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ANATOMICAL AND VASCULAR ANOMALIES COMPLICATING THE COURSE OF DOUBLE OUTLET RIGHT VENTRICLE IN AN INFANT

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Abstract: *Congenital heart diseases are an important cause of mortality and morbidity in pedatric patients. Fallot Type Double outlet right ventricle is a rare heterogenous congenital heart disease characterized by a complex group of lesions similar with Tetralogy of Fallot. Surgical approach depends on the anatomy of associated lesions, therefore in order to perform a successful correction, a careful review of the anatomy and a detailed diagnostic is needed due to complexity of intracardiac anomalies. The main diagnostic tool used nowadays for the diagnosis of double outlet right ventricle is echocardiography exam, a non-invasive examination that can provide information about anatomical features, but it will not allow the full description of the coronary anatomy. Therefore, preoperative assessment should include, extensive imaging assessment (echocardiography exam, angio-CT exam) for a correct management of surgical strategy prior to surgical correction.*

INTRODUCTION

Congenital heart diseases are an important cause of mortality and morbidity in pediatric patients, with a reported incidence in European countries of approximately 8 cases/1000 births.(1) Double outlet right ventricle (DORV) is a rare heterogenous congenital heart defect with an incidence of 1-3% in live births, characterized by a complex group of lesions in which both great arteries arise mainly from the right ventricle, associated with discontinuity of the interventricular septum of different size and morphology.(2) The different variants of DORV are described mainly by the relationship of the great vessels, the type of ventricular septal defect and the presence or absence of pulmonary stenosis.(3)

One of the most common variants of DORV is Fallot Type, characterized by similar lesions seen in Tetralogy of Fallot: subaortic interventricular communication, dextroposition of the aorta with overriding of the ventricular septum and pulmonary stenosis at different levels (infundibular, valvular, supravalvular).(4)

The clinical presentation of patients with DORV is very variable and it depends on the anatomy (mainly the severity of pulmonary stenosis) or of associated defects. It can range from early onset of cardiac failure with or without cyanosis, but in some patients, signs and symptoms can appear in the first years of life or even in childhood. The Fallot type DORV presents with different degrees of cyanosis, mainly depending on the pulmonary stenosis. Due to complex associated lesions, one major challenge in DORV cases is to describe the appropriate anatomy and morphology of the defects for a successful surgical approach. We present a case of a two-year-old boy diagnosed in postnatal period with DORV associated with complex defects that complicated this case.

CASE PRESENTATION

The two-year old boy was diagnosed in the first weeks of life with Fallot Type DORV with subaortic ventricular septal defect, severe pulmonary infundibular and valve stenosis, hypoplasia of pulmonary valvular ring, pulmonary trunk and branches, associated with anomalous origin and course of a major coronary artery branch.

On review of his medical history, at 3 months old, the patient was admitted to our clinic due to multiple hypoxic crisis, diaphoresis and difficulty with feeding. Physical evaluation revealed growth failure, muco-tegumentary cyanosis (oxygen saturation of 80%) and III/VI systolic ejection murmur audible on the left side of the sternum.

A transthoracic echocardiography was performed which revealed normal atrioventricular connections with enlarged right cavities that led to compression of left cavities and subaortic ventricular septal defect. The great arteries were both arising mainly from the right ventricle, the aorta was overriding the ventricular septum (figure no. 2A). Right ventricular outflow obstruction was observed, with severe pulmonary infundibular and valvular stenosis (peak gradient of about 83 mmHg), hypoplasia of pulmonary valve (z score of -2.73) and pulmonary trunk (z score of -1.97) and pulmonary branches (z score of -2.93 for right pulmonary artery, respective -3.23 for left pulmonary branch) - (figure no. 2B).

Due to anatomical features, namely the presence of the pulmonary branches' hypoplasia, corrective surgery was not feasible, therefore palliative procedure was required. In 2017 the patient benefited from the construction of a systemic-to-pulmonary Blalock-Taussig arterial shunt.

