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CASE REPORT
Congenital heart diseases

Critical value of the balloon occlusion test of a coronary fistula in a patient with pulmonary atresia and intact ventricular septum (RCD code: I-1C.4; II-2A.1)

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Abstract

We present the case of a boy with pulmonary atresia (PA), intact ventricular septum (IVS), and a coronary fistula between the right ventricle (RV) and a single left coronary artery (SLCA). In the newborn period, the child was operated on using a right Blalock–Taussing shunt (RBTS). At the age of 6 months, he was admitted to the department of Paediatric Cardiology due to hypoxaemia and cardiac catheterisation was performed before qualification for cardiac surgery. During haemodynamic evaluation of the patient, we conducted the balloon occlusion test of the coronary fistula and demonstrated that coronary circulation depends on the wide fistula and high pressure in the RV. We decided not to perform embolisation of the fistula and qualified the child for bidirectional Glenn palliation without RV decompression. The balloon occlusion test in patients with coronary fistulas plays a critical role and its result can be crucial for further management of the patient. JRCD 2019; 4 (3): 55–58

Key words: rare disease, pulmonary atresia with intact ventricular septum; single coronary ostium; coronary fistula; balloon occlusion test

Case presentation

We present the case of a 6-month-old boy with a complex heart defect: pulmonary atresia (PA), intact ventricular septum (IVS), anomaly of coronary arteries with a coronary fistula between the right ventricle (RV) and a single left coronary artery (SLCA).

After delivery of the child, prostaglandin E1 (alprostadil) infusion was initiated and on the 3rd day of life, a right Blalock-Taussig shunt (RBTS) connecting the brachiocephalic trunk with right pulmonary artery was performed in order to provide adequate pulmonary flow. The postoperative period was complicated by congestive heart failure and required milrinone and dopamine infusions. Within two weeks, the condition of the patient stabilised with an oxygen saturation (SaO_2) of 82%. The child was discharged on a low antiplatelet dose of aspirin (3 mg/kg). During follow-up, the main complication was progressive cyanosis with low systemic SaO_2 of 72–75%. At the age of 6 months, he was admitted for haemodynamic evaluation prior to bidirectional Glenn palliation without RV decompression.

Patient management

On physical examination the patient was in moderate condition with significant central cyanosis seen on mucous membranes and fingernails. On auscultation, a continuous murmur with intensity of 2–3/6 on the Levine scale was found, indicating the presence of a fistula as well as patent RBTS. Blood pressure was 80/50 mm Hg. Electrocardiogram (ECG) revealed normal sinus rhythm with severe right ventricular (RV) hypertrophy and ST-segment abnormalities (Figure 1). Chest radiography revealed that the cardiothoracic ratio was significantly increased, up to 0.66 with elevated

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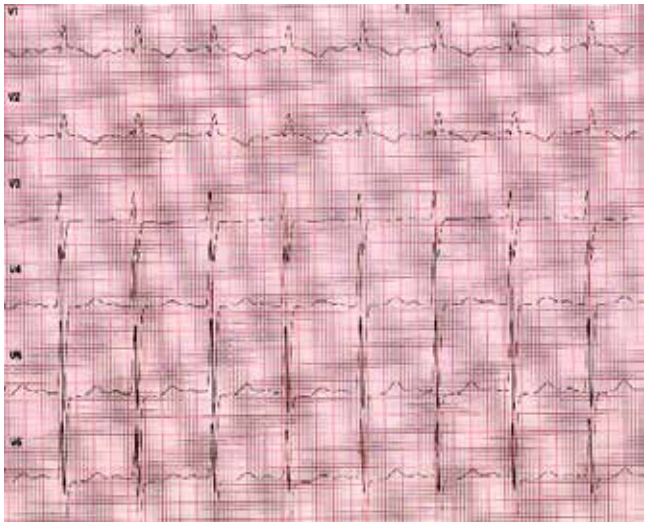

Figure 1. Trace 50 mm/s, amplitude 5 mm. Regular sinus rhythm, 150/min. Right ventricle hypertrophy: prominent Q and R waves in V1; very deep S waves in V4-V6. ST-T depression up to 2mm in V5-V6. Incomplete right bundle branch block

