Cardiology in the Young

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Hybrid subxiphoid pulmonary valve replacement in a Toddler: focus on technical tips

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Brief Report

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Abstract

Pulmonary valve replacement in small children is rarely needed; when performed, it is usually done surgically. Herein, we describe a hybrid technique for pulmonary valve replacement through a small subxiphoid incision in a Toddler without utilising the cardiopulmonary bypass machine. The procedural technique is thoroughly explained. Collaboration between paediatric cardiology and cardiovascular surgery teams achieved optimal outcome with minimised invasiveness.

Introduction

Severe pulmonary valve regurgitation (whether in the conduit or native right ventricle outflow tract) is often well tolerated in small children, and valve replacement is rarely indicated during the first decade of life. However, when necessary, exclusive transcatheter options are often not feasible due to the patient's small size.[1,2] In this context, a hybrid approach emerges as an appealing option for achieving valve replacement with the advantage of avoiding cardiopulmonary bypass.[1–3] Unlike the adult population, experience in small children is lacking and the outcomes are sometimes suboptimal, probably related to the procedure technique in the light of the small patient's size.[1–5]

Case presentation

The patient is a 2-year-old male, weighing 10.7 kg, with pulmonary hypertension and a history of repaired truncus arteriosus, including the placement of a 13-mm right ventricle to pulmonary artery pulmonary homograft conduit. The surgical repair was complicated by severe bilateral branch pulmonary artery stenosis, which necessitated immediate stent placements. Additionally, the conduit valve failed and became severely regurgitant within 2 months, with evidence of increased conduit size. Subsequently, the patient developed stenosis of the common left pulmonary vein which is believed to be contributing to his pulmonary hypertension. He underwent multiple cardiac catheterisations to rehabilitate the pulmonary arteries and the left pulmonary veins. Also, he was started on pulmonary hypertension treatment. Despite optimisation of feeding via a gastric tube, the patient continued to experience failure to thrive. His echocardiogram revealed significant dilation of the right ventricle and a severely regurgitant, dilated right ventricle to pulmonary artery conduit. The decision was made to replace his pulmonary valve to improve haemodynamics and promote better somatic growth.

Technique

Attempted access to the femoral veins was unsuccessful, with angiography revealing bilateral femoral vein occlusion. Access was then obtained via the right internal jugular vein. The conduit systolic pressure was elevated, measuring 50 mmHg, corresponding to 70% of the systemic systolic pressure. There was an 8 mmHg gradient across the conduit and a 12 mmHg gradient across the left pulmonary artery stent. Angiograms demonstrated a dilated conduit, with the largest diameter measuring 17 mm and the smallest diameter measuring 13 mm (Figure 1 *a* and *b*). A small 4–5 cm vertical subxiphoid incision was made. The diaphragmatic surface of the right ventricle was carefully dissected out. A retractor was applied. Two pledgeted 5-0 Prolene purse string sutures were placed on the predetermined diaphragmatic surface of the right ventricle away from the tricuspid valve. An additional 0.5 cm right paramedian incision was made below and to the right of the subxiphoid incision through which 6 Fr sheath was tunnelled under the skin with the tip at the opened subxiphoid area. Through the purse string, a 7 Fr sheath was inserted in the right ventricle through the purse string sutures. Then, through this sheath, a 0.035" Amplatzer super stiff wire was placed in the left pulmonary artery. The sheath was removed, and the wire was passed through

