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☐ CASE REPORT ☐

# A Minimally Invasive Approach for the Treatment of Mid-Aortic Syndrome in Takayasu Arteritis

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A 61-year-old woman who presented with claudication and dyspnea on exertion was found to have severe calcified narrowing of the descending aorta and severe insufficiency of the aortic valve. These findings were compatible with Takayasu arteritis. To treat these hemodynamic abnormalities, extra-aortic bypass surgery combined with replacement of the aortic valve and ascending aorta-to-hemiarch replacement was performed through a separated upper hemi-sternotomy and limited median laparotomy. We present our successful surgical experience with this case.

Key words: 1. Takayasu arteritis

- 2. Aortic bypass surgery
- 3. Minimally invasive surgery
- 4. Thoracic aorta

## Case report

A 61-year-old woman visited the emergency department because of severe dyspnea and claudication in both legs. The patient had been diagnosed with Takayasu arteritis (TA) 30 years previous. Angiotensinreceptor blockade had been prescribed to control the patient's renovascular hypertension. The patient's pulse pressure was elevated (152/50 mm Hg), and both femoral arteries were barely palpable. On examination, a significant diastolic murmur was heard at the left parasternal second space. In the initial laboratory findings, the patient's glomerular filtration rate was decreased (44 mL/kg/min/1.72 m<sup>2</sup>, Modification of Diet in Renal Disease equation), while inflammatory markers were within the normal range

(C-reactive protein level, 0.41 mg/dL; erythrocyte sedimentation rate, 17 mm/hr). The ankle-brachial index (ABI) was 0.57 and 0.55 on the right and left sides, respectively. Computed tomography (CT) showed dilation of a porcelain ascending aorta (45 mm) with severe calcification and multifocal luminal narrowing of the descending aorta (Fig. 1). Echocardiography revealed severe regurgitation of the aortic valve (AV) and a left ventricular indexed diastolic diameter of 64 mm. Based on these findings, the decision was made to perform surgical correction.

The incisional approaches were similar to those reported previously [1]. In brief, a median upper sternotomy was made for the AV and ascending aortic procedures (Fig. 2A). In addition, a small midline laparotomy (6 cm) was made to perform bypass anasto-

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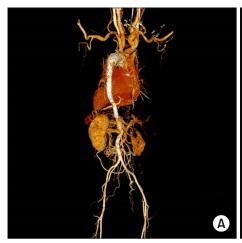




Fig. 1. Preoperative computed tomography. (A) Multifocal luminal narrowing in the descending aorta. (B) The dilated ascending aorta with severe calcification of the aortic wall.



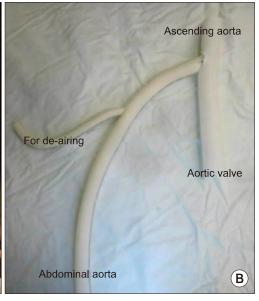


Fig. 2. Intraoperative findings. (A) Double incisions for the surgical approach (upper hemi-sternotomy and mini-laparotomy). (B) A preliminarily shaped hand-sewn graft.

mosis at the abdominal aorta. A newly designed graft was prepared by connecting 2 differently shaped grafts (Hemashield graft; Boston Scientific, Boston, MA, USA) to reduce the cardiac ischemia time, saving time for additional anastomosis (Fig. 2B). Single arterial and venous cannulation was used at the innominate artery and right atrial auricle, respectively. After aortotomy, the 3 AV leaflets were observed to be thickened and retracted. While lowering the body temperature to 25°C, the native AV was resected. Under hypothermic circulatory arrest, hemiarch replacement was performed with the assembled graft. Thereafter, whole-body perfusion was restored. During the re-warming of the patient's body, AV re-

placement was performed using a mechanical prosthesis (ATS open pivot bileaflet mechanical heart valve prosthesis, 24 mm; ATS Medical Inc., Minneapolis, MN, USA) with a continuous suture technique using 3 sets of Prolene 3-0 sutures (Ethicon Inc., Johnson & Johnson, New Brunswick, NJ, USA). After weaning the patient from cardiopulmonary bypass, the previously connected 20-mm graft was passed through the diaphragm from the anterior mediastinum to the intra-abdominal space (Fig. 3A, B). In the peritoneal space, the graft was positioned to pass through the transverse mesocolon. A graft to the abdominal aortic anastomosis was achieved with side biting of the aorta. The cardiopulmonary bypass, cardiac ischemia,

