

Stent Implantation in the Right Ventricular Outflow Tract to Pulmonary Artery in a Child with Pentalogy of Cantrell: Case Report

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Introduction

Pentalogy of Cantrell is a syndrome that was first described in 1958. It is characterized by 5 main malformations: defect in the lower sternum, defect in the anterior pericardium, defect in the upper abdomen, deficiency of the anterior diaphragm, and heart defects.¹ The syndrome is rare, with an incidence of 1 in 65,000 live births.¹ It is characterized by midline defects resulting from defective development of the septum transversum.² Other congenital heart lesions associated with pentalogy of Cantrell may include atrial septal defect, pulmonary valve stenosis, tetralogy of Fallot, dextrocardia, anomalous pulmonary venous connection, tricuspid atresia, and truncus arteriosus.² Children with pentalogy of Cantrell constitute a population with high morbidity and mortality.² In this case report, we demonstrate a hemodynamic procedure performed in a patient with this rare congenital anomaly.

Case report

A premature newborn (gestational age of 36 weeks) was diagnosed with pentalogy of Cantrell, presenting absence of the lower portion of the sternum bone, with partial herniation of the heart through the anterior thoracic wall, diastasis of the rectus abdominis muscles in the supraumbilical anterior abdominal wall, with insinuation and partial herniation of the liver (Figures 1 and 2), in addition to complex heart disease. Echocardiogram and angiotomography demonstrated double inlet left ventricle and double outlet right ventricle, with malposition of the great arteries and important infundibular and pulmonary valve stenosis. Shortly after birth, the patient developed hypoxemia requiring orotracheal intubation and mechanical ventilation with high settings, without significant improvement.

Keywords

Congenital Heart Defects; Treatment Outcome; Catheterization

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Manuscript received February 28, 2024; revised April 11, 2024;

accepted April 24, 2024

Editor responsible for the review: Karen Saori Shiraishi Sawamura

DOI: <https://doi.org/10.36660/abcimg.20240029i>

The patient was transferred to a pediatric cardiology reference center at the age of 21 days, in a severe hypoxemic state (saturation of 50% to 60% with a fraction of inspired oxygen of 100%), without prostaglandin infusion, and closed ductus arteriosus. An attempt was made to reopen the ductus arteriosus with a prostaglandin infusion, but it was not successful. Limited prognosis was discussed with family members due to complex heart disease and comorbidities. Therapeutic strategies were also discussed with the clinical team, and the possibility of surgically performing a systemic-to-pulmonary shunt was ruled out due to the anatomy and cardiac herniation. Therefore, the decision was made to take the 24-day-old patient weighing 2.2 kg to the cardiac catheterization room for stent implantation in the ventricular outflow tract to the pulmonary trunk.

Interventional procedure

The right femoral vein was punctured with the aid of ultrasound, and a 4/5 F slender transradial introducer was positioned. A 4 F Judkins right catheter was used on a 0.014" × 190 cm Balance Heavyweight guidewire to perform angiography in the main ventricle, confirming the diagnosis of infundibular and pulmonary valve stenosis, associated with malposition of the great arteries and hypoplasia of the pulmonary arteries (Figure 3). The 0.014" × 190 cm Balance Heavyweight guidewire was positioned in the inferior branch of the right pulmonary artery. The venous introducer was replaced with a 6 F transradial introducer. Palmaz Blue 5.0 × 18 mm and Palmaz Blue 6.0 × 15 mm stents were sequentially implanted in the ventricular outflow tract in a



Figure 1 – Patient with partial herniation of the heart and liver.

