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Tápanes Daumy, Hiram; Díaz Ramírez, Francisco; Peña Fernández, Maylin; Senra Reyes, Lysmara
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Late surgical correction of anomalous origin of the left pulmonary artery and ventricular septal defect

Hemitruncus is defined as anomalous origin of one branch of the pulmonary artery from the ascending aorta with the other branch arising normally from the pulmonary trunk. (1) This acyanotic heart defect with increased pulmonary blood flow with manifestations of severe pulmonary artery hypertension (PAH) and heart failure should be corrected at early stages of life. We describe the case of a 9-month-old infant with late diagnosis of anomalous origin of the left pulmonary artery emerging from the aorta, associated with ventricular septal defect, who underwent successful corrective surgery.

A 9-month-old infant was admitted with heart failure. She had a history of hospitalizations due to recurrent pneumonias. She had signs of protein-energy malnutrition, marked hypernea, subcostal and low intercostal retractions, harsh vesicular breath sounds and rhonchi. Cardiovascular auscultation revealed loud P2, ventricular gallop, and an early and midsystolic murmur grade III/VI heard at Erb's point radiated to the left and right paraesternal borders. An early diastolic murmur of pulmonary regurgitation was heard at the pulmonic area (Graham Steell murmur). The amplitude of peripheral pulses was low. Chest X ray showed a cardiothoracic ratio of 0.68, bilateral hilar congestion, increased pulmonary blood flow specially in the right pulmonary field and peripheral vessel oligemia in the left pulmonary field. The electrocardiogram showed sinus rhythm, right axis deviation (+120°), increased R wave amplitude in V1-V2 and deep S waves in V5-V6 (right Sokolow-Lyon index). The echocardiogram defined the anomalous origin of the left pulmonary artery emerging from the ascending aorta, with severe PAH with suprasystemic pulmonary artery pressures, biventricular dilatation and moderate left ventricular dysfunction with ejection fraction of 43%. A diagnostic cardiac catheterization was performed which confirmed the severity of PAH estimated by transthoracic echocardiography (Figures 1 A and B). Vasoreactivity testing demonstrated a discrete reduction of pulmonary artery pressures and pulmonary resistance index. The patient underwent corrective surgery considering the presence of refractory heart failure and severe but potentially reversible PAH. The procedure consisted of the reinsertion of the left pulmonary artery in the main pulmonary artery and closure of the ventricular septal defect with a fenestrated patch. During the postoperative period, the institutional protocol for PAH was initiated, which includes intravenous high-dose sildenafil and epoprostenol associated with furosemide, spironolactone, captopril and carvedilol for left ventricular dysfunction. The patient evolved with favorable outcome and is followed-up at the outpatient clinic. A computed tomography angiography performed after 6 months of surgery confirmed the effective surgical correction (Figure 2 A and B) and serial echocardiograms show normalization of ventricular dimensions, pulmonary artery pressures and left ventricular function.

The anomalous origin of one of the pulmonary arteries from the aorta was initially described by Fraentzel in 1868, and is commonly associated with other congenital heart defects as tetralogy of Fallot, ventricular septal defect, transposition of the great vessels and subvalvular aortic stenosis. (2)

The largest case series published on this heart disease is attributed to Kutsche and Van Mierop in 1988, who described 108 patients, 89 with anomalous origin of the right pulmonary artery and 19 of the left pulmonary artery emerging from the aorta. (3) Other authors report that the anomalous origin of the right pulmonary artery is 4 to 8 times more frequent than the involvement of the left pulmonary artery. (1)

The embryological development of these varieties of hemitroncus is different: anomalous origin of the right pulmonary artery is thought to be secondary to incomplete or delayed leftward migration of the right sixth aortic arch, while the defect of the left pulmonary artery is considered as failure in the development of the left sixth aortic arch and persistence of the fifth aortic arch. (4)