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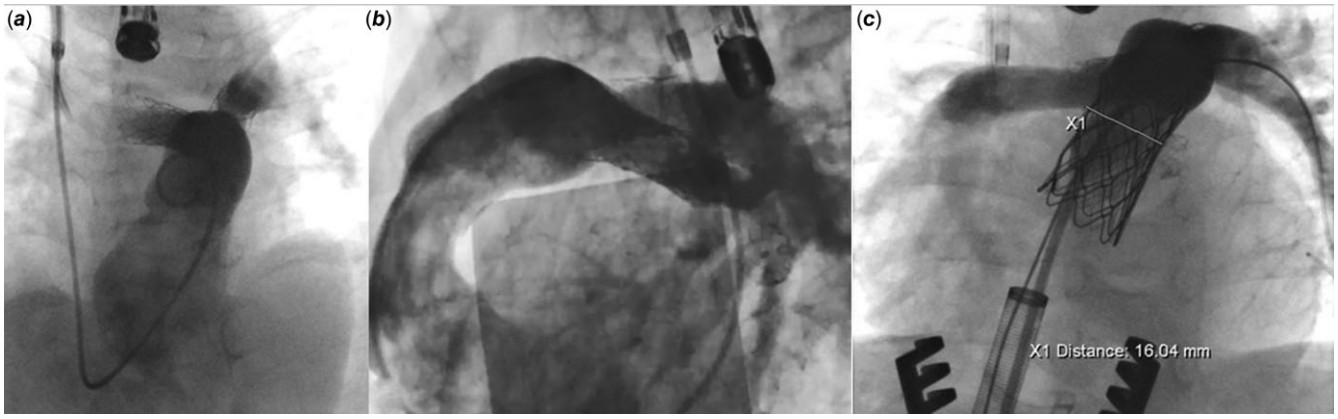


Figure 1. Angiogram ((*a*) frontal projection and (*b*) lateral projection) shows dilated right ventricle to pulmonary artery conduit with free regurgitation. (*c*) Angiogram post-Melody valve replacement shows the valve is well positioned with no regurgitation. Notice the excellent alignment of the sheath with the conduit when the sheath was placed through a right paramedian incision.

the tunnelled sheath. Over the wire, a serial dilation of the skin tunnel and the right ventricle access site was carried out with the final placement of a 22 Fr 33 cm DrySeal sheath (Figure 2). The coronary compression test using a 16 mm balloon was negative. As such, the Melody valve was planned to be delivered using a 16 mm balloon.

The carrot of an 18 mm Medtronic Ensemble II delivery system was cut, and the balloon was removed with the aim to utilise the sheath of the Ensemble system (Figure 3 *a*). A 16 × 40 mm BIB balloon was inserted inside the sheath of the Ensemble delivery system. A TPV 22 Melody valve was crimped over the 16 mm BIB balloon (Figure 3 *b*), and a supporting 4010 Palmaz XL stent was mounted and crimped over the valve (Figure 3 *c*). Then, the balloon-valve-stent unit was pulled inside the sheath of the Ensemble system. The tip of the balloon was kept slightly outside the Ensemble sheath and minimally inflated to smooth the transition between the wire and the Ensemble system (Figure 3 *d*). Through the DrySeal sheath, the Ensemble system was inserted and advanced over the wire. After confirming proper positioning with angiography, the valve and stent unit were deployed by sequential inflation of the inner and outer balloons. The final angiogram demonstrated a competent valve (Figure 1 *c*). Pressure measurements revealed a 10 mmHg gradient across the valve, a 5 mmHg gradient across the left pulmonary artery stent, a right ventricle systolic pressure of 61 mmHg and an end-diastolic pressure of 10 mmHg, and a distal pulmonary artery pressure with a mean of 32 mmHg. The patient was discharged home after a 3-day uneventful hospital stay.

One month later, a scheduled cardiac catheterisation was performed. The occluded right femoral vein was recanalised, and balloon angioplasty was carried out on the stents in the left common pulmonary vein and branch pulmonary arteries. After recanalising the occluded right femoral vein, haemodynamics revealed a right ventricular systolic pressure of 65 mmHg (78% of systemic) and an end-diastolic pressure of 11 mmHg. There was no gradient across the Melody valve, a 20 mmHg gradient across the right pulmonary stent, no gradient across the left pulmonary artery stent, and an elevated mean distal pulmonary artery pressure of 43 mmHg. The increase in the mean distal pulmonary artery pressure could be related to the worsening stenosis in the pulmonary vein and the administration of contrast during the recanalisation of the occluded femoral vein prior to measuring haemodynamics. At the 4-month follow-up, the patient showed improved growth, gaining 800 g, and his pulmonary valve conduit remained competent with no evidence of stenosis.

