

Pan-aortic hybrid treatment of mega-aorta syndrome

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Hybrid procedures combining traditional open and newer endovascular techniques are increasingly used to treat complex aortic disease. We present a novel approach for total aortic replacement, including hybrid repair of the arch and thoracoabdominal aorta, in a patient with “mega-aorta syndrome.” A two-stage approach using a valve-sparing aortic root replacement, total arch replacement (stage I elephant trunk), and left carotid-axillary bypass was used to treat the root, proximal-mid arch, and left subclavian aneurysmal pathology. This was followed by a hybrid distal arch/Extent II thoracoabdominal aneurysm repair 3 months later. After 15 months follow-up, the patient remains asymptomatic with an intact repair, no endoleak, and normal ventricular and aortic valve function. This case demonstrates a novel “pan-aortic” hybrid approach for repair of extensive thoracic aortic disease. (*J Vasc Surg* 2011;53:1398-401.)

Hybrid procedures combining traditional open and newer endovascular techniques are increasingly utilized to treat complex aortic disease. We present a novel approach for total aortic replacement, including hybrid repair of the arch and thoracoabdominal aorta, in a patient with “mega-aorta syndrome.”

CASE REPORT

A 69-year-old male with a known abdominal aortic aneurysm developed chest pain radiating to his midupper back. CT angiography revealed a 5.5 cm ascending and transverse arch aneurysm, a 6.1-cm Extent II thoracoabdominal aortic aneurysm (TAAA), and a left subclavian artery aneurysm (Fig 1). Shortly thereafter, the patient began to have pain and bluish discoloration of the first three digits on his left hand, consistent with emboli from the subclavian aneurysm, and anticoagulation was begun with warfarin.

The patient was subsequently referred to our center for the management of his “mega-aorta syndrome.” An echocardiogram was obtained demonstrating normal left ventricular function and a competent trileaflet aortic valve. He had normal renal function with a serum creatinine of 0.8. A two-stage repair was planned, and the patient was taken to the operating room for stage I after a preoperative cardiac catheterization demonstrated no significant coronary artery disease.

At the first operation, the patient underwent David-V valve-sparing aortic root replacement, total arch replacement (stage I elephant trunk procedure), and left common carotid to left axillary artery bypass to treat his ascending, arch, and left subclavian

aneurysmal pathology. This was performed using standard invasive hemodynamic monitoring, including a pulmonary artery catheter and intraoperative transesophageal echocardiography (TEE), as well as electroencephalography to guide the duration of cooling for deep hypothermic circulatory arrest needed for total arch replacement.

The left common carotid to left axillary bypass was performed first with an 8 mm polytetrafluoroethylene (PTFE) graft. A large left subclavian artery aneurysm was found with significant thrombus present. The distal anastomosis of this bypass was to normal caliber axillary artery beyond the aneurysm. After right axillary cannulation and median sternotomy, total arch replacement was performed using a modified Mt Sinai technique as previously described.¹ The innominate and left common carotid arteries were individually anastomosed to the limbs of a multibranch Dacron graft; the left subclavian artery was ligated at its origin from the arch to treat the subclavian aneurysm in conjunction with the carotid-axillary bypass. The arch was replaced using a 28-mm Vascutek Dacron Siena graft with radiologic markers (“Dumbo graft”) to facilitate second stage endovascular repair (Vascutek Terumo, Ann Arbor, Mich). A 15 cm elephant trunk was fashioned with the proximal anastomosis just proximal to the ligated left subclavian artery. Lastly, a David-V valve-sparing aortic root replacement was performed with a 32-mm Vascutek Valsalva graft (Vascutek Terumo), as the aneurysmal process extended to below the level of the sinotubular junction. The patient received six units of packed red blood cells, 16 units of fresh frozen plasma, one unit of cryoprecipitate, and six units of platelets. The operative time was 8 hours and 10 minutes. The patient tolerated the procedure well and was discharged home after a 10-day postoperative stay.

