty are not available either, but most interventional cardiologists

prefer the use of stents in adults, since these devices are associated with a greater reduction of the pressure gradient and lower rates of recoarctation and aneurysm formation (as there is no need to produce the tears in the vessel that are required when the balloon is employed alone).^{23,24} There is a wide range of these devices, which includes Genesis, EV3, Cheatham-Platinum and Atrium stents (an advantage of the latter is that they do not have to be mounted on the balloon). To position them, some centers resort simply to the stability afforded by the balloon-in-balloon catheter, although most also employ rapid ventricular pacing. We will have to wait for the COAST (Coarctation Of the Aorta Stent Trial) study, which used the Cheatham-Platinum stents (the recruitment period ended >1 year ago), and the COAST II study, which will use coated stents, as these studies will provide essential information on the use of stents in this condition.

Pulmonary Artery Stenosis

Stenosis of the pulmonary arteries can occur in any segment of the pulmonary vascular tree and generally develops in the context of other complex heart diseases (tetralogy of Fallot, Williams syndrome, etc.). Intervention is indicated when symptoms are detected and/or there is a narrowing of the vessel caliber of >50% and/or an increase in right ventricular pressure >50 mmHg.⁷ The pressure gradient across the lesion does not appear to be useful in these cases as the changes in pressure produced by stenosis are mitigated by the redistribution of pulmonary blood flow and could mask the true pressure gradient across the lesion.

Available devices in transcatheter treatment are high-pressure balloons, cutting balloons, and stent implantation. The success rate with cutting balloons is 92% in small-caliber vessels,²⁵ but the follow-up of these series was incomplete and the long-term results are unknown. The major problem with the utilization of cutting balloons in adults, as with drug-eluting stents, is the lack of devices measuring over 8 mm and 5 mm, respectively, which frequently limits their possible use. The development of bioabsorbable stents could be an interesting option in this field.

VASCULAR COMMUNICATIONS (COLLATERALS, FISTULAS, AND DUCTUS)

Patent Ductus Arteriosus

Patent ductus arteriosus can be classified into 5 groups according to its anatomy (Krichenko's classification; type A, the

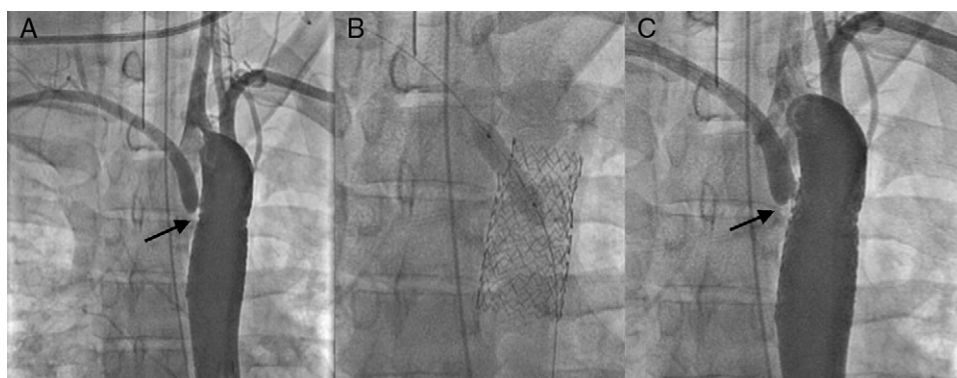


Figure 5. Aortic coarctation in a patient with anomalous origin of the left subclavian artery from the thoracic aorta. A: following stent implantation into the coarctation, a reduction in the diameter of the subclavian artery ostium is observed. B: a 0.014" guide wire is advanced through the stent and angioplasty is performed with an 8-mm balloon. C: final outcome, with an opening of the cells of the stent toward the subclavian artery ostium and improvement in arterial clamping.

conical ductus, is the most common type), and into 3 groups based on its hemodynamic impact (small, with no impact; moderate, with left ventricular volume overload; and large, with an increase in pulmonary vascular resistances) (Fig. 6). Closure of the ductus is indicated when there is left ventricular volume overload with dilation or previously known endarteritis, and even in the case of small, asymptomatic patent ductus arteriosus to prevent endarteritis⁷ (Table 5). If closure is considered, a prior CT-angiography is of great help in determining the size and anatomy in adults. In very small openings (<2.5 mm–3 mm), closure can be performed with coils; in larger openings, a success rate at 1 year of 98% has been demonstrated with the Amplatzer[®] Duct Occluder I device (St. Jude Medical; Minneapolis, Minnesota, United States). The advantages of the Amplatzer[®] Duct Occluder II are that it requires a catheter of smaller diameter for deployment and that it has 2 symmetrical discs, which allow for implantation using either a pulmonary artery or an aortic approach.

