Early spontaneous closure of large arterial ducts in two term neonates with Ebstein anomaly after failed attempts of transcatheter closure

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Abstract

In neonatal Ebstein's anomaly of the tricuspid valve, prolonged ductal patency in patients without anatomic pulmonary valve atresia can be deleterious. Circular shunts may develop in patients with different degrees of pulmonary and tricuspid insufficiency. Closure of the arterial duct may result in haemodynamic improvement in particular scenarios. The transcatheter approach is a valuable closure alternative despite some technical difficulties in large-sized arterial ducts and low birth weight neonates. Herein, we report on two consecutive term newborns with Ebstein's anomaly and large arterial ducts in whom mechanical stimulus of the arterial duct during failed attempts of transcatheter closure led after two days to definitive closure followed by good clinical outcomes.

The unique features of neonatal Ebstein's anomaly of the tricuspid valve are the presence of anatomic or functional pulmonary atresia, the patency of the arterial duct, and the variability of pulmonary vascular resistance.[1] Promoting early ductal closure after ruling out or relieving anatomic pulmonary obstruction has been promoted to improve neonatal survival.[1–3] There is no opportunity to determine the timing and the occurrence of spontaneous closure or diminution in the size of the patent arterial duct.[4,5] Transcatheter ductal closure is an interesting option with several advantages but can be technically challenging in low birth weight neonates with large and tortuous arterial ducts due to the limited available sizes of low-profile approved devices.[6–9] Herein, we report on two consecutive term neonates with Ebstein's anomaly and large arterial ducts in whom mechanical ductal stimulus during failed attempts of transcatheter closure led to subsequent early and definitive closure with good clinical outcomes.

Cases presentation

After ruling out anatomic pulmonary obstruction, two consecutive neonates with Ebstein's anomaly were approached transvenously from the femoral vein at our institution in an attempt to close a haemodynamically significant arterial duct. Procedures were performed under general anaesthesia, heparinisation, prophylactic antibiotics, and fluoroscopic control. Baseline aortography was first performed to delineate the ductal anatomy. The tricuspid valve was carefully crossed using a 2.7 Fr Progreat® microcatheter (Terumo Corp. Japan) in combination with a 0.014-inch HI-TORQUE PILOT™ 50 guidewire (Abbott, USA) that were both used as a tutor for the delivery catheter. The clinical and procedural characteristics of both patients are detailed in Table 1.

Case No. 1

An 8 mm Amplatzer™ Vascular Plug II (AVP II) (Abbott, USA) was selected to close the arterial duct (pulmonary end: 6.3 mm, length 8.9 mm) of a 9-day-old patient (3 kg) that was delivered through a 5-Fr JR 3.0 Launcher™ coronary guide catheter (Medtronic Inc., USA). The device was removed before release for an important residual shunt and device instability (Fig 1). The catheter was upgraded to 6-Fr during which the patient experienced two episodes of ventricular tachycardia requiring cardioversion. The delivery attempt of 10 mm AVP II into position was very laborious, and the procedure was aborted after important access-related bleeding.

Case No. 2

A microvascular plug-9Q (MVP-9Q) (Medtronic Inc., USA) was selected to close the arterial duct (pulmonary end: 7.6 mm, length 16.1 mm) (Fig 2) of a 14-day-old patient (2.8 kg) that was delivered through a 5-Fr JR coronary catheter (Cordis Corp., USA). The device immediately migrated to the pulmonary artery trunk after deployment. The unreleased device was judged

Table 1. Patients' clinical, procedural, and follow-up data.