After a 3-month recovery period, the patient was brought to the operating room for the second stage of his repair. Follow-up CT scan prior to this operation demonstrated an intact first stage repair and further expansion of the Extent II TAAA to a diameter of 7 cm (Fig 2). Second stage repair included extra-anatomic four-vessel visceral debranching via midline laparotomy using a custom designed multibranch Dacron graft as previously described;² the proximal right common iliac artery served as the inflow for the abdominal debranching graft. The sequence of visceral anastomoses was left renal artery, superior mesenteric

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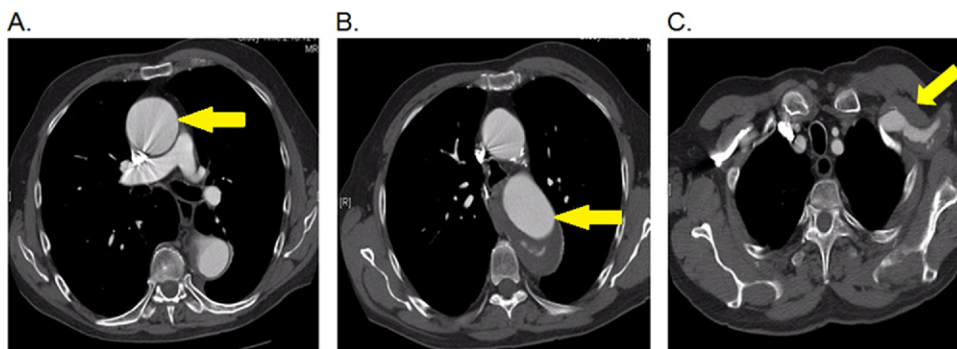


Fig 1. Preoperative computed tomography angiography demonstrating (A) 5.5 cm ascending and transverse aortic arch aneurysm (*arrow*); (B) 6.1-cm Extent II thoracoabdominal aneurysm (*arrow*); and (C) left subclavian artery aneurysm (*arrow*).

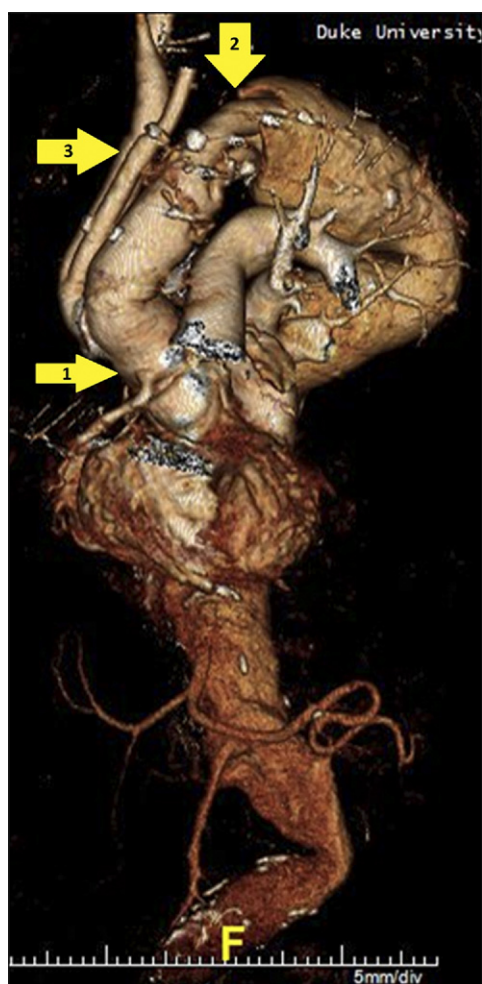


Fig 2. Three-dimensional reconstruction computed tomography angiography after stage I repair demonstrating intact aortic root (*arrow one*) and total arch replacement with elephant trunk (*arrow two*). The innominate and left common carotid arteries have been individually reimplemented into a multi-branch Dacron graft (*arrow three*); the left subclavian artery has been ligated proximally and bypassed distally (*patent bypass not included in image field of view*) to exclude the symptomatic subclavian aneurysm. Note residual distal arch/Extent II thoracoabdominal aortic aneurysm (TAAA).

artery, celiac axis, and right renal artery with individual visceral artery ischemic times of approximately 8 minutes. Lumbar drain placement for cerebrospinal fluid drainage, continuous recording of somatosensory and motor evoked potentials, and intraoperative TEE, in addition to standard invasive hemodynamic monitoring, were used for the second stage repair.