Over time, the patient was periodically monitored. At the age of 1.8 years, the patient was readmitted in our clinic with

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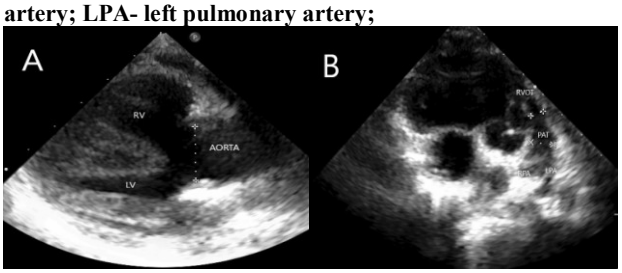
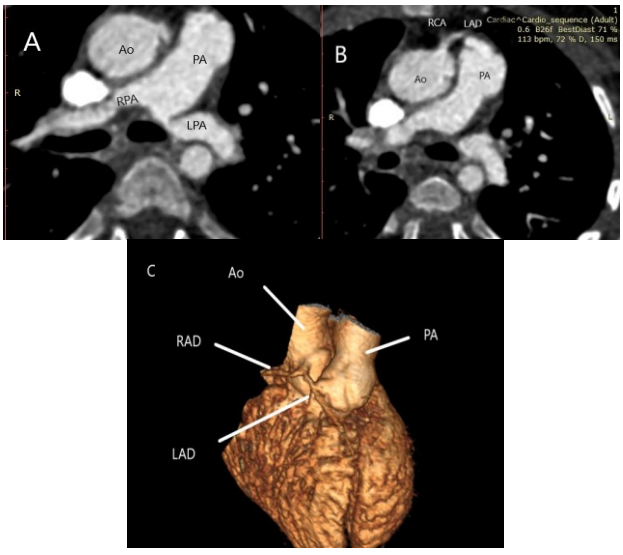
signs and symptoms of hypoxemia. Central and peripheral cyanosis was present with low oxygen saturation of 68-70% associated with moderate limitation of physical activity and growth failure. Transthoracic echocardiography revealed restrictive aorto-pulmonary shunt.

Figure no. 1. Transthoracic echocardiograms (parasternal long-axis view and apical five chambers view) of our patient with Fallot-Type double-outlet right ventricle A) Sub-aortic ventricular septal defect and the aorta overriding interventricular septum. B) Severe sub-pulmonary and pulmonary valve stenosis with hypoplasic pulmonary artery trunk and pulmonary artery branches; RV- right ventricle; LV- left ventricle; RVOT- right ventricle outflow tract; PAT- pulmonary artery trunk; RPA- right pulmonary artery; LPA- left pulmonary artery;



To obtain better view of the aorto-pulmonary shunt, vascular anatomy of pulmonary trunk, arterial branches and also coronary arteries, a preoperative angiographic computer tomography was performed. The angio-CT exam revealed mild stenosis of right pulmonary branch, kinking of left pulmonary branch and anomalous origin of left anterior descending artery that emerged from right coronary artery with an abnormal course across right ventricular outflow tract.

Figure no. 2. Cardiac computed tomography angiography exams of our patients with Fallot-Type double-outlet right ventricle A) Pre-operative CT-image reveals mild stenosis of right pulmonary branch and kinking of left pulmonary branch. B) Pre-operative CT exam in transverse plane reveals the left anterior descending artery origin from the right aortic sinus C) Pre-operative volume-rendered CT exam shows the left anterior descending artery origin from the right aortic sinus. In addition, this image shows the left anterior descending artery crossing the right ventricle outflow tract. Ao- aorta; PA- pulmonary artery; RCA- right coronary artery; LAD- left anterior descending artery;



The case was discussed with the surgeons in order to perform secondary corrective surgery. Due to severe pulmonary infundibular stenosis and abnormal course of a major coronary artery branch, the classic corrective surgery was not feasible and the selected surgical approach was to insert an external Contegra conduit between the right ventricle and pulmonary artery trunk. Postoperatively, the patient developed sinus bradycardia that required temporary pacing, but normal sinus rhythm recovered within several days post cardiac surgery. The patient was discharged after three weeks.

DISCUSSIONS

Double outlet right ventricle was first described in 1793 as a partial form of transposition of great vessels, but the "double outlet right ventricle" term was first used in 1962, when this disease was described.(1) However, despite the fact that diagnostic tools encountered significant progress, DORV is still a pathology less understood that can present itself as a single condition or it can be associated with multiple cardiac or extracardiac defects.

First of all, according to Walters et al, the definition of DORV is still a controversial problem between authors due to similarities with other congenital heart diseases such as Tetralogy of Fallot and Transposition of Great Arteries.(5) According to Congenital Heart Surgery Nomenclature and Database Project, DORV is defined as a discordant ventriculo-arterial pathology in which valve origin of the great arteries arise from the morphological right ventricle, either entirely or more than 50% - the "50% rule".(5) Other authors include in this definition other criteria such as the loss of normal fibrous continuity between the mitral and aortic valve due to the fact that in DORV, aortic valve is both structurally and anatomically detached from the left ventricle.(6) Based on the current classification, there are described four types of DORV: the VSD type, Tetralogy of Fallot type, Transposition of the Great Arteries type and remote type.

