Hybrid Strategy. A Novel Method for the Management of Complex Congenital Heart Diseases

Bicuspid aortic valve is a congenital cardiac defect, involving a wide range of presentations, from aortic valve stenosis with severe cardiac failure in the newborn, to aortic dissection in the adult, or remaining unnoticed throughout life without requiring any treatment.

We present the case of a preterm patient with prenatal diagnosis of bicuspid aortic valve with severe stenosis. On admission, physical examination shows tachypnea and retraction, pale-cyanotic coloring, heart rate of 187 beats per minute, single S1 and S2, 2/6 systolic aortic murmur, symmetric brachial and femoral pulses, prolonged capillary filling >3 seconds, blood pressure 65/49 mm Hg and SpO_2 78%. Chest X-ray presents cardiomegaly and pulmonary edema. Mechanical respiratory assistance is initiated and prostaglandins are administered. Color Doppler echocardiography shows critical aortic stenosis with severe left ventricular dysfunction (Figure 1A).

At 18 hours of life, combined cardiac catheterization and aortic valvuloplasty with 6.0 × 20 mm Tylshak Mini hybrid balloon is performed through right carotid access (dissection) (Figure 1B). On the fourth day the echocardiogram evidences 16 mm Hg peak transaortic gradient with mild aortic regurgitation, and wide interatrial communication and patent ductus.

Hemodynamic instability with multiple organ failure (kidney failure, intraventricular hemorrhage and supraventricular tachycardia) persists from the fifth to the thirteenth day of life. Without clinical improvement, bilateral cerclage with polytetrafluoroethylene (PTFE) band is performed (Figure 1C) and the hybrid procedure is completed 48 hours later (reopening of the sternotomy and 1.7 mm Type E Krichenko stent implantation in the ductus).

At 5 days of treatment, the echocardiogram reveals decreased cardiac diameters, biventricular hypertrophy with improved function in both ventricles, and increased gradient through the cerclages and at the left ventricular outflow tract level due to improved function.

Three months after the hybrid procedure, percutaneous cerclage removal (Figure 2A and 2B) and ductal stent occlusion (Figure 2C) are carried out.

Neonatal critical aortic stenosis, in which the immature myocardium faces an abrupt increase in afterload, generates greater wall stress with left ventricular dilation, instead of the compensatory hypertrophy encountered in older children. The increased end-diastolic volume and pressure alter coronary flow producing diastolic dysfunction. (1,2) There are problems in the immature neonatal myocardium, a) structural: myocyte precursors replicate and increase in number (hyperplasia), but not in size (hypertrophy), myofibrils are disorganized, there is scant presence of the T tubule system and mitochondrial sarcoplasmic reticulum, high DNA concentra-

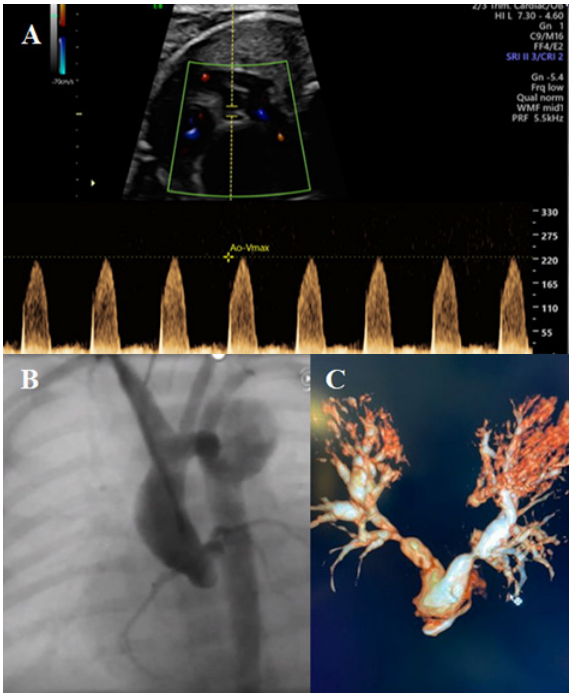

Fig. 1. Doppler echocardiography showing critical aortic stenosis (A). Angiography through carotid access post aortic valvuloplasty (B). Angiographic reconstruction of surgical cerclage in both pulmonary branches (C).

