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IMAGES IN PEDIATRIC CARDIOLOGY

Midaortic Syndrome Presenting as Neonatal Hypertension

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Abstract We describe a case of mid-aortic syndrome presenting as systemic hypertension in infancy and early childhood. Angiography of the descending and abdominal aorta is the diagnostic test of choice to confirm the diagnosis of mid-aortic syndrome. Severity of hypertension is one of the major factors in determining the timing of intervention. Because of variability in the anatomic extent of mid-aortic syndrome, management options need to be individualized.

Keywords Midaortic syndrome · Neonatal hypertension

The term "midaortic" syndrome (MAS) is often used to describe segmental narrowing of the abdominal aorta and ostial stenoses of its major branches. Approximately 80% of patients have renal artery stenosis, 25% have involvement of the celiac axis, and 70% have multiple intra-abdominal vessel involvement [2]. Herein we present the findings of an abdominal aortic angiogram and anatomical images (presurgery and postsurgery) (Figs. 1–3) from a case of MAS diagnosed in neonatal period.

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Surgical treatment has been successful in children and adolescents who have systemic hypertension due to MAS. However, symptomatic MAS in newborn and infants are almost universally fatal. Surgical repair of MAS later in childhood include aorto-aortic bypass, aortic resection with interposed grafting, patch aortoplasty, reanastomotic resection of renal arteries into prosthetic grafts, and bypass grafting of the stenosed renal and visceral arteries with autologous conduits [1, 3–5]. Patch aortoplasty repair is preferred for infants with long-segment disease who have

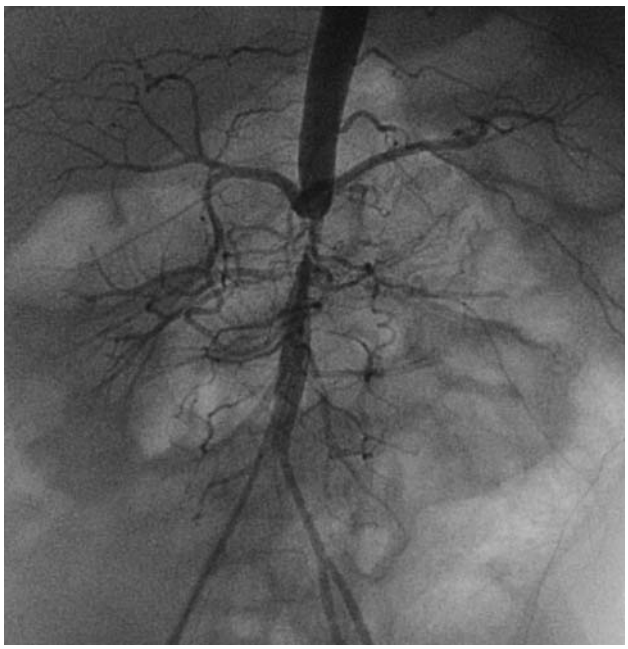

Fig. 1 Abdominal aorta angiography demonstrating severe long-segment coarctation of the abdominal aorta and stenoses of multiple visceral branches including both renal arteries. There is also severe hypoplasia of the common iliac arteries

