Pulmonary Artery Banding for Functional Regeneration of End-Stage Dilated Cardiomyopathy in Young Children

World Network Report

We report the worldwide experience of surgical pulmonary artery banding (PAB) treatment of end-stage left ventricular dilated cardiomyopathy (LV-DCM) with preserved right ventricular function. Pediatric LV-DCM is a leading cause of cardiac death, especially if the left ventricular end-diastolic diameter exceeds a z score of +5. Heart transplantation (HTx) is the only viable life-saving option, but is limited by donor availability and an unpredictable long-term survival. PAB was introduced on the following hypotheses: (1) geometric rearrangement of LV dimension is achieved by reestablishing the interventricular septal position with gradual restoration of LV ejection fraction; (2) cardiac improvement with potential for regeneration reciprocal to the patient's age is promoted by PAB-induced right ventricular hypertrophy. Fifteen centers of 11 countries from America, Asia, and Europe introduced PAB in an effort to diminish the need for LV assist device support and HTx, and especially to enable functional recovery.

The Figure shows the flowchart of 70 patients who received PAB between March 2006 and 2017. Nine patients (mean age 159±101 days) underwent PAB placement after complex open-chest procedures including: mitral valve repair (n=4); mitral valve replacement (n=1), anomalous left coronary artery arising from the pulmonary artery repair (n=1), epicardial pacemaker placement (n=1), fenestrated atrial septal defect closure (n=1), and repair of a left-sided partial anomalous pulmonary venous return (n=1). All but the latter patient recovered following a mean intensive care stay of 25±33 days and 54±32 days to discharge. The child with partial anomalous pulmonary venous return died 8 months after corrective surgery, the LV-DCM did not respond to PAB, and HTx listing was declined. Experience with these 9 patients supports that PAB can serve as a recovery strategy to wean patients with LV-DCM after open-heart surgery.

PAB was selectively performed without operative mortality in 61 patients (n=31 female) using a short open-chest approach. Given the different diagnostic tools among the institutions, retrospective record analysis classified DCM as idiopathic (n=44), chronic myocarditis (n=9), and associated with LV noncompaction (n=8). The mean age at PAB was 266±310 days and 87±162 days (n=28) from diagnosis. Five patients required cardiopulmonary resuscitation before admission and 2 had extracorporeal membrane oxygenation before PAB. In 8 patients, PAB was performed as bridge to early HTx with the aim to avoid a LV assist device. Despite this, in 1 center, 2 of 4 patients treated with PAB required a LV assist device; both subsequently died. The other 6 patients underwent HTx successfully within the first 4 weeks after uneventful PAB; 2 of them despite exhibited improved function.

It is noteworthy that, of the remaining 53 patients (mean age 274±320 days), 6 patients died (11%). Two of them, with morphological LV noncompaction, deteriorated and died several months after percutaneous transcatheter PAB, despite initial functional recovery. Five patients (9%) showed no evidence of PAB response. Four

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The pressure gradient (n=34) had percutaneous pulmonary debanding after death and transplant rate, 27 of the recovered patients (12.5%) had functional recovery. Eight of the 42 patients showed significant improvement, but not yet full recovery, because their brain-type natriuretic peptide (n=34). The functional class improved from a median of IV (III–IV) to I (I–II). Median brain-type natriuretic peptide increased across the PAB form 41±14 at discharge to 70±22 mm Hg before debanding.

PAB-world-network reports a new indication for an established surgical procedure along with encouraging short- to medium-term outcomes in a pediatric population with end-stage LV-DCM. Technically, PAB is simple, safe, effective, and affordable. It is a promising treatment strategy, especially in countries where transplant is not a realistic option. Introduction of PAB to the therapeutic arsenal represents a paradigm change in managing pediatric end-stage LV-DCM.

ARTICLE INFORMATION

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Data Sharing: The data, analytic methods, and study materials will not be made available.

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Disclosures

None.

APPENDIX

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