Open repair of long-segment aortic atresia complicated by uncontrolled hypertension

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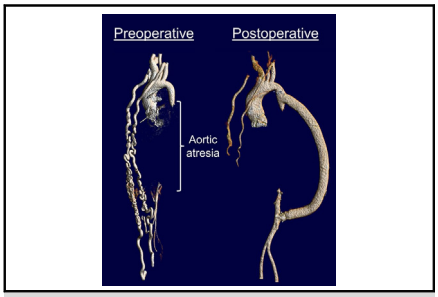


Descending infrarenal bypass of congenital long-segment aortic atresia in a 35-year-old woman.

CENTRAL MESSAGE

Descending infrarenal aortic bypass can safely treat rarely encountered forms of middle aortic syndrome. A multidisciplinary approach optimizes diagnosis, follow-up, and lifestyle counseling.

Video clip is available online.

Middle aortic syndrome (MAS) is a rare vascular anomaly, comprising only 0.5% to 2.0% of cases of aortic stenosis.[1] We present a case of MAS characterized by long-segment aortic atresia with extreme collateralization manifesting as poorly controlled hypertension in an otherwise well-developed adult. We performed descending infrarenal extra-anatomic bypass to treat both aortic malformation and hypertension, exemplifying how established surgical techniques can be modified to safely treat complex and rarely encountered forms of MAS. Permission was obtained from the patient for publication of study data; institutional review board approval was not required.

CLINICAL SUMMARY

A 35-year-old woman presented with hypertension refractory to labetalol 400 mg 3 times daily and nifedipine 90 mg daily, intermittent headaches, chest pain, and dyspnea. Computed tomography showed total occlusion of the thoracic aorta 5 cm distal to the left subclavian artery, compensated by massively dilated internal thoracic arteries (10 mm) with collateralization to both femoral arteries (Figure 1). After evaluation by pediatric and adult congenital cardiology, she was referred to cardiothoracic surgery for definitive repair.

With the patient in left lateral decubitus, a fourth interspace, muscle-sparing thoracotomy was performed. A 16-mm graft was anastomosed end-to-side to the descending thoracic aorta with running 5-0 polypropylene suture. The graft was wrapped in bovine pericardium to prevent pulmonary communication and minimize risk of graft infection. Through a left, posterolateral retroperitoneal incision, a 14-mm graft was anastomosed end-to-side to the infrarenal aorta. The thoracic graft was tunneled through the left posterior diaphragm and pressurized to complete the graft-to-graft anastomosis without kinking (Figure 2, *A*). Her brachial-femoral gradient was reduced from 40 mm Hg before repair to 5 mm Hg after repair (Video 1).

The patient was discharged on postoperative day 10 on labetalol 400 mg. Within 2 months, she was normotensive without taking antihypertensive agents. One year postoperatively, the patient was symptom-free and remained normotensive. Her surgical graft was intact with growth of her native thoracic and abdominal aorta to nearly normal anatomic caliber and complete regression of her internal thoracic arteries (Figure 2, *B*). When she first presented to

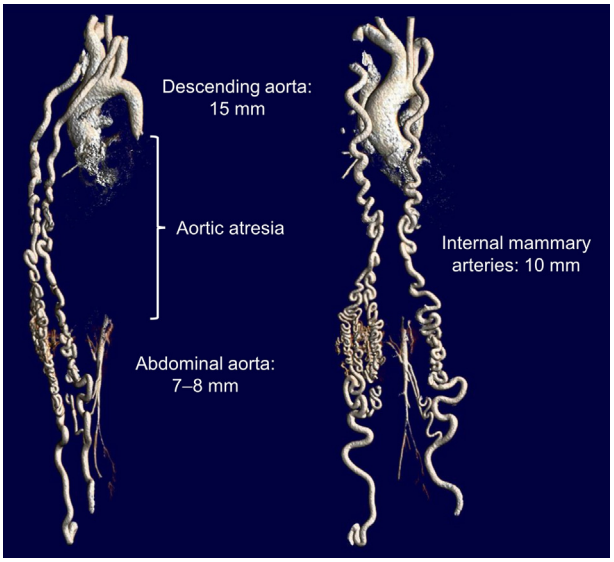


FIGURE 1. Preoperative computed tomography scan of long-segment aortic atresia with a descending aortic diameter of 15 mm and an abdominal aorta that reconstitutes juxtarenally from massively dilated internal thoracic arteries.

our clinic, she was distressed that the new diagnosis of MAS would prevent her from safely having children in the future. Now, 2 years postoperatively, she is pregnant under the care of her local obstetrician.

DISCUSSION

MAS is a heterogeneous malformation with syndromic (eg, Alagille syndrome and Williams syndrome), congenital, and acquired etiologies. Patients are primarily diagnosed in childhood. In a 30-year experience of 53 patients with MAS, only 3 (5.7%) were diagnosed during adulthood.[2] Congenital lesions are most commonly seen in pediatric patients, whereas acquired MAS is far more common in adults. In a series of 143 adult patients with MAS, 76.9% had Takayasu arteritis, 19.6% had atherosclerosis, and only 1 patient had congenital MAS.[3] Intraoperative inspection of our patient's aorta showed robust, healthy tissue without cobblestoning or atherosclerotic plaques, and, upon restoration of flow, her aorta rapidly remodeled to normal diameter within 1 year. These observations suggest a late-presenting, congenital etiology.

