Title:
Revalidation to single ventricle pathway with single ventricular assist device: proof of concept.

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Glossary of Abbreviations:
- total anomalous pulmonary venous drainage, TAPVD
- atrioventricular septal defect, AVSD
- ventricular assist device, VAD
- bidirectional cavo-pulmonary shunt, BCPS
- extracorporeal membrane oxygenation, ECMO
- polytetrafluoroethylene, PTFE

Central Picture Legend:
- Ventricular assist device and shunt in a patient with single ventricle.

Central Message:
- Ventricular assist device implantation improved pulmonary vascular resistances, ventricular function, and atrioventricular valve regurgitation inpatient with single ventricle.
The association of obstructed total anomalous pulmonary venous drainage (TAPVD) and single ventricle condition carries a guarded prognosis, [1,2] with a mortality of up to 90% within the first year of life.[2] In recent times, long-term circulatory support of single ventricle conditions has found more success as a bridge to transplant, but not as a bridge to recovery. We present a case of heterotaxy syndrome, unbalanced atrioventricular septal defect (AVSD), severe common atrioventricular valve regurgitation, reduced ventricular function, and TAPVD in whom ventricular assist device (VAD) implantation resulted in a gradual improvement in ventricular function, atrioventricular valve regurgitation and pulmonary vascular resistances, leading to the successful weaning of the support leading to reassumption of the single ventricle pathway rather than heart transplant.

Clinical Summary

This male newborn was diagnosed with heterotaxy syndrome, dextrocardia, unbalanced AVSD, hypoplastic right ventricle, moderate common atrioventricular valve regurgitation, and obstructed TAPVD to the left superior vena cava (Video). IRB approval and informed consent were waved due to the nature of this study. Due to poor prognosis associated with the constellation of findings, the parents decided to opt for compassionate care. The patient survived and at three months, goals of care were rediscussed. He underwent a bidirectional cavo-pulmonary shunt (BCPS), a TAPVD repair and pulmonary artery banding to preserve some antegrade flow. Two days after the surgery, the patient was extubated. On post-operative day three, he had a cardiac arrest in cardiac intensive care unit and was placed on extracorporeal membrane oxygenation (ECMO) during resuscitation. He was weaned off ECMO on post-operative day eight, but experienced repeated desaturations and dependency on mechanical ventilation. At this stage, the take down of the BCPS to a systemic to pulmonary artery shunt
was deemed necessary but was high risk due to ventricular dysfunction, severe atrio-ventricular valve regurgitation and labile pulmonary vascular resistance. On post-operative day 11, it was decided to proceed with the elective addition of VAD. At 5.3 kg, the BCPS was taken down, and a 6 mm atrial cannula was secured into the right atrium. An 8 mm polytetrafluoroethylene (PTFE) graft, secured to a 5/6 LV apical vent cannula, was sutured to the ascending aorta. A 4 mm PTFE graft was used to create a systemic to pulmonary shunt using the Laks technique with proximal end on the outflow graft. PediMag (Abbott, US, CA) centrifugal VAD support was initiated (Figure). The main pulmonary artery was divided. The atrioventricular valve repair could not be performed because of his dextrocardia anatomy. Over time, the patient's ventricular function normalized, and common atrioventricular valve regurgitation improved significantly to mild range. At the age of six months, eight weeks after VAD implantation and BCPS takedown, the VAD was explanted with cardiopulmonary bypass. The 8 mm PTFE graft was clamped and divided between the origin of the Laks shunt and the outflow cannula leaving a small segment of the 8 mm PTFE graft from which the 4 mm Laks shunt was attached. Patient recovered well from this operation with normal ventricular function and mild atrio-ventricular regurgitation at discharge. The patient returned at 14 months of age for an elective BCPS and did well with no post-operative complications. At the age of 2 years, he is thriving while awaiting Fontan completion.

Discussion

We hereby report successful VAD support of a patient with heterotaxy, unbalanced AVSD, obstructed TAPVD, atrio-ventricular regurgitation and poor ventricular function as a bridge to recovery after failure of BCPS circulation. Despite the overall growth and progress made in VAD support of patients with single ventricles, outcomes reported with this strategy have been
suboptimal. A previous study demonstrated high mortality in stage 1 and stage 2 patients compared to stage 3 patients; 6-month survival was 30% for stage 1, 40% for stage 2, and 95% for stage 3 [3]. The team from Gainesville reported better outcomes with various constructions of single ventricular support, which inspired us to use a Y branching of the outflow graft cannula in our case. They performed VAD insertion in 15 patients with functionally single ventricle. Ten patients survive (67%) and achieved heart transplant, and five patients died (33%).[4]

Sustained improvement of atrio-ventricular valve regurgitation with VAD support has been described in adults with mitral valve regurgitation but not in patients with single ventricle [5]. We believe that the observed improvement in ventricular function and atrioventricular valve regurgitation following VAD implantation in this patient is a proof of the concept that patients with failing single ventricular circulation can be bridged to recovery by temporary VAD support. We believe that this concept may open avenues of treatment.


Figure legend: Bidirectional cavo-pulmonary shunt (BCPS) Take Down and Ventricular Assist Device implantation: The BCPS was taken down, and a 6 mm atrial cannula was secured into the right atrium. An 8 mm polytetrafluoroethylene (PTFE) graft, secured to a 5/6 LV apical vent cannula, was used as an outflow cannula, and was sutured to an opening made in the ascending aorta. A 4 mm PTFE graft was used to create a systemic to pulmonary shunt using the Laks technique. PediMag (Abbott, US, CA) VAD support was initiated.

Video legend: Clinical summary of the patient.