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Long-term results of complex treatment of middle aortic syndrome in children

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Middle aortic syndrome (MAS) characterized by segmental narrowing of the thoracic and/or abdominal aorta, often associated with stenoses of renal and visceral arteries, is an uncommon cause of arterial hypertension. Management strategy depends on the center experience and the anatomical forms of MAS.
The aim: presentation of single center experience of complex interventional and surgical therapy in children with MAS.
Material/Methods
Between 1994 and 2013, 26pts (aged 3-17 yrs, median 11.6) with severe arterial hypertension (AH) resistant to multidrug therapy and diagnosis of MAS underwent interventional and surgical treatment. Twenty five pts had narrowing of thoracic and/or abdominal aorta (length of stenosis 2–12cm, minimum diameter 1.5–6mm), one aortic atresia below the origin of stenosed left renal artery. Renal arteries stenosis coexisted in 12pts, coeliac trunk stenosis in 11pts, superior mesenteric artery in 10pts. Aotic narrowing was treated with stents in 24pts, aortic thoraco-abdominal bypass in one. Additional transcatheter (balloon angioplasty of renal arteries – 7pts, coeliac trunk – 2pts, mesenteric artery – 3pts, stenting of coeliac trunk – 1pt) and surgical (kidney autotransplantation – 12pts) procedures for primary treatment of MAS were performed.
Results
There was significant improvement ($p<0.001$) in pre versus post stent aortic narrowing diameters (4.2±2.8 mm vs 12.4±3.2mm) and systolic gradient (45±5.2 mmHg vs 12±5.1mmHg). Renal function after autotransplantations were normal. During mean 9.2±2.5 years follow-up additional procedures - elective stent redilation of stents implanted to aorta (8pts), additional stent implantation to aorta due to small in-stent aneurysm formation (pts) and progression of narrowing (2pts), balloon angioplasty of aorta due to neoitimal hyperplasia (7pts), balloon renal artery angioplasty (1pt), redilation of stent in truncus coeliacus (1pt) were performed
At the latest clinical follow-up 5pts had no antyhypertensive treatment, all other had better control of AH on the lower doses of medications.

Conclusions:
1.Complex interventional and surgical treatment of children with middle aortic syndrome allows for better control of arterial hypertension.
2.Aortic obstruction related to disease can be treated successfully with the stent implantation.
3.Continuous follow-up of patients with middle laortic syndrome is required for recognition of indications for additional interventions.