Severe congenital aortic stenosis (CAS) often requires treatment in the early stages of life. Initial treatment is surgical commissurotomy or balloon valvuloplasty. Sometimes these approaches lead to aortic regurgitation. Once it occurs the only solution is valve replacement with mechanical prostheses or pulmonary autograft. Mechanical prostheses require anticoagulation and have a significant incidence of reintervention and mortality. (1) Replacement with pulmonary autograft does not require anticoagulation but is a more aggressive surgery involving a second valve and its long-term outcome has not yet been established. (2) “Bicuspidization” of unicuspid aortic valves has been proposed as a reconstructive surgery (3). Our hypothesis is that this concept is also applicable to a unicuspid valve that has become insufficient after treatment of the stenosis surgical or percutaneous. We describe a case in which “bicuspidization” was used in this scenario.

We present the case of a child diagnosed with CAS at birth. A commissurotomy was performed 14 days after birth. The gradient disappeared but moderate to severe aortic regurgitation appeared. He was reoperated at 33 months to rebuild the valve. Apparently, the fused tissue was divided between the valve cusps, tricuspidazing the valve and placing shortening sutures on the free leaflet edges.

Postoperative echocardiography revealed grade 2 regurgitation, with progressive center jet over the next two years, presenting severe regurgitation with left ventricular dilation at 4 years. Cardiac resonance imaging showed a 50 mm end-diastolic diameter and an ejection fraction of 50%. The patient had profuse sweating with exercise as symptom of heart failure, and was referred to our center.

Valve repair surgery was indicated and planned. Adherences were dissected by median sternotomy and cardiopulmonary bypass was initiated. A transverse aortotomy was performed. The valve was a unicuspid aortic valve reconstructed by tricuspidization. The two comissures of the right leaflet had insufficient height and the effective height of the three leaflets (4) was limited. The right leaflet was hypoplastic.

The valve was bicuspidized. The rudimentary right leaflet was resected and a new commissure was created in front of the left /noncoronary leaflet, connecting the remaining tissue of the left and noncoronary leaflets to the new commisure, with two patches of heterologous pericardium. The leaflets were adjusted achieving an effective height of 9 mm.

Intraoperative echocardiography showed a competent valve with planimetric area of 1.6 cm². The postoperative period was uneventful and the child was transferred to the general ward after two days and discharged four days later, recovering his usual activities in 4 weeks. A year later he continues to be well.

The echocardiography at one year shows a competent valve with 34 mm end-diastolic and 21 mm end-systolic ventricular diameters and normal ejection fraction (71%). (Figures 1 and 2; Videos 1 and 2)

Treatment of CAS aims to eliminate the transvalvular gradient. After initial surgical or interventional procedure, significant valve regurgitation occasionally appears. Traditionally the only solution to this problem has been valve replacement. Valve reconstruction has not been applied uniformly or successfully, which is not surprising when in most cases the underlying anatomy is unicuspid. This is characterized by commissural and leaflet hypoplasia, (5) an anatomic condition leading to poor hemodynamic function.

Valve reconstruction has an important advantage in restoring hemodynamic valve function; it involves...
a low rate of endocarditis, limiting surgery to the diseased valve and allowing growth of the ventricular outflow tract together with the somatic growth of the patient.

We had already described the concept of “bicuspidization” of unicuspid valves, creating a new commissure with normal height and adding pericardial tissue to replace the lack of native valve tissue (3). We have recently proposed a technical modification that acquires a more symmetrical bicuspid design (6).

The case we report demonstrates that this concept also applies to regurgitant valves due to CAS treatment. The initial functional result is close to that of a normal valve. As with any novel approach mid-term results are still limited and no data on long-term durability is available. Even if it was limited to 5 or 10 years, this would allow the child time to grow before applying a more definitive solution. Furthermore, a new valve reconstruction could be performed if the somatic growth of the child caused the used pericardial patches to become “small”.

The reconstruction of the aortic valve should be the technique of choice if significant aortic regurgitation occurs after CAS surgical or interventional treatment.

