Total Aortic Replacement in a Patient With Mega Aorta Syndrome
— A Case Report —

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A 64-year-old woman without manifestations of Marfan syndrome was referred for the close investigation of a pulsatile abdominal mass. Computed tomographic scans and magnetic resonance images revealed aneurysmal dilatation of the entire aorta, including the ascending aorta and extending to the bifurcation of the aorta, as well as tortuousness of the thoracoabdominal and abdominal aorta. Digital subtraction angiography also showed aneurysmal dilatation of the entire aorta and trivial aortic regurgitation. However, aortic annular dilatation was not found by echocardiography or aortography. The entire aorta was replaced in two stages. First, graft replacement of the ascending aorta, except for the sinus segment, and the aortic arch was performed using an elephant trunk technique under hypothermic cardiopulmonary bypass with selective cerebral perfusion. Twelve weeks later, the remaining aorta, including the descending aorta and extending to the common iliac artery on the right side, and to the common femoral artery on the left side, was replaced with a partial cardiopulmonary bypass using femoral artery and vein cannulation.

We believe that patients with mega aorta syndrome are best treated by total aortic replacement. The results in the present case indicate that the elephant trunk technique is useful for extensive aortic replacement in stages, and greatly facilitates the second stage operative procedures.

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MANY patients with aortic dissection, mega aorta syndrome, and Marfan syndrome show aneurysmal involvement of multiple aortic segments1–3. In these patients, total aortic replacement is considered to be the ideal treatment, and a multi-stage approach to extensive aortic aneurysms is currently required. However, exposure of the distal end of a previously inserted graft for aortic clamping is usually difficult, if not impossible. In 1983, Borst and colleagues4 reported an elephant trunk technique that could be used to simplify and facilitate a subsequent downstream operation.

In this paper, we report a patient with mega aorta syndrome who was treated by total aortic replacement in two stages using the elephant trunk technique.

CASE REPORT

A 64-year-old woman was referred to us for the close investigation of a pulsatile mass of the abdomen. On auscultation of the chest, there was no cardiac murmur. Her blood pressure was 140/70 mmHg, and her pulse rate was 72/min. She had a history of

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hypertension. The pulsatile mass was $6 \times 6$ cm in size and was palpable on the left side of the umbilicus. Computed tomographic scans and magnetic resonance images showed aneurysmal dilatation of the entire aorta to over 6 cm in diameter, including the ascending aorta and extending to the bifurcation, as well as tortuosity of the thoracoabdominal and abdominal aorta, which contained massive mural thrombi. Trivial aortic regurgitation and aneurysmal dilatation of the ascending aorta were also revealed by digital subtraction angiography (Fig. 1). Based on these findings, a diagnosis of mega aorta syndrome was made, and total aortic replacement in two stages was scheduled. In the first-stage operation, graft replacement of the ascending aorta and the aortic arch with a 24-mm Hemashield Dacron graft (Meadox Medicals Inc, USA), except for the sinus segment, was performed through a mid-ternal incision, and hypothermic cardiopulmonary bypass (CPB) with selective cerebral perfusion was used as a support method. Cold crystalloid cardioplegic solution was administered into the aortic root to achieve cardiac standstill after the ascending aorta was cross-clamped just proximal to the brachiocephalic trunk. The aortic cross-clamping was released when the rectal temperature reached 22°C, and the aortic arch was opened. The descending aorta was then completely transected 2 cm distal to the left subclavian artery. For a distal anastomosis, a modified elephant trunk technique was employed during systemic low flow perfusion with an open aortic technique, and approximately 6 cm of the distal free end of the tube graft was left in the distal aneurysm. Following a proximal anastomosis just distal to the aortic valve commissures, arch vessels were reconstructed with separate grafts (10-mm and 8-mm Hemashield Dacron grafts, Meadox Medicals Inc, USA) during a rewarming CPB. Total CPB time was 210 min. Myocardial ischemia lasted 130 min, and selective cerebral perfusion lasted 110 min. The total time required for all of the operative procedures was 535 min. Twelve weeks later, in the second-stage operation, graft replacement of the remaining aorta, including bifurcated graft replacement to the common iliac artery on the right side and to the common femoral artery on the left side, was performed with the aid of a partial CPB using femoral artery and vein cannulation. The aorta was approached through a left antero-lateral thoracotomy in the 4th intercostal space and a spiral incision extending from the 8th intercostal space to the left hypogastric region. Aortic cross-clamping was easily performed about 2 cm distal to the previous distal anastomosis and at the level of the 8th intercostal space, where a narrow segment was present. The aneurysm was then incised and the graft was located within the lumen. There was no thrombus around the graft. Another graft (a 24-mm tube Hemashield Dacron graft, Meadox Medicals Inc, USA) was connected to the end of the previous graft without difficulty, and the origins of the
5th and 6th intercostal arteries were then performed. Finally, the blood flow to the lower extremities was restored with a bifurcated graft (a 20×9-mm bifurcated Hemashield Dacron graft, Meadox Medicals Inc, USA) which was anastomosed to the end of the second graft. All surgical procedures in the second-stage operation, including the anastomosis between the bifurcated graft and the right common iliac artery, were performed via the retroperitoneal space. The total operation time was 895 min with a partial CPB time of 204 min.

