

CASE REPORT

A 3 Year Old Girl with L-Transposition of The Great Vessels and Severe Left Sided AV Valve Insufficiency

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L-transposition of the great vessels (L-TGA) is a rare but complex congenital heart disease that could represent a challenge for patients and their caring physicians. Patients with L-TGA could go undetected for years if they have no other associated cardiac defects; however, they could present with complete heart block with time. In its simplest and connection analysis, corrected transposition of the great vessels entails atrio-ventricular discordance and ventriculo-arterial discordance, meaning the left atrium connected to the right ventricle, and the right ventricle gives rise to the aorta. The right atrium is connected to the left ventricle and the left ventricle gives rise to the pulmonary artery. As a result, the circulation is corrected with the pink blood being ejected to the aorta through the RV and the blue blood being ejected to the pulmonary artery through the LV. There can be associated lesions with L-TGA, the most important of which is ventricular septal defects,

sub-valvar pulmonary stenosis and deformation of the tricuspid valve (Systemic AV valve). Complete heart block can occur with those patients at a rate of 1% per year.

NM is a 3 1/2 year old toddler who is known to have L-TGA with Ebstenoid malformation of the tricuspid valve with severe insufficiency. Her right ventricle started to fail. There was pulmonary hypertension documented by cardiac catheterization with the systolic pulmonary artery pressure of 70 mmHg. The choices at that time were either to repair the valve and keep the anatomy the same or perform the double switch procedure. The decision was to proceed with the double switch approach. In this approach the surgeon did arterial switch procedure and atrial switch (Senning procedure). The patient had her sternum opened after surgery and packed over night on mechanical ventilation. The next day, her chest pack was removed and the sternum was closed. Extu-

bation was performed within 6 hours of her chest closure. Her hemodynamics continued to be very stable and was able to feed orally the following day. An echocardiogram was performed at post-operative day number 4 and demonstrated both ventricles with normal systolic function and complete resolution of her tricuspid regurgitation. This patient needs close follow-up in the future of her biventricular systolic function, baffle obstruction or leak and arrhythmias. Performing double switch procedure is a huge achievement for our institution since it entails a certain level of quality of care provided to our patients.