Modified Bex-Nikaidoh Procedure

Objective

The Bex-Nikaidoh procedure is a surgical option for transposition of the great arteries (TGA) or double outlet right ventricle (DORV) with ventricular septal defects (VSD) and pulmonary stenosis (PS). Abnormal coronary anatomy with posterior looping around the great vessels and papillary muscle crossing the VSD are relative contraindications. We present a patient with DORV and TGA with PS, VSD, crossing papillary muscle and aberrant coronary anatomy who underwent modified Bex-Nikaidoh procedure.

Case Video Summary

A 22-month-old with DORV, TGA, and subpulmonic VSD presented for planned operation. Shortly after birth a balloon atrial septostomy was performed for inadequate mixing. He was stabilized and discharged with plans for surgery after optimized growth. CTA heart showed 2 coronary arteries with coronary #1 from the left sinus of Valsalva containing the LAD and coronary #2 from the posterior sinus of Valsalva giving rise to RCA and LCX and LCX with a retro-pulmonic course. Echo showed anterior papillary muscle of the tricuspid valve straddling the VSD with infundibular septal attachments, precluding a Rastelli repair.

Therefore, a modified Bex-Nikaidoh procedure was performed. After right atriotomy, papillary muscle position was confirmed and isolated. Coronary buttons were harvested and mobilized to facilitate aortic root harvest. After aortic root harvest, pulmonary root was inspected and deemed too small for transfer. The pulmonary root was opened, and leaflets excised for incorporation into the neo-left ventricular outflow tract (LVOT). The neo-aortic root was translocated. VSD was closed with bovine pericardial patch creating the neo-LVOT. The divided papillary muscle was re-implanted into VSD patch. Coronary buttons were implanted. Right ventricle to pulmonary artery conduit was created using aortic homograft. The tricuspid valve was repaired, and atrial septal defect (ASD) closed with fenestrated bovine pericardial patch. The patient weaned from cardiopulmonary bypass with normal biventricular function and discharged on post operative day 27. Discharge echo showed normal biventricular function, trivial neo-aortic insufficiency, mild tricuspid regurgitation, no residual VSD and small residual ASD.

Conclusions

A modified Bex-Nikaidoh operation can successfully be performed in patients with DORV, TGA, VSD and PS despite relative contraindication of posterior looping coronary artery and papillary muscle crossing the VSD.

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