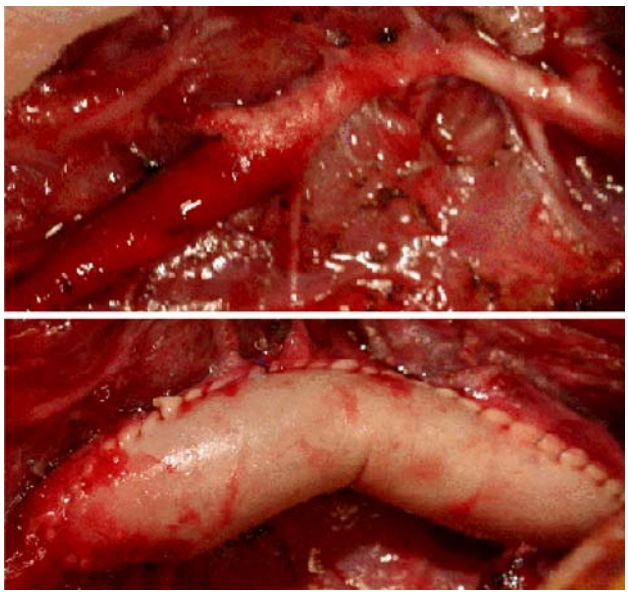

Fig. 2 Long-segment abdominal coarctation before surgery (top) and postsurgical reconstruction of abdominal aorta (bottom)

not completed their growth and development. Bypass grafting and aortoplasty are equally effective for older children and young adults. In selected children with renal artery involvement, favorable results have been reported with aortic and renal artery reconstruction using patch aortoplasty [5].

In our patient, posterior homograft angioplasty from the descending thoracic aorta to the left iliac artery was performed. Successful aortic reconstruction in very young infants is achievable with this technique (Fig. 2). This technique avoids injury to the visceral branches and allows growth of the abdominal aorta (Fig. 3). Unfortunately, renal artery stenosis is not corrected following successful aortic reconstruction. Hence, infants with MAS with renal artery involvement are not likely to have significant reduction in antihypertensive medicine requirements following patch aortoplasty.

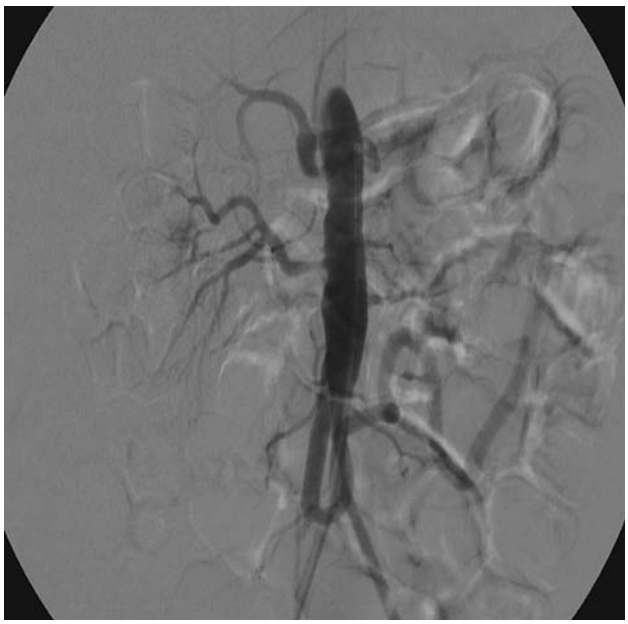

Fig. 3 Abdominal aortogram at follow-up showing tortuous but patent abdominal aorta, occlusion of the superior mesenteric artery, enlarged inferior mesenteric artery, proximal narrowing of the right renal artery, and poor visualization of the left renal artery

References

1. Barral X, de Latour B, Vola M, Lavocat MP, Fichtner C, Favre JP (2006) Surgery of the abdominal aorta and its branches in children: late follow-up. J Vasc Surg 43:1138–1144
2. Bergamini TM, Bernard JD, Mavroudis C, et al. (1995) Coarctation of the abdominal aorta. Ann Vasc Surg 9:352–356
3. Delis Kt, Gloviczki P (2005) Middle aortic syndrome: from presentation to contemporary open surgical and endovascular treatment. Perspect Vasc Surg Endovasc Ther 17:187–203
4. Stanley JC, Criado E, Upchurch GR Jr, et al. (2006) Pediatric renovascular hypertension: 132 primary and 30 secondary operations in 97 children. J Vasc Surg 44:1219–1228
5. Zeltser I, Parness IA, Ko H, Holzman IR, Kamenir SA (2003) Midaortic syndrome in the fetus and premature newborn: a new etiology of nonimmune hydrop fetalis and reversible fetal cardiomyopathy. Pediatrics 111:1437–1442