Duct stenting in an ELBW infant with aortic arch interruption

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ABSTRACT

Aortic arch interruption is a rare cardiac malformation. In some cases, it is part of a more complex congenital heart disease. Survival of extremely low birth weight infants with this cardiac disease is very rare. Surgical correction is the only treatment. Ductal stenting as a bridge procedure in small affected infants is a good alternative.

Keywords: Aortic arch, ductal stent, low birth weight

INTRODUCTION

In the setting of a critical obstruction of the left heart, the right ventricle has to supply both the pulmonary and the systemic circulation through an open ductus arteriosus. Furthermore, coronary and cerebral perfusion depends on retrograde perfusion into the ascending aorta through the arterial duct. The ductus arteriosus remains open and it is often larger than normal, with the diameter ranging between 6 and 9 mm. The stent diameter should be between 7 and 9 mm, mostly 8 mm, without kinking. Early duct obstructions should be ruled out. It is not uncommon to require two or three stents need to be placed in the telescope technique. Self-expandable stents or balloon-expandable stents may be used. The use of self-expandable stents in this indication is more recent as it has a lower radial force but may slip.

CASE REPORT

An extremely low birth weight male neonate was admitted to our hospital after birth. He was born with cesarean section after a 35^{+2} week twin gestation with a birth weight of 1090 g. One month previously, he had been diagnosed on fetal echocardiography with possible interruption of the aortic arch. The mother had received betamethasone. He was intubated immediately after delivery and transferred to our cardiac unit for the

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continuation of care and initiation of prostaglandin and dopamine. The initial echocardiogram confirmed the diagnosis of interrupted aortic arch type B with left-sided aortic arch and aberrant right subclavian artery and small membranous ventricular septal defect: ascending aorta was origin to right and left common carotid arteries and also to left subclavian artery [Figure 1]. The descending aorta and right subclavian artery were supplied by the pulmonary artery through a wide patent ductus arteriosus.[3] His clinical course in the neonatal unit was initially stable, but 3 weeks after admission, he developed necrotizing enterocolitis.[4] Ductal stenting was decided as a palliative intervention that would enable the discontinuation of prostaglandin.[5,6] At the age of 1 month with a body weight of 1180 g, the infant was transferred to the catheterization laboratory for attempting ductal stenting. Under general anesthesia, the right femoral vein was percutaneously cannulated with a 5 French short (pediatric) sheath. No heparin was administered after vascular access was obtained. Initially, a balloon atrial septostomy was performed. Pulmonary aortography demonstrated an 8 mm long and 6 mm wide patent ductus connecting directly with the thoracic descending aorta. The duct was accessed from the femoral vein, using a 4F Swan (Wedge) catheter and a 0.014" straight, 320 cm long guidewire. The

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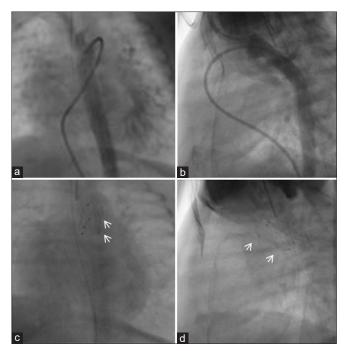


Figure 1: Arteriography of the descending thoracic aorta through the duct before duct stenting in anteroposterior (a) and lateral (b) view. X-ray illustration of the stent *in situ* (arrows) in anteroposterior (c) and lateral (d) view

wire was stabilized in the descending aorta. Over the wire, an 8 mm/12 mm sinus-superflex-DS (Optimed) self-expandable stent was advanced, introduced, and deployed in the duct. The total fluoroscopy time was 7 min [Figure 1]. Prostaglandin was discontinued gradually after stenting. The saturation in the lower extremities was constantly 95%. Enterocolitis eventually retreated and weight gain was satisfactory the following 3 months reaching a body weight of 2.4 kg. [Figure 2]. However, his neonatal course was complicated by episodes of sepsis with negative cultures. He underwent a surgical correction of his condition at the age of 5 months. After the operation, he was very difficult to ventilate and was placed on high-frequency ventilation. Unfortunately, 1 month after the operation, he developed severe desaturation and bradycardia, suffered a cardiac arrest, and died despite adequate resuscitation measures.

DISCUSSION

Aortic arch interruption is a rare cardiac malformation. In some cases, it is part of a more complex congenital heart disease. Survival of extremely low birth weight infants with this cardiac disease is very rare or probably unique. Therefore, duct stenting and atrial septum manipulations including stenting were performed in the vast majority of patients with conscious sedation and meticulous anesthesiology management. Newer self-expandable stents CE-certified for duct and



Figure 2: Two-dimensional echocardiogram of the pulmonary artery and duct after stenting. Short axis view at the level of pulmonary artery (a) displaying the stent (arrows). Echocardiogram of the stent (b) and color flow imaging within the stent (c) revealing normal flow with no aliasing

coarctation stenting in neonates with hypoplastic left heart syndrome further optimized the interventional approach. A hybrid palliative approach has been used in low birth weight neonates with interrupted aortic arch and ventricular septal defect. This is a safe and reproducible approach that can be considered in high-operative-risk neonates with multiple risk factors. (8-10)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands 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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