After the debranching portion of the procedure was completed, a total of five Zenith TX2 (Cook Medical Inc, Bloomington, Ind) components (36 mm × 20.2 cm proximal device; 38 mm × 20.2 cm second proximal device; 38 mm × 20.2 cm third proximal device; 38 mm × 15.2 cm fourth proximal device, and 38 mm × 13.6 cm distal device) were deployed extending from the proximal portion of the elephant trunk graft down to the aortic bifurcation. Completion arteriography demonstrated no endoleak, complete exclusion of the arch/thoracoabdominal aneurysm, and excellent patency of the visceral debranching graft. Estimated blood loss during this procedure was 2.5 L, and the operative time was 5 hours and 26 minutes; 151 mL of contrast were utilized. Fig 3 demonstrates the completed two-stage repair.

The patient's postoperative course was complicated by acute tubular necrosis requiring temporary hemodialysis. He left the intensive care unit on postoperative (POD) day 6 and was discharged to home on POD 34 ambulating, tolerating an oral diet and off dialysis. The patient continues to do well with normal functional status, as well as normal renal function, now 15 months following his second stage procedure. His most recent CT scan performed 1 year postoperatively demonstrates no endoleak and significant reverse remodeling of the thoracoabdominal aorta (Fig 4); follow-up echocardiography demonstrates normal left ventricular function with no insufficiency of his spared aortic valve.

DISCUSSION

Endovascular approaches have been increasingly utilized for the treatment of thoracic aortic pathology. The short-term safety and midterm efficacy of these procedures have been generally demonstrated in the treatment of descending thoracic aneurysms, dissections, and traumatic transections.³ Given these favorable results, surgeons have begun to combine open and endovascular techniques via so-called hybrid repairs to treat more complex aortic disease.^{4,5}

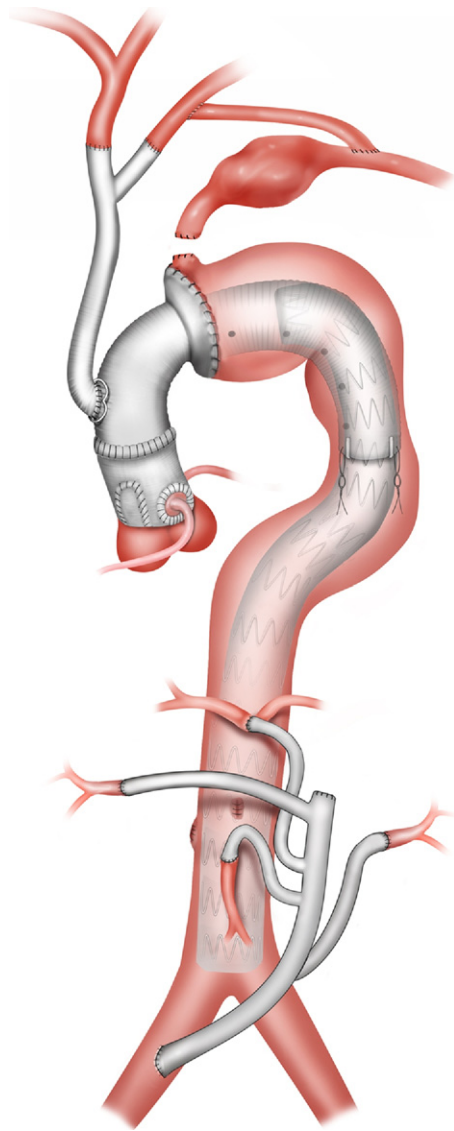


Fig 3. Illustration of completed stage I and stage II repair.

