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Bivalvation Procedure with Plicated Interatrial Septum Remnant for Common Atrioventricular Valve Regurgitation in Neonate with Asplenia Syndrome

Imoto Y., Matsuba T., Ogata Y., Yamamoto H., Matsumoto K., Arata K., Imagama I., Kanda H., Toyokawa K., Tateishi N., Nagatomi S.

Cardiovascular and Gastroenterological Surgery, Kagoshima University Graduate School of Medical and Dental Sciences, Kagoshima, Japan

Background: Regurgitation of common atrioventricular valve (CAVV) with moderate grade or more in a neonate with asplenia syndrome is a formidable problem, especially when complicated with TAPVC with pulmonary venous obstruction. Morphological understanding is very difficult in neonatal CAVV, and repair technique has not been established.

Case: The patient was a 7-day-old boy with asplenia syndrome complicated with single ventricle with CAVV, infracardiac type TAPVC, pulmonary atresia and PDA. Echocardiography showed severe CAVV regurgitation from the central portion. He presented with failure to thrive. Operation was performed with conventional cardiopulmonary bypass with aortic and bicaval venous cannulae. Performed procedures were as follows; PDA ligation, Blalock-Taussig shunt (EPTFE 3.5 mm), patch augmentation of the pulmonary bifurcation, anastomosis of the common PV to the atrium, ligation of the vertical vein, and CAVV repair. Saline test in CAVV revealed significant regurgitation, but responsible valve lesions were unclear. Meticulous techniques for valve repair as are used in older patients seemed unrealistic, and we performed plication of the interatrial septum remnant (IASR) expecting the bridge of IASR would improve coaptation of the central portion of the CAVV and also protect against valve prolapse. Postoperative echocardiography showed mild CAVV regurgitation. He underwent bidirectional Glenn procedure at the age of one year, and total cavopulmonary connection (TCPC) at 3 years old. Eight years have passed after TCPC, but his CAVV regurgitation remains mild and no significant stenosis is noted.

Conclusion: Bivalvation procedure with plicated IASR is a useful technique for CAVV regurgitation in neonates with asplenia syndrome.