Carlos Porras1, Hans-Joachim Schäfers1, Ignacio Zabala1, Fernando Cabrera1, María Ángeles Izquierdo2, Miguel Such1

Aortopulmonary Window Associated with Tetralogy of Fallot

The aortopulmonary window (APW) is a rare congenital malformation which occurs in 1 out of each 100,000 live births, representing 0.15% of all congenital heart defects. It is defined as an abnormal communication between the aorta and the pulmonary artery and / or right pulmonary branch, above the two separate normally formed semilunar valves. (1) It may occur alone, but it is commonly associated with another congenital heart disease. It varies in size and location, resulting in different classifications, those of Robinson and Mori being the most commonly employed at present.

This communication produces a large left - right (L-R) shortcircuit similar to a large patent ductus arteriosus (PDA), leading to congestive heart failure and early development of pulmonary hypertension. Surgical treatment is indicated at diagnosis. The prognosis of untreated patients is poor with 40% mortality in the first year of life. (2)

We describe the case of a male patient, with a diabetic mother, born at term by cesarean section with a birth weight of 3090 g. At 9 days old he is referred to cardiology as a result of a heart murmur. Tetralogy of Fallot is diagnosed and he continues with cardiovascular ambulatory follow-up with atypical evolution, tachypnea, radiological cardiomegaly, dilated left heart chambers and echocardiographic pulmonary hy-
perflow signs, under oral furosemide treatment.

He is admitted to the clinic on September 5, 2011 at the age of 7 months with respiratory distress, subcostal and intercostal retractions, prolonged expiration, wheezing, tachypnea, tachycardia, systolic murmur in every foci, S3 at the apex and 92% oxygen saturation with ambient air. Two days later he is transferred to intensive care due to general malaise, fever, reticular rash, bilateral crackles, and hepatomegaly. Samples are taken for virological examination. Decompensated congestive heart failure by an infectious respiratory process is presumed. Oseltamivir, ceftriaxone, spironolactone, furosemide, digoxin, and enalapril treatment is started.

Echocardiography shows pink tetralogy of Fallot with mild pulmonary infundibular and valve stenosis with 36 mmHg gradient, pulmonary hyperflow, severe dilation of the left heart chambers, moderate right ventricular hypertrophy, persistence of the left superior vena cava (LSVC), dilated coronary sinus, right aortic arch, and adequate biventricular function.

Forty-eight hours later, the patient is admitted under assisted mechanical ventilation (AMV) due to clinical condition deterioration with positive virological test for respiratory syncytial virus together with pneumonia. After 15 days of antibiotic treatment, with the infectious process controlled, the patient remains on AMV; and because of diagnostic uncertainty due to dilation of the left chambers and pulmonary hyperflow data, cardiac catheterization is indicated.

On September 22, 2011, cardiac catheterization reports tetralogy of Fallot with mild infundibular and valve stenosis with a 40 mm Hg gradient, large type 1 APW, moderate pulmonary hypertension distal to the window, right aortic arch, normal coronary arteries, persistence of the LSVC with absent innominate vein, dilated coronary sinus, and severe dilation of the left heart chambers (Figure, 1).

On September 24, 2011, catheter-related sepsis is found, with culture positive for coagulase negative and methicillin resistant staphylococcus and Candida albicans. Treatment is initiated with vancomycin and fluconazole. After controlling the infection he undergoes corrective surgery on October 17, 2011, weighing 4150 g.