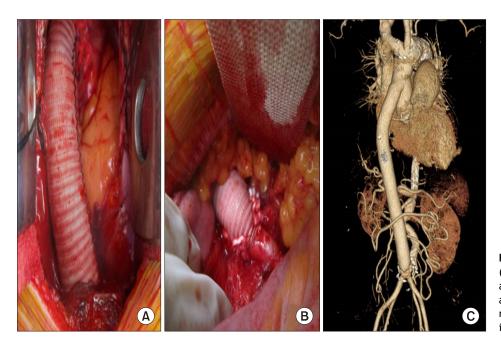


Fig. 3. The results of graft bypass. (A) Replacement of the ascending aorta. (B) Bypass graft anastomosed at the abdominal aorta. (C) Postoperative computed tomography showing well-maintained blood flow.

and circulatory arrest times were 97, 73, and 8 minutes, respectively. The patient's postoperative course was uneventful, without any complications. The patient was transferred to the general ward on postoperative day 1 and discharged on postoperative day 10. Postoperative CT showed well-maintained graft patency and visceral organ perfusion (Fig. 3C). The postoperative ABI was 1.13 on the right side and 1.12 on the left side. The glomerular filtration rate increased to 71 mL/kg/min/1.72 m<sup>2</sup>. During 17 months of follow-up, the patient was free of any symptoms of heart failure, claudication, or end-organ dysfunction, and there were no notable complications.

## Discussion

TA is a form of chronic granulomatous arteritis that mainly affects the aorta and its major branches. In this patient, regurgitation of the AV was found, as well as severe calcified narrowing of the descending aorta and dilatation of the ascending aorta with calcification. The characteristic pathologic findings of TA are hypertrophy of the ascending aorta, fibrosis of the adventitia, thinning of the media, disappearance of elastic fibers, and inflammation of the vascular wall [2,3]. In this case, histological examination of the ascending aortic wall showed non-specific aortitis in the adventitia and media. These findings

suggested that the disease of the ascending aorta was due to TA. Aortic regurgitation can also develop with TA, and is a risk factor for mortality. It is a fairly common complication of TA, as significant aortic regurgitation has been reported to occur in 18% of TA patients in Korea [4]. In cases of aortic regurgitation due to TA, the valve itself has no evidence of inflammation, and it is characterized by valvular thickening, fibrosis, and hyaline or myxoid changes [2,3]. In this case, the AV was thickened and retracted. The results of the pathologic analysis showed fibromyxoid degeneration with calcification. A relationship between aortic regurgitation and degenerative or rheumatic changes cannot be ruled out, but in this case, aortic regurgitation seems to have been due to TA.

In patients with TA, the presence of narrowed points in the thoracoabdominal aorta is related to the presentation of hypertension in the upper extremity and claudication in the lower limb [1]. For these patients, bypass graft interposition using a small conduit has been often used; however, this approach has been revealed to be a significant risk factor for graft occlusion [1,5,6]. In order to improve the patency of the bypass graft, the ascending aorta has been considered to be the best proximal inflow site. In the present case, the patient's native ascending aorta was severely calcified and atherosclerotic,

and there was multifocal luminal narrowing in the distal aortic arch and descending aorta. Therefore, replacement of the ascending aorta and hemiarch was regarded as the ideal option to offer a clear-inflow conduit without the risk of embolic stroke prior to the construction of the extra-anatomic bypass.

Matsuno et al. [7] reported that performing an extra-anatomic aortic bypass between the ascending aorta and the abdominal aorta in patients with middle aortic syndrome had several advantages. Blood flow bypass can be achieved through an extra-aortic graft despite diffuse tubular narrowing of the descending aorta, avoiding the substantial risks posed by extensive replacement of the descending aorta. In addition, it was possible to correct AV regurgitation and ascending aortic dilatation by a single operation in the present case. Reducing the extent of surgical incisions might have positively affected the patient's perioperative pain level and pulmonary function. In conclusion, a double-minimal-incision approach may be a reasonable surgical option to treat patients with TA who present with similar conditions.

### Conflict of interest

No potential conflict of interest relevant to this article was reported.

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