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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 7.6years (9 days to 20years). Among them, 11 (85%) had a genetic disorder Williams Beuren syndrom in 2, neurofibromatosis in 1, metabolic disease in 1, Takayashu 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.