Coronary Artery Fistulas

Coronary artery fistulas are congenital malformations in which a direct communication is established between a coronary artery and a cardiac chamber or pulmonary structure. These defects usually arise from the right coronary artery and generally drain into the right chambers (coronary sinus, atrium, or ventricle).²⁶ In fistulas of considerable size and in those of medium size that cause documented ischemia, arrhythmias, or ventricular dilatation, percutaneous closure is indicated, once the course has been delineated and the potential for complete closure has been confirmed by angiography and CT-angiography⁷ (Table 6). With transcatheter closure, complete occlusion has been observed in up to 80% of patients, with minimal complications during the procedure. Nevertheless, during long-term follow-up, the rate of adverse events was 15%, mostly due to myocardial ischemia²⁷; these events were most frequent in fistulas draining into the coronary sinus. Direct comparisons between surgical and transcatheter closure have not been documented, but the outcomes appear to be similar.

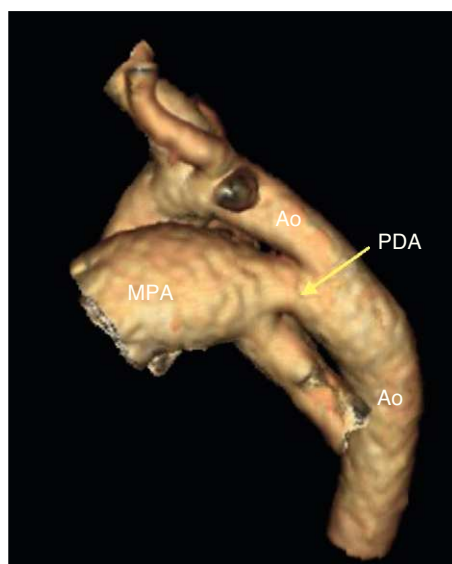


Figure 6. Computed tomography angiography and 3-dimensional reconstruction of a patent ductus arteriosus. Ao, aorta; MPA, main pulmonary artery; PDA, patent ductus arteriosus.

Table 5

Indications for Transcatheter Intervention for Patent Ductus Arteriosus According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for transcatheter intervention for patent ductus arteriosus	
Class I	When left chambers are dilated Presence of pulmonary hypertension Net left-to-right shunt Previous endocarditis
Class IIa	It is reasonable to consider closure of small asymptomatic ductus with transcatheter devices
Contraindications for interventional procedures in patent ductus arteriosus	
Surgical management is preferred in cases of large ductus or with distorted anatomy that cannot be treated by means of device closure	
Closure is not indicated in patients with pulmonary hypertension and right-to-left shunt	

Table 6

Indications for Transcatheter Intervention in Persistent Coronary Artery Fistulas According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for transcatheter intervention in persistent coronary artery fistulas	
Class I	Large fistulas should be closed regardless of the symptoms, following delineation of their course and confirmation of the possibility of complete closure Small fistulas should be closed in cases of documented myocardial ischemia, arrhythmias, ventricular systolic or diastolic dysfunction not attributable to other causes, or endarteritis
Contraindications for interventional procedures in coronary artery fistulas	
Closure of small asymptomatic fistulas with no hemodynamic impact is not recommended	

Collaterals

Systemic-to-pulmonary vein collaterals (frequent in patients who have undergone the Fontan procedure due to the increase in pulmonary resistances) and pulmonary arteriovenous malformations can produce arterial desaturation and paradoxical embolism. In contrast, systemic-to-pulmonary collaterals develop in heart diseases associated with pulmonary hypoperfusion (pulmonary atresia with ventricular septal defect [VSD], Eisenmenger syndrome, etc.) and produce volume overload in the systemic ventricle, in addition to increasing the risk of hemoptysis in these patients.

