Right Ventricular Outflow Tract Perforation and Stenting for a Premature Neonate with Pulmonary Valve Atresia and Double Outlet Right Ventricle

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Abstract

Pulmonary valve atresia (PVA) is a fatal pediatric cardiac condition and commonly associated with other anomalies such as double outlet right ventricle (DORV). There is a disagreement over the best surgical strategy for treating PVA in conjunction with DORV. Prematurely born neonates are considered high-risk group for early pulmonary artery shunt surgery as the staged repair is found to be associated with higher interstage mortality, while the primary repair may lead to frequent postrepair reinterventions due to the presence of associated developmental anomalies. In this report, we present the successful results of using percutaneous transcatheterization in a high-risk patient; a 6-h premature neonate who had DORV, PVA with other anomalies including situs inversus, levocardia, D-malposed great arteries without patent ductus arteriosus. This rapid interventional catheterization offered to the neonate has an immense advantage of lifesaving and the protection against the remolding effect on the right ventricle that usually develops when waiting for the palliative procedures or surgery to take place.

Keywords: Pulmonary valve atresia, right ventricular outflow obstruction, stent

Introduction

Pulmonary valve atresia (PVA) is a unique cyanotic congenital heart disease, commonly associated with other cardiac anomaly like double outlet right ventricle (DORV).[1] Neonates with PVA accompanied by DORV without patent ductus arteriosus (PDA) often present with critical clinical findings immediately after birth that needs an urgent cardiac intervention.[2,3] The clinical presentations of this condition include shock, cyanosis, respiratory distress, metabolic acidosis, and feeding difficulty.[4] Patients with pulmonary atresia and intact ventricular septum usually need a series of cardiac interventions some in the first days of life including (balloon valvotomy, balloon septal valvoplasty, tenting, or shunting) and later on a number of surgeries are needed (Glenn, Fontan, or Hybrid procedures).[5] Since the optimal surgical approach to correct this defect is still controversial.[6,7] We report a successful result of using percutaneous transcatheterization in a high-risk patient; a 6-h premature neonate who had DORV and PVA together with other anomalies to enhance the knowledge about the improved outcomes of pulmonary valve perforation with right ventricular outflow tract (RVOT) stenting in neonate.

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Case Report

A 6-hour-old female neonate was born after 35 weeks of gestation with a birth weight of 2050 g. After prolonged resuscitation for primary apnea, the baby maintained to be deeply cyanotic. The baby was admitted to the "Congenital Interventional Cardiology Department," Ibn Al-Bitar Tertiary Center for Cardiac Surgery. Prenatally, the mother had no antenatal care with uneventful pregnancy with negative history of fever, rash, drug, or X-ray exposure during this pregnancy and no previous history of abortion and negative family history for congenital heart disease. On admission, the neonate did not have dysmorphic features, with heart rate = 156 BPM, respiratory rate = 68 CPM, temperature = 35.9 C°, and the

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admission-oxygen-saturation (SPO2) =36%. Echocardiography confirmed complex congenital heart anomalies (situs inversus, levocardia, DORV, D-malposed great artery, and PVA without PDA), the patient was initially started on prostaglandin E1 infusion (5 ng/kg/min) as trial to maintain the patency of ductus arteriosus but later on confirmation of absent PDA the infusion stopped after 24 h from admission. The patient was kept on intravenous fluid and received heparin and aspirin and evaluated preoperatively by serial blood tests and Echo study, send for Chest X-ray and a cross-matched blood was prepared.

Under general anesthesia, cardiac catheterization was performed. The right femoral vein access was inserted by venous sheath 5F. Initial hemodynamic assessment for the right ventricle (RV), aorta, and both atria which confirmed the presence of membranous PVA with small collaterals in absence of a septal defects in addition to situs inversus, DORV, and D-malposed great arteries [Figure 1a and b]. Aorto-cineangiogram was done for the confirmation of collateral's position and size with absent PDA [Figure 1c-e].

