Atypical secondary hypertension due to mid-aortic syndrome

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A 17-year-old girl was referred to our Unit for resistant hypertension (180/110), despite full-dose treatment with amlopidine, ramipril, and hydrochlorothiazide. Physical examination showed an abdominal bruit and the absence of the pedilial pulse; thigh and leg muscles were normally developed. Angio-computed tomographic (angioCT) scanning revealed hypoplasia of the suprarenal portion of the aorta (5 mm diameter) that was normal in the infrarenal portion (12 mm diameter). The hypoplasia caused obstruction of the celiac trunk and of the superior mesenteric artery; at that level, the vascularization was warranted by the hypertrophy of the Riolano arcade through the inferior mesenteric artery that had a 7 mm diameter. The vascularization of the iliac–femoral axis was warranted by collateral circles on the thoraco-abdominal wall connecting the internal mammary arteries and the epigastric arteries, both markedly hypertrophic. We made a diagnosis of mid-aortic syndrome, a rare congenital disease of unknown aetiology, characterized by narrowing of the abdominal aorta and stenosis of its major branches. Symptoms depend on the severity of the lesions and the organ perfused by arteries originating from the narrowed aorta. Severe to malignant renovascular hypertension can result in heart and/or renal failure; leg claudication is frequently associated. Treatment is aimed at blood pressure control, renal function preservation, and surgery delay until adulthood. Surgical reconstruction is the preferred strategy. The patient received a by-pass between the thoracic descending aorta and the infrarenal abdominal aorta with reimplantation of the renal arteries and of the superior mesenteric artery and a by-pass between the prosthesis and the splenic artery. Six months after surgery, the patient is normotensive under ramipril 5 mg; angioCT scanning shows normal function of the by-pass and dramatic reduction in collateral circles.

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