

CASE REPORT

Shock – manifestation of pulmonary atresia with aorto-pulmonary collaterals and single ventricle physiology in a 2-day neonate

Kovacicova L¹, Skrak P¹, Zahorec M¹, Masura J²*Intensive Care Unit, Pediatric Cardiac Center, Bratislava, Slovakia. lkovacicova@yahoo.com*

Abstract: Background: The combination of pulmonary valve atresia and ventricular septal defect accounts for about 2 % of cases of congenital heart disease. Most of the cases have intracardiac anatomy of Tetralogy Fallot and present with cyanosis in neonatal age.

Objectives: To report a case of a newborn with rare combination of pulmonary atresia, ventricular septal defect and single ventricle physiology presenting with shock very early following birth.

Methods and results: We describe a newborn infant who developed shock with severe metabolic acidosis and respiratory distress several hours following birth. Cardiac ultrasound showed pulmonary atresia and ventricular septal defect with single ventricle physiology. Cardiac catheterisation revealed major aorto-pulmonary collaterals with an excessive pulmonary blood flow. Resuscitative measures resulted in hemodynamic stability. However, due to unfavorable prognosis, a decision not to undertake surgical palliation was made. Withdrawal of intensive care led to rapid demise of the patient.

Conclusions: We report a case of a newborn with pulmonary atresia, ventricular septal defect and single ventricle physiology in whom an excessive flow through major aorto-pulmonary collaterals led to shock and death very early following birth (*Fig. 1, Ref. 5*). Full Text (Free, PDF) www.bmj.sk.

Key words: pulmonary atresia, major aorto-pulmonary collaterals, single ventricle, pulmonary circulation, shock.

Congenital heart diseases affect more infants than any other type of birth defect – occurring in 8 to 10 out of 1,000 live newborns. The combination of pulmonary valve atresia and ventricular septal defect accounts for about 2 % of cases of congenital heart disease (1). Most of these cases have intracardiac anatomy of Tetralogy Fallot and present with cyanosis in neonatal age (2).

We report an unusual case of a newborn with pulmonary atresia, ventricular septal defect and single ventricle physiology presenting with shock several hours following birth.

We focus on key clinical issues for the caring physician – clinical diagnosis, initial resuscitation and decision making process for further management.

Case report

A 3.180 g male neonate was born on 40 week of normal pregnancy. Apgar scores were 9 and 10 at 1 and 10 minutes, respectively. Initial physical examination was normal apart from cheilognathopalatoschisis. Thirty-two hours after delivery, the clinical condition deteriorated; baby became pale, tachypneic, and tachycardic. Sepsis was suspected and therapy with antibiotics

was initiated. Blood gases analysis revealed severe metabolic acidosis (base deficit of 16 mEq/L). Sodium bicarbonate was administered, and baby was transferred to neonatologic intensive care unit. There, despite an aggressive therapy with sodium bicarbonate, metabolic acidosis worsened. Inherited metabolic disease was suspected and metabolic screening performed. Arterial oxygen saturation ranged between 95–99 %, however respiratory effort was increasing and the baby was intubated. Due to a heart murmur and abnormal heart configuration on a chest radiograph, the baby was examined by pediatric cardiologist and then transferred to Pediatric Cardiac Center.

In our center, the initial examination revealed sinus tachycardia and systemic hypotension. Laboratory examination showed severe lactic acidosis (plasma lactate, 31.7 mmol/L), and renal and hepatic function impairment (creatinine, 173 mol/L; aspartate aminotransferase, 3.96 IU/L; alanine aminotransferase, 2.57 IU/L). Ultrasound examination revealed heterotaxy, IDD (situs inversus, D-loop ventricles, D-loop transposition of the great arteries), non-obstructive supracardiac type of total anomalous pulmonary venous drainage, common atrium, mitral atresia, severely hypoplastic left ventricle, ventricular septal defect, pulmonary atresia, aorto-pulmonary collaterals, hepatic veins draining into atrium. Pulmonary arteries were not detected.

