

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

## World Network Report

**W**e 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,<sup>1,2</sup> especially if the left ventricular end-diastolic diameter exceeds a z score of +5.<sup>3</sup> 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<sup>4</sup>: (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<sup>5</sup> 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±101days) 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.

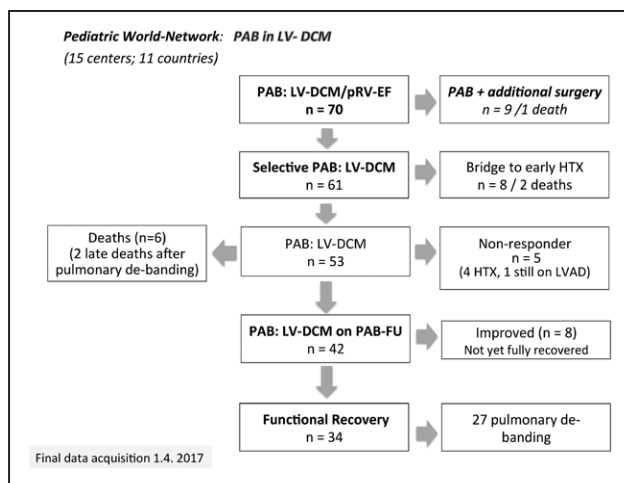
It is noteworthy that, of the remaining 53 patients (mean age 274±320 days), 6 patients died (11%). Two of them, with morphological LV noncompaction, deteriorated and died several months after percutaneous transcatheter PAB, despite initial functional recovery. Five patients (9%) showed no evidence of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

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**Figure. Seventy patients with left ventricular dilated cardiomyopathy and preserved right ventricular ejection fraction: treatment by a surgical pulmonary artery banding (PAB).**

Nine patients received PAB after additional open-heart surgery. In 61 patients, PAB was performed by a selective open-chest approach; from 42 PAB responders, detailed FU analysis was performed (see text). DCM indicates dilated cardiomyopathy; EF, ejection fraction; FU, follow-up; HTx, heart transplantation; LV, left ventricle; PAB, pulmonary artery banding; and pRV, preserved right ventricle.

of these had subsequent HTx, and one is still awaiting HTx after being placed on a LV assist device.

Forty-two patients were followed for a mean  $38 \pm 28$  months (range, 3 months to 11 years). Following surgical PAB, intensive care stay was  $12 \pm 7$  (ventilation time  $2 \pm 2$  days); in-hospital stay was  $38 \pm 25$  days. Thus far, the majority ( $n=34$ ) has experienced functional recovery. None has required relisting for HTx. LV end-diastolic diameter decreased from  $46 \pm 6$  mm (z score  $7 \pm 2$ ) to  $37 \pm 8$  mm (z score  $4 \pm 2$ ) at discharge and  $35 \pm 6$  mm (z score  $1 \pm 2$ ) at the last follow-up. The corresponding LV ejection fraction increased from pre-PAB of  $20 \pm 8\%$  ( $n=34$ ) to  $44 \pm 12\%$  after 3 to 6 months ( $n=34$ ) and  $60 \pm 10\%$  ( $n=34$ ). The functional class improved from a median of IV (III–IV) to I (I–II). Median brain-type natriuretic peptide values obtained before PAB, after 3 to 6 months, and at the last follow-up ( $n=20$ ) decreased from 3046 pg/mL (interquartile range, 990–4911 pg/mL) to 183 pg/mL (interquartile range, 75–250 pg/mL) and 29.5 pg/mL (interquartile range, 18.5–46.5 pg/mL), respectively. At the end of the observation period, 34 of these patients recovered. Eight of the 42 patients showed improvement, but not yet full recovery, because their mean LV end-diastolic diameter measured  $43 \pm 7$  mm (12.5% z score decrease) and LV ejection fraction measured  $25 \pm 10\%$  (+39% increase). Considering overall death and transplant rate, 27 of the recovered patients ( $n=34$ ) had percutaneous pulmonary debanding after  $14 \pm 9$  months without relapse. The pressure gradient

increased across the PAB from  $41 \pm 14$  at discharge to  $70 \pm 22$  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

The online-only Data Supplement is available with this article at <http://circ.ahajournals.org/lookup/suppl/doi:10.1161/CIRCULATIONAHA.117.029360/-DC1>.

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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