A novel method of creation of a fenestration in nitinol occluder devices used in closure of hypertensive patent arterial ducts

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ABSTRACT

Test occlusion with a balloon is done to predict operability of large hypertensive patent ductus arteriosus (PDA). If the fall in the pulmonary artery pressures is inadequate, a complete closure is not desired. To create a predictable premeasured fenestration in a nitinol occluder device used for closing hypertensive PDA. A large nitinol occluder device was punctured with an 18G needle to advance a 0.035˝ stiff guide wire through the occluder before loading it into the delivery system. The occluder with the guidewire was then deployed across the PDA. A coronary guide catheter was later threaded through the guidewire into the fabric of the device, which was still held by the delivery cable. A coronary stent was deployed across the fenestration in the occluder to keep it patent. An 8-year-old boy with Down syndrome and hypertensive PDA was hemodynamically assessed. Even though there was a fall in the pulmonary vascular resistance index and pressures on test occlusion, the pulmonary artery pressures were labile with fluctuations. A customized fenestration was made in a 16 mm muscular ventricular septal defect occluder (MVSO) with a 4.5 mm bare-metal coronary stent. The pulmonary artery pressures remained at half of the aortic pressures after the procedure. This fenestration model precisely and predictably fenestrated a large occluder device used to close a hypertensive large PDA. Long-term patency of these fenestrations has to be assessed on the follow-up, and may be improved through larger fenestrations, systemic anticoagulation and use of covered stents.

Keywords: Device closure, Down syndrome, fenestration, hypertensive patent ductus arteriosus

INTRODUCTION

Interventional treatment of patients with very large patent ductus arteriosus (PDA) and severe pulmonary hypertension (PAH) is challenging. Occurrence of the pulmonary vascular disease is accelerated in patients with Down syndrome and advancing age.[1,2] Test occlusion of the PDA with balloon or occluder device is often utilized in the catheterization laboratory to decide operability of large hypertensive ducts.[3,4] In patients, who demonstrate clear left to right flows on echocardiography across the ducts and lack of oxygen desaturation in lower limbs, if the pulmonary artery pressure shows a significant fall and separation from aortic pressure, the ducts are considered operable. These patients may still need to be continued pulmonary vasodilators.[5,6] Inadequate fall in pulmonary artery pressures and minimal separation from aortic pressures are indicators of inoperability.[7] When children with Down syndrome are sedated for

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DOI: 10.4103/0974-2069.171399

How to cite this article: Singhi AK, Sivakumar K. A novel method of creation of a fenestration in nitinol occluder devices used in closure of hypertensive patent arterial ducts. Ann Pediatr Card 2016;9:53-8.

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cardiac catheterization, they may have labile pulmonary artery pressures with wide swings during agitation or airway occlusion.[2]

Prefabricated customized fenestrations and on table fenestrations have been utilized in nitinol atrial septal occluders in hypertensive atrial septal defects.[8-11] Surgical fenestrations of patches were also utilized for closing ventricular septal defects (VSDs) and they have also proven to be effective.[12,13] Fenestration of an occluder device or partial banding of the PDA may be a desirable strategy in very large hypertensive ducts with borderline operability.[5] The literature about partial banding of PDA for hypertensive ducts is very sparse as the procedure involves thoracotomy and collapsing the left lung, which alters the hemodynamics. Hypertensive ducts will need a device with retention skirts on either side like a muscular ventricular septal occluder to prevent reverse embolization of the device into the descending aorta during episodes of pulmonary hypertensive crises.[7,15] We present a novel model of customized on-table fenestration of a large muscular ventricular septal occluder device, and our experience of closing a hypertensive duct with such a fenestrated device, after approval from the institutional ethical committee and detailed informed consent from parents of the patient.

CASE REPORT

An 8-year-old boy with Down's syndrome was diagnosed to have a very large PDA in the early neonatal period. His birth weight was 2.5 kg. He had recurrent respiratory tract infections all throughout his infancy and early childhood that decreased considerably after 4 years of age. He was treated with decongestive medications in infancy, which was discontinued at 2 years. He had poor weight gain. At the time of presentation to us, he was malnourished weighing 21 kg with a Z-score of –1.5.[16]

Pulse oximetry showed oxygen saturation in the upper limb and lower limb of 94-95% and there was no difference between the two. Cardiovascular system examination showed mild cardiomegaly, short systolic murmur, and a loud pulmonary component of the second heart sound. The chest X-ray showed minimal cardiomegaly, dilated central pulmonary arteries, and a minimal plethora [Figure 1]. Electrocardiogram showed rightward QRS axis with biventricular hypertrophy. There was a very large 10 mm PDA shunting left to right predominantly without any gradient from the aorta to the pulmonary artery and very minimal left atrial and ventricular enlargement [Figure 2, Movie 1].