The most common type of DORV is Fallot-Type, with similar lesions seen in Tetralogy of Fallot, therefore the most important features are the position of the intraventricular communication and the severity of pulmonary outflow restriction. In most cases of Fallot-type DORV, the intraventricular communication is situated below the origin of the aortic valve.(4) This type of ventricular septal defect is called "subaortic" due to the absence of sub-aortic septum. Physiologically, the amount of oxygenated blood that enters the systemic circulation is ensured by the subaortic intraventricular communication, that directs the blood flow right towards the aorta. This type of ventricular septal defect is associated with either subpulmonary or valvular stenosis due to an abnormal outlet septum and right ventricle hypertrophy.(5) Usually, in the postnatal period, most patients have satisfactory arterial oxygenation saturation, therefore surgical treatment is not necessary. However, if pulmonary stenosis is severe, an amount of deoxygenated blood is directed into aorta and pulmonary blood flow decreases, therefore patients can present with hypoxemic spells.(4)

The main diagnostic tool used nowadays for the diagnosis of Fallot type DORV is echocardiography exam. One of the main advantages is that echocardiography is a non-invasive examination that can provide information about atrio-ventricular and semilunar valves, ventricular morphology and function, can establish the interrelationship between the major arteries and associated defects.(7) Staged approach is preferred due to complexity of intracardiac anomalies.

Some studies reported that primary corrective surgery may be high risk in the first months of life due to high reintervention rates and increased mortality.(8) Therefore, in

some cases, palliative surgery is considered a safer alternative at first (depending on the anatomy), followed by a staged corrective surgery. Regarding survival aspects, Oladunjoye et al reported that initial palliative surgery followed by total repair was associated with similar survival rates than primary corrective surgery.(9) In our case, because of the frequent hypoxic spells, the patient benefited in the first months of life from a palliative approach: a systemic-to-pulmonary shunt. This surgical approach was preferred due to the anatomical features (hypoplastic pulmonary branches).

Regarding the next steps, preoperatively, in order to ensure the success of surgical correction, some key aspects had to be identified. First of all, the echocardiography exam should identify the size and morphology of interventricular communication in order to ensure that systemic blood flow is unobstructed.(10) In our case, the ventricular septal defect was localized in a subaortic position, therefore the surgical approach was focused to establish a continuity between left ventricle and aortic valve by closing the ventricular defect with a patch while creating a pathway to direct left ventricle flow into the aorta.

Secondly, another important modification associated with DORV and subaortic ventricular septal defect in Fallot Type DORV, is the obstruction of right ventricle outflow tract. Usually, the reconstruction of right ventricular outflow tract is performed by a transannular patch associated with complete sub pulmonary muscular resection if sub pulmonary stenosis is present. But prior to corrective surgery, surgeons should assess if coronary artery anomalies that cross the right ventricle outflow tract are present in order to avoid coronary damage during muscle resection. Despite the fact that studies report a small incidence of coronary artery anomalies in DORV (2-14 %) (11,12,13), a complete preoperative assessment of coronary anatomy should be performed for a successful surgical approach. Detection of some coronary artery anomalies, such as abnormal course across the right ventricular outflow, is possible from echocardiography exam, but it does not allow the full description of the coronary anatomy. Cardiac catheterization is considered a valuable tool for determining the accurate coronary anatomy; however, it is an invasive procedure with risks in pediatric patients.(14) This is emphasized by the fact that some coronary anomalies cannot be detected intraoperatively in some patients with previous surgeries due to pericardial adhesions.(11)

Studies focused on evaluating non-invasive methods that can identify a detailed coronary anatomy, related that the accuracy of angio-CT exam is almost 97% in describing coronary anomalies in pediatric population.(13) In our case, CT exam revealed that left anterior descending artery emerged from right coronary artery with an abnormal course across right ventricular outflow tract.

In our case, the classic corrective surgery was not feasible, and the surgical technique was based on inserting a right ventricle-pulmonary artery conduit to preserve the normal coronary artery flow. Therefore, preoperative assessment should include, besides the echocardiography exam, angio-CT exam for a better visualization of coronary anatomy and pulmonary artery branches.

CONCLUSIONS

Double outlet right ventricle represents a rare disease characterized by a group of associated heart defects. Due to the fact that surgical approach depends on the anatomy of associated lesions, in order to perform a successful correction, a careful review of the anatomy and a detailed diagnostic is needed.

In this report we aimed to emphasize the crucial role of an extensive imaging assessment (echocardiography, angiography) prior to surgical correction for a correct management of surgical strategy.

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