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Outcomes of Modified Konno Procedure in Obstructive Hypertrophic Cardiomyopathy in Children

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Objectives:
Modified Konno procedure provides definitive relief of obstruction in children with complex congenital left ventricular outflow tract (LVOT) stenosis. The objective was to evaluate its results in children with obstructive hypertrophic cardiomyopathy (OHCM).
Methods:
We retrospectively analyzed the records of the 68 consecutive patients who underwent the modified Konno ventriculoplasty for OHCM in our center between 1991 and 2015.
Results:
The median age at surgery was 8 years (1.2 month – 19 years). Familial history of HCM was present in 27 (40%) children. Idiopathic HCM and RASopathy were the most frequent causes, with 47 (70%) and 19 (28%) patients, respectfully. 36 (53%) patients were NYHA III or IV. 58 (95%) patients were on β-blocker therapy. Mean septal thickness was 22.8±7.5 mm (median z-score: +15.9). Mean maximal LVOT gradient was 94±39 mmHg. An associated procedure was necessary in 25%. Following procedure, in-hospital mortality rate was 5/68 (7%) and was associated with age at procedure less that 1-year, low weight, Noonan syndrome and right ventricular outflow tract obstruction. Occurrence of post-operative complete heart block (CHB) was 13% and was not significantly associated with any baseline characteristic. Median follow-up time was 5.3 years. Freedom from death, heart transplantation, resuscitated sudden cardiac death and implantable cardioverter-defibrillator appropriate shock in hospital survivors was 70% at 20-year follow-up and was associated to the presence of an associated lesion requiring a supplementary concomitant procedure (HR=8.78, p=0.01) and to baseline septal thickness (HR=1.19, p=0.01) in multivariate analysis. 20-year cumulative incidence of reintervention was 24% and was associated to pre-procedure mitral valve dysplasia in multivariate analysis (HR=4.75, p=0.049). All patients had long-standing complete relieve of LVOT obstruction, with a mean residual gradient of 11±6.6 mmHg. At last evaluation, functional status was dramatically improved, with 81% of patients being NYHA I. Yet 22% had signs of diastolic impairment, which was associated to baseline septal thickness (HR=1.12, p=0.04).
Conclusions:
The Modified Konno procedure allowed definitive obstruction relief in pediatric OHCM and dramatic improve of functional status, at the cost of a high rate of CHB. Event-free survival after surgery in this pediatric population with severe hypertrophy was good.