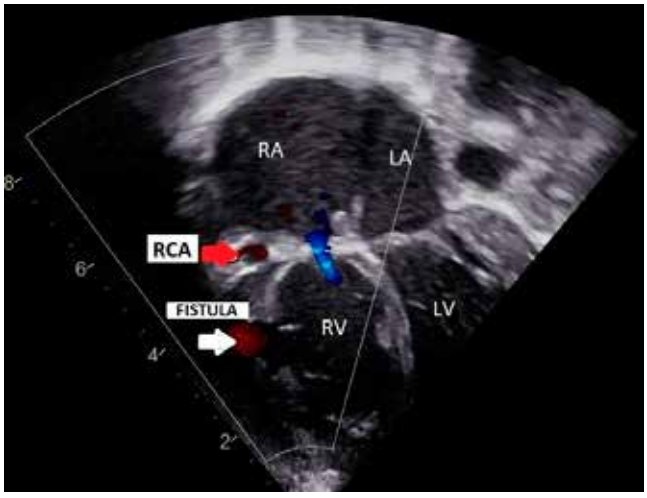

Figure 2. Transthoracic echocardiogram. 4 chamber view. Fistula (white arrow) connecting right coronary artery (RCA – red arrow) with right ventricle. Tricuspid valve regurgitation (blue flow). RA – right atrium; RV – right ventricle; LA – left atrium; LV – left ventricle

apex and signs of decreased pulmonary blood flow. Polycythaemia accompanied by an extremely elevated NT-proBNP level (25 000pg/ml) was seen in laboratory tests.

Transthoracic echocardiography (TTE) revealed a PA/IVS with right atrial and RV enlargement and hypertrophy, large atrial septal defect with right-to-left shunt, and decreased inflow through the tricuspid valve (TV) with 2nd degree insufficiency. The gradient pressure through the insufficient TV was 92 mmHg, indicating a suprasystemic RV pressure. Pulmonary arteries were confluent with the patent RBTS. Left ventricular (LV) contractility was within normal range (ejection fraction by the Simpson method = 58%). Additionally, we observed a wide (5mm in diameter) fistula connecting the LCA with the RV (Figure 2). Upon examination, anatomic outflow of the RV was atretic, hence the blood could flow

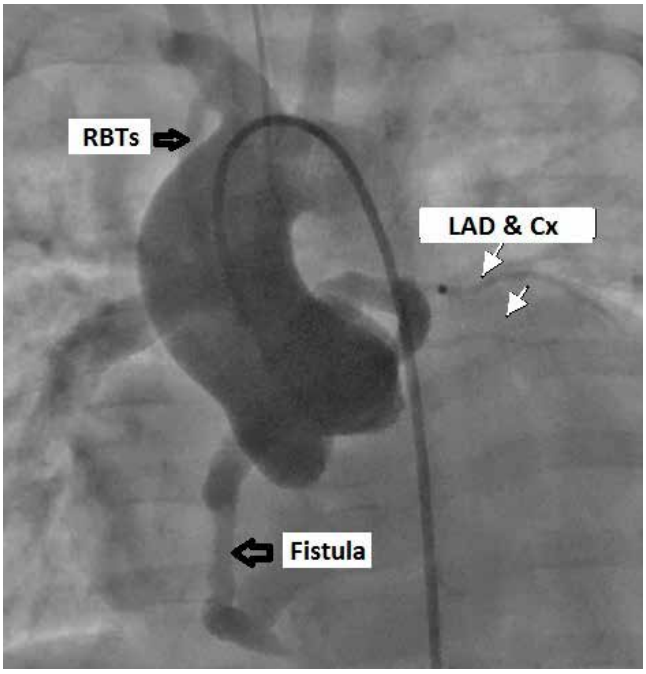

Figure 3. Aorta angiography. (CAU 40˚, Long Axial Oblique 30˚). Widened single coronary artery dividing into narrow LAD and Cx as well as wide, long and tortuous fistula with outlet into RV. Patent arterio-pulmonary anastomosis (RBTs) connecting common brachiocephalic trunk with right pulmonary artery. RA – right atrium; RV – right ventricle; LA – left atrium; LV – left ventricle, LAD – left anterior descending artery, Cx – circumflex artery

only through the fistula, sinusoidal connections, and the insufficient TV. Inflow through the TV was significantly impaired by the high RV end-diastolic-pressure (RVEDP). It was suspected that RV filling could be partially dependent on the fistula.

