Three-Patch Aortic Root Reconstruction With Extended Left Main Coronary Artery Patch Augmentation in Neonates and Infants

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Left main coronary artery (LMCA) stenosis is present in approximately 5% of patients with congenital supravalvular aortic stenosis (SVAS) (Fig. 1)[1] and is associated with an increased risk of sudden cardiac death.[2] However, patients undergoing coronary artery intervention at the time of SVAS repair are at the highest risk of experiencing major adverse cardiac events.[3] Literature reports of surgical techniques and outcomes of concomitant coronary artery repair in these high-risk patients are diverse and inconsistently described. We have recently adopted a standardized surgical technique for management of this complex pathology by combining extended LMCA patch augmentation with a 3-patch aortic root reconstruction (Brom's technique). In this report, we describe our contemporary surgical technique of 3-patch aortic root reconstruction with extended LMCA patch augmentation for patients with congenital SVAS with ostial LMCA stenosis and bilateral outflow tract obstruction. Institutional review board approval was obtained for retrospective review of patient charts.

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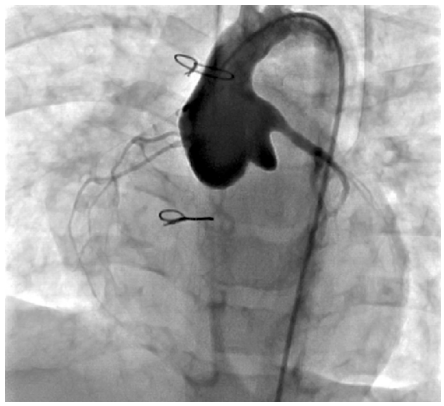
Keywords: Supravalvar aortic stenosis, William's syndrome, Coronary artery



Three-patch aortic root reconstruction with left main coronary artery patch augmentation.

Central Message

Three-patch aortic root reconstruction with left main coronary artery (LMCA) augmentation can be performed safely in neonates and infants with supravalvular aortic stenosis and ostial LMCA stenosis.

TECHNIQUE

Please see the supplementary video for an example of the operative technique. Operations are performed via median sternotomy using cardiopulmonary bypass. After preliminary dissection and bicaval cannulation, right-sided obstructive lesions are repaired prior to aortic cross-clamping, including right ventricular outflow tract (RVOT) patch augmentation, resection of obstructing infundibular muscle bundles, main pulmonary artery (PA) augmentation, and branch PA augmentation from hilum to hilum when needed. The exception is patients with well-placed branch PA stents, which are instead dilated with a nasal speculum.

After reconstruction of the right-sided obstructive lesions, cardioplegic arrest with 20 mL/kg of del Nido cardioplegia is administered after aortic cross-clamping. The ascending aorta is then divided just above the sinotubular junction. Vertical incisions are then placed into the noncoronary sinus and then the right coronary sinus to the left of the right coronary artery. The incision in the left coronary sinus is then carried out onto the LMCA past the site of fibrous ostial obstruction, nearly to the left main bifurcation. A total of 10–12 mm triangular patches of glutaraldehyde-fixed autologous pericardium or pulmonary homograft are then fashioned and used to reconstruct the sinuses. The left coronary sinus patch contains an extra extension for augmentation of the LMCA. After completion of the aortic root reconstruction, the ascending aorta and arch are augmented anteriorly under a period of deep hypothermic circulatory arrest or regional perfusion, when indicated (6 of 8 patients), terminating with re-anastomosis of the augmented ascending aorta with the sinotubular junction of

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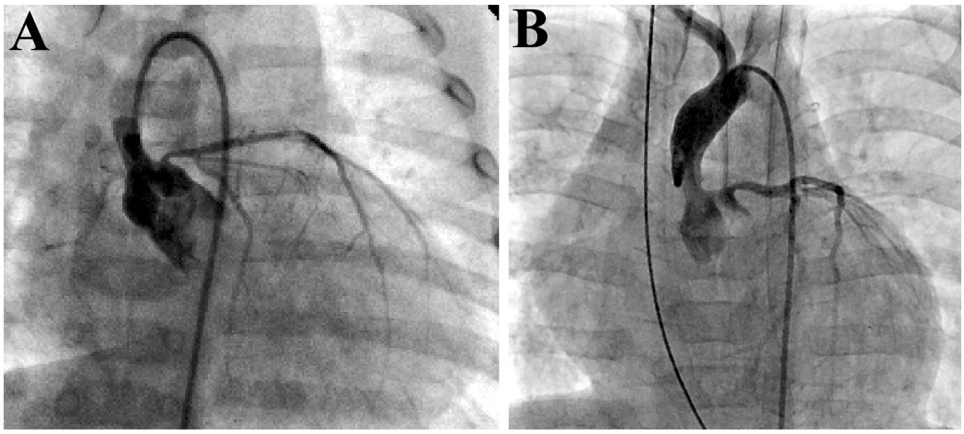


Figure 1. (A, B) Preoperative aortic root angiograms demonstrating supravalvular aortic stenosis with left main coronary artery stenosis.

the reconstructed aortic root. The cross-clamp is then removed and the patient is weaned from cardiopulmonary bypass following de-airing maneuvers. Echocardiography is performed to confirm satisfactory repair of all lesions without residual outflow tract gradients. Heparin is initiated 6 hours postoperatively at 10U/kg/hr and titrated to a therapeutic range as tolerated. Patients are then managed with oral aspirin after institution of an enteral diet.

