Experience with Coarctation of the abdominal aorta (Mid-Aortic Syndrome) in children

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Objective: The aim of this study was to describe and analyze the features and outcomes of children and adolescents diagnosed with coarctation of the abdominal aorta.

Methods: Demographics, clinical data echocardiographic measurements, MRI and/or CT-scan imaging, angiography if available, surgical and/or medical treatment data were analyzed retrospectively.

Results: Thirteen patients (6 males) were diagnosed with mid-aortic syndrome (MAS), at the mean age of 7.6 years (9 days to 20 years). Among them, 11 (85%) had a genetic disorder Williams Beuren syndrome in 2, neurofibromatosis in 1, metabolic disease in 1, Takayasu in 1, arterial dysplasia in 1, diabetes in 1, deletion 1p36 and cardiomyopathy in 2, severe statural and neurological impairment in 1 and short-gut syndrome in 1. High blood pressure was observed in all cases, except one neonate and 2 infants with cardiomyopathy and severe LV systolic dysfunction. Hyppoplasia of aorta was supra-renal in 2 cases, infra-renal in 5 and global in 6; renal arteries were involved in 5 cases and mesenteric arteries in 5. Eleven patients received medical antihypertensive therapy (85%), 3 underwent percutaneous (23%) and 5 surgical (38.5%) aortic and renal angioplasty; only one case had neither medical nor interventional therapy. Median follow-up is 5 years (mean 8.8 years).

Outcome was favourable in 8 cases, while 2 had uncontroled high blood pressure and one right heart failure. Two patients died (15.3%) because of renal and cardiac severe impairment.

Conclusion: MAS is frequently associated with genetic disorders and mainly complicates with HBP. Angioplasty is required in more than half of the cases. Renal involvement probably impacts the most on prognosis.