Open repair in chronic type b dissection with connective tissue disorders

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Introduction

Aneurysms associated with inherited connective tissue disorders (CTD) constitute a specific but important issue in thoracoabdominal aortic aneurysm (TAAA) surgery. In this respect, Marfan syndrome and Ehlers-Danlos syndrome (EDS) type IV represent the most significant disorders causing aneurysmal dilatation of the thoracic aorta. Marfan syndrome typically causes aortic root dilatation, aortic valve insufficiency and dissection, whereas EDS can generate dissection or aneurysm formation in almost every major artery, including the thoracoabdominal aorta. The majority of patients with Marfan syndrome first suffer from ascending thoracic aortic aneurysm, which requires surgical repair. Marfan patients are significantly younger than patients with degenerative aneurysms (1). Therefore, the physical condition of younger Marfan patients is much better than the non-Marfan group (2). Despite this, there is a higher incidence of dissection and rupture during TAAA surgery in Marfan patients and the recommended threshold for repair is thus an aneurysm diameter of 5.0-5.5 cm (3).

The second most common cause of TAAA formation is chronic type B dissection. In patients having surgery for TAAA, up to 24% suffer from chronic dissections (4). Conversely, 20-40% of all patients with aortic dissections will require subsequent aortic replacement for aneurysmal degeneration, irrespective of initial medical or surgical treatment at the time of acute presentation (5). The presence of blood flow in the false lumen is the most significant risk factor for increased aortic diameter, with a mean growth rate of 3.3 mm/year (as compared to 1.4 mm/year diameter shrinkage in patients with a thrombosed false lumen). Position along the aorta also influences growth rate with thoracic aortic aneurysms expanding significantly faster than AAAs (4.1 and 1.2 mm/year, respectively) (6). The growth rate of post-dissection aneurysms can be comparable to that of degenerative thoracic aneurysms or faster. Taking these considerations into account, the combination of chronic dissection, aneurysm formation and connective tissue disease poses major challenges regarding indication for treatment, choice of treatment, and clinical outcome.

Post dissection versus degenerative TAAA

Post dissection thoracoabdominal aneurysms can develop following a DeBakey type I (extensive Stanford type A) or DeBakey type III (Stanford type B) aortic dissection. Such late aneurysmal degeneration of the outer wall of the false lumen occurs in 30-45% of patients. One of the main differences between degenerative and post dissection aneurysms is patency of intercostal and lumbar arteries: in the latter group, almost all segmental arteries are patent, whereas the majority of segmental vessels are occluded in atherosclerotic aneurysms (7). In post dissection aneurysms, the important segmental vessels commonly originate from the true lumen. This is advantageous for technical reasons because reimplantation of segmental arteries is safer in the stronger true lumen aortic tissue than in the thinner walled false lumen tissue.

Patients with connective tissue disease presenting with post dissection TAAA have a past history of type A or type B dissection. In post type A dissection with longitudinal extension, arch involvement is always present, and repair of this aortic segment is required first. Post type B TAAA in connective tissue patients mostly originates close to or

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at the level of the left subclavian artery, necessitating more proximal cross clamping.

With respect to surgical techniques, post dissection aneurysms are considered not different from degenerative aneurysms, except for certain technical aspects described below. A threshold of 5 cm maximal diameter is used as indication for surgery in connective tissue diseased TAAA. In addition, intercostal, visceral and renal arteries are treated fundamentally differently: selective bypasses are used instead of button reimplantation in order to prevent button aneurysms in the long-term.

Surgical technique

Positioning and incision

Following introduction of the intrathecal catheter for cerebro spinal fluid (CSF) drainage, the patient is positioned on a vacuum mattress in a right lateral decubitus position with the shoulders turned to the right (70-80°) edge of the table, the abdomen approximately 60° and the left hip at 30° to allow access to the left groin (*Video 1*). Double lumen endotracheal intubation allows deflation and collapse of the left lung, which is carefully retracted in order to avoid lung damage and bleeding.

The level of the proximal clamp position is prepared depending on the extent of the aneurysm. Usually, the clamp is positioned just distal to the left subclavian artery; however if necessary, the clamp can be placed between the left carotid and left subclavian arteries. Adequate preparation of this proximal clamp position is of crucial importance and includes the following steps: identification of the vagus and recurrent laryngeal nerve to avoid injury, and transection of the ligamentum arteriosum to ameliorate access to the inner curve of the distal arch and allow digital encircling of the aorta. If more proximal clamping is required, access can be made easier by opening the pericardium dorsal to the phrenic nerve and approaching the aortic arch over the pulmonary artery. This maneuver offers access to the inner curve of the arch opposite the left carotid artery. Gentle finger dissection at the dorsal and cranial level of the aortic arch will free the tissue step by step, ultimately offering a safe clamp position. Following this, the aorta is dissected from the esophagus and completely freed to avoid later prosthetic-esophageal fistula formation. Proximal and medial aortic side branches, such as the left bronchial arteries, can be clipped and divided.