Cardiac catheterization

During cardiac catheterization on conscious monitored sedation with ketamine, the pulmonary arterial and aortic pressures were similar both in room air breathing and on oxygen [Figure 3a]. Hypoventilation due to sedation led to mild systemic desaturation which was confirmed by sampling the pulmonary veins for oximetry after a transseptal puncture. The absence of right to left shunt across the duct was confirmed on oximetry. On vasodilator testing with oxygen, there was significant increase in pulmonary blood flow, which caused reduction in pulmonary vascular resistance through the pulmonary artery pressures remained similar to aortic pressures [Table 1]. The large tubular duct measured 10 mm on lateral view aortogram and was similar in size to the descending thoracic aorta below the duct [Figure 4, Movie 2]. Test occlusion with a 16 mm muscular VSD device (Cera muscular VSD occluder, Lifetech Scientific, Shenzhen, and PRC) caused significant reduction in pulmonary artery pressure and its separation from aortic pressures [Figure 3b]. A larger device was chosen to allow for elongation of the device since the duct was 20 mm long and tubular, but the occluder device had a length of 8 mm only. During the test occlusion, the pulmonary artery pressures were considerably labile and fluctuated during times of agitation of the child and airway occlusion during sedation.

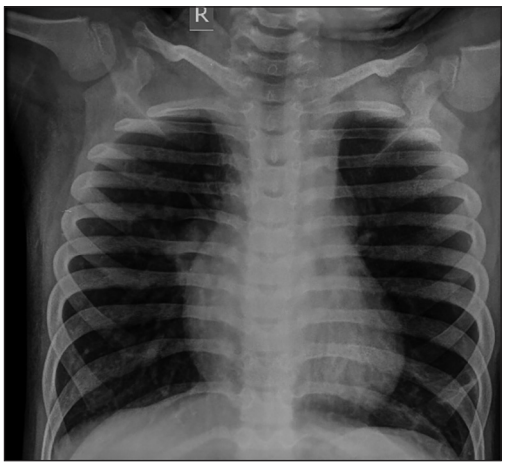

Figure 1: Chest X-ray showing mild cardiomegaly and very minimal pulmonary plethora

Table 1: Hemodynamics

SITE	Pressure resting state	Pressure on oxygen	Post device occlusion
RA	Mean 10	Mean 8	Mean 8
PA	92/58 (75)	94/59 (74)	65/34 (49)
LA	Mean 10	Mean 10	
Ao	95/61 (77)	95/60 (78)	134/90 (108)
Qp/Qs	1.1	4.7	
PVR WU	25.2	3.8	
PVR Index	20.6	3.2	
PVR/SVR	0.9	0.2	

RA: Right atrium, PA: Pulmonary artery, Ao: Aorta Qp-pulmonary blood flow; Qs-systemic blood flow; PVR WU – pulmonary vascular resistance in Wood units; SVR WU – systemic vascular resistance in Wood units

The factors that suggested operability included young age of the patient, cardiomegaly on X-ray, left to right shunt on echocardiogram, increase in shunt ratio on oxygen, fall of pulmonary vascular resistance on oxygen and significant fall in pulmonary artery pressures on test occlusion. However, the chromosomal abnormality, high resting pulmonary vascular resistance values and labile nature of the pulmonary artery pressures made us decide on the partial closure of the PDA with a fenestrated device. A test occlusion with an occlusion balloon could not be done in this child, since the occlusion balloon either occluded partially the descending aorta or the pulmonary artery; and failed give true hemodynamic information. Hence, the muscular ventricular septal occluder device was used to test occlude the duct. In view of significant fall of pulmonary artery pressure, it was decided to close the hypertensive duct with the same device but with a customized on-table fenestration in the fabric of the device.