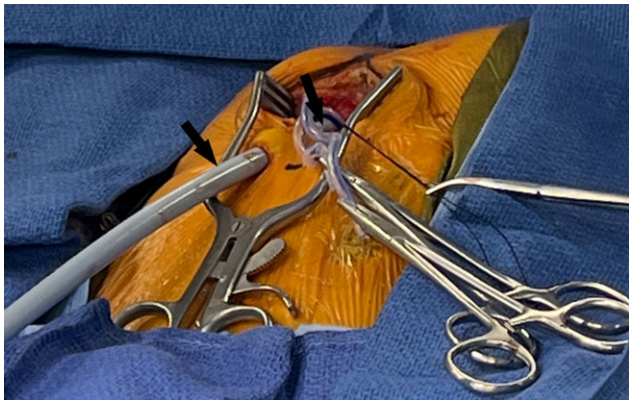


Figure 2. The subxiphoid incision with the tunnelled sheath (black arrows) inserted through a small right paramedian incision.

Discussion

In small children, elevated conduit pressure can worsen the regurgitation caused by the conduit valve insufficiency, especially when the conduit is dilated, leading to an earlier presentation.[2] In the presented case, the indication for valve replacement is the right ventricle dilation accompanied by a refractory failure to thrive, likely related to the severe regurgitation of the conduit.

Early failure of the right ventricle to pulmonary artery conduit is common, especially in small children.[6] One proposed aetiology is conduit compression by the sternum, which can deform the valve.[2,6] In the presented case, additional aetiologies may include the pressurised conduit, which probably led to dilation of the conduit valve annulus and accelerated leaflet deterioration.

Due to the small size, an exclusive transcatheter Melody valve replacement was not an option.[1–3] Nevertheless, there were two possible exclusive surgical approaches. The first is conduit replacement; however, the conduit valve could again fail early. The second option is to place a valve inside the existing conduit via a surgical approach. The valve's frame would protect against

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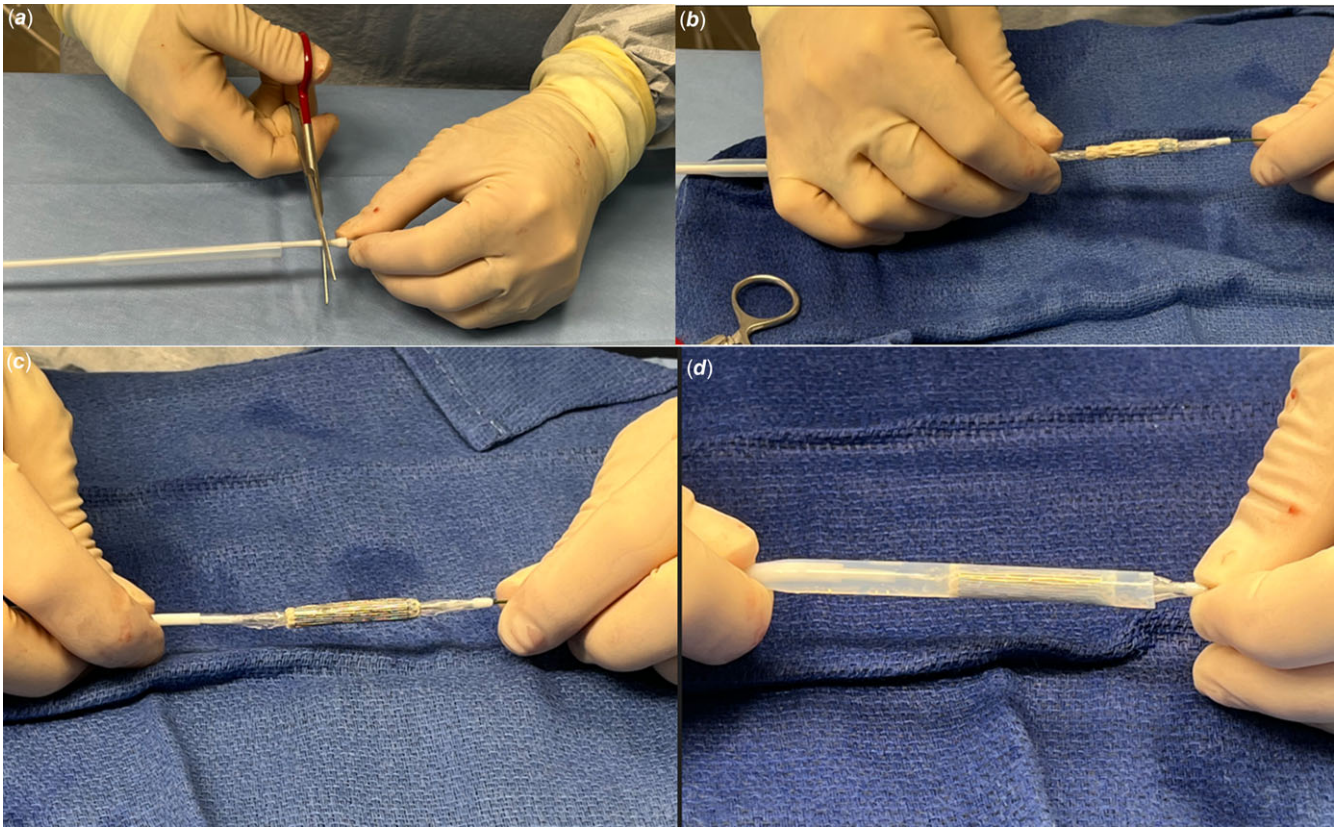