Preparation of more distal sequential clamp positions, either at the level of the diaphragm or higher, only requires a small opening of the parietal pleura covering the aorta, allowing sharp or digital encircling of the aorta. Opening of the parietal pleura should be performed as close to the aortic wall as possible and under direct vision of the accessory hemiazygos vein, which runs parallel and lateral to the aorta. The ideal plane for the cross clamp is between the accessory hemiazygos vein and the aortic wall, avoiding significant venous bleeding when transecting the aorta. Damage of intercostal arteries should also be avoided and therefore dissection repair is best performed at the level of a costo-vertebral junction where no intercostal arteries are located. If possible, several clamp positions are prepared, allowing step by step opening of the aneurysm. This strategy reduces blood loss and contributes to decreased spinal cord ischemia.

The costal margin is divided and access to the aorta is achieved by either a retroperitoneal or transperitoneal approach, depending on the surgeon's preference. In any case, the abdominal aorta is approached laterally from the descending colon and dorsal to the spleen and left kidney, which is rotated anteriorly. The diaphragm can be completely divided but we prefer to cut the anterior, muscular section only, which prevents injury to the branches of the phrenic nerve. This limited opening of the diaphragm also offers better postoperative pulmonary function compared to complete division.

The surgical strategy in post dissection aneurysms is similar to the above-described technique; however, some different approaches can be considered. First, sequential clamping, if feasible, enables stepwise aortic reconstruction and limits the potentially substantial blood loss caused by profound back bleeding from intercostal arteries owing to the distal perfusion; blocking with balloon catheters or oversewing these arteries require significant amounts of time and therefore sequential steps are preferred. Second, reversing the direction of aortic reconstruction from distal to proximal avoids insufficient perfusion of visceral, renal and segmental arteries due to changed flow patterns induced by retrograde perfusion. Type B dissections cause a wide range of morphologic changes with different false and true lumens, entries and re-entries with subsequent visceral and renal circulation via separate pathways. Cross clamping the proximal descending aorta and starting retrograde aortic perfusion can change these alternative perfusion channels to the visceral organs, potentially causing malperfusion and subsequent ischemia, especially in those cases with extremely narrow true lumens.

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Video 1 also shows a post dissection TAAA in a young Marfan patient. There is an exceptionally small true lumen, causing malperfusion and severe liver function disturbances. In these cases we start the reconstruction at the distal part, either with a bifurcated graft to the iliac arteries or a tube graft to the aortic bifurcation. The graft is subsequently cannulated for arterial inflow and extra corporeal circulation commenced. Following this, the renal and visceral vessels are reconstructed and thoracic and distal arch repair are done as last steps. Treatment and handling of the dissected layers require utmost care because severe complications can develop if the true lumen is not correctly identified and controlled. For example, the true lumen is often so much smaller that with compression, it may even be disregarded altogether. If the aorta is not entirely encircled and cross clamped, significant bleeding through the incompletely clamped true lumen occurs as soon as the aorta is opened. Another potential danger of overlooking the small true lumen is leaving intercostal arteries without reattachment.

Organ protection

Our surgical protocols for post dissection and degenerative TAAA surgical repair are similar and consist of extra corporeal circulation, selective perfusion of the visceral and renal arteries, CSF drainage and monitoring motor evoked potentials (MEPs) (8). In types I, II and III TAAA, extracorporeal circulation is installed by means of the femoral artery and femoral vein or left femoral artery and pulmonary vein (or left atrium) cannulation. Our tubing system consists of four integrated catheters which are connected with 12 to 15 French balloon inflatable catheters for selective perfusion of the celiac axis, superior mesenteric artery and both renal arteries (9). Alternatively, the kidneys can be protected by means of continuous perfusion with cold Ringer's lactate (10).

Impact of CTD

The largest experience of TAAA repair in Marfan patients comprises 137 with confirmed and 163 with suspected Marfan syndrome (11). The 30-day mortality rate was 4.3% and freedom from repair failure was significantly better in patients with confirmed (90% at 10 years) Marfan syndrome than in suspected (82% at 10 years, P=0.001). Out of these 300 patients, 31 had descending thoracic aortic aneurysms (DTAA) and 178 suffered from TAAA. Surgical mortality was less than 6% and major complications included renal failure in 6% and neurologic deficit in 4%. The authors concluded that operative treatment of aortic pathology in Marfan patients provides excellent results and long-term survival. Dardik *et al*. (12) demonstrated that patients with CTD have similar peri-operative and long-term survival rates after TAAA repair when compared to patients without CTD. Patients with CTD suffered from more extensive aneurysms (type I and II) and had an increased risk of paraparesis. In CTD patients, long-term CT scan surveillance showed no relative increase in aortic size and persistent freedom from recurrent aortic events. The postoperative cumulative 5-year survival rate of TAAA patients with and without CTD was similar (53%). In our experience (2,13), mortality did not occur and major complications like paraplegia, renal failure, stroke and myocardial infarction were not encountered. At 38 months follow-up, all patients were alive and returned to work. Kalkat *et al*. (14) obtained a similar outcome, justifying the conclusion that surgical repair of DTAA and TAAA provides excellent results in patients with Marfan syndrome.

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