The pathophysiology of PAH in this congenital heart disease is due to two mechanisms: one of the pulmonary artery branches is subjected to systemic pressure from the ascending aorta and the remaining artery receives the entire output of the right ventricle. (1)

Estimating pulmonary artery pressures and pulmonary vascular resistance index is crucial to define the possibility of surgical correction. In the presence of ventricular septal defect, the traditional calculations by the Fick method may fail and other variables should be analyzed, as manifestations of heart failure, pulmonary venous flow velocity entering the left atrium and the presence of cardiomegaly in the chest X-ray. In these cases, the estimation of pulmonary artery flow and pulmonary vascular resistance index should be calculated for each pulmonary artery with cardiac

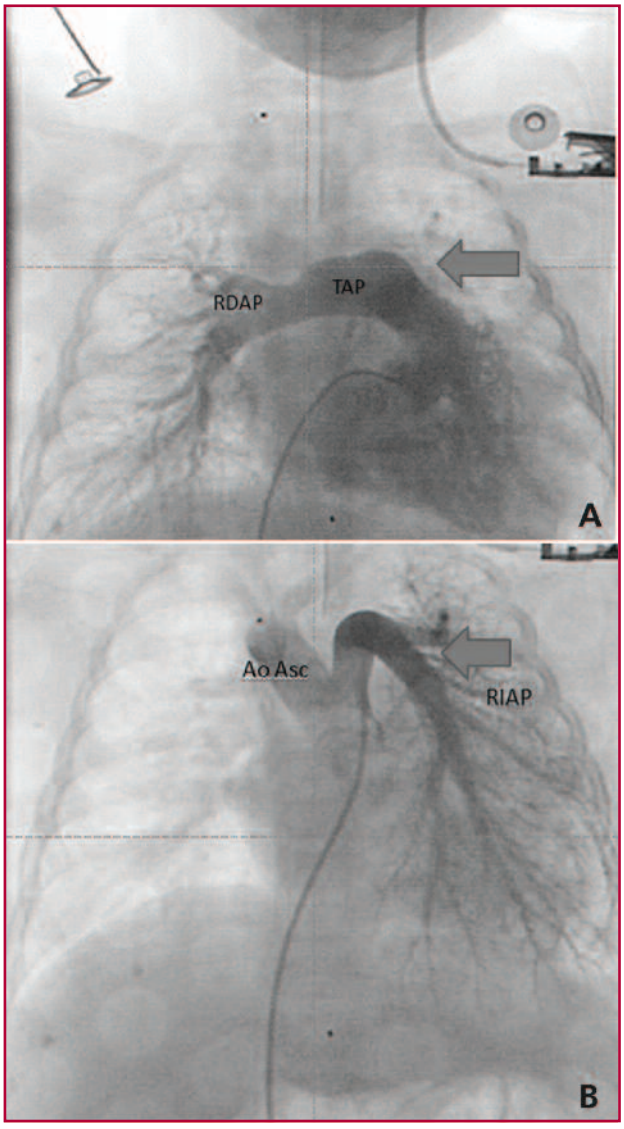

Fig. 1. A. Coronary angiography image showing the pulmonary artery trunk (TAP) emerging from the right ventricle. The arrow shows absence of the left pulmonary artery branch. RDPA: Right pulmonary artery **B.** Coronary angiography image with the left pulmonary artery (RIAP) emerging from the ascending aorta (Ao As). The arrow shows the left pulmonary artery branch.