For the closure of small-caliber collaterals, coils have been used for years, with good results; moreover, these devices have been improved. On the one hand, by the development of controlled-release coils and, on the other, by the creation of coils compatible with MRI, a technique that is being increasingly used to study the physiology and anatomy of congenital heart disease. In contrast, larger vessels are usually closed with the Amplatzer[®] Vascular Plug (St. Jude Medical); the Vascular Plug II, whose development has greatly facilitated the closure of collaterals that are more complex and distal along the vascular tree²⁶ (Fig. 7), and the recent introduction of the Vascular Plug IV have enabled the use of smaller catheters (4 Fr) which are deployed with greater ease and accuracy.

SEPTAL DEFECTS

Patent Foramen Ovale

Patent foramen ovale is not considered a congenital disease as such, but a normal variant that is present in 25% of the population.

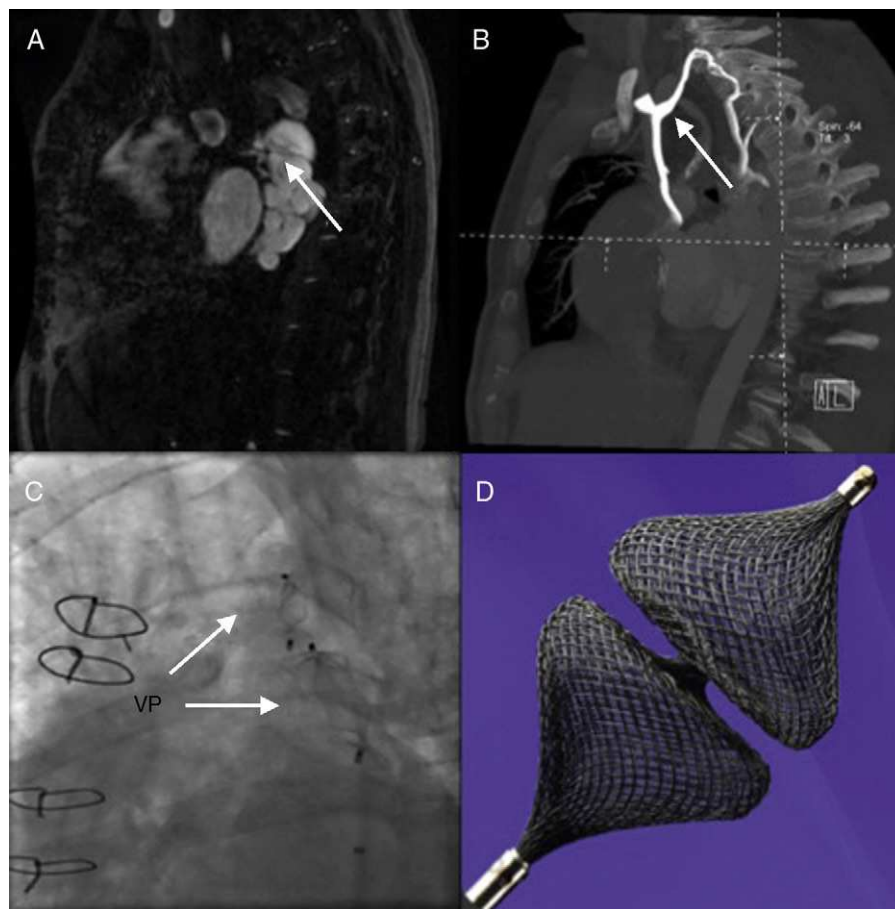


Figure 7. Closure of a venovenous collateral in a patient with left ventricular hypoplasia plus Fontan operation. A: cardiac magnetic resonance image showing a large collateral arising from the azygos vein and draining into the right inferior pulmonary vein. B: 3-dimensional reconstruction of computed tomography angiography image delineating the complete course of the collateral. C: fluoroscopic view of the implantation of 2 Vascular Plug II devices (St. Jude Medical) at the level of the aforementioned collateral. D: Vascular Plug IV device (St. Jude Medical). VP, Vascular Plug.