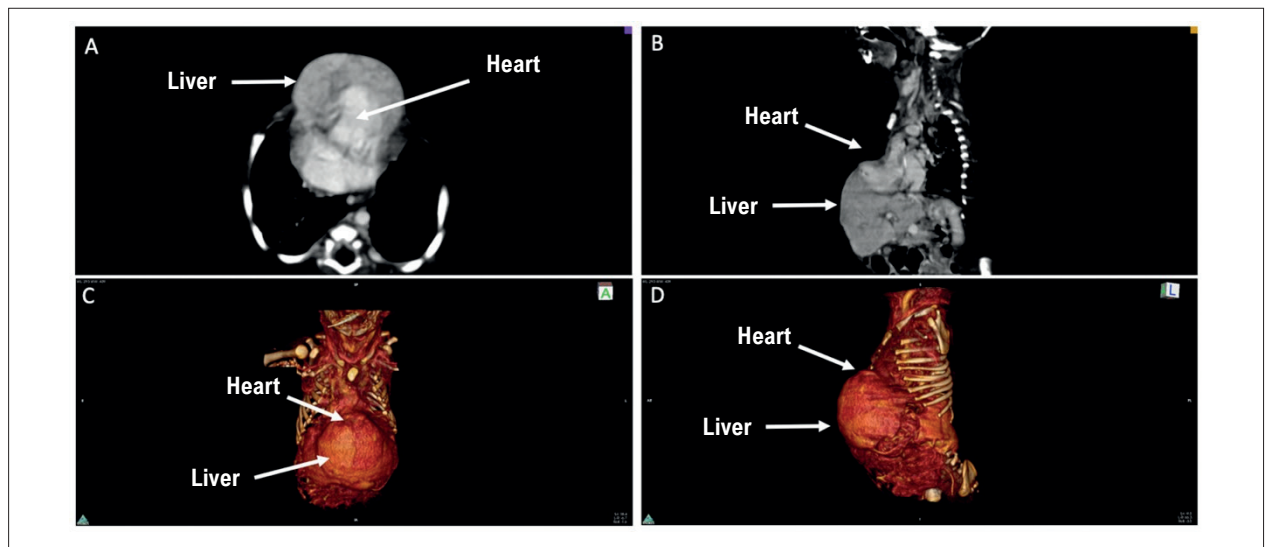


Figure 2 – CT angiography. A and B) CT angiography showing herniation of the heart and liver; C and D) 3D reconstruction demonstrating herniation of the heart and liver.

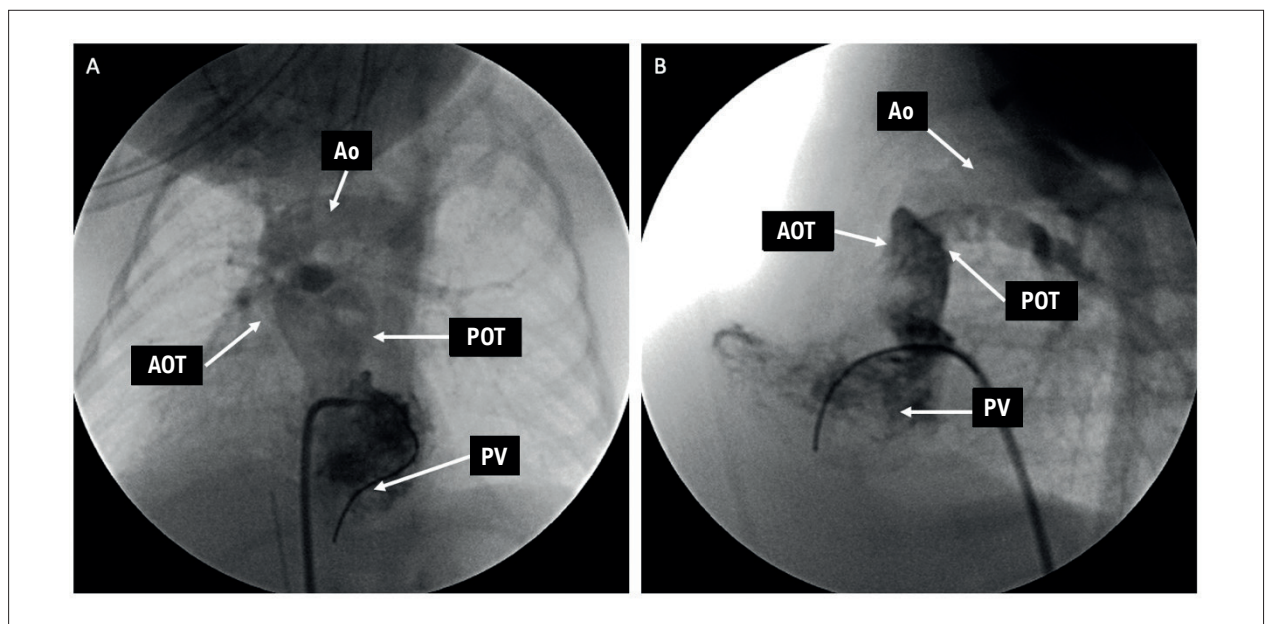


Figure 3 – Angiography and ventriculography. A) Angiography in cranial PA showing significant subvalvular and pulmonary valve obstruction; B) Left lateral ventriculography showing partial herniation of the heart and stenosis in the pulmonary outflow tract. Ao: aorta; AOT: aortic outflow tract; POT: pulmonary outflow tract; PV: pulmonary vein.

telescopic manner. Control cineangiography was performed (Figure 4). The catheter and introducer were removed, and compressive occlusive dressing was applied after manual hemostatic compression.