Perforation of pulmonary valve (PV) membrane was done using percutaneous transluminal coronary angiography wire Cordis® 0.014 inch (0.36 mm) that clinched a guide catheter to be positioned at the RVOT. A stable position was achieved by placing wire in distal end of the left pulmonary branch, the selected delivery sheath replaced the diagnostic catheter, then a stent and balloon was placed over the stiff wire and at intended position in RVOT, the balloon was inflated. Sequential dilatation was achieved by starting with a 4.5 mm coronary balloon. Selective pulmonary angiography was done to provide correct localization of the stent [Figure 2a], the RV angiogram showed a great improvement in the blood flow [Figure 2b-d], and oxygen saturation postintervention improved (SpO_2 increased to 91%). An antegrade pulmonary flow was achieved [Figure 2].

Postintervention, the patient was cared for in an intensive care unit where heparin infusion started and continued for 2 days in a dose of 28 unit/kg/h eventually replaced by oral administration of 3 mg/kg/day aspirin which should be continued till the time of Glenn operation which was planned at 8 months. Echocardiographic follow-up examinations were done after 6, 12, 24, and 48 h, on day 5, as well as 1 week and 2 weeks postoperatively. On discharge, the oxygen saturation was 91% and the patient was kept on oral aspirin. After discharge, the patient was regularly followed at monthly intervals and at 7 month of age found in good general condition and achieved normal growth parameters (weight and length on 10th centile, head circumference on 25th centile).

Discussion

This 6-hour-old female neonate represents a cardiac emergency case as she was presented with gasping breathing, cyanosis, and desaturation immediately after delivery, subsequently diagnosed with PVA and DORV. Due to the absence of PDA, early and timely intervention to establish pulmonary flow is lifesaving.[8] In order to be compatible with life, this type of congenital heart disease must be accompanied by septal defects.[9] Rogoff and Anthony had reported a 25-year-old female who had DORV and PVA to survived without intervening as she found to have two ventricular septal defects that allow blood mixing.[10] In the absence of septal defects, this maldevelopment causes significant symptoms immediately after delivery as the small size collaterals from aorta to pulmonary artery where insufficient to maintain pulmonary blood flow to the lungs,[11] as we noted in the current report.

