

A Successful Correction of a Truncus Arteriosus in an 8-Year-Old Girl with Functional Pulmonary Stenosis by a Particular Opening of the Truncal Valve

Karima Hami, Rida Ajaja, Younes Cheikhaoui

Congenital Cardiology Unit, Cheikh Zaid Hospital, Abulcassis University, Rabat, Morocco

Email address:

Hami7esp@yahoo.com (Karima Hami)

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Abstract: *Background:* Truncus arteriosus is a rare congenital heart malformation requiring repair in the neonatal period. Without correction, the majority of patients die within the first year of life. However, this case shows that some patients remain operable even at an advanced age. *Case presentation:* We report the case of an 8-year-old girl with type 2 truncus arteriosus with pulmonary vascular protection by a particular opening of the truncal valve partially obstructing the origin of the pulmonary artery in systole and causing a reduction in pulmonary flow. The initial evaluation based on the clinical presentation, the electrocardiogram and the echocardiography showed a significant elevation of the pressures of the right ventricle with a functional stenosis at the origin of the pulmonary artery. Hemodynamic exploration demonstrated that the pulmonary arterial hypertension is reversible, thereby the patient was successfully operated. *Conclusion:* The objective of this observation is to demonstrate that truncus arteriosus referred late does not eliminate the possibility of complete surgical repair, which underlines the importance of an exhaustive evaluation of these patients based on several arguments.

Keywords: Congenital Heart Defect, Truncus Arteriosus, Pulmonary Hypertension

1. Introduction

Truncus arteriosus (TA) is a rare congenital cyanotic heart condition representing less than 3% of all Congenital heart diseases CHD, characterized by a ventricular septal defect (VSD), a single arterial outflow trunk arising from the both ventricles overriding the defect, supplying the systemic, coronary and pulmonary circulation. Truncus is capped by a single semilunar valve which is frequently structurally abnormal and cusps poorly supported leading to varying degree of truncal incompetence [1]. This defect requires repair within the first weeks in life; without surgical repair in early infancy, death is ineluctable in the majority of cases (80%) before 1 year of age [2]. However, rare survivals of non operated TA with reversible pulmonary arterial hypertension were seen in isolated case reports [3, 4].

2. Case Report

A 8-year-old girl was diagnosed recently to have type II truncus arteriosus based on Collette and Edwards' classification. She was minimally symptomatic in the neonatal period and early childhood, she has a mild cyanosis and New York Heart Association (NYHA) functional class I. Her geographical location in a rural environment did not allow an early diagnosis which was only made 1 year ago, due to the worsening of her functional class becoming Dyspnea stage II of the NYHA.

On physical examination, she was thin, with finger clubbing. On auscultation, there was an ejection systolic murmur at the left parasternal edge. The oxygen saturation was 90% on room air. Blood investigation was normal except a high level of hemoglobin (17.9 g/l) Electrocardiogram revealed right ventricular (RV) hypertrophy. Chest

radiograph (figure 1) showed an enlarged heart with prominent pulmonary trunk and increased pulmonary vascularity.



Figure 1. Chest X-ray showing cardiomegaly, dilated pulmonary artery, and pulmonary plethora.

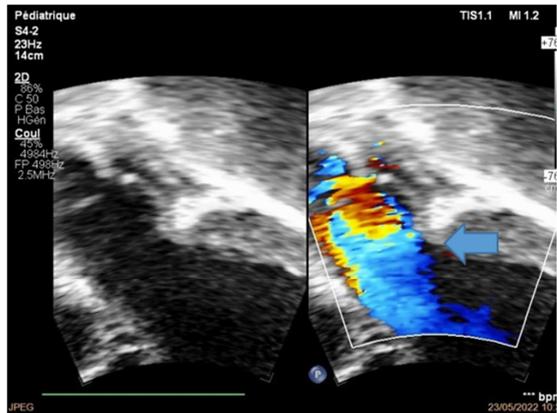


Figure 2. Trans-thoracic echocardiogram: Interposition of the truncal valve on systole at the origin of the PA (blue arrow).



Figure 3. Peroperative finding: truncal valve closing the origin of the Pulmonary artery in systole (arrow).

Trans-thoracic echocardiogram showed type II TA based on Collette and Edwards’ classification (Figure 2). The left ventricle (LV) was severely dilated with mild LV systolic dysfunction, ejection fraction estimated at 42%. The truncal annulus was dilated with mild regurgitation. The right ventricle (RV) systolic pressure was 116 mmHg with severe RV hypertrophy. Pulmonary flow was moderately accelerated in systole, with a gradient of 29/49 mmHg due to the interposition of the posterior truncal valve at the origin of the PA.

Invasive hemodynamic study revealed no significant rise in PA pressure. Calculated pulmonary vascular resistances (PVR) were 6 Wood units significantly dropped to 2 Wood units after oxygen and nitric oxid (NO).

Table 1. Oxymetry data obtained at cardiac catheterization.

	Fio 21%	Fio 2100% + NO 20ppm
QP I (L/min/m ²)	11.93	14.78
QS I (L/min/m ²)	3.29	4.07
QP/QS ratio	1.36	3
PVRI (Uw.m ²)	6.45	2

QPI, pulmonary blood flow indexed; QSI, systemic blood flow indexed; PVRI, pulmonary vascular resistance index.

