



Double Switch Procedure for CCTGA with VSD and Subpulmonary Stenosis

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Clinical Image

We describe the problem of Congenitally Corrected Transposition of Great Arteries (ccTGA) with VSD and sub pulmonary stenosis. The surgical management strategy of double switch procedure using anen-bloc aortic root translocation and the Senning procedure is presented on the basis of a case of a one-year-old boy presenting with ccTGA, a restrictive ventricular septal defect and severe sub pulmonary stenosis (left ventricular outflow tract obstruction, Figure 1). Due to accompanying lesions in addition to ccTGA, decision for surgical correction was made by our interdisciplinary congenital cardiac team. The operative procedure includes insertion of the heart lung machine by cannulating the upper and lower caval veins directly, as well as the aortic arch. On total cardiopulmonary bypass, the aortic and pulmonary roots are mobilized through the sub arterial conus muscle and harvested. Extensive mobilization of the coronary arteries as a critical step of the procedure is carried out. The Bex-Nikaidoh aortic translocation procedure entails posterior incision into the conal septum and through the hypo plastic pulmonary valve annulus, opening up the stenotic left ventricular outflow tract [1]. The aortic root is then relocated in a posterior direction above the left ventricular outflow tract. The VSD closure is then performed. The right ventricular out flow tract and right ventricle to pulmonary artery continuity are re-established using a conduit like bovine jugular vein graft. In the herein presented case, the technical modification of en-bloc rotation (180 degrees) for anatomical correction of ventriculo-arterial discordance is shown, which includes excision and rotation of both outlet valves en-bloc. The reason for choosing this strategy was the fact that pulmonary valve annulus was only mildly hypoplastic and the pulmonary valve itself functionally intact. In order to correct double discordance of ccTGA, the atrial switch is then performed using the Senning operation [2], which entails creation of a systemic venous channel from both the inferior and superior caval veins to the inlet valve of the morphologically right ventricle on the patients left side. The pulmonary venous channel is then redirected around the systemic venous channel (also called baffle), and after coursing a c-shape pattern reaches the mitral valve and inlet into the morphologically left ventricle on the patient's right side of the body. Complex TGA is defined as TGA in the presence of a ventricular septal defect and/or subvalvular, valvular or supra valvular pulmonary stenosis or left ventricular outflow tract obstruction (Figure 2). ccTGA includes double discordance, meaning discordance of the atrio-ventricular and the ventriculo-arterial connections. Indication for surgery in patients with ccTGA is based primarily on accompanying lesions, including atrioventricular valve regurgitation, outflow tract obstruction, ventricular septal defect and others. In a recent series of 63 patients treated between 1997 and 2016, freedom from death and any clinically relevant cardiac event after anatomic correction of ccTGA was 95% and

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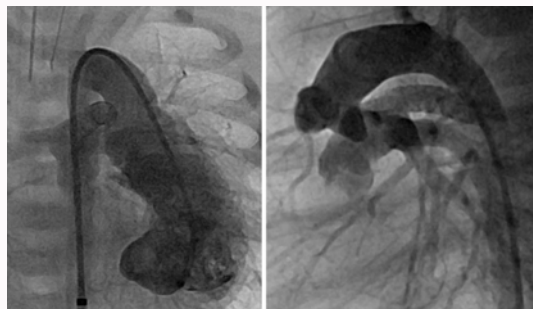


Figure 1: Congenitally Corrected Transposition of Great Arteries (ccTGA) with situs solitus of atria and I-looping of the morphologic right ventricle, which is on the patient's left body side (ventriculography left image). Transposed great vessels with stented ductus arteriosus (right image).

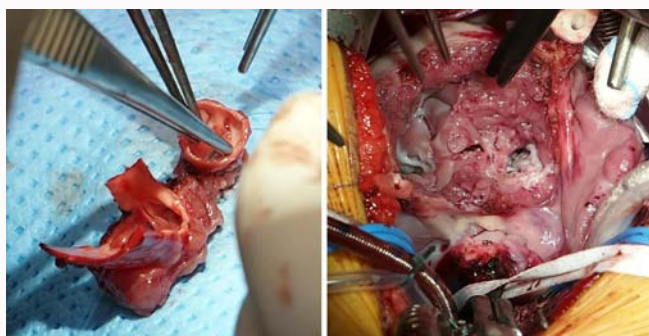


Figure 2: Intraoperative view of aortic and pulmonary root translocations using the en-bloc rotation technique. Left image: both outlet valves are excised and trimmed en bloc and subsequently rotated and sutured to muscular rims of conus muscle after treatment of subvalvular stenosis (right image).

71%, respectively, after 15 years of follow-up [3]. The late functional state and quality of life in this cohort of patients is promising, as well as echocardiographic findings at late follow-up which show preservation of function of the morphologically left ventricle in 93% after 15 years. Also for dTGA, satisfying long-term survival and other clinical outcomes have been reported [4]. Dilatation of the neo-aortic root and neo-aortic regurgitation is observed over time and contributes to a growing number of readmissions of grown-ups with congenital cardiac defects.

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