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SPECIAL EDITION

Controversies in Surgery for Thoracic Aorta

Thoracoabdominal aortic replacement in patients aged 50 and younger

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Abstract

Objective Open repair of a thoracoabdominal aortic aneurysm (TAAA) has been regarded as one of the most invasive procedures in cardiovascular surgery. Conversely, endovascular technology currently enables the repair of the thoracoabdominal aorta, and this approach is less invasive. However, the long-term durability of this method of endovascular repair remains unknown. This investigation retrospectively analyzed the long-term durability of thoracoabdominal aorta repair in patients aged 50 and younger.

Patients and methods Since 1995, 100 patients aged 50 and younger underwent thoracoabdominal aortic replacement at our institution. There were 63 males, and the average age was 38 ± 7. Ninety-six had aortic dissection as an aortic pathology. Marfan syndrome was found in 61 patients, Loeys–Dietz in 10