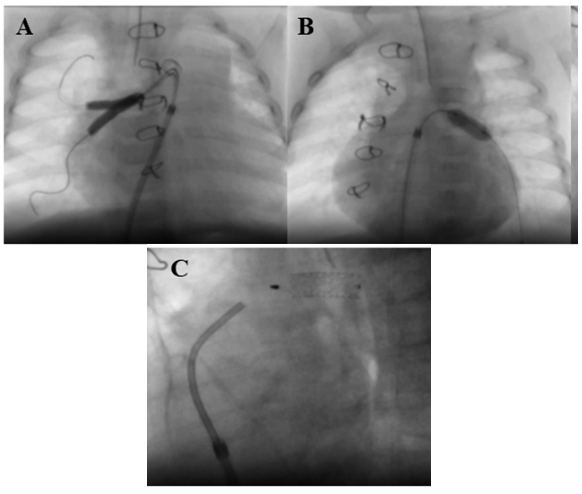

Fig. 2. Angiography. Dilation of both pulmonary branch cerclages (A and B). Percutaneous ductal stent occlusion (C).

tion and predominance of non-contractile elements; b) biological, such as lower sarcoplasmic reticulum calcium uptake and lower density of alpha and beta receptors; and c) metabolic, as the preferred use of glucose as energy source. (2)

The hybrid approach in refractory heart failures of different etiology is a therapeutic alternative, consisting of bilateral cerclage in pulmonary branches, generation of a non-restrictive interatrial communication and ductal stent implantation. (3,4) Pulmonary cerclage increases contractility on the right ventricle (RV) (Anrep effect), producing hypertrophy and the capacity of myocyte regeneration in an immature heart, and improved right ventricular diastolic filling. (3,4) Biological and genetic changes at the level of the fibers shared by both ventricles (co-hypertrophy) may restore electromechanical synchrony between the two ventricles and ventriculoarterial coupling. (5)

The hemodynamic evolution post valvuloplasty can sometimes be unfavorable due to abnormal ventricular remodeling, with unexpected clinical consequences. The hybrid approach is a very useful novel technique for different pediatric heart failure scenarios which cannot be managed from a clinical point of view, or as bridge to transplantation. This approach activates various physiological mechanisms, producing a relevant balance between pressures, flows and resistances, corroborated by computational studies of flow dynamics. (6)

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web).

Ethical considerations

Not applicable.

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From Cardiopulmonary Arrest to Extracorporeal Membrane Oxygenation in Pulmonary Thromboembolism: An Inter-Hospital Work

The pulmonary thromboembolism (PTE) is a prevalent entity that involves patients with a wide range of ages and comorbidities. It can affect young patients without relevant comorbidities, and cause a great impact in terms of morbidity and mortality. High-risk PTE implies the highest mortality, especially in those who present with cardiopulmonary arrest (CPA). In addition, a significant percentage of patients show severe symptoms or hemodynamic decompensation on admission or during their progress. The indicated strategy in these patients is the immediate reperfusion. Nowadays, the most widely supported strategy is the systemic thrombolysis, leaving surgical treatment –either surgical embolectomy or venoarterial extracorporeal membrane oxygenation (VA-ECMO)– as a second option when medical treatment fails or is contraindicated. However, data from experienced centers suggest that surgical techniques are safe and effective. The following is a case report of a patient with massive PTE and CPA, in which two centers worked together in pursuit of ventricular support as a rescue therapy.