Historically, treatment of the “mega-aorta syndrome” was revolutionized following development of the two-stage elephant trunk technique by Borst in 1983.⁶ This technique markedly improved results for two-stage aortic replacement, although conventional two-stage repair still presented many challenges related to two major operations as well as the interval risk of aortic rupture between stages. Recently, the advancement of endovascular aortic stent grafting has allowed surgeons to further decrease the risk of “mega-aorta” repair whereby stage I elephant trunk procedures are followed by second stage repairs consisting of endovascular stent grafting of the arch and/or descending aorta using the elephant trunk as proximal landing zone, as was done in this case. This approach has the advantages of a lower risk second stage endovascular repair as well as requiring a shorter interval of recovery between stages, thus

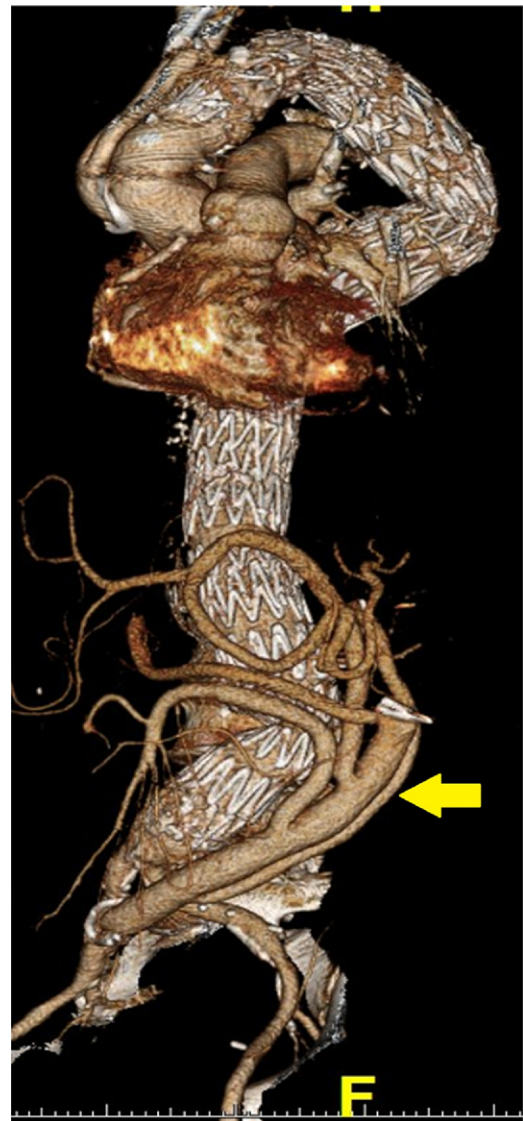


Fig 4. Three-dimensional reconstruction computed tomography angiography 1 year after stage II repair demonstrating intact repair with aneurysm exclusion, no endoleak, and patent grafts (*arrow*).

minimizing interval risk, and has been associated with excellent outcomes.^{7,8}

The current report describes a novel approach to total aortic replacement for “mega-aorta syndrome,” including hybrid repair of the arch and thoracoabdominal aorta. In addition, a valve-sparing aortic root replacement was performed to completely replace the proximal portion of the patient’s aortic disease yet allow him to retain his normally functioning trileaflet aortic valve. To the best of our knowledge, a planned, staged hybrid approach has not been used to repair the entire aorta; specifically, visceral debranching combined with stented elephant trunk has not been described. Further, valve-sparing root replacement has not been reported in conjunction with hybrid aortic replace-

ment. One prior case of hybrid pan-aortic replacement has been reported, but this was not a planned approach as it was performed 18 years after initial repair of a type A dissection and involved five stages to complete.⁹

In the current case, second stage open repair was a potential option, but given the patient's age >65 years, which has been associated with increased morbidity and mortality after conventional TAAA repair,¹⁰ as well as our extensive institutional experience with the use of hybrid abdominal debranching procedures,^{2,4} we elected to proceed with a hybrid second stage repair. We fully recognize, however, that low rates of mortality have not been uniformly reported for hybrid thoracoabdominal repair.¹¹ Renal failure was a notable complication of this second procedure, although the patient left the hospital off of dialysis and now has normal renal function. Renal failure has been a reported complication in these procedures and its etiology likely multifactorial given the contrast required during the procedure and the mandatory warm ischemic time during the period of renal debranching.⁵ In the present case, the visceral debranching and endovascular portions of the second stage were performed concurrently, whereas we now prefer to stage these operations for renal protective reasons. The University of Michigan group has recently reported excellent outcomes using a staged approach to hybrid TAAA repair.¹²

In summary, we herein describe a novel hybrid repair of "mega-aorta syndrome." Hybrid repair of pan-aortic aneurysms is feasible and merits further investigation compared with standard open repair in appropriately selected patients.

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