Nevertheless, this defect has been recognized as a mediator of various diseases, such as paradoxical embolism, orthostatic oxygen desaturation observed in platypnea-orthodeoxia syndrome, decompression sickness in divers, and migraine, among others. The clinical guidelines of the American College of Cardiology/American Heart Association recommend its closure only if a second episode of cerebral embolism is observed in a patient who is receiving medical treatment. The probably indiscriminate implantation that prevailed until 2006 led the US Food and Drug Administration to withdraw its approval of these devices and, since then, they are only approved for compassionate use and as part of research protocols. This is a question that arouses controversy among cardiologists and neurologists and, in turn, has renewed the interest of the biomedical industry in the development of innumerable products, ranging from bioabsorbable to easily retrievable devices (Fig. 8).

Atrial Septal Defect

ASDs constitute 22% to 30% of the cases of congenital heart disease in adults and are more prevalent among women (2:1). These defects represent a highly heterogeneous condition and are often more complex than they appear to be. Closure is indicated in those of the ostium secundum type, which show dilatation of the right chambers without irreversible pulmonary hypertension (Table 7). Ostium primum, coronary sinus, and sinus venosus

defects should be repaired surgically. The most widely used device in the world is the Amplatzer® Septal Occluder (St. Jude Medical), which features an extensive range of different sizes, a simple and easily recaptured release system, and delivery sheaths that can usually be smaller than those required with other devices. As occurs with the foramen, there is a wide range of devices, but few are suitable for the closure of ASDs measuring >24 mm. Nevertheless, they offer the advantage of not being associated with cases of erosion, a rare complication that has been observed in less than 0.05% to 0.3% of Amplatzer® devices and appears to be related to an oversized device or defects directly related to the aorta or left atrium. The overall success rate with complete closure of the defect is over 95%. Closure can be performed under fluoroscopic, transesophageal echocardiographic, or intracardiac echocardiographic guidance, depending on the characteristics of the defect and the operator's experience.

Ventricular Septal Defect

VSD encompass multiple anatomic variants and are uncommon in the adult population without congenital heart disease (Fig. 9). Muscular defects, whether congenital or acquired (caused by trauma or infarction), and postoperative and perimembranous VSD are considered to be treatable with percutaneous closure. Transcatheter closure is an attractive option for patients with congenital heart disease who have undergone multiple surgical