Based upon the finding of high arterial oxygen saturations (96 %) in a patient with single ventricle physiology, an excessive pulmonary blood flow was suspected. To decrease the ratio of pulmonary blood flow to systemic blood flow (Q_p/Q_s), ventilatory strategy was aimed to achieve a high partial level of car-

¹Intensive Care Unit, Pediatric Cardiac Center, Bratislava, Slovakia, and
²Department of Cardiology, Pediatric Cardiac Center, Bratislava, Slovakia

Address for correspondence: L. Kovacicova, MD, PhD, Pediatric Cardiac Center, Limbova 1, SK-833 51 Bratislava, Slovakia.
Phone: +421.2.59371729, Fax: +421.2.54792317



Fig. 1. The angiogram showed multiple aorto-pulmonary collateral arteries arising from different levels of the aorta, diminutive pulmonary trunk and severely hypoplastic native pulmonary arteries connected to collaterals.

bon dioxide in the blood (permissive hypercapnia). Sedation was maintained using benzodiazepines and atracurium was used for neuromuscular blockade. An inotropic support with digoxin and dopamine was initiated. Therapeutic measures led to hemodynamic stabilization, resolution of metabolic acidosis and organ recovery. At the age of 9 days, the baby underwent cardiac catheterization which revealed multiple major aorto-pulmonary collaterals (MAPCAs) with excessive pulmonary blood flow (Fig. 1). Surgical intervention was contemplated but rejected because of complexity of cardiac anatomy and unfavorable prognosis. Supportive care continued, mechanical ventilation and infusion of dopamine were discontinued. Baby expired 25 hours after extubation.

Discussion

In the first week of life, many congenital heart diseases present in a surprisingly limited ways. Signs and symptoms include cyanosis and congestive heart failure or shock. Lesions that manifest with congestive heart failure and shock are those that are ductal dependent for systemic blood flow (hypoplastic left heart syndrome, interruption of aortic arch, coarctation of the aorta, severe aortic stenosis).

Patient with pulmonary atresia and ventricular septal defect is often symptomatic early in the neonatal period and the most common presentation is cyanosis. On rare occasions, an infant develops heart failure with signs of increased pulmonary blood flow through systemic-to-pulmonary collaterals (1). This does not usually occur until the pulmonary arterioles have involuted from the fetal state, most commonly after 4 to 6 weeks of age. In our patient, an enormous blood flow from systemic to pulmonary

circulation through collaterals resulted in shock, systemic hypoperfusion, severe lactic acidosis, and renal and hepatic function impairment very early after birth. To stabilize the hemodynamics, an aggressive resuscitation measures were required, including those reducing pulmonary blood flow.

Patients with pulmonary atresia, major aorto-pulmonary collaterals and single ventricle physiology have been traditionally considered inoperable. Palliation with perspective of Fontan type circulation is the possible therapeutic approach (3, 4); however, such approach is problematic. The recent study from Stanford University presents results of surgical treatment of fourteen patients with major aorto-pulmonary collaterals and single ventricle physiology (5). The patients were completely unifocalized to shunts, and in addition, underwent generally accepted single ventricle palliation procedures. The mortality of 54% showed that this was very high risk group of patients; a particularly poor outcome was reported when anomalous pulmonary venous return was a part of complex heart lesion. In view of unfavorable cardiac anatomy and prognosis, it was decided not to undertake surgical palliation in our patient. When a decision not to operate on patient was made, a withdrawal of dopamine infusion and mechanical ventilation led to rapid demise.

Conclusions

An excessive flow through major aorto-pulmonary collaterals in a neonate with pulmonary atresia, ventricular septal defect and single ventricle physiology led to shock and death very early following birth. The decision not to undertake a surgical palliation was made because general medical experience predicts a very poor outcome of such approach.

References

1. O'Leary PW, Mair DD, Edwards WD, Julsrud PR, Puga FJ. Pulmonary atresia and ventricular septal defect. 864–879. In: Moss and Adams' (Eds): Heart Disease in Infants, Children, and Adolescents. Philadelphia; Lippincott Williams & Wilkins, 2001.
2. Tchervenkov CI, Roy N. Congenital Heart Surgery Nomenclature and Database Project. Pulmonary Atresia-Ventricular Septal Defect. *Ann Thorac Surg* 2000; 69: S97–105.
3. Ko Y, Nakamura Y, Nomura K, Yamashiro F. Bidirectional cavopulmonary shunt using the azygos vein. *Jpn J Thorac Cardiovasc Surg* 2005; 53 (4): 213–216.
4. Miyaji K, Nagata N, Matsui H, Miyamoto T, Kitahori K. Successful Fontan procedure for asplenia with pulmonary atresia and major aortopulmonary collateral arteries. *J Thorac Cardiovasc Surg* 2003; 126 (5): 1648–1650.
5. Reinhartz O, Reddy VM, Petrossian E, Suleman S, Mainwaring RD, Rosenthal DN, Feinstein JA, Gulati R, Hanley FL. Unifocalization of Major-Aortopulmonary Collaterals in Single-Ventricle Patients. *Ann Thorac Surg* 2006; 82 (3): 934–939.

Received January 28, 2008.

Accepted March 28, 2008.