To determine the importance of the fistula for coronary circulation and to qualify the patient for surgery, heart catheterisation was performed. During the procedure, we observed moderate desaturation (SaO_2 60%) in the superior vena cava (SVC) and in the aorta (SaO_2 85%), and increased saturation in the RV (SaO_2 82%) indicating the presence of a bidirectional shunt through the fistula (Table 1). We confirmed the suprasystemic pressure in the RV with a significantly elevated RVEDP (15 mm Hg).

Angiography of the ascending aorta revealed an abnormally wide SLCA dividing into a narrow left anterior descending artery (LAD) and circumflex artery (Cx) with an ectatic, long, and tortuous fistula (Figure 3).

Selective angiography of the fistula showed a wide (5mm in diameter) outlet along with many proximal and distal branches of the right coronary artery (RCA) at the end of the abnormal vessel (Figure 4).

Right ventriculography revealed tiny sinusoidal connections supplying the interventricular septum and bidirectional flow through the fistula (Figure 5). Pulmonary flow was provided by the patent but narrow RBTS, as well as aortopulmonary anastomoses (mostly from the right intramammary artery – RIMA) (Figure 4). We decided that the fistula outlet was the only suitable place for an interventional closure without obstructing flow through the branches

Table 1. Pressures and saturations measured during heart catheterization revealed suprasystemic RV pressure with significantly increased end-diastolic pressure and saturation indicating on the shunt from the aorta through the fistula into the right ventricle

	SaO_2 (%)	PaO_2 (mm Hg)	Pressures (mmHg)		
			systolic	diastolic	mean
SVC	60	36,3	–	–	–
RA	57	34,3	13	9	9
RV	82	52,1	92	15	–
PV	100	357,7	–	–	–
LA	–	–	11	10	9
LV	85	54,9	70	9	–

SVC – superior vena cava; RA – right atrium; RV – right ventricle; PV – pulmonary vein; LA – left atrium; LV – left ventricle; SaO_2 – oxygen saturation; PaO_2 – oxygen partial pressure

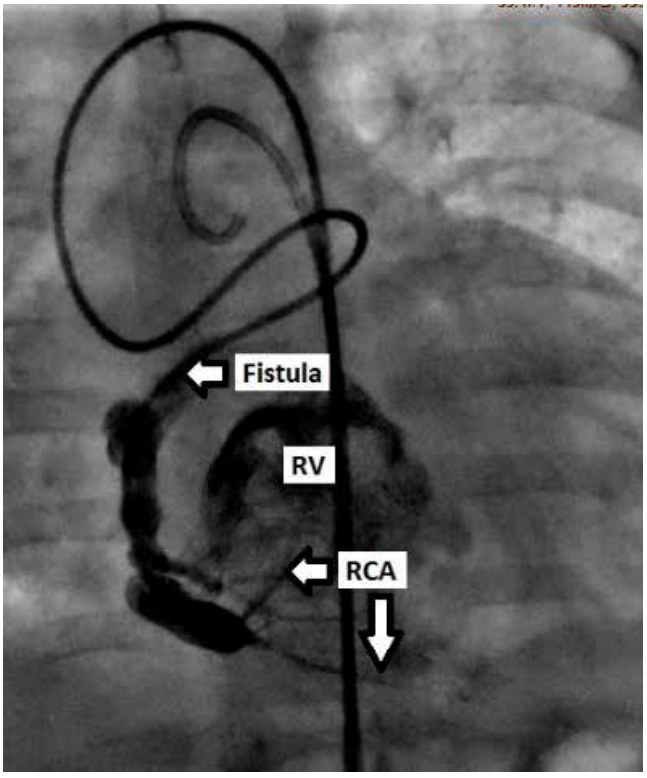

Figure 4. Selective fistula angio (antero-posterior). Wide and tortuous fistula dividing into proximal and distal branches of RCA and the outlet into right ventricle

of the RCA. Before the final decision, we performed the balloon occlusion test of the fistula. Deep ST-segment depressions appeared on the ECG during balloon inflation. We concluded that coronary circulation is partially RV-dependent and that decompression during biventricular repair would be life threatening. The child was qualified for a palliative Glenn operation (connection of the SVC with the right pulmonary artery). The postoperative period was uneventful, with the patient having an SaO_2 of 78%. He was discharged within two weeks.