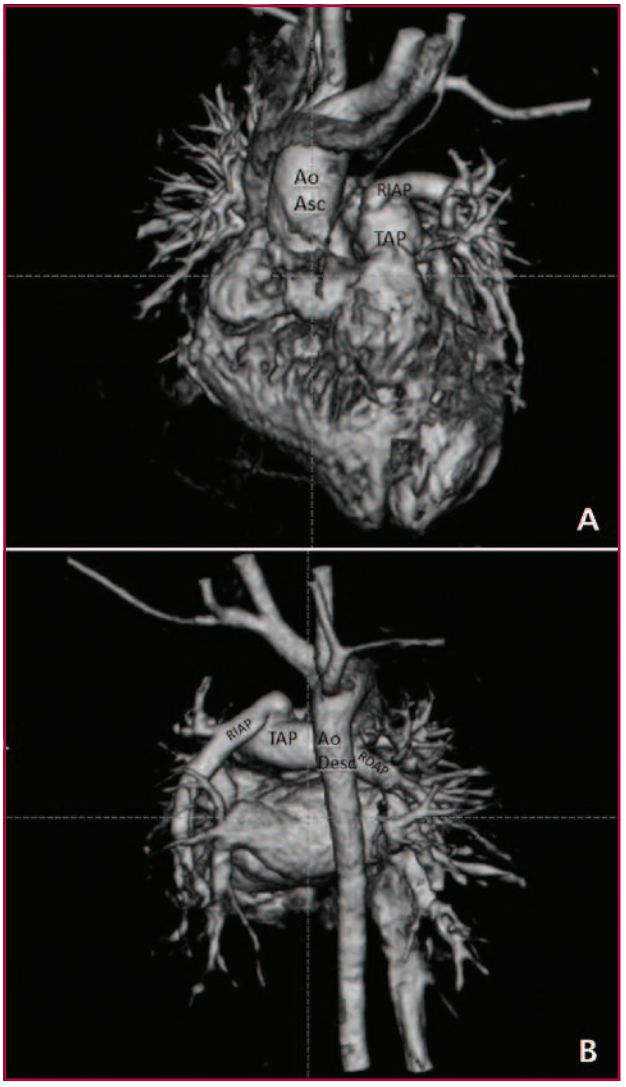

Fig. 2. A. Volume reconstruction of computed tomography coronary angiography (oblique view), showing the surgical correction with the left pulmonary artery branch emerging from the pulmonary artery trunk. Ao Asc: Ascending aorta. RIAP: Left pulmonary artery. TAP: Pulmonary artery trunk. **B.** Volume reconstruction of computed tomography coronary angiography (posterior view), showing the surgical correction with native right pulmonary artery branch emergence and the corrected origin of the left pulmonary artery from the pulmonary artery trunk. As Desc: Descending aorta. RDAP: Right pulmonary artery RIAP: Left pulmonary artery.TAP: Pulmonary artery trunk

magnetic resonance imaging.

Several techniques are described with favorable outcomes in terms of survival. Nathan et al. reported survival of 93% at 20 years in 16 patients operated on, and with almost no need of reinterventions. However, early and timely diagnosis is crucial in this rare congenital heart defect to prevent the development of irreversible PAH, as surgery is not recommended in these cases and is cause of death. (5)

The success achieved with corrective surgery in this patient is very important for our institution, considering that the diagnosis was late and nitric oxide was not available for the treatment of PAH in the postoperative period.

Hiram Tápanes Daumy[1], Francisco Díaz Ramírez[1], Maylin Peña Fernández[2], Lysmara Senra Reyes[1]
1 Cardiocentro Pediátrico "William Soler". Havana, Cuba.
2 Hospital Pediátrico "Juan Manuel Márquez". Havana, Cuba.
Hiram Tápanes Daumy. Cardiocentro Pediátrico "William Soler"
Ave. 43 Nº 1418 esquina A Calle 18 - CP 11900
Ciudad de La Habana, Cuba
e-mail: hiram@cardiows.sld.cu, hiramtapanes@infomed.sld.cu

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Efficacy of multielectrode Array catheter for ventricular tachycardia ablation in a patient with electrical storm

Ventricular arrhythmias are one of the major causes of mortality in patients with coronary artery disease. In this group of patients, ventricular tachycardia (VT) is produced by a reentrant mechanism due to the presence of a fibrotic scar, and may appear even decades after myocardial infarction (MI). (1) Despite the important advances in the treatment of MI, the risk of VT in the population with coronary artery disease is still an important issue in clinical practice. Antyarrhythmic agents and implantable cardioverter defibrillator (ICD) devices are the cornerstone in the management of ventricular arrhythmias. (2) These therapies are far from being infallible and may present significant adverse events. (3) Patients implanted with an ICD may experience significant physical and psychological deterioration due to repetitive delivery of shocks.