Fenestration technique

The 16 mm muscular VSD occluder was punctured with an 18G needle, and a 0.035" exchange length guidewire was passed through the puncture fenestration. A short 8F dilator was passed over the wire to make the fenestration larger to allow passage of a 5 French right coronary guiding catheter (Launcher, Medtronic Inc., Minneapolis, Minnesota) [Figure 5]. An exchange length 0.035"Terumo Glidewire was passed through the fenestration in the device, and a 5F Judkins right coronary guiding catheter was taken through the fenestration over the wire. A 10 French long Mullins sheath (Cook Medical, Bloomington, MA) was placed in the descending aorta through the ductus from femoral vein. The whole assembly of the occluder device with the coronary guide catheter and 0.035" guidewire was then loaded into a 9F short sheath and introduced into the 10F Cook Mullin sheath. Even though the 16 mm muscular VSD occluder needed only an 8F delivery system, a larger sheath was chosen since the occluder with the coronary guide catheter and guidewire passing through the device was larger in its profile. The VSD device was deployed in the usual way with a stable position [Figure 6a, Movies 3 and 4]. Through the 5F judkins right coronary guiding catheter, a 4 mm baremetal stainless steel coronary stent, which was 20 mm long (Prolink LP, Vascular concepts, Halstead, Essex, UK) was deployed across the fenestration at 16 atmospheres. The stent expanded to a diameter of 4.5 mm at 16 atmospheres. Post-procedure fluoroscopy and an angiogram showed good device and stent position with good descending aortic flows [Figures 6b and 7; Movie 5]. Color Doppler echocardiogram after the procedure confirmed a small left to right shunt flow through the fenestration in the occluder. The patient was started on dual pulmonary vasodilators, sildenafil, and bosentan. On a 6-month follow-up, he is asymptomatic with moderate PAH on echocardiogram.

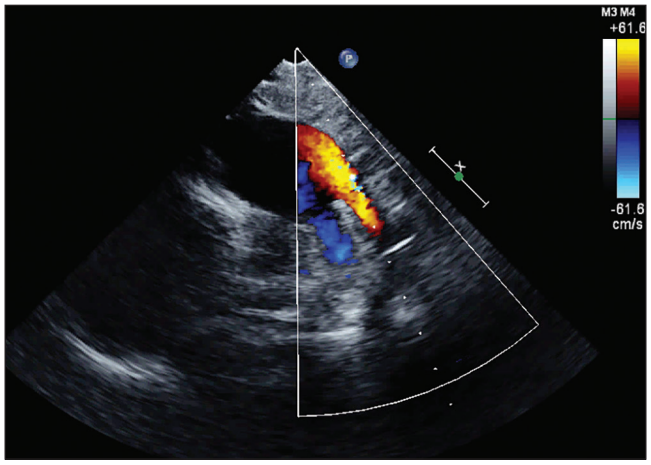

Figure 2: Color Doppler echocardiogram shows laminar left to right flows across a large patent ductus arteriosus in parasternal short axis view [Movie 1]

DISCUSSION

Hypertensive duct in an older patient with Down syndrome and recurrent airway obstruction poses a

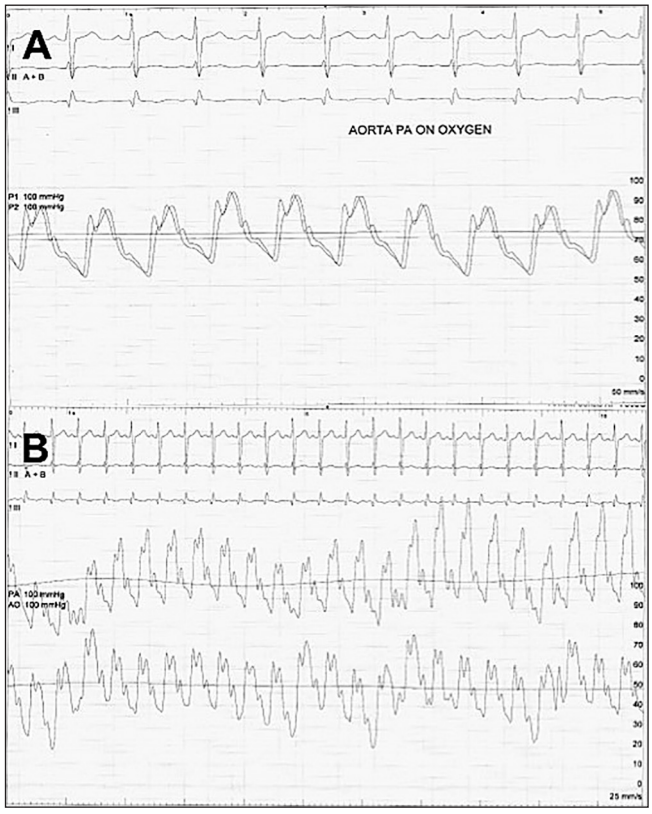

Figure 3: Simultaneous aortic and pulmonary artery pressure tracing on oxygen (a) Severe pulmonary arterial hypertension with no differences in systole and diastole. Following test occlusion with the device, there was a good fall of pulmonary artery pressures (b) And separation from the aortic pressures