Although the patient recovered uneventfully for 9 days, paraparesis, rectal and urinary incontinence suddenly developed after severe abdominal pain and back pain without hypotensive attack on the 10th postoperative day. These symptoms have improved with physical therapy and the patient is presently walking by herself. Normal rectal and bladder function returned 3
months after the second operation.

Postoperative digital subtraction angiography performed 70 days after the second-stage operation revealed successful reconstruction of the total aorta (Fig. 2).

DISCUSSION

In patients with surgically treated aneurysms of the aorta, more than half of the patients with aortic dissection, mega aorta syndrome, or Marfan syndrome show involvement of multiple aortic segments.1–3 It is generally accepted that these patients are best treated by extensive or multiple operations to prevent the aneurysm from rupturing and to prolong their lives.

Crawford and colleagues7 reported favorable mid-term results in 53 patients who had been treated with total aortic replacement. Although aggressive replacement of the entire aorta for patients with diffuse aneurysmal disease or multiple aneurysms of the aorta was recommended, they indicated that one of the surgical problems related to total aortic replacement in stages is the difficulty and dangerousness of the proximal aortic clamping in the second-stage of surgery, and of the anastomosis of the descending thoracic or thoracoabdominal graft to the distal end of a previously inserted ascending and transverse aortic arch graft. The standard elephant trunk technique, first described by Borst,2 has provided a solution to this difficult surgical problem. Svensson4 demonstrated that 56 patients who had been treated using the modified elephant trunk technique underwent the second stage operative procedure with a 30-day survival rate of 96%. However, he also demonstrated that torn aorta at the distal suture line was a fatal complication of the standard elephant trunk technique in 2 patients. The fatal complication resulted from tearing of the aortic wall because of the great tension exerted on the wall by the needle during suturing at a difficult angle. The modified technique involves inverting the proximal part of the graft into the distal part of the graft and then placing it in the descending thoracic aorta during anastomosis, as opposed to pushing the invaginated end of the graft down into the descending thoracic aorta just before completing the distal anastomosis in end-to-end fashion in the standard technique (Fig. 3). Since the inverted graft in the descending thoracic aorta provides excellent exposure of the distal anastomosis and plenty of room to manipulate the needle, there is no need to use excessive torsion on the needle to suture through the graft and the aortic wall within the confines of the crevice between the graft and aortic lumen. In addition, the modified technique enables a greater area of surface contact area between the graft and the aortic wall after the inverted tube graft is withdrawn. Thus, it is possible to reduce the risks of tearing the aorta and of bleeding at the distal anastomosis.

Several complications related to the length of the distal free end of the graft, such as kinking and obstruction of the graft, embolization, and paraplegia, have been reported. Although we used a relatively short distal free end (about 6 cm) of the graft in this patient, a moderate length of about 10 cm for the distal free end of the graft may be preferable, since it has been reported that a long distal free end extending to the diaphragm can lead to paraplegia and paraparesis.2,8 On the other hand, if the distal free end was too short, clamping it and attaching the second graft would be difficult in the later operation. Judging from our experience, a distal free end slightly longer than 6 cm should make it much easier to clamp the graft after opening the distal aneurysm without clamping the previous anastomosis or proximal aorta.

In addition to a spiral incision, a left anterolateral thoracotomy in the 4th intercostal space was used to expose the proximal descending aorta in our second-stage operation. When the spiral incision from the 7th intercostal space extending to the left hypogastric region in the lateral position is used, it may be possible to replace the descending, thoracoabdominal, and abdominal aorta through a single incision, which may help to reduce the duration of the operation and to limit hemorrhage.

Delayed paraparesis, including rectal and urinary incontinence, developed on postoperative day 10 in this patient. Delayed neurological deficits are considered to be due to factors that exaggerate subclinical ischemic states which arise during the operation, or to the underlying aortic disease.
including hypotension, air embolism, and thrombotic occlusion of the spinal arteries due to extension of thrombosis in the intercostal arteries. Therefore, to prevent not only immediate but also delayed neurologic deficits, care should be taken to reattach as many patent intercostal and lumbar arteries to the graft as possible.

In conclusion, aggressive replacement of the entire aorta is recommended for patients with diffuse aneurysmal disease or multiple aneurysms of the aorta. The modified elephant trunk technique is especially useful for extensive aortic replacement in stages, and improvements in the survival rate for the second-stage procedures can be expected with this technique.

REFERENCES