Follow-up

During the 12-month follow-up, the boy was in good condition with moderate cyanosis. He is expected to undergo heart catheterisation before cardiosurgical consultation regarding further palliative treatment.

Review of literature

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare and heterogenous heart defect with pulmonary valve or muscular outlet tract atresia and usually presents with hypoplastic RV [1].

There are different treatment options (palliative procedures vs biventricular correction) depending on the TV diameter, RV end-diastolic volume (RVEDVol), as well as abnormal coronary circulation. There are a number of different protocols evaluating the likelihood of safe correction based on echocardiography and heart catheterisation with angiography for analysis of the coronary circulation [2]. In the presented case, the main problem was presence of coronary anomalies which included abnormal origin, course, divisions and connections.

Fistulas are the result of persistent sinusoidal connections in the primitive heart. They provide abnormal connections between the coronary arteries and heart chambers (atria or ventricles) in addition to systemic and pulmonary vessels. The most frequent type of fistula is the connection between the RCA and RV accompanied by the steal phenomenon and ineffective myocardial perfusion. In patients with PA/IVS, high pressure in the RV provides additional flow through the coronary sinusoidal connections.

The majority of patients with PA/IVS usually present with hypoplastic RV, high RVEDP, as well as high systolic pressure (even su-

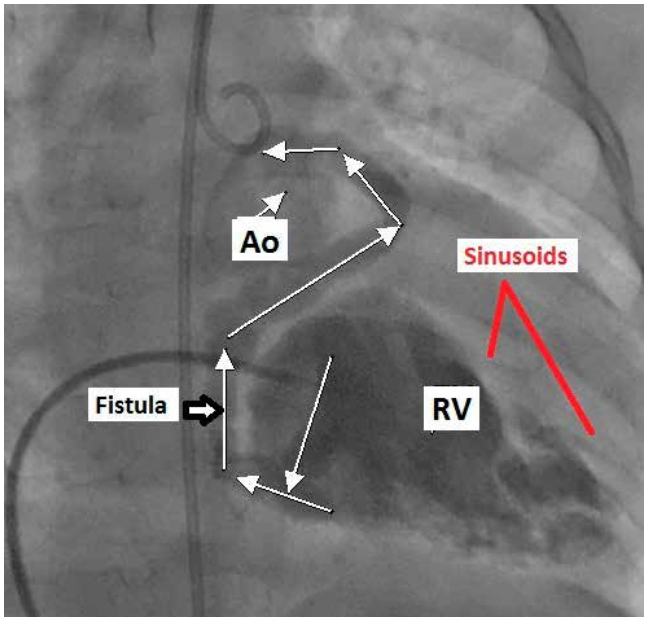

Figure 5. RV ventriculgraphy (antero-posterior). RV hypertrophy with pulmonary outlet tract atresia and the flow through the fistula into the aorta as well as through the tiny sinusoids

prasystemic pressures can be present) in the RV [3]. In these cases, abnormal sinusoidal connections may develop between the RV and coronary arteries, even in 30–60% of patients with partially or fully dependent coronary flow. The fistulas are usually not present in patients with normal/enlarged RV [4]. In our case, development of a large RV could be dependent on increased inflow through the fistula.

The correction of PA/IVS with pulmonary atresia reduction and RV decompression may cause a subsequent reduction in blood flow in the coronary arteries and myocardial ischaemia in the postoperative period. The balloon occlusion test may be useful in the evaluation of coronary circulation and its result can be used to decide if it is safe to close the fistula with a coil or vascular plug [4,5].

Significant ischaemia occurred during the balloon occlusion test in our patient (inversion of T waves). Because of this, we did not close the abnormal connection and decided to qualify the patient for the palliative Glenn procedure, instead of biventricular correction.

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