Because of the clinical, radiological and ultrasound data, of the Left to right shunt limited by this functional stenosis due to the interposition of the truncal valve at the origin of the pulmonary artery (PA), supported by the catheterization data, a decision of complete repair was made. Surgery was performed via a standard median sternotomy and pericardiotomy. The establishment of cardiopulmonary bypass included ascending aorta, superior and inferior vena cava cannulation and core cooling to 25°C. Circulatory arrest was achieved by fibrillation, then the cardiopulmonary bypass and aortic cross-clamping was applied. Once the truncus was transected, the PA orifice and the coronary arteries were identified.

Intraoperative findings were consistent with preoperative imaging: The truncal valve was tricuspid, The truncus root was very dilated and overriding into the RV (>50%).

An autologous pericardial patch was used to create the RV outflow tract with the placement of a homograft. The distal edge of the homograft was cut at an angle and then incised to enlarge the anastomotic suture line. The VSD closure was performed using a Dacron fenestrated patch. At the end of the procedure, a trans-esophageal echocardiography was performed to check the VSD closure, and the patency of pulmonary conduit.

In the early post-operative period, the brief use of NO was required because of the pulmonary arterial hypertension. Patients was electively ventilated for 24 h and then gradually weaned of the mechanical ventilator support, she left intensive care after 48 h.

The ultrasound control before the discharge has being reassuring, it showed a good systolic function of the 2 ventricles, a moderate truncal valve regurgitation and a laminar flow on the RV-PA conduit.

3. Discussion

The age of repair of the truncus arteriosus has a major impact on the outcome, thereby, most survivals non-operated patients with truncus arteriosus develop pulmonary vascular disease early in life. Urban [5] demonstrated that pulmonary arterial hypertension crisis are found 6 times more in children operated after the age of 3 months than those operated before.

The management of children with a TA presenting after one year is challenging and not standardized, it requires a rigorous preoperative evaluation looking for the signs of left-right shunt, based on the clinic, chest radiography and echocardiographic data, which will be supplemented by a hemodynamic evaluation if doubt persists about the state of pulmonary resistance. The clinical features suggestive of operability include history of recurrent respiratory infection, minimal desaturation on exercise, cardiomegaly on chest radiography and left ventricular volume overload [6].

The relation between pulmonary resistance and surgical risk was only studied for those with presence of both right and left PA, and it was higher in patients with indexed pulmonary vascular resistances PVR (iPVR) >8 Wood units.m² pre-operatively [7]. In fact, the PVRi of 6–8 Wood units.m² was widely accepted as a cut-off for operability in patients with non-restrictive left to right shunts [8].

In Gouton's work [9], in which 33 children were operated late, catheterization was performed in 15 children, the other children were operated only on the clinical, radiological and echocardiographic arguments of persistence of a left-to-right shunt. A cut-off of 88% was retained as an operability parameter.

Chen [10] reported his retrospective study including fifty patients, repaired for TA, among which thirty patients older than one year, there was no significant difference in postoperative morbidity and mortality with those operated early.

In our patient, echocardiography showed dilation of the left atrium and ventricle, the functional stenosis caused by the particular opening of the truncal valve in the trunk of the pulmonary artery. We decide to complete this assessment by the cardiac catheterization which demonstrated a significant drop in pulmonary vascular resistance after administration of NO and oxygen by mask for 10 min. However, many authors have questioned the validation of the oxygen test as a parameter for evaluating the reversibility of pulmonary resistance and also the long-term outcomes [9].

In the other hand, the correlation between the pulmonary arterial pressure and the size of the main pulmonary artery was reported in previous studies in children [11, 12]. However, in the absence or hypoplasia of the pulmonary artery, the size of bilateral PAs was used.

Zhu [13] reported that 22,4% of patients presenting with large pulmonary arteries were more predisposed to have the pulmonary hypertension crisis after repair.

This case is probably the only of its kind because of the protection of the pulmonary vascular bed by the flap opening

of the truncal valve which creates a pulmonary functional obstacle in systole. Surgical strategies to prevent right ventricular decompensation in the immediate post operative phase and probably in the mid and long-term due to pulmonary hypertension include creation of a patent foramen ovale or a fenestrated patch closure of the ventricular septal defect.

Both these foramen serve as the site for significant right to left shunt whenever the right ventricular pressure becomes suprasystemic secondary to severe pulmonary vascular hypertension. This serves to maintain the cardiac output at the expense of systemic desaturation. The use of an unidirectional valved patch for closure of VSD is a good strategy in patients with borderline operability [14]. It allows right to left shunt in case of very high pulmonary artery pressure and prevents any left to right shunting or RV failure in the middle and long term. Novick [15] demonstrates that the use of Flap Valve Double Patch Closure of Ventricular Septal Defects allows a reduction in mortality in children with increased pulmonary resistance.

The immediate postoperative prognosis is linked to the onset of arterial hypertension crisis, which generally evolves favorably with NO and Sildenafil. After discharge from the hospital, Sildenafil is continued for about 6 months at least. [6]. Concerning the reoperation, the short and long-term prognosis of patients operated late is similar to that of patients repaired early, involving the need to change the pulmonary conduit and to repair the truncal valve if necessary [16, 17].

The long-term results in these patients operated late are not well known since the pulmonary vascular disease is often progressive despite surgical correction in a significant proportion of cases. long-term follow-up including cardiac catheterization is necessary [6].

4. Conclusion

This case demonstrated the importance of rigorous assessing patient's operability in older children. This patient has a particular form of opening of her truncal valve as a clapet which enormously reduces the systolic flow towards the pulmonary arteries, which allows a protection of the pulmonary vascular bed against fixation of pulmonary resistance.

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