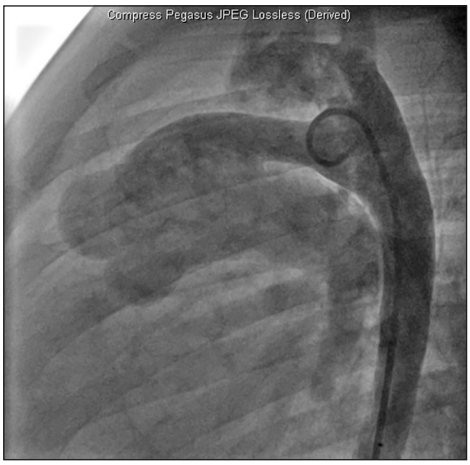

Figure 4: Lateral view aortogram showed a very large 10 mm tubular patent arterial duct which was similar in size to the descending thoracic aorta [Movie 2]

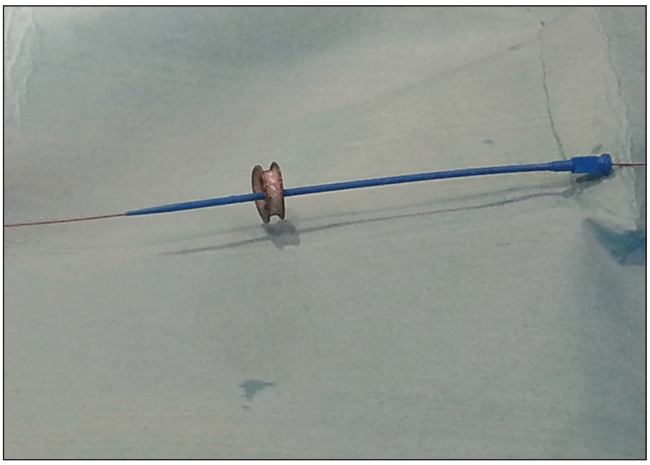

Figure 5: A central puncture was made through the fabric of a 16 mm muscular ventricular septal occluder device with 18G needle, and an 8F dilator was passed over a 0.035˝ wire through the device

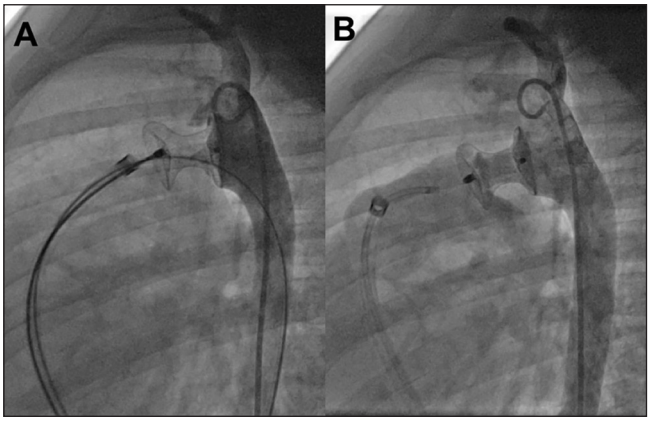

Figure 6: Fluoroscopic lateral view (a) A stable position of the 16 mm Muscular ventricular septal defect device across the patent ductus arteriosus while a combination of 5F right coronary guiding catheter with guide wire is passing through the center of the device. Descending aortic angiogram in lateral view (b) Filling of pulmonary artery through the stent [Movie 4]

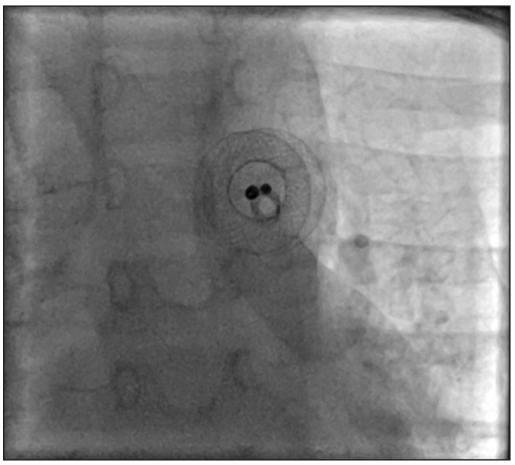

Figure 7: Fluoroscopic image in anteroposterior view showing opened coronary stent orifice enface in the middle of the occluder [Movie 5]

significant challenge in management. Decision-making regarding operability is conventionally based on demonstration of clear left to right shunt on clinical examination, chest X-ray, echocardiogram, and pulse oximetry of lower limbs. Test occlusion is often utilized to predict operability by noting the reduction of pulmonary artery pressures and its separation from aortic pressures. Postocclusion pulmonary artery systolic pressures more than half of aortic systolic pressures have been identified to be a predictor of persistent PAH after complete device occlusion in hypertensive ducts.[4] The pulmonary artery pressures may be very labile in patients who have airway issues, respiratory infection and on agitation and exercise. Such labile episodes may predispose to suprasystemic pulmonary artery pressures and aortic embolization of the occluder. Patients with high pulmonary vascular resistance are also treated with pulmonary vasodilators to remodel the vasculature on long-term.[5,6]

In spite of dual pulmonary vasodilator therapy, patients with labile elevations of pulmonary artery systolic pressures may experience serious clinical events during such rebounds. In patients with shunt lesions like atrial or VSDs or PDA with severe PAH, it is intuitive to either surgically band these hypertensive ducts or fenestrate the nitinol occluder devices used in cardiac catheter interventions.[8-12] Atrial septal occluder devices used in closure of secundum atrial septal defects are fenestrated either in factory (customized fenestrated devices, Occlutech, Helsingborg Sweden) or on table in the catheterization laboratory by tearing the fabric with surgical blades or electrical bipolar cautery perforation.[14] VSDs in hypertensive situations are also fenestrated in surgical theaters by fenestrating the patches or creating an unidirectional valve on the polytetrafluoroethylene patches.[7-11] Even though surgical banding of the hypertensive duct is a reliable way of reducing the duct size, it carries a risk of collapsing the left lung during

the surgery that markedly worsens the borderline hemodynamics.

Our novel model of fenestration of the device will provide predictable communication between the aorta and pulmonary artery since the lumen of the stent fenestration can be predetermined. A small stent placed within the occluder should offer resistance for transmission of the aortic pressures to the pulmonary vascular bed and thereby enable reduction of pulmonary artery pressures. The resultant reduction of shear stress and circumferential stretch on pulmonary arterioles should reduce the vascular resistance in the pulmonary circuit. The challenge remains on maintaining the patency of the stent over long periods. If the hemodynamics and pulmonary vascular resistance improves on long-term follow-up and the stent flows persists, placing coils or small vascular plugs within the stents can easily close these stent lumen.

Limitations of this model

We conceptualized this novel model of on-table fenestration of the nitinol occluder device in our patient who showed large labile fluctuations in the pulmonary artery pressures on test occlusion. This fenestration, if it remains patent, will ensure a pop off for elevated pulmonary artery pressures. The serious limitation of this model will be the long-term patency of this small coronary stent with a 4 mm lumen across the occluder device. Optimal oral anticoagulation with warfarin may ensure prolonged patency of these stents. Additional strategies to improve the patency of these stents may be deploying larger stents with a diameter of 5-7 mm. If the polyester fabric fibers of the device protrude into the stent lumen, it may predispose to early thrombosis in spite of oral anticoagulation. If there is such an occurrence, it could be prevented by use of polytetrafluoroethylene covered stents. This strategy should be adopted for more patients to study its long-term impact on the management of hypertensive ducts. In view of uncertainty about the long-term patency of these fenestrations, equal emphasis also has to be given for continuing pulmonary vasodilator therapy for an indefinite period until the next catheterization hemodynamics show favorable pulmonary remodeling.

CONCLUSION

Treatment of very large hypertensive ducts in grown-up children especially with Down syndrome poses a significant clinical challenge. If the test occlusion of the duct results in minimal reduction of the pulmonary artery pressures, fenestration of the occluder device with a coronary stent will provide a predictable and precise controlled pop off for the pulmonary circulation in the event of a rebound PAH. Long-term patency of these fenestrations has to be assessed on the follow-up, and may be improved through larger fenestrations, systemic anticoagulation and use of covered stents.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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