Complete Repair in Total Atrioventricular Canal Defect with Cyanosis

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Atrioventricular septal defects account for 4% of congenital cardiac malformations and over 50% of cardiac defects seen in Down syndrome[1]. Clinical presentation is marked by congestive heart failure early in infancy. Cyanosis is rarely found in infants and suggests irreversible pulmonary hypertension or associated cardiac defects as tetralogy of Fallot, double outlet right ventricle[2], Ebstein anomaly[3], persistent left superior vena cava draining in the left atrium (Barbero Marcial, personal communication). Children with Down´s syndrome is particularly difficult to assess because they often suffer from upper airways obstruction[4], which may contribute to the increased pulmonary vascular resistance determined at cardiac catheterization. This association of factors becomes a challenge for operability and, we will report one such case.

Moderate cyanosis (room air oxygen saturation 75%) was presented in a 7-month-old-girl with Down syndrome and total atrioventricular septal defect (AVSD). Despite cyanosis, she still presented some signs of pulmonary overflow as failure to thrive, dyspnea, hepatomegaly, and a left sternal border systolic murmur. Cardiac catheterization showed Qp/Qs = 1,67 and pulmonary vascular resistance of 6,5 U/m^2.

Complete repair was made with two-patch technique and a small persistent foramen ovale (PFO) was maintained opened. After cardiopulmonary bypass (CPB) the O_2 sat was 80% (FiO_2 100%, NO 20 ppm). Open lung biopsy was made in the third postoperative day (POD) when the thorax was closed and revealed Heath-Edwards grade III of pulmonary hypertension.

Nitric oxide was maintained until 6th POD and calcium channel blocker was introduced with improved O_2 saturation. After a long postoperative period marked by congestive heart failure and infection, she was discharged without cyanosis, in functional class II (NYHA).

Case Report

The patient was a 7 month-old-girl with Down syndrome and a history of cardiac murmur since birth, tachypnea and failure to thrive. She was previously hospitalized for a bronchopneumonia, when cyanosis was noted (O_2 sat 80%-85%). Echocardiogram showed total AVSD with a large ventricular septal defect (VSD), common AV valve with moderate regurgitation and an ostium primum (OP) septal defect. Systolic pulmonary artery pressure was estimated by echo in 53mmHg.

On physical examination, there was a moderate cyanosis (O_2 sat 70%-80%), a loud systolic murmur in the left sternum border and hepatomegaly.

Electrocardiogram revealed synus rhythm with signs of right ventricular overload. The chest X-ray showed moderate cardiomegaly and increased pulmonary vascularity. Pulmonary wedge angiogram was normal (Fig 1), pulmonary vascular resistance was 6,5 U/m^2, with a Qp/Qs 1,67(FiO_2= 100%).

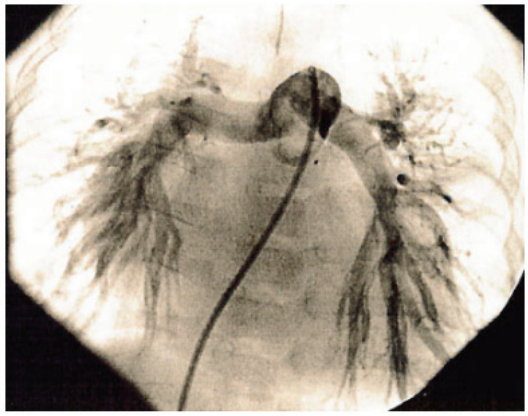

***Fig. 1** - Preoperative pulmonary angiogram.*

Complete surgical repair was made using 20°C hypothermic cardiopulmonary bypass. The large and typical VSD was closed with bovine pericardium. AV valve

Key words

Complete atrioventricular septal defect, cyanosis, surgical repair.

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was divided without any residual regurgitation, and ostium primum ASD was closed with autologous pericardium. A small PFO (4mm) was maintained due to the high pulmonary artery pressure. After cardiopulmonary bypass (CPB), TEE showed any valve regurgitation, VSD and ostium primum ASD closed, and a small right-to-left shunt through the PFO. In sinus rhythm, the O2 sat was 80% (FiO2 100%; NO 20ppm). The sternum was maintained open and a PA catheter was inserted through the right ventricle.

The sternum was closed on the 3rd PO, and a left pulmonary biopsy was made due to persistent high PA pressure (systemic level) with pulmonary hypertension crisis and hypoxemia. The pulmonary biopsy showed grade III Heath-Edwards classification (Fig 2).

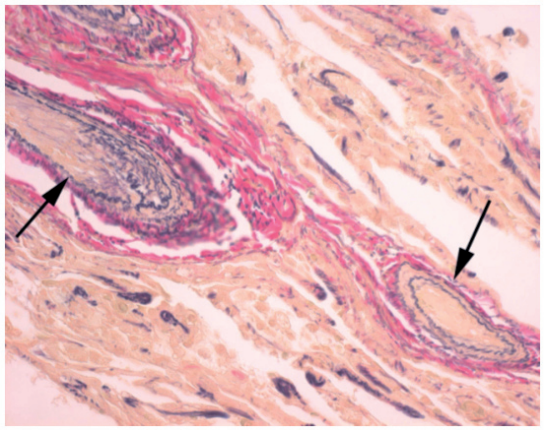


Fig. 2 *- Postoperative lung biopsy. Microphotography of lung biopsy showing two intra-acinar arterioles with complete obstruction by cellular proliferation (dark arrows). Miller color to elastic fibers. Objective 40x*

Nitric oxide was maintained until 6th PO and calcium channel blocker (nifedipine) was introduced with improve of O2 sat, 96%.