We present the case of a 39-year-old male patient, with no risk factors or cardiovascular history, who reported an Achilles tendon surgery 45 days prior to his consultation. He came to the Emergency Department after having experienced an episode of sudden dyspnea associated with syncope without prodromes and traumatic brain injury (TBI) at home. During his stay in the Emergency Department, he presented a new syncopal episode. A Doppler transthoracic echocardiography (TTE) was performed (Figure 1) which showed dilatation of the right chambers; this, together with the recent history of trauma surgery, led to the suspicion of PTE. He immediately experienced a CPA, so advanced cardiopulmonary resuscitation maneuvers were performed and, based on his history of TBI, it was decided to start a percutaneous treatment by thromboaspiration and local thrombolysis. The patient progressed to hemodynamic instability so, after ruling out intracranial hemorrhage by computed tomography, systemic

thrombolytics were administered. However, the patient remained in refractory shock despite the administration of vasoactive drugs at maximum doses. Contact was established with a High Complexity Center and the mobile ECMO team was activated. Once the coagulopathy was stabilized, the patient received venoarterial ECMO at the first center and was subsequently transferred to the High Complexity Center. On admission, the patient was hemodynamically unstable, received ventricular and respiratory support and required maximum doses of noradrenaline, vasopressin and milrinone; laboratory tests showed acute kidney and hepatic injury, metabolic acidosis with hyperlactacidemia and marked coagulopathy. The values of high-sensitivity troponin T and NT-proBNP were 6700 ng/L and 480 pg/mL, respectively. The electrocardiogram (Figure 2) showed sinus tachycardia and the chest X-ray showed cardiomegaly and overt signs of bilateral flow redistribution. During the first 24 hours the patient remained in a mixed type of shock (both cardiogenic and vasoplegic) requiring vasoactive drugs, with multiorgan failure and marked coagulopathy; then, post cardiac arrest care, hemodynamic support, and internal correction with ventricular support were implemented. After the critical fibrinogen values were stabilized, an anticoagulant treatment with sodium heparin administered by continuous IV infusion was initiated and adjusted to anti-factor Xa values. His progress was favorable, the vasoactive drugs were reduced, and right ventricular function improved according to the TTE, so at 72 hours the ECMO weaning test was performed, and it was decided to withdraw circulatory support with no complications. The patient remained under orotracheal intubation, with intact neurological response and good urinary volume with response to intravenous diuretics (despite elevated serum creatinine levels with a peak of 10 mg/dL, but without requiring hemodialysis); therefore, 7 days after admission, the patient was successfully extubated. Subsequent exhaustive kinetic motor and respiratory rehabilitation was performed; antiphospholipid syndrome and other thrombophilias were ruled out; negative fluid balance with forced diuresis at the expense of furosemide was performed with good response, so the patient was transferred to his center of origin to continue with the rehabilitation in the general ward, with an ECG evidencing increased voltages (Figure 2) and a TTE showing completely normalized biventricular function.

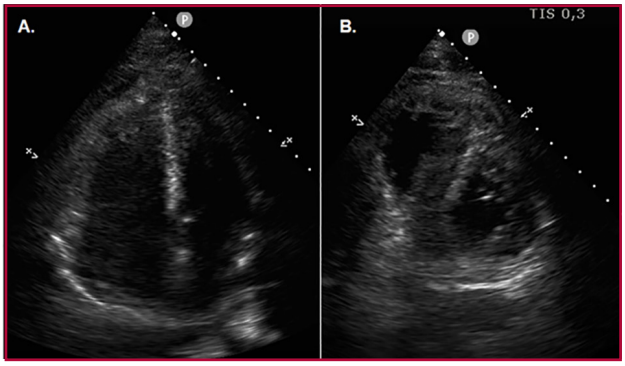

Fig. 1. Transthoracic echocardiogram on admission. A. End-systolic apical four-chamber view. Marked dilatation of right chambers. B. Parasternal short axis. Right ventricular enlargement with flattening of the interventricular septum.