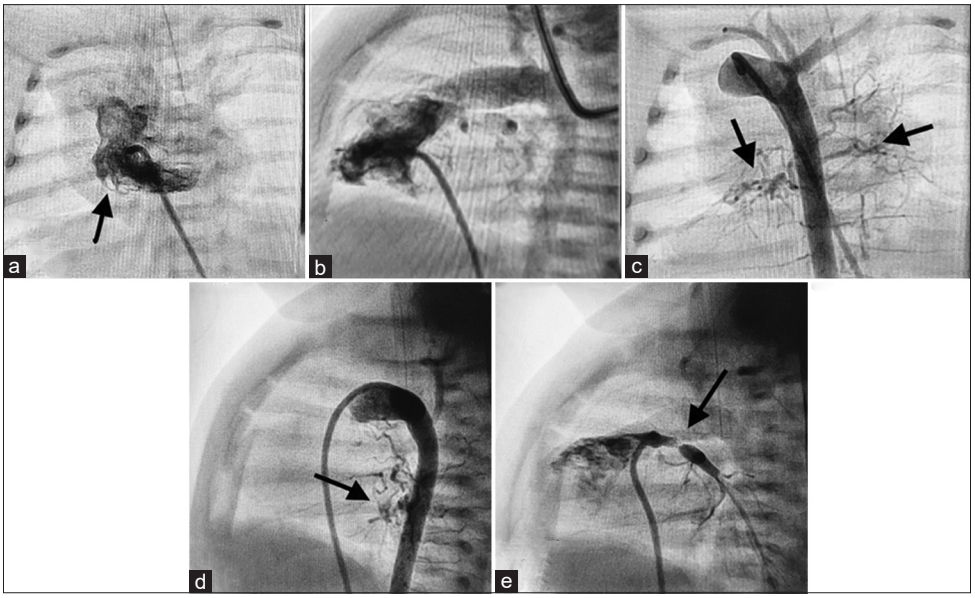

Figure 1: Catheterization findings before perforation and stenting. (a) Right ventricular angiogram showing a blind pouch to pulmonary valve (arrow). (b) An angiogram from the right ventricle to the aorta shows a double outlet right ventricle and a nonvisualized pulmonary artery. (c) (Frontal view) + (d) (lateral view): Angiography showing collaterals from aorta to pulmonary were present (arrows). (e) Right ventricular angiogram showing a blind right ventricular outflow tract (arrow)

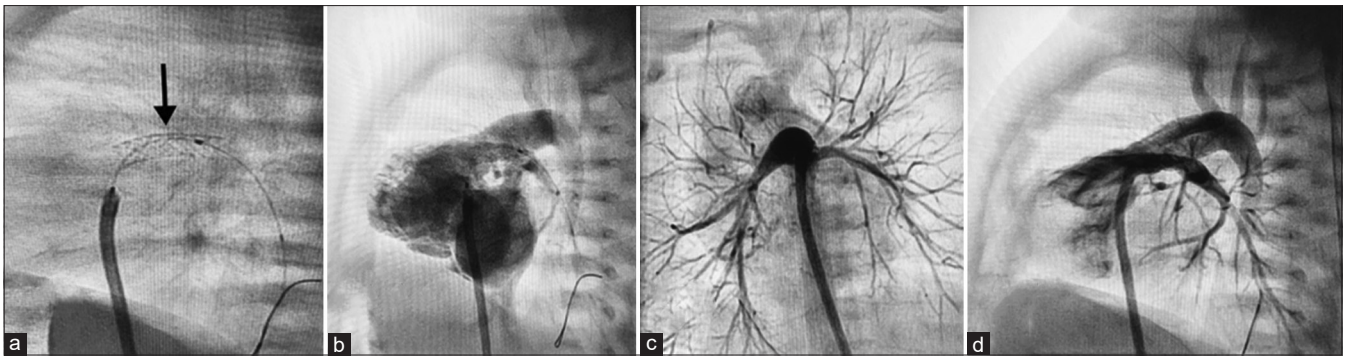

Figure 2: Catheterization findings after perforation and stenting. (a) Right ventricular angiogram showing outflow tract stent implantation (arrow), and empty right ventricle. (b) Full blood flow across the pulmonary valve postright ventricular outflow tract tract stenting. (c) (Frontal view) and (d) (lateral view): Full blood flow through the pulmonary arteries and lung postright ventricular outflow tract stenting

The management of children with PVA usually depends on size of RV; however, in the current case, the patient had DORV and rudimentary LV, so in such case, the management and prognosis depend on atretic pulmonary artery grade, anatomy and the other associated anomalies.[12] The complexity and heterogeneity of this disorder made the recommendation of a single procedure that is effective for all patients is not possible.[13]

Many physicians consider palliative surgery is the only intervention for DORV with PVA, while others found that in the presence of aortopulmonary collaterals perforation of the pulmonary valve as early as possible should be executed to prevent RV remolding.[14] This procedure is technically difficult because the exaggerated balloon dilation can lead to balloon rupture inside the stent and subsequent obstruction of the stent.[15] In addition, it carry nonnegligible risk of mortality.[14] However, the encouraging results of RVOT stenting make it a valid alternative to palliation surgery (aortopulmonary shunt) in high-risk group.[15]

Conclusion

This case report shows the effectiveness and safety of RVOT stenting for PVA accompanied by DORV. The adopted approach allowed the increase of pulmonary vascular performance, development of pulmonary vasculature, and the progression in bilateral pulmonary artery development to prepare the neonate to the second stage of palliative surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for images and other clinical information to be reported in the journal. The patient's parents understand that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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