Control transthoracic echocardiography revealed a set of well positioned stents in the pulmonary outflow tract, covering the entire infundibulum and pulmonary valve, with an estimated maximum gradient of 21 mmHg and significant reflux, in addition to preserved flow to the pulmonary arteries (Figure 5).

After catheterization, the patient received a prophylactic dose of enoxaparin (1 mg/kg/dose, once daily) for 2 days, and was subsequently switched to acetylsalicylic acid 5 mg/kg/day. During the days following catheterization, the patient progressed with significantly improved saturation (70% to 89%) with a reduction in the fraction of inspired oxygen (to 65%) and other ventilatory settings. During hospitalization, the patient developed sepsis. Multidrug-resistant *Serratia marcescens* was isolated in blood and urine cultures, and the patient died at 2 months of age.

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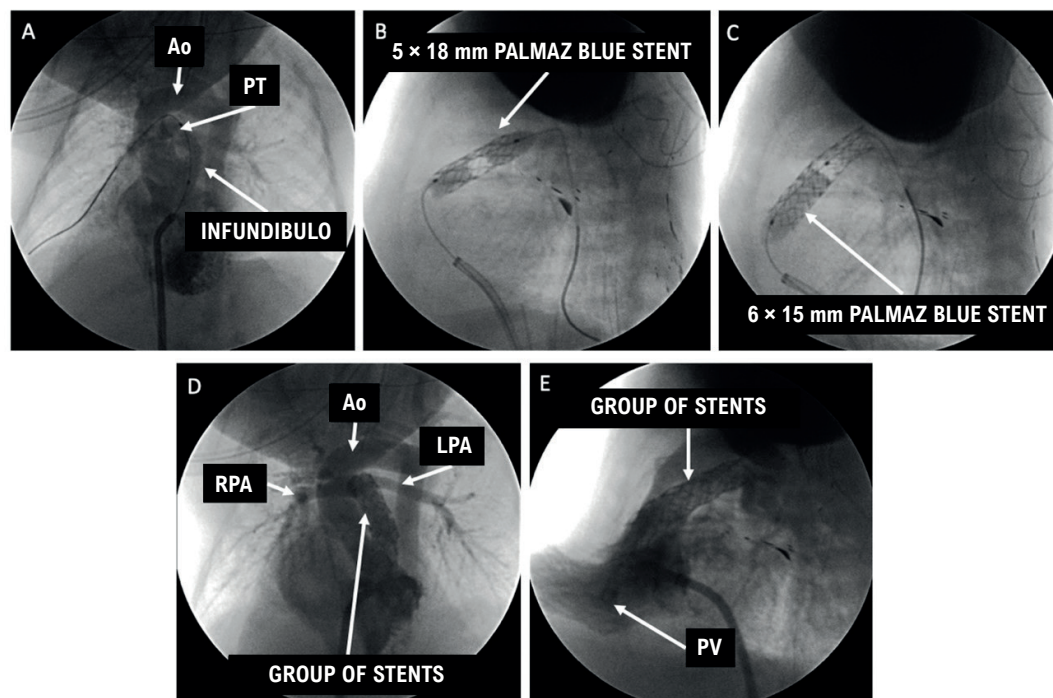


Figure 4 – Percutaneous intervention. A) Positioning a 0.014" 190 cm Balance Heavyweight guide in the inferior branch of the right pulmonary artery and supported by the same progressed stent. B) PALAMZ BLUE 5x18mm stent implanted in the pulmonary outflow tract; C) PALAMZ BLUE 6x15mm stent implanted telescoping the previously implanted stent; D) Ventriculography in cranial PA showing significant improvement in anterograde flow to pulmonary arteries; E) Ventriculography in left profile showing the entire infundibulum covered by the stent set. Ao: aorta; LPA: left pulmonary artery; PT: pulmonary trunk; PV: pulmonary vein; RPA: right pulmonary artery.