She presented postoperative bronchopneumonia treated with cefepime and vancomycin, and uremia, and ascites responsive to high doses of diuretics.

After two previous unsuccessful attempts, she was finally extubated on the 20th POD. Vasoactive drugs were suspended on the 19th POD, and she was discharged from Pediatric ICU 27th POD.

In the 20th POD, after fever episodes, hemoculture and catheter tip cultures were positive for candida albicans, and amphotericin B was maintained for 21 days.

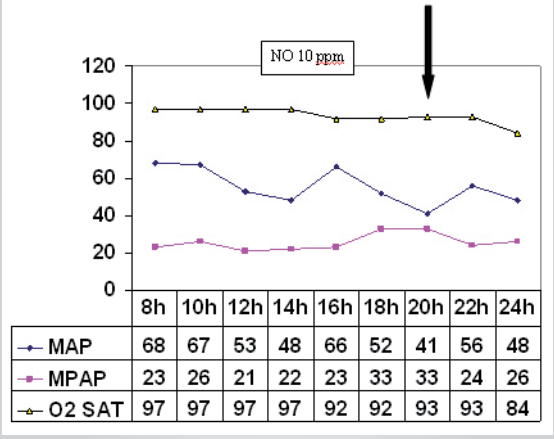
She was discharged from hospital 48 days after operation without dyspnea or cyanosis, or failure to thrive. She was using nifedipine, furosemide, and L-tyroxin.

Discussion

Preoperative assessment of the hypertensive pulmonary circulation and operabilility of these cases are frequently difficult[5]. The clinical history is important, cardiac failure may appear to improve as pulmonary vascular resistance increases. The chest radiography is plethoric when the pulmonary vascular resistance is low to permit a high blood flow,and the development of severe obstructive disease leads to peripheral pruning and hypertranslucent appearance in association with dilation of the hilar and proximal vessels. Cardiac catheterization is used to study the pulmonary wedge angiogram and to measure the pulmonary vascular resistance.

Cyanosis and/or a pulse oximeter reading of ≤ 92% was encountered in 31% of 77 neonates with Down syndrome[6], and was associated with other cardiac defects as tetralogy of Fallot (11%) and tetralogy of Fallot and AVSD (2%). The presence of moderate cyanosis (O_2 sat 80%-85%) in patients with complete AVSD is suggestive of pulmonary vascular obstructive disease and operability should be extensively studied preoperatively. Cardiac catheterization with pulmonary wedge angiogram, Qp/Qs and pulmonary vascular resistance calculation with and without pulmonary vasodilators (O_2 ,NO, prostaglandin) is mandatory. This patient still had symptoms of cardiac failure (tachypnea,failure to thrieve,hepatomegaly) and cardiac catheterization showed normal pulmonary wedge angiogram, Qp/Qs = 1,67, and PVR = 6,5 U/m^2. Despite elevated pulmonary vascular resistance and cyanosis, the presence of signs of congestive heart failure encourage the surgical repair.

Complete repair was made maintaining a small PFO (4mm) opened to decompress the right heart. Nitric oxide (10 ppm), 100% FiO_2, and milrinone was administered since coming off bypass and were substituted by calcium channel blocker to control postoperative pulmonary hypertensive crisis (Table1). Open lung biopsy was



	8h	10h	12h	14h	16h	18h	20h	22h	24h
MAP	68	67	53	48	66	52	41	56	48
MPAP	23	26	21	22	23	33	33	24	26
O2 SAT	97	97	97	97	92	92	93	93	84

MAP- pressão arterial média (mmHg); MPAP- pressão arterial pulmonar média (mmHg); O2 sat – saturação de oxigênio.

Graphic 1 *- Graphic representation of pulmonary hypertensive postoperative crises on the 5th POD.*

made in the third PO when the sternum was closed, but in cases with suspect of pulmonary vascular disease biopsy should be indicated as a criteria of operability[7,8,9]. In this case, despite cyanosis there was a favorable postoperative clinical evolution. Regression of pulmonary hypertension might be attributed to potential growth of pulmonary vascular bed[10], however long-term follow-up and non-invasive monitoring of pulmonary artery pressure

is mandatory.

Similar congenital heart defects like VSD in patients with higher than 6U/m2 pulmonary vascular resistance index should also be considered for surgical repair, when there are still signs of left-to-right shunt. In the literature, long-term follow-up after VSD closure in patients with a previous pulmonary vascular resistance greater than 6U/m2 showed good outcome[11] in 31 (79%) with significant reduction of pulmonary artery pressures. A wider overview of these patients concerning clinical and laboratory data prior to surgical intervention can guide to the most appropriate management.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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