Median sternotomy is performed. A large pericardial patch is resected and treated with glutaraldehyde, and a large dilated left atrium and type 1 APW is found (Figure 2). The three caval veins, the aorta close to the proximal end of the innominate artery (distal to the APW) and the right superior pulmonary vein are cannulated. Immediately after the initiation of extracorporeal circulation (ECC) a tourniquet is applied to both pulmonary branches. The anterior aspect of the APW in its pulmonary end is incised under moderate hypothermia, aortic cross clamping, and antegrade cardioplegia, continuing up to the posterior aspect, totally separating the aorta from the pulmonary trunk. Priority is given to direct closure of the aorta with 5-0 prolene. Pulmonary arteriotomy is extended to the proximal end, valve commissurotomy is accomplished and Hegar dilators are passed towards the ring and branches. Right atriotomy is performed, the infundibular stenosis is resected through the tricuspid valve and the interventricular communication (IVC) is closed with subaortic pericardial patch and 5-0 prolene continuous suture. The pulmonary artery is closed with a pericardial patch and prolene 6-0, followed by atriotomy closure, deaeration maneuvers and aortic unclamping. After weaning from ECC, modified hemofiltration is initiated extracting 220 cm3. ECC
time: 165 minutes and aortic cross-clamping time: 104 minutes. The patient leaves the operating room with open chest, and adrenaline and milrinone infusion. Forty-eight hours later, the chest is closed and 10 days after the patient is extubated. He is discharged from hospital at 21 days postoperatively.

After 22-months follow-up the patient is asymptomatic, with no cardiac medication, and a weight of 13 kg. The echocardiogram shows closed IVC without residual shunt, mild pulmonary valve stenosis without hemodynamic repercussions, with normal aortic flow and ventricular diameters.

The aortopulmonary window can occur alone, but in 10% of cases it is associated with other lesions (3, 5). The variation in terms of location and size gives rise to different classifications. The most commonly used are Richardson’s classification that divides it into type 1, 2 and 3 and Mori’s classification that divides it into proximal, distal and total.

Type 1 or proximal: the defect is located between the ascending aorta and the pulmonary trunk above the Valsalva sinuses. Type 2 or distal: the defect is more distal between the ascending aorta and the pulmonary trunk involving the origin of the right pulmonary artery. Type 3 or total: the right pulmonary artery arises from the ascending aorta.

Both the type of associated lesion as the APW size affects the patient’s clinical condition. In infants with isolated APW, signs and symptoms of heart failure usually develop early and its presentation is similar to a large ductus or IVC. The continuous heart murmur is heard only in less than half of the cases (when the defect is small) and the ECG and chest X-ray are nonspecific, evidencing a large L-R short circuit with cardiomegaly and pulmonary hyperflow. Echocardiography should distinguish the presence of two separate semilunar valves, which allows differentiating the APW from the trunkus arteriosus. It is vital to define the distance between the proximal border of the defect and the valves, the position of the coronary arteries, and the distance between the distal border and the pulmonary branches. (6).

Surgical experience in APW is limited since it is a very rare defect. Mortality ranges from 8 to 30%, depending on the age at surgery, pulmonary vascular resistance, associated lesions and patient condition. (2, 4, 5, 7, 8) Once diagnosed, it should be corrected as early as possible.

Simple ligation or division has relatively high frequency of complications, such as recanalization, bleeding and narrowing of the aorta or pulmonary artery. Currently, this technique should be reserved for very young severely ill patients, in whom ECC would be very deleterious and in type 1 isolated, small size APW; located distantly from the pulmonary artery and the semilunar valves. This type of APW would also be candidate for hemodynamic closure. (9)

In most reports transaortic closure with simple patch is preferred. In our patient with large APW associated with tetralogy of Fallot we preferred the complete separation of the arteries, with incision in the pulmonary end, favoring direct closure of the aorta without patch, with good visualization of coronary arteries and semilunar valves, based on the technique described by Van Son. (10) In our case, due to the associated disease we also had to explore and expand the pulmonary trunk with a pericardial patch.

We believe that the technique of choice in these cases is the complete separation of the APW, favoring primary closure of the aorta and using the pericardium for pulmonary artery closure. The correction of APW with associated complex lesion can be performed simultaneously with low morbidity and mortality.

