Long-term survival and need of reintervention after treatment of critical valvular aortic stenosis in the neonate; a complete national study in Sweden

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Objective: To examine long-term survival after treatment of critical valvular aortic stenosis during the neonatal period in a complete national cohort with surgical valvotomy being the preferred method of initial treatment.

Methods: All children in Sweden treated due to critical aortic stenosis during the neonatal period from Jan 1st 1994 to Dec 31st 2016 were included. Patient files were analyzed and cross-checked against the Swedish National Population Registry as of Nov 21st 2017 allowing for reliable and complete data on survival. The diagnosis was confirmed by reviewing echo-studies and echo-reports. Critical aortic stenosis in the neonate (younger than 30 days of age) was defined as a valvular stenosis with duct-dependent systemic circulation or depressed left ventricular function (fractional shortening ≤ 27%).

Primary outcome was all cause mortality and secondary outcomes were the need of re-intervention and aortic valve replacement.

Results: Sixty-one patients were identified (50 boys, 11 girls) with gestational age median 39 weeks (range 26-42). All medical files were retrieved with no patient lost to follow-up. Initial treatment was surgical valvotomy (n=52), balloon valvotomy (n=6), closed transventricular valvotomy (n=2) and Ross procedure (n=1). Median age at initial treatment was five days (range 0-26). There were no 30 day mortality but four late deaths at five and 10 months and at two and 21 years of age. Re-intervention was needed in 34 patients at median 3.4 months (range 4 days-17.3 years). Freedom from re-intervention was 64, 52, 48, 44% at 1, 5, 10, 15, 20 years respectively. Aortic valve replacement was needed in 22 patients (36%): Ross procedure (n=11), mechanical prosthesis (n=6), homograft (n=3), biological prosthesis (n=2). In two patients conversion to a single-ventricle palliation was performed and two further patients had a heart transplantation.

Conclusion: Surgical valvotomy is a safe and reliable initial treatment method in these critically ill neonates, but the need of reintervention is high with a freedom from re-intervention of 52% at 10 years of age. In 10 out of 11 patients who reached the age of 18 years during the study the native aortic valve had been replaced.