Between 2011 and 2017, this technique was applied to 8 patients (age range 2 days to 18 months) with SVAS and ostial LMCA stenosis documented by cardiac catherization and direct intraoperative inspection, all of whom required concomitant

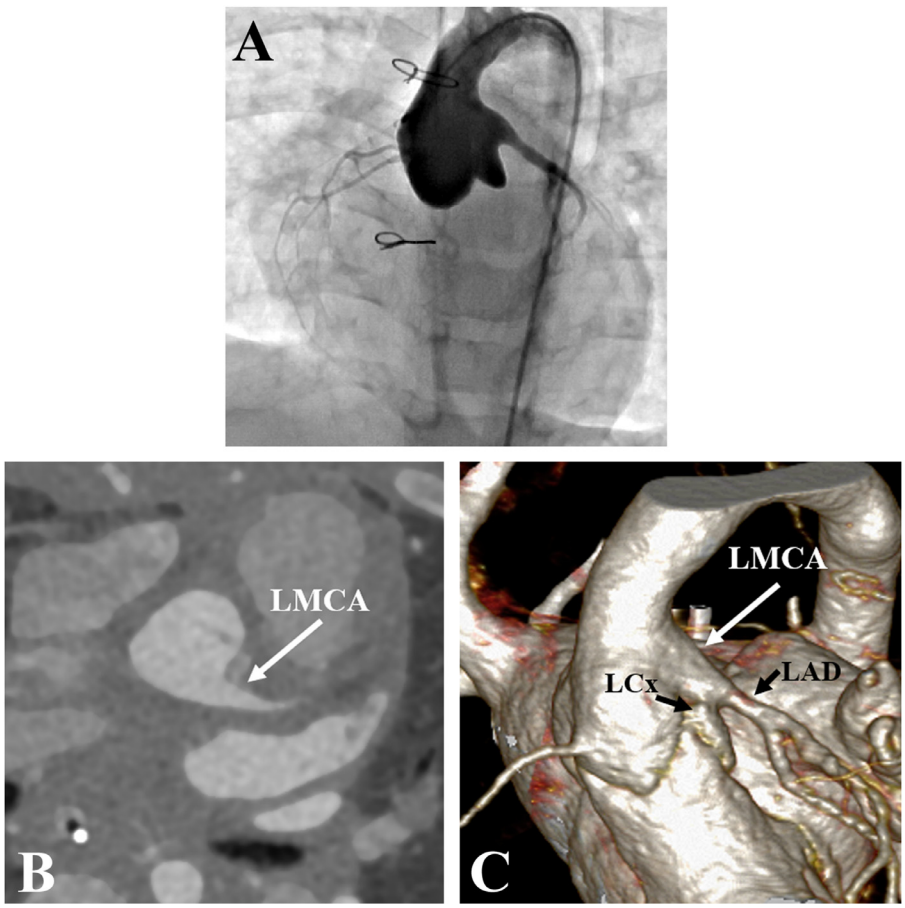


Figure 2. (A) Postoperative aortic root angiogram and (B, C) computed tomography angiography demonstrating successful repair of the obstructive aorta and coronary artery lesions.

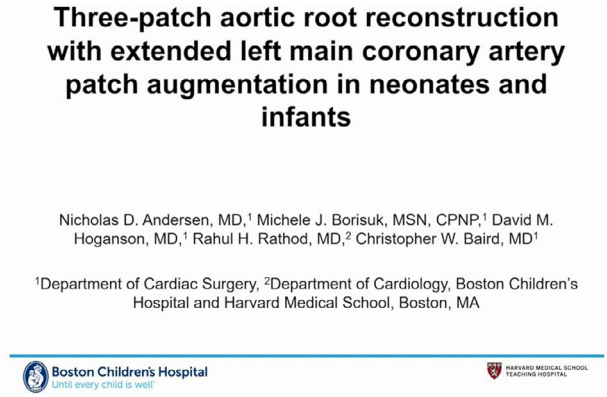
pulmonary artery and/or RVOT augmentation for biventricular outflow tract obstruction. All patients survived the initial procedure and postoperative echocardiography demonstrated a reduction in the SVAS gradient from a mean of 57 ± 28 mm Hg preoperatively to <10 mm Hg in all cases. Good flow was seen in the coronary arteries and no patient exhibited more than mild aortic insufficiency. All patients underwent catheterization or computed tomography angiography in the follow-up period, demonstrating unobstructed coronary artery blood flow in all cases (Fig. 2). One patient required re-operation for RVOT augmentation due to residual subpulmonary and/or infundibular stenosis and also possessed severe peripheral pulmonary stenosis with severe pulmonary hypertension and right ventricular dysfunction. This patient ultimately died 66 days after the index procedure after experiencing arrhythmia, cardiac arrest, and multisystem organ failure following ECMO cannulation. However, good flow in the LMCA had been visualized on all prior echocardiography and cardiac catheterization studies.

DISCUSSION

Patients with William's syndrome undergoing coronary artery intervention or combined right and left ventricular outflow tract procedures are at the highest risk of major adverse cardiac events, reported at 32% and 21%, respectively, in the Society of Thoracic Surgeons database.[3] As a result, complete repair may be deferred until later in life or indefinitely. Although technically challenging, we recommend complete repair of all obstructive outflow tract and coronary artery lesions at the time of the index procedure, including in neonates and infants, to minimize the need for postoperative intervention and sudden cardiac death. Our current approach to these highest risk patients with LMCA stenosis is to employ the 3-patch aortic root reconstruction with extension of the left coronary sinus patch into the LMCA nearly to the bifurcation. In our experience this technique was associated with excellent survival to hospital discharge, and no residual aortic or coronary lesions identified on follow-up imaging (Fig. 2).

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Video 1. Video of surgical technique.

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