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Left main coronary artery stent placement in a 7.0 kg infant with Williams Syndrome

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Abstract

We report the case of a 9-month-old male with Williams syndrome who underwent patch augmentation of supra-aortic stenosis and pulmonary artery stenosis, and required emergent drug-eluting left coronary artery stenting on post-operative day 1 for severe left ventricular dysfunction related to myocardial ischemia.

Keywords

coronary artery disease; williams syndrome; percutaneous coronary intervention

Introduction:

Coronary artery disease is rare in children. Percutaneous coronary intervention is rare in children and infants, as stent implantation is not considered standard therapy due to technical difficulty, concern related to procedural safety, and lack of long-term efficacy. Despite these limitations, there are instances when percutaneous coronary intervention may be a life-saving option. There are published case reports^{1–6}, and series^{7–9}, predominantly in older children and adolescents, detailing technical procedural success. We report the case of an infant with Williams syndrome who underwent successful emergent stent placement in the left main coronary artery while on veno-arterial extracorporeal membrane oxygenation (VA ECMO) on post-operative day 1 following three patch repair of supra-aortic stenosis.

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Compliance with Ethical Standards:

Conflict of Interest: The authors have no financial conflicts of interest to disclose related to this case report.

Informed consent: Informed consent was obtained prior to all procedures detailed in the above case report.

Case Presentation:

A 9-month-old male, weighing 7.0 kg, presented for cardiac consultation for evaluation of a murmur. He was born at 36 weeks gestation by cesarean section following a pregnancy complicated by twin gestation, preeclampsia, and fetal bradycardia. His medical history was significant for a two week neonatal intensive care unit (NICU) admission for supplemental oxygen requirement and nasogastric tube feedings to support adequate growth. His infancy was complicated by poor somatic growth and increasing tachypnea. His echocardiogram at his initial cardiology visit revealed moderate-to-severe aortic stenosis with a mean outflow gradient of 43 mmHg, moderate-to-severe left ventricular (LV) hypertrophy with preserved systolic function, severe right pulmonary artery stenosis, and patent proximal coronary arteries. His initial electrocardiogram showed no signs of acute or chronic ischemic heart disease. A cardiac catheterization revealed severe supravalvar aortic stenosis with an associated 73 mmHg peak-to-peak systolic gradient and severe bilateral branch pulmonary artery hypoplasia with associated gradients of 52–54 mmHg, and patent coronary arteries with proximal stenosis of the left main coronary artery noted. He was diagnosed with Williams syndrome clinically, and the diagnosis was confirmed by chromosomal microarray analysis.

The patient underwent three-patch surgical repair of his supravalvar aortic and bilateral pulmonary arterioplasty, including tissue resection and sharp debridement around the ostium of the right and left coronary arteries. His operative findings included a substantial amount of tissue around both coronary ostia, which were debrided. The left coronary os accepted a 2 mm probe intra-operatively. Following a brief initial period of hemodynamic stability in the cardiac intensive care unit, he suffered a bradycardic arrest and was emergently placed onto VA ECMO on post-operative day one. A peri-arrest echocardiogram showed total akinesis of the left ventricle. Due to concern regarding myocardial ischemia as the etiology of his acute decompensation, he was taken emergently to the cardiac catheterization lab.

Angiography confirmed a widely patent right coronary artery; however, the left main coronary artery was found to be severely stenotic (Figure 1). The proximal diameter, just beyond the ostium, was less than one millimeter (mm) as compared to a distal diameter of 2.0 mm immediately before the bifurcation into the left anterior descending and circumflex coronary arteries. After multidisciplinary discussion, the decision was made to proceed with stent placement. A 4 Fr x 75 cm Performer Guiding sheath (Cook Medical) was positioned in the ascending aorta and the tip of a 4 Fr pigtail catheter (Merit Medical) was advanced and extruded from the sheath in a manner to allow for variable angulation of the catheter tip. The orifice of the left coronary artery was successfully cannulated using a 0.014" Whisper guide wire (Abbott Vascular). The wire entered a marginal branch, however due to the technical difficulty in cannulation, it was not repositioned. The pigtail catheter was then removed and a 2.25 × 8 mm Resolute Integrity zotarolimus-eluting coronary stent (Medtronic) was advanced over the wire and implanted across the proximal left coronary artery. The proximal portion of the stent was post dilated with a 3.0 × 20 mm Quantum Maverick balloon catheter (Boston Scientific). Post intervention angiography revealed improved left coronary artery caliber with no luminal injury or evidence of thrombus (Figure 2).