Figure 3. The steps of loading the Melody valve with the supporting stent. (*a*) Cutting the "carrot" of the 18 mm Ensemble system. (*b*) A 16 mm BIB balloon is inserted inside the sheath of the Ensemble system and the Melody valve is crimped on the balloon. (*c*) The supporting (Palmaz XL) stent is mounted and crimped over the Melody valve. (*d*) The distal part of the balloon is extended outside the sheath and partially inflated to smooth the transition with the Ensemble.

compression and likely prolong the valve's longevity.[2] However, this approach would require full sternotomy, cardiopulmonary bypass, and possibly a prolonged recovery, especially in light of pulmonary hypertension.[2,3] Contrary, the hybrid approach allowed placing a competent valve inside the pre-existing conduit with minimised invasiveness.

A few technical tips are worth mentioning, as we believe they facilitate the procedure and mitigate risks. First, the tunnelled 22 Fr DrySeal sheath provided stability to the sheath, reducing the risk of inadvertent sheath loss or forward displacement.[2] Notably, the sheath was kept in the same position just a couple of centimetres inside the right ventricle cavity, which aided in valve introduction, allowed for angiography, and minimised the risk of haemodynamic instability as the DrySeal sheath manipulation was not needed. The sheath was intentionally inserted through the right paramedian incision, rather than the previously reported left, so that its trajectory mirrored that of the conduit (Figure 1 *c*), facilitating easier advancement of the valve.[2] The right paramedian incision was appropriate for this patient based on the orientation of his conduit. Different conduit orientations would require adjustments to the location of the incision. Second, modifying the Ensemble system added another layer of safety. Since the valve is covered with the sheath of the Ensemble, the risk of dislodgment during advancement is reduced. Moreover, the valve can be safely removed if advancement proves difficult or if wire positioning needs to be adjusted or is lost. Third, slightly inflating the balloon smoothed the transition from wire to the Ensemble sheath, mitigating the risk of the Ensemble getting caught on cardiac structures, similar to the functionality of the "carrot" of the Ensemble system. Lastly, stenting the conduit as a single step simplified the procedure and, more importantly, mitigated the previously reported risk of stent dislodgment.[1,7]

Conclusion

Hybrid pulmonary valve replacement approach can be considered in small children, particularly in high-risk cases. This method allows for valve replacement with reduced invasiveness and a smoother recovery. However, more information is needed regarding the longevity of the Melody valve when used in young children.

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Competing interests. None.

Ethical standard. No specific ethical approval from Institutional Reviews Boards is necessary for this type of publication. The authors assure that all patient data provided in this case report are anonymised.

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