Rubén A. Toledo, Pablo R. Pérez Caram, Alejandra Segovia, Jorge Gómez, Maria S. Olivieri, Florencia Jeréz
Sanatorio Infantil San Lucas, San Miguel de Tucumán, Argentina.
e-mail: rubenatoledo@yahoo.com.ar

REFERENCES

Nosocomial Infective Endocarditis Caused by Extended-Spectrum Beta-Lactamase-Producing Klebsiella pneumoniae

Nosocomial infective endocarditis (IE) caused by extended-spectrum beta-lactamase-producing Gram-
negative bacilli is a severe and rarely reported complication. Associated with health care, it arises from invasive procedures that cause bacteremia, with an increase in recent years in the number of cases due to the growing frequency and complexity of diagnostic and therapeutic maneuvers. (1) In a recent prospective, observational, multicenter study, IE associated with health care accounted for 34% of all recorded episodes. Most of these cases were caused by Staphylococcus aureus, while Gram-negative bacilli accounted for < 2%. (2) Infective endocarditis by Klebsiella pneumoniae is rare, as evidenced by the small number of cases described in the literature, and has a bad prognosis. (3) Nosocomial infections by extended-spectrum beta-lactamase-producing (ESBL) K. pneumoniae are an emerging entity and represent a major health problem. The description of a health care-associated IE case on the aortic valve due to ESBL-producing K. pneumonia was considered of interest because of its low frequency and high mortality.

A 53-year-old male patient with type II diabetes is hospitalized for decompensated heart failure. Trans-thoracic echocardiography (TTE) reveals severe bicuspid aortic valve stenosis with severe ventricular dysfunction. The patient evolves with fever 48 hours after admission, without apparent infective focus. Blood and urine cultures are performed and empirical antibiotic treatment is initiated. Subsequently, antibiotics are suspended for negative cultures. Forty-eight hours later he presents with fever associated with hypotension. New cultures are performed and empirical antibiotic treatment with piperacillin-tazobactam and vancomycin is reinitiated. In 3/3 blood cultures ESBL-producing K. pneumoniae is isolated only sensitive to imipenem, meropenem and colistin. Due to this finding, the antibiotic treatment is changed to imipenem. Transesophageal echocardiogram showed aortic ring abscess and aortic valve vegetations. Valve replacement surgery with mechanical prosthesis and perivalvular abscess drainage is performed. The postoperative echocardiogram shows a normally functioning mechanical prosthesis. The patient evolves afebrile and hemodynamically stable. After 5 weeks of postoperative imipenem antibiotic treatment with negative culture control, the patient is discharged. Twenty days later the patient consults for febrile episodes and is rehospitalized due to hemodynamic decompensation. Blood culture (5/5) bacteriological rescue of ESBL-producing K. pneumoniae is obtained with the same antibiotype of the first episode, and an pseudoaneurysm of the prosthetic ring with left ventricular communication is observed by TTE. (Fig. 1 y 2) The patient evolves with cardio-respiratory arrest within a tachycardia / ventricular fibrillation context, unresponsive to resuscitation maneuvers.

Infective endocarditis episodes of non-HACEK Gram-negative bacilli are uncommon (1.8% of all etiologic agents) affecting in most cases the native valve. (4) E. coli, followed by P. aeruginosa and K. pneumoniae were found in order of frequency. The latter group evidenced the highest in-hospital mortality (40%). Most IE episodes are related to health care, intravascular devices being the most common source of infection (29%). Among endocarditis complications there is increased frequency of paravalvular involvement and abscess formation, and a predominant presence of comorbidities, mainly of diabetes. (5) Regarding K. pneumoniae as a cause of IE, this is unusual given the poor adherence to heart valves, and there are few reports of IE caused by this microorganism. The most important review showed that the aortic valve was the most affected (74%) with a mortality rate of 38% for the native valve and 57% for the prosthetic valve, whereas mortality in cases of early and late prosthetic valve endocarditis was 80% and 40%, respectively. (3) In this review, the overall mortality of K. pneumoniae endocarditis was 49%. So far there are four cases reported in the international literature on ESBL-producing K. pneumoniae IE. (6) Our patient had a heart condition with poor prognosis aggravated by IE that led to his death. Although this entity is rare, the increased consumption of antibiotics and increased contact with health care may favor the development of these cases, although it is necessary to have a high clinical suspicion to make the diagnosis.
A 72-year old male hypertensive patient, with type 2 diabetes mellitus was hospitalized on November 23, 2007 for acute abdomen. He underwent laparotomy with enterostomy and evolved with nosocomial pneumonia. On December 1st, 2007, he was reoperated for obstruction distal to the suture with interloop abscesses, requiring bowel resection and meropenem and amikacin antibiotic therapy. Fever persisted (December 23, 2007) due to vascular catheter. On January 21, 2008, septic arthritis is diagnosed, surgical drainage is performed and treatment with vancomycin / trimethoprim-sulfamethoxazole is started.