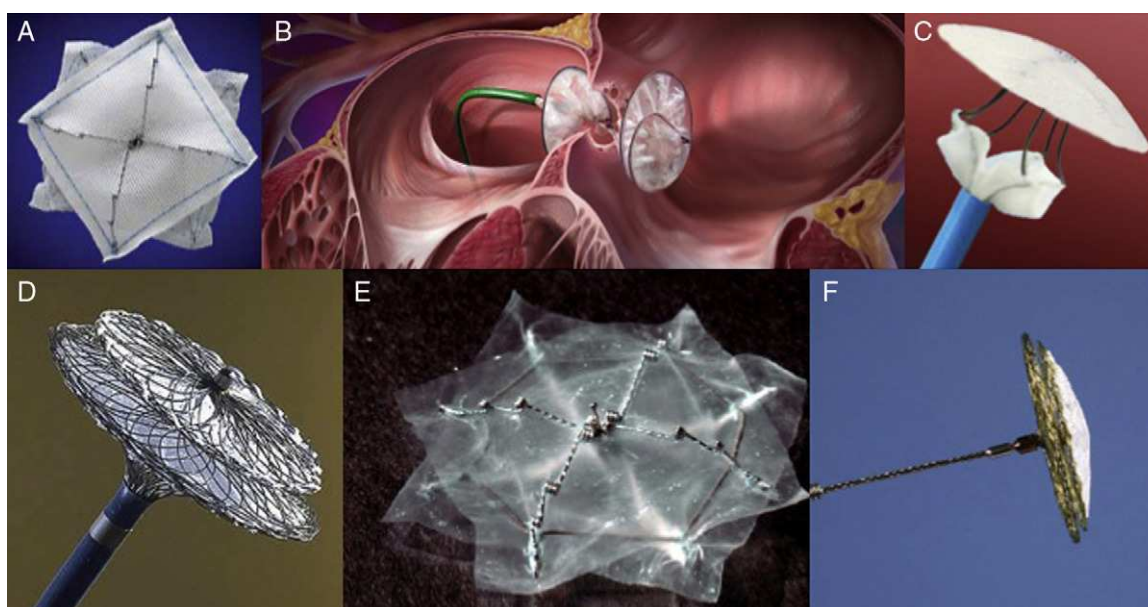


Figure 8. Different devices for atrial septal defect and patent foramen ovale closure. A: CardioSEAL® (NMT Medical; Massachusetts, United States). B: Helex® (Gore Inc.; Arkansas, United States). C: STARFlex® Septal Occluder (NMT Medical). D: Amplatzer® Patent Foramen Ovale Occluder (St. Jude Medical). E: BioSTAR® Bioabsorbable Septal Occluder (NMT Medical). F: Figulla® Patent Foramen Ovale Occluder (Occlutech; Sweden).

interventions and have a VSD or a residual VSD. Closure is indicated in patients with significant hemodynamic overload without irreversible pulmonary hypertension and in those who have developed endocarditis⁷ (Table 8). There are discrepancies in the definition of perimembranous VSD. Despite a success rate of 84%, atrioventricular block has been reported in 2% to 5% of cases (a slightly higher incidence than that of surgical series).²⁸ Currently, the closure device for perimembranous VSD has not been approved by the US Food and Drug Administration. Nevertheless, the new version of the Amplatzer® device for perimembranous VSD was

implanted in humans for the first time in 2011 and, to date, some 20 have been placed, with excellent results; most notably, there have been no reports of atrioventricular block.²⁹ However, medium- and long-term follow-up will be necessary before investigators can conclude that the incidence of block is lower with this new device and its use becomes widespread.

POSTOPERATIVE RESIDUAL DEFECTS

Patients treated surgically for complex congenital heart disease may have residual lesions (defects left intentionally in hopes of a certain benefit) or complicated lesions (undesired postoperative complications) that require a second intervention. In most cases, since the patients have undergone multiple interventions, the initial approach usually involves transcatheter techniques. Many of these cases require special resourcefulness to select the best strategy and device.

Fontan Fenestration

On occasion, patients who undergo the Fontan procedure require fenestration between the intracardiac/extracardiac conduit and the systemic atrium that enables decompression of the pulmonary venous circuit. Over the long term, these residual lesions can produce a significant right-to-left shunt, which leads to hypoxia and favors paradoxical embolism. In most cases, closure does not differ from that of an ASD but coated stents may sometimes be employed to close the fenestration.³⁰

Conduit Obstruction

Extracardiac conduits, which are mostly utilized to connect the subpulmonary ventricle with the pulmonary artery, can be subject to obstructions due to calcification, tissue proliferation, or extrinsic compression. This restenosis is mostly produced in the anastomotic regions and can be resolved with angioplasty alone or stent implantation.

Table 7

Indications for Percutaneous Closure of Atrial Septal Defects According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for percutaneous closure of ASDs	
Class I	Right atrial and ventricular dilatation with or without related symptoms
Class IIa	Paradoxical embolism or platypnea-orthodeoxia syndrome
Class IIb	Presence of net left-to-right shunt with pulmonary artery pressure less than two thirds of the systemic pressure or pulmonary vascular resistances less than two thirds of the systemic vascular resistances and positive response to pulmonary vasodilators or to test occlusion of the defect
Criteria for percutaneous closure	
ASD with a minimum diameter >5 mm and <40 mm on echocardiographic study	
Adequate rims (>5 mm) from the defect toward adjacent structures, including superior and inferior venae cavae, coronary sinus, atrioventricular valves, and pulmonary veins	
Contraindications for percutaneous closure	
All the septal defects that are not ostium secundum defects, including ostium primum, sinus venosus, and coronary sinus defects	
Percutaneous closure should be avoided in septum that is markedly aneurysmal or has multiple fenestrations, in which a scarcity of surrounding tissue is observed	
Other options should be considered in cases of nickel allergy or contraindication for antiplatelet therapy	

ASD, atrial septal defect.