	Case no. 1	Case no. 2
Gender	Female	Male
Gestational age	37 weeks	38 weeks
Presenting clinical feature	Low oxygen saturation	Mild respiratory distress Low cardiac output
CHD	Ebstein Anomaly Type B Functional pulmonary atresia Mild PVI Severe TVR (3 m/s) Right to left atrial shunt	Ebstein Anomaly Type C No antegrade pulmonary flow Moderate PVI Severe TVR (2.5 m/s) Right to left atrial shunt
Disease progression	Antegrade pulmonary flow at DOL 2 with oxygen and iNO therapy	Noninvasive ventilation Supraventricular tachycardia at DOL 4 (Adenosine/ Amiodarone) VAP
Indication of arterial duct closure	Volume overload Risk of circular shunt	Volume overload Risk of circular shunt No anterograde pulmonary flow
Age at intervention (days)	9	14
Weight at intervention (kg)	3	2.8
Height at intervention (cm)	48	52
Ductal Type	Fetal	Fetal
Arterial duct aortic end (mm)	8.4	9.1
Arterial duct pulmonary end (mm)	6.3	7.6
Ratio of Arterial duct pulmonary end to left pulmonary artery	1.57	1.41
Arterial duct length (mm)	8.9	16.1
Venous Access (Fr)	6.6	5
SI-SO time (min)	70	65
Fluoroscopy time (min)	23.6	26.1
Device Delivery	Transvenous	Transvenous
Attempted closure devices	AVP 8 mm - AVP 10 mm	MVP-9Q
Procedural incidents	VT requiring cardioversion 40 ml bleeding	Immediate device migration to PA trunk
Procedure outcome	Procedure abortion Indication for surgical closure	Procedure abortion Wait and see Postponed surgical closure

(Continued)

Table 1. (*Continued*)

	Case no. 1	Case no. 2
Delay to postoperative spontaneous arterial duct closure (days)	2	2
Follow-up period (months)	15	3
Final clinical outcome	Alive, Weight 9.5 kg, Height 94 cm, SpO2 95%, No HF symptoms TTE: No PVI, Stable moderate TVR (2.5 m/s), Normal IVC, No R-to-L atrial shunt	Alive, Weight 4 kg, Height 55 cm, SpO2 85%, No HF symptoms TTE: No PVI, Stable moderate TVR (2.5 m/s), Normal IVC, R-to-L atrial shunt

AVP: Amplatzer vascular plug; CHD: congenital heart disease; DOL: day of life; HF: heart failure; iNO: inhaled nitric oxide; IVC, inferior vena cava; MVP: microvascular plug; PA: pulmonary artery; PVI: pulmonary valve insufficiency, SI-SO: sheath in sheath out; TTE: transthoracic ultrasound; TVR: tricuspid valve regurgitation; VAP: ventilator-associated pneumonia; VT: ventricular tachycardia.
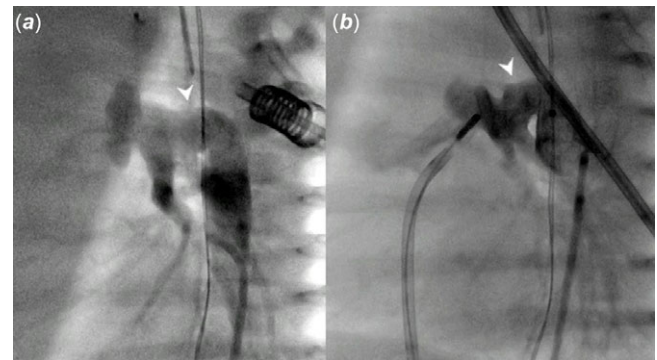

Figure 1. Case. 1. Lateral angiography showing a large arterial duct (*a*). Failure to close a using 8 mm Amplatzer™ Vascular Plug II with a significant para-device residual leak (*b*).
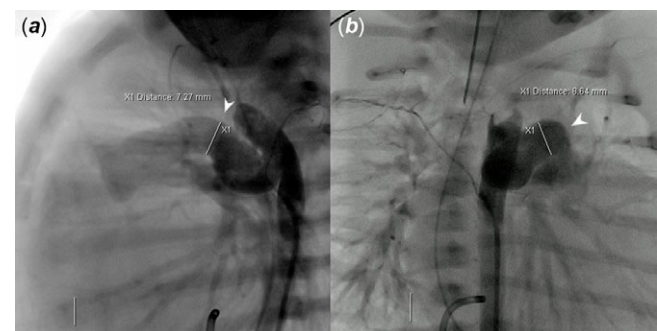

Figure 2. Case. 2. Lateral (*a*) and right anterior oblique (*b*) angiographies showing a very large arterial duct in 2.8 kg term newborn with Ebstein's anomaly.

too small and thereby was re-sheathed and removed. The procedure was aborted in the absence of a larger low-profile occlusion device in the armamentarium.