The infant was decannulated from VA ECMO five days after the procedure and the left ventricular systolic function improved from essentially no motion at the time of catheterization to an ejection fraction of 39% (4 chamber bullet method) two weeks later. Stent thromboprophylaxis was initiated with aspirin (5 mg/kg/day) at the time of the procedure and clopidogrel (0.2 mg/kg/day) was added once the infant was decannulated from VA ECMO. The infant had a prolonged hospital stay of three months and was discharged home on aspirin, clopidogrel, digoxin, Lasix, spironolactone, and metoprolol. He continued on dual anti-platelet therapy for one year, when his clopidogrel was discontinued. Repeat cardiac catheterizations at four weeks, six weeks, ten weeks, and seven months after coronary stent implantation revealed a widely patent left coronary artery without evidence neointimal proliferation or in-stent thrombus formation (Figure 3). For a period of approximately 16 months following his surgery and coronary intervention he was able to thrive and develop; however, he did develop progressively worsening congestive heart failure as his biventricular systolic and diastolic function deteriorated. He developed multi-system organ failure due to his long-standing cardiac dysfunction, and ultimately expired 20 months after his initial surgery and catheterization. Throughout the remainder of his life, his stent remained patent by echocardiographic imaging (Figure 4).

Discussion:

This case demonstrates the feasibility of percutaneous coronary artery stent implantation in a critically ill, post-operative infant with an acute myocardial infarction. The infant developed cardiovascular collapse shortly after surgical repair due to severe narrowing of the left main coronary artery. Stent implantation allowed for improvement of left ventricular function. To our knowledge, this patient is one of the youngest and smallest patients with Williams syndrome to undergo coronary stent implantation. There has been only one younger patient with Williams syndrome reported to have undergone coronary stenting while on ECMO.¹ This patient, reported by Upadhyay et. al, underwent coronary stent at 6 months of age following arrest and emergent cannulation to VA ECMO (eCPR) while obtaining access in the cardiac catheterization lab, and had not undergone prior surgeries. This child had evidence of endomyocardial fibroelastosis (EFE), suggesting long-standing ischemic insult to the myocardium, as opposed to our case who suffered an acute decompensation shortly after surgery. Although previous reports demonstrate technical success in the post-operative period, our case is the closest to surgery as the procedure was performed on post-operative day one, while on VA ECMO.

There are currently no published or accepted guidelines for the prophylaxis of coronary artery stent thrombosis in the pediatric age group, let alone infants. Based on the adult-based guidelines and the experience of others in this age group, we chose to proceed with dual anti-platelet therapy. However, while the infant remained supported on VA ECMO, clopidogrel was held, as the risk of significant bleeding was deemed to be too great. The patient did not suffer any bleeding-related complication while on dual agent prophylaxis. The patient demonstrated left main coronary patency on serial repeat catheterizations and echocardiograms for the remainder of his life.

Percutaneous coronary intervention is rare in children and infants, as limitations include technical difficulty, concern related to procedural safety, and lack of long-term efficacy. The small femoral and coronary arteries limit sheath and stent size respectively. However, in vitro testing of the coronary stent used in this case, demonstrates a maximum diameter of 3.0 mm. This is the mean left main coronary artery diameter for children with a body surface area of $\sim 1.1 \text{ m}^2$. While this patient ultimately succumbed to complications from his chronic systolic and diastolic heart failure, if the patient had survived into late childhood or early adolescence, his stent would have been of adequate size according to coronary artery Z-score data.

In conclusion, pediatric coronary artery stent implantation remains a rare procedure; however published reports demonstrate its feasibility and technical success with a low rate of procedure-related complications, particularly in the immediate post-operative period. Although the long-term outcome of coronary artery stent implantation in infants is unknown, advances in stent technology may allow for a longer period of clinical efficacy removal than previously believed.

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Figure 1: Straight A/P angiography demonstrating a severely stenotic left main coronary artery (LMCA), with a proximal diameter of less than 1 mm. The white arrow denotes the area of stenosis.

Figure 2:
Straight AP angiography post-stent placement and dilation demonstrates a widely patent left main coronary artery without extravasation of dye, thrombus, or evidence of endothelial injury. The white arrow denotes the patent area of stented coronary artery.

Figure 3:

Angiography 7 months post procedure with AP camera angulated RAO 30 degrees. The left main coronary remains patent without evidence of intimal stenosis, and no significant collateralization to distal left coronary system. The white arrow denotes the patent area of stented coronary artery.

Figure 4: Parasternal short axis view demonstrating stable stent placement and color Doppler flow in diastole, indicating stent patency. The white arrow denotes the patent left main coronary artery.