The location and extent of the aortic lesion and involvement of visceral branches is an important consideration when developing a surgical plan for patients with MAS. Our patient's complete atresia from the midthoracic to juxtarenal aorta was exceptionally rare. In our own experience of 110 adolescent and adult patients with aortic coarctation, only 2 exhibited long-segment disease.[4] In a systematic review of 630 pediatric patients with MAS, distal thoracic aortic involvement was present in only 3%, with no reported cases of midthoracic lesions.[1] Her extent of disease warranted both high thoracic and abdominal exposure. Although not minimally invasive, our 2-incision approach was more limited than a traditional thoracoabdominal incision, potentially reducing associated pain, dysfunction, and bleeding from large intercostal collaterals. Axillary femoral extra-anatomic bypass was considered but not pursued, due to concerns with discomfort, adequacy of flow, and durability in a young, thin patient. Visceral artery involvement is common in MAS, with 66% of patients exhibiting renal artery stenosis.[1] Our patient's renal

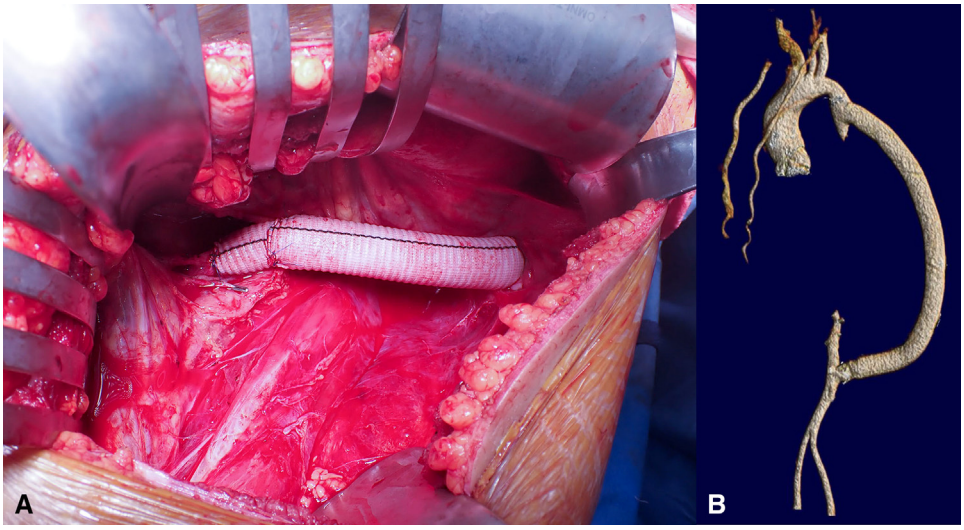
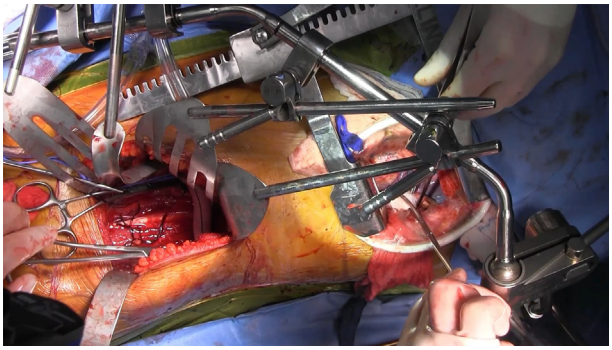


FIGURE 2. A, Intraoperative photograph through the retroperitoneal incision demonstrating diaphragmatic tunneling of the thoracic graft with anastomosis to the abdominal graft. B, Computed tomography of the completed repair 14 months postoperatively with a descending aortic diameter of 17 mm, abdominal aortic diameter of 13 mm, and complete regression of internal thoracic artery collateralization.



VIDEO 1. Descending infrarenal aortic bypass for long-segment aortic atresia. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00300-0/fulltext.

arteries were of normal caliber: a single 5-mm right renal artery, and 2 3.5-mm left renal arteries.

In the presence of additional cardiac lesions warranting concomitant intervention, Yamamoto and colleagues[5] demonstrated the flexibility of aortoaortic bypass in their series of 6 patients with MAS. In 1 patient requiring concomitant coronary artery bypass grafting and aortic root replacement, they performed ascending abdominal aortic bypass via median sternotomy, providing necessary exposure for the additional components of the operation.

Although established guidelines on postsurgical follow-up and long-term management for patients with MAS are limited, we demonstrate that successful treatment of rare aortic malformations can be achieved with multidisciplinary evaluation, careful surgical planning, and thoughtful postoperative lifestyle counseling that includes family planning for young, female patients.

Conflict of Interest Statement

Dr Roselli has consulting relationships with Cook, CryoLife, Gore, Medtronic, and TerumoAortic. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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