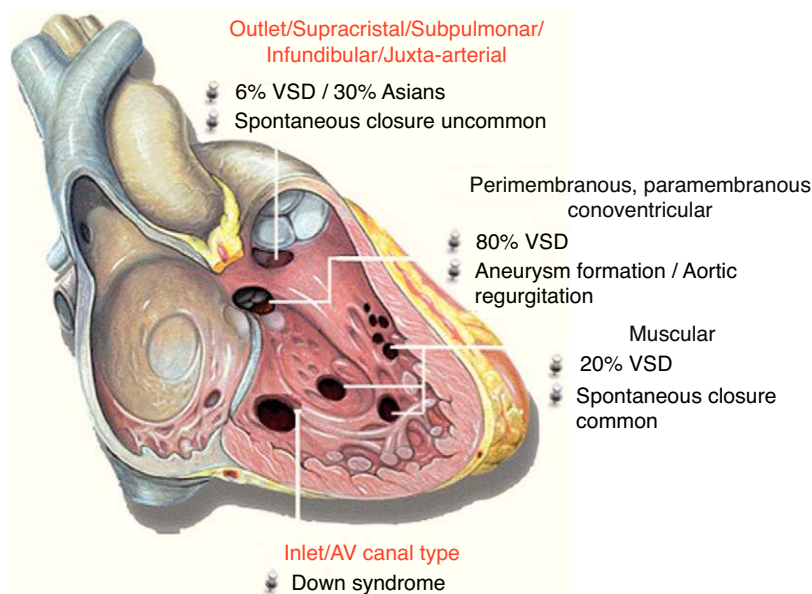


Figure 9. Illustration of the different types of ventricular septal defects. Types of ventricular septal defects: in red, percutaneous closure not recommended and, in black, closure is a viable option. AV, atrioventricular; VSD, ventricular septal defect.

Table 8

Indications for Percutaneous Closure of Ventricular Septal Defects According to the 2008 Guidelines of the American College of Cardiology/American Heart Association⁷

Indications for percutaneous closure of VSDs	
Class I	QP/QS>2 or signs of left ventricular volume overload
Class I	History of infective endocarditis
Class IIa	QP/QS>1.5 and when pulmonary arterial pressure is less than two thirds of the systemic pressure or the pulmonary vascular resistances are less than two thirds of the systemic vascular resistances or there is left ventricular systolic or diastolic dysfunction
Criteria for percutaneous closure	
Only type IV VSD or muscular defects are subject to percutaneous closure (IIb), although there is extensive experience with type II or perimembranous VSD	
VSD following infarction for which surgery has been ruled out or in cases of postoperative residual shunt	
Adequate rims (>4 mm) from the defect toward the adjacent structures, including the aortic, pulmonary, mitral, and tricuspid valves	
Contraindications for percutaneous closure	
All septal defects that are not muscular, such as type I or subpulmonary defects, type III defects, or atrioventricular canal defects. There are doubts concerning type 2 or perimembranous defects	
Perimembranous defects with aortic valve prolapse or a markedly aneurysmal septum should be avoided	
Other options should be considered in cases of nickel allergy or contraindication for antiplatelet therapy	

VSD, ventricular septal defect.

Interatrial Dehiscences Following the Mustard or Senning Procedures

The dehiscences observed in the Mustard or Senning procedures usually lead to desaturation, and their closure is very similar to that employed in Fontan fenestrations. In most cases, ASD closure devices or coated stents are used. In complete occlusion, guide wires for chronic coronary occlusion, or even radiofrequency guide wires, can be used to restore patency.

CONCLUSIONS

Over the last 3 decades, the growth of interventional cardiology for the treatment of adult congenital heart disease has been spectacular. The continuous development of new devices and the introduction of valves for percutaneous placement have constituted major steps forward in the quality of the care received by these patients. Advances in both the techniques and the technology used have enabled novel complex interventions to be performed with high success rates and a lower incidence of complications. Close collaboration among pediatric cardiologists, imaging specialists, surgeons, and interventional cardiologists is required to obtain optimal results in these patients.

CONFLICTS OF INTEREST

None declared.

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