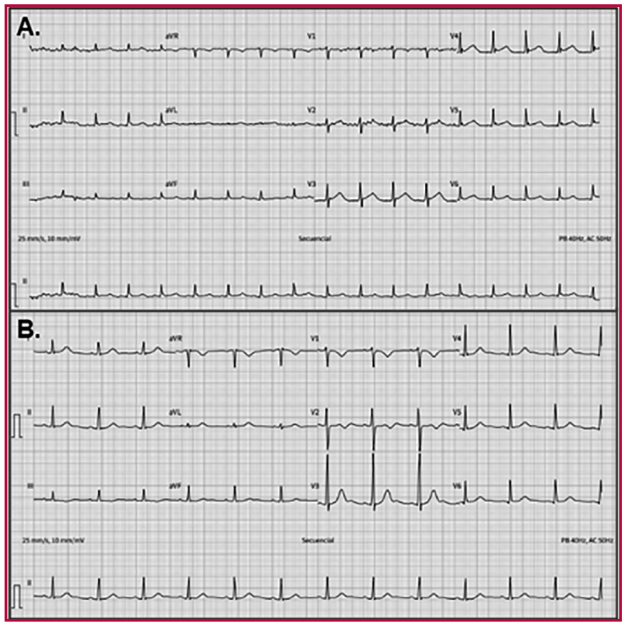

Fig. 2. A. Electrocardiogram on admission showing sinus tachycardia. B. Electrocardiogram at discharge showing sinus rhythm, normal heart rate with increased voltages in general and a more marked difference in right precordial leads.

The PTE is a life-threatening condition, so it is a cardiovascular emergency, with an annual incidence of 70 cases per 100 000 inhabitants. (1) It is the third leading cause of cardiovascular death, following myocardial infarction and stroke. (2,3) Mortality from massive pulmonary embolism is around 30%, while in those who experience cardiopulmonary arrest it can be near 95%. (4) Current clinical practice guidelines recommend immediate reperfusion therapy in patients with high-risk PTE: in patients with hemodynamic decompensation, systemic thrombolysis is recommended, leaving invasive methods (surgical or percutaneous embolectomy or ventricular support) in a second place whenever the former is contraindicated or fails. (5) However, in High Complexity Centers with wide experience in surgical methods (embolectomy and VA-ECMO), by implementing such methods mortality has been drastically reduced in patients with massive PTE, including those who have experienced cardiorespiratory arrest, to whom ECMO is indicated with a mortality rate near 25%. (2,3) Therefore, in a setting like ours, it is essential to create multidisciplinary and multicentre teams

to allow, firstly, early detection of high-risk PTE patients and, secondly, the implementation of action strategies according to each patient, such as referral to high complexity and experienced centers, and, in case of being a candidate, to perform ventricular support or surgical embolectomy when indicated. Thus, by establishing interhospital networks which quickly identify and respond to these needs, the morbidity and mortality associated with this high-risk subgroup could be reduced. Furthermore, a paradigm shift in the management of these patients could be considered, since surgical techniques could be implemented as the initial therapy in the centers where they have been developed, instead of using the rescue therapy, as they do at present, thus achieving a favorable impact on these patients' progress. These could become worldwide guidelines in the future, which in our setting are not yet reproducible. However, with the potential existence of reference centers that centralize referrals of patients requiring surgical salvage therapy from surrounding centers, in the future it could be implemented as the initial therapy according to the experience.

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web).

Ethical considerations

Not applicable.

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Dysphagia – an Uncommon Presentation of a Rare Pacemaker Implant Complication

We present a case of a 63-year-old patient admitted to the emergency room (ER) for complaints of dysphagia. He reported a left-sided neck distension which later affected his capacity to eat appropriately. These symptoms appeared after a pacemaker implantation for second-degree atrioventricular block, two weeks prior. There were no symptoms of heart failure, syncope, pre-syncope, palpitations, dizziness, constitutional symptoms, or fever. The physical examination showed a cervical asymmetry, with a distended left side, associated with local warmth, and discrete asymmetry between the right and left arms. The rest of the physical examination was unremarkable. The patient has a history of schizophrenia, depression, and dyslipidemia. Following an invasive procedure, the authors suspected an infection related to the procedure as the point of origin.

The patient underwent a computed tomography (CT) scan that reported thrombosis of the left internal jugular vein, with occlusion of the left brachiocephalic and subclavian veins (shown in Fig. 1). There was no evidence of collections/abscesses. A brief transthoracic echocardiography was performed in the ER, which rose the suspicion of thrombus/endocarditis. The subsequent transesophageal echocardiogram documented a thrombus adherent to the pacemaker leads. There was an equivocal image of vegetation. Blood cultures, blood panel with C-reactive protein (CRP) and procalcitonin (PCT) were collected. The patient was also scheduled for a repeat CT for evaluation of pulmonary embolism.