The patient is admitted to our hospital on January 29, 2008 with sepsis for meticillin-resistant Staphylococcus aureus (MRSA). He presents with the following intercurrent diseases: MRSA sepsis with multiple embolic foci, right iliopsoas abscess and sternoclavicular and knee septic arthritis. An abdominal CT scan (Figure 1A) reveals mycotic aneurysm with contained partial rupture. Endovascular resolution is decided with an Excluder endoprosthesis (Fig. 2) and prolonged antibiotic therapy.

The patient progresses well, and is discharged with vancomycin-sulfamethoxazole + trimethoprim - rifampicin on March 6, 2008. He is in his fourth asymptomatic year with clinical and tomographic control (Figure 1B) and neomycin treatment for life.

Mycotic aneurysms were described by Osler in association with infective endocarditis in 1851. Their true incidence is unknown and it is estimated to be about 0.65% to 1.3% of all aneurysms. (1) Reports are more numerous due to the increase in elderly patients, imaging methods and knowledge of the disease. (2)

These aneurysms are the result of bacteremia and subsequent embolization which causes plaque overinfection. Rarely, the healthy wall is colonized through the vasa vasorum, resulting in the formation of aneurysms. Other pathways are osteomyelitis penetrating directly or through the lymphatic system to the aorta, causing necrosis of the wall with false aneurysm formations and rupture.

The most common microorganisms are Staphylococcus and Salmonella species (28-71% and 15-24%, respectively), with Streptococcus pneumoniae in the third place.

Diagnostic suspicion is based on a pulsatile mass in the context of persistent sepsis with no clear focus, and positive blood cultures (50-85%). Negative blood cultures do not rule out the disease, and in this case imaging becomes very important. Angiography findings are: wall disruption, swelling of adjacent soft tissue, or presence of a perivascular mass.

Early diagnosis and therapy with broad-spectrum antibiotics associated with surgical or endovascular treatment directly impact on survival. Endovascular treatment is a good alternative to surgery, as it is minimally invasive and reduces cardiopulmonary, neurological and renal complications in critically ill patients. (2, 3)
In 1998, Semba et al., were the first to report successful stent treatment in three patients, followed by other studies with similar results. These works have highlighted the advantage of this procedure compared with surgery [it avoids large incisions, anticoagulation, extracorporeal circulation (in case of thoracic aorta involvement), aortic cross-clamping and hemoderivative transfusions]. Moreover, it shortens hospitalization with fast social reinsertion.

A meta-analysis evaluating survival with endovascular treatment, reported 89.6 ± 4.4% at 30 days and 82.2 ± 5.8% at 2 years survival rates. Furthermore, the only significant independent predictors of persistent infection after endovascular treatment were aneurysm rupture and fever at the time of the procedure. (5)

In conclusion, endovascular treatment of mycotic aneurysm could be a valid alternative to surgical treatment, reducing morbidity and mortality in patients with multiple comorbidities and critical condition.

Cristian S. García, José C. Santucci, Ricardo A. Costantini, Juan Manuel Telayna
MT SAC
Hospital Universitario Austral.
Hemodynamics and Catheterization Unit
Av. Juan Domingo Perón 1500 – (B1629AHJ) Pilar – Buenos Aires
– Argentina
Tel. 0230-4482893

REFERENCES

REV ARGENT CARDIOL 2014;82:318-319 - http://dx.doi.org/10.7775/rac.v82. 14.3244