The evaluation of Double Inlet Left Ventricle with Ventriculoarterial Discordance: Four years experience from a single center

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Objectives:
Double inlet left ventricle is the most common form of single or common ventricle. The restriction of the bulboventricular foramen (BVF) and subaortik obstruction are important complications in these patients. We undertook a review of the data of patients with double inlet left ventricle (DILV), ventriculoarterial discordance (VAD).

Patients and Methods:
Thirty-one patients diagnosed as DILV were evaluated between 2009 and 2013. The patient records reviewed retrospectively.

Results:
Twenty-four of 31 patients have functionally single ventricle, VAD and systemic outflow through a BVF; no patient had outflow obstruction at birth. Thirteen were female (54%), 11 were male (45%). Two patients have atretic pulmonary valve, 6 have pulmonary stenosis, 16 have pulmonary hypertension. One of the patients had arcus, isthmus hypoplasia at initial evaluation, where coarctation developed in another patient after pulmonary banding operation (PAB). Fourteen patients were operated and 3 patients planned to be operated, 2 patients were inoperable and 5 patients were lost follow up. Among the operated patients initially shunt operation was performed in 3, PAB in 9, bidirecctional cavapulmonary connection (BCPC) in 2 patients. Initial palliation with PAB was performed in association with other procedures and the median age of initial palliation was 1,75 (0,5 to 2,5) months. The median follow-up duration was 7,5 (0,13-51) months. During follow up BCPC were completed in 6 of the patients (BCPC was performed as initial operation in 2) and the median age was 12,5 (8 to 156) months during BCPC operation. BVF restriction was not reported in our patients at admission or after PAB procedure. Arcus reconstruction and pulmonary banding was planned in one patient at admission and the coarctation developed on follow up was repaired in another patient 5 months after banding operation. Two patients died during follow up.

Conclusion:
Norwood operation or pulmonary artery banding with aortic arch repair can be performed as the initial palliation in DILV, VAD patients. The latter procedure and early BCPC after palliative banding can be the procedure of choice to prevent early BVF restriction.