Anticoagulation was immediately started with low molecular weight heparin (LMWH). Blood cultures and PCT were normal. There was no evidence of pulmonary embolism on the new CT scan.

After treatment with LMWH for a week, there

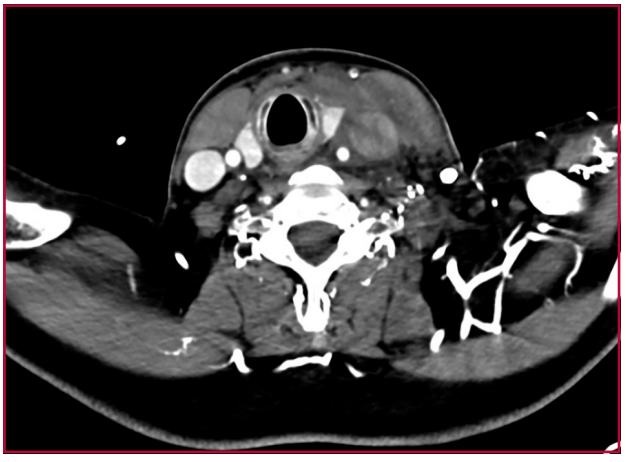

Fig. 1. CT scan – axial plane. Carinal shift due to jugular vein thrombosis and ipsilateral edema.

was a complete resolution of the thrombus (shown in Fig. 2). The patient was also seen in the immunohemotherapy clinic, and thrombophilia was discarded. After a 12-month follow-up, the patient remains asymptomatic and generally well.

Reported permanent pacemaker complications are mostly related to the risk of infection and thrombosis and embolic events. Other rarer complications also described are beyond the scope of this scientific letter. (1) Serious thrombotic events related to pacemaker implantation have been described, with an incidence from 0.6% to 3.5%. These serious events include heart failure presentation and pulmonary thromboembolism. (2) Nevertheless, clinically asymptomatic thrombus appears to be much more frequent, with an incidence of up to 35-45% in the same cohorts. (3) The symptomatic cases can be present in the acute, subacute or delayed setting, varying from days to years after lead implantation. (2,4) In the acute setting, there seems to be a hypercoagulable state and endothelial trauma that favors thrombosis. (5) There is no consensus on a therapeutic strategy and follow-up of the patient with symptomatic lead-induced thrombosis. There are some cases when oral versus intravenous medical therapy is discussed, as well as mechanical thrombectomy and thrombolysis, mostly in acute cases. (2)

We report on a case of an atypical presentation of thrombosis with the most prominent symptom being dysphagia. Early echocardiographic evaluation and therapy can be essential to prevent, avoid and improve clinical outcomes in these patients with no need for interventive action. We aim to bring to the reader's attention that acute pacemaker thrombosis is an entity that requires medical community awareness to an early diagnosis and prevent worse outcomes. Atypical presentations should not be disregarded and time from an intervention should raise our suspicion of its connection. Not everything attached to a lead or related to a recent intervention is a vegetation.

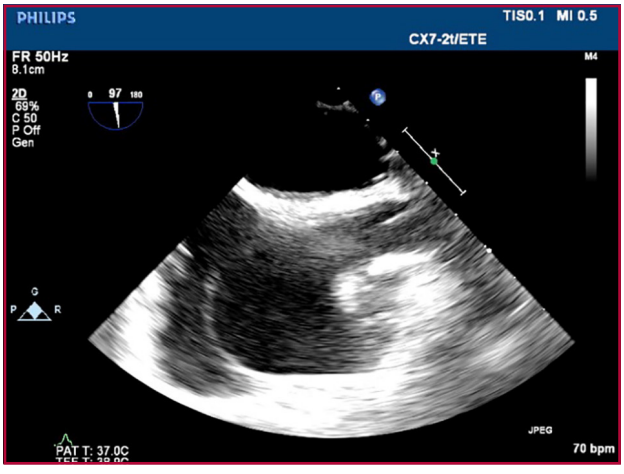

Fig. 2. Transesophageal echocardiography bicaval plane. Post anticoagulation image of the pacemaker leads, without evidence of thrombus

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website).

Ethical considerations

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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