Catheter ablation of VT may have a significant impact in patient's quality of life by alleviating symptoms, reducing the requirement of antiarrhythmic drugs and the number of ICD shock deliveries. However, this procedure may result difficult because the arrhythmia is poorly tolerated in many occasions and hemodynamic decompensation make collection of point-by-point electrograms impossible.

The use of the multielectrode Array catheter as a diagnostic tool is becoming more frequent in our environment. (4) This noncontact device consists of 64 electrodes deployed on an inflatable mesh that is suspended in the selected heart chamber to treat the arrhythmia. The catheter records electrograms from multiple sites within a single cardiac chamber and is capable of determining the precise site of origin of the arrhythmia from a single run of tachycardia. We report the case of a 69-year-old male patient treated in our hospital, with history of hypertension, dyslipidemia, and former smoking. His usual creatinine levels were 1.4 mg/dl (creatinine clearance measured by MDRD of 56 ml/min/1.73 m2). Eighteen-years ago he presented myocardial infarction of the inferior wall and received medical treatment. In 2011 he complained of progressive angina and underwent myocardial revascularization surgery requiring implantation of four bypass grafts. At that time, the echocardiogram revealed severe left ventricular dysfunction with abnormal contraction in the inferior and posterior wall. Six months later he was admitted to the emergency department with sustained VT with hemodynamic instability. An ICD device was implanted. During follow-up, three appropriate shocks were delivered due to VT: Amiodarone 400 mg/day and carvedilol 25 mg bid were initiated.

The patient was admitted to the emergency department due to nausea, dizziness, abdominal pain and dyspnea of one-week duration. He reported two shock ICD deliveries. He denied fever or fever-like symptoms. He did not complain of angina and did not modify his usual medication.

He was evaluated by a cardiologist due to heart failure and signs of low cardiac output. At the physical examination, the patient looked severely ill, with heart rate of 140 beats per minute, tachypnea, hypotension, poor distal perfusion and oxygen saturation of 75% breathing room air. He did not present fever. Crackles were heard over both lung bases and middle fields.

The electrocardiogram showed a regular tachycardia of 140 beats per minute with wide QRS complexes (Figure 1), that was interpreted as sustained VT with hemodynamic instability. As the ICD was programmed to treat VT with heart rates >150 beats per minute, therapy with external cardioversion with a 200 J biphasic shock was decided.

The laboratory tests were within normal ranges. The chest X-ray showed signs of pulmonary congestion. Interrogation of the ICD device showed that the patient had presented three appropriate shocks to treat VT within the past 48 hours.

Furosemide was administrated and a negative fluid balance was achieved; amiodarone and lidocaine were given intravenously. Coronary angiography was performed to rule out acute coronary syndrome, and no new coronary artery stenoses were observed. Blood and urine cultures were negative. The patient presented several episodes of VT with hemodynamic instability and other symptomatic, short-lasting episodes despite the treatment. During hospitalization, three ICD shocks were delivered in the range of VT detection.

A diagnosis of electrical storm in a drug-refractory patient with hemodynamic instability was made and VT catheter ablation was indicated using a multielectrode Array catheter. Ventricular tachycardia was induced by programmed stimulation from the right ventricle (RV) (Figure 2) causing hemodynamic instability and was rapidly reverted by electrical shock. Substrate mapping and propagation mapping could be performed using a very few tachycardia beats, allowing the identification of the site of arrhythmia origin. A scar was identified in the anterolateral region of the RV. The area of slow tachycardia conduction was de-