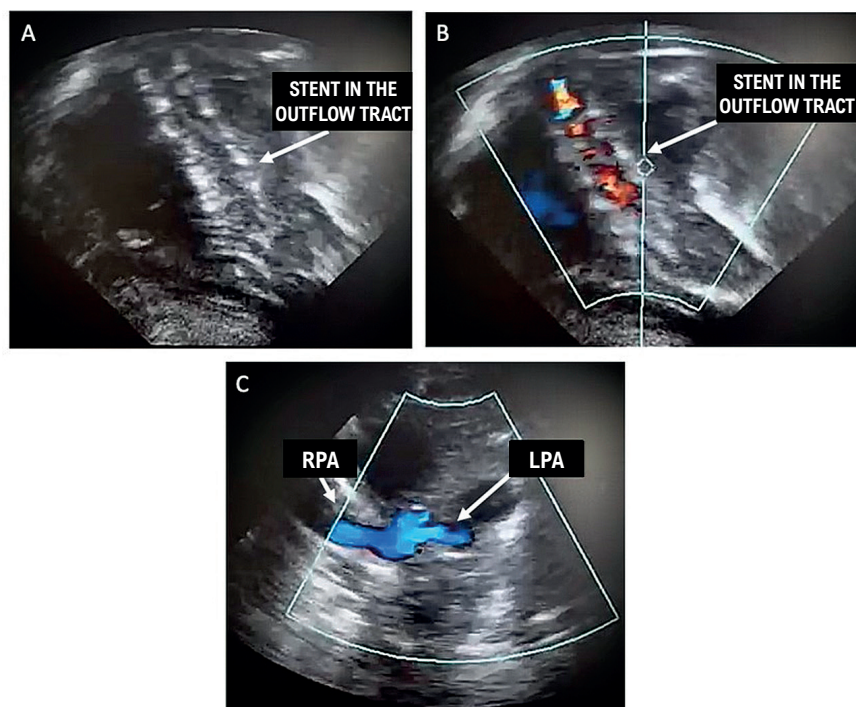


Figure 5 – Angiography and ventriculography. A) Angiography in cranial PA showing significant subvalvular and pulmonary valve obstruction; B) Left lateral ventriculography showing partial herniation of the heart and stenosis in the pulmonary outflow tract. LPA: left pulmonary artery; RPA: right pulmonary artery.

Discussion

Patients with pentalogy of Cantrell and cardiac malformations who require surgical intervention have worse prognosis.² In this case report, we demonstrate a rare case of percutaneous treatment with stent implantation in the ventricular outflow tract to alleviate critical obstruction to pulmonary flow in a patient with double inlet left ventricle, double outlet right ventricle, malposition of the great arteries, and pentalogy of Cantrell.

To date, few case reports have been published on percutaneous treatment of congenital heart disease in patients with pentalogy of Cantrell. McMahon describes the case of a patient with double outlet right ventricle and pulmonary outflow tract obstruction with successful stent implantation in the ventricular outflow tract.³ Galeczka describes percutaneous atrial septal defect closure procedure in a patient,⁴ while Tanaka describes a pulmonary valvuloplasty performed by ventricular puncture in a patient with pentalogy of Cantrell and ectopia cordis.⁵

Percutaneous stent implantation in the pulmonary outflow tract ensures better anterograde flow to the pulmonary arteries, improving the patient's saturation and promoting the growth of the pulmonary arteries. This is a palliative measure, with the aim of optimizing the patient's clinical conditions in order to plan surgical correction of the heart disease at an appropriate age and weight.

Percutaneous stent implantation in the right ventricular outflow tract is a widely used strategy in patients with tetralogy of Fallot.⁶⁻⁸ Possible complications of the procedure include definitive impairment of pulmonary valve function, stent fracture, stent embolization, restenosis of the right ventricular outflow tract, and arrhythmia.⁹ The most common mechanism of restenosis of the right ventricular outflow tract is obstruction due to an infundibulum not covered by the stent, generally due to the implantation of a stent that is shorter than ideal. In the reported case, as the first implanted stent did not cover the entire infundibulum, the decision was made to implant a second telescoped stent to cover all the musculature of the infundibulum. Stent implantation in the right ventricular

outflow tract results in greater growth of the pulmonary arteries than modified Blalock-Taussig surgery.¹⁰

Conclusion

This report demonstrates a rare case of percutaneous treatment with stent implantation in the ventricular outflow tract to alleviate critical obstruction to pulmonary flow in a patient with double inlet left ventricle, double outlet right ventricle, malposition of the great arteries, and pentalogy of Cantrell.

Author Contributions

Conception and design of the research, analysis and interpretation of the data and writing of the manuscript: Lombardi JG, Calamita PC, Gardenghi G; acquisition of data and critical revision of the manuscript for intellectual content: Lombardi JG, Calamita PC, Tannus LM, Barreto MRP, Rosado LEP, Gardenghi G.

Potential Conflict of Interest

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

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This study was approved by the Ethics Committee of the CEP/HUGO (GO) under the protocol number 77435824.0.0000.0033. All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.

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