Both patients were sent back to the neonatal ICU for surveillance and a re-discussion of the management strategy. However, follow-up ultrasound showed a spontaneous and progressive diminution in size until complete ductal closure two days after

the procedure with good immediate and follow-up outcomes (Table 1).

Discussion

The spiral of clinical deterioration observed in some neonates with Ebstein's anomaly has been attributed to ductal patency and circular shunts.[1–3] In the absence of anatomic pulmonary obstruction or after relief of outflow obstructions, early limitation of ductal patency ductal was paired with improved survival in neonates with Ebstein's anomaly.[3] Functional pulmonary atresia happens because the pulmonary blood pressure exceeds the pressure that the small and ineffective functional right ventricle can generate, and the leaflets of the pulmonary valve are unable to open. Therefore, as long as the pulmonary vascular resistance remains high and pulmonary insufficiency remains minimal, systemic oxygenation is maintained through the opened arterial duct and little systemic steal occurs. Yet, when the pulmonary vascular resistance drops, a large arterial duct will jeopardize the diagnosis of functional pulmonary atresia. A large arterial duct will also increase the pulmonary artery pressures, impedes the opening of the pulmonary valve leaflets, and increases the systemic steal. Medically induced pulmonary vasodilation may not always result in the excursion of the pulmonary valve leaflets and the diagnosis will be delayed. In addition, the evaluation of the accurate functional capacity of the right ventricle is very challenging in the presence of systemic pulmonary artery pressures mediated by a large arterial duct. On the other side, in neonates with pulmonary valve insufficiency, ductal patency mediates dangerous circular shunts, leading to intolerable ventricular volume load, significant systemic steal, and haemodynamic instability. In addition, a large left-to-right ductal shunt may compromise cardiac output as increased left atrial pressures limit the right-to-left atrial flow and right-sided decompression.

In all these scenarios, the limitation of ductal patency appears clinically reasonable and beneficial. Transcatheter ductal closure is an appealing minimally invasive straightforward alternative when compared to medical therapy in term neonates or surgical ligation.[10] However, this percutaneous approach and manipulation can be technically difficult in low birth weight neonates. The relative mismatch of currently available introducers, plugs, or occluders to the small anatomic dimensions makes the effective closure of very large ducts very challenging. The implantation of multiple coils or occlusion devices is risky and can be associated with a higher risk of device embolization.[5] Others and we have reported spontaneous ductal closure after unsuccessful attempts at transcatheter device closure of arterial ducts in preterm infants.[7–9] Here, we report, for the first time, the same event in two consecutive term newborns with Ebstein's anomaly. Congenital interventionists are also familiar with different degrees of temporary ductal spasms secondary to mechanical stimulation during cardiac catheterizations.[11,12] However, in these two term newborns, we did not observe ductal spasms or changes in the ductal size whether during manipulation or at the end of the procedure and the postoperative ductal closure was permanent without any evidence of recanalization during follow-up. It appears that mechanical stimulation of the ductal inner wall from transcatheter wire/device manipulation and/or the acute stretch provoked by the occluder deployment within the lumen may initiate the process of spontaneous ductal closure even in term neonates. Therefore, a watchful observation for a minimum of 48 hours after a failed closure procedure might avoid surgical ligation of the arterial duct and related morbidity and complications.[8,9]

Conclusion

Spontaneous and stable closure of large arterial ducts was seen in two consecutive term neonates with Ebstein's anomaly two days after mechanical stimulation during failed transcatheter attempts. We conclude that transcatheter closure should be attempted in similar cases requiring early ductal closure. Surgical ligation can be postponed a couple of days after failed transcatheter interventions when clinically possible. Further well-conducted studies seem interesting to evaluate whether mechanical stimulation of the ductal wall can be a useful alternative to initiate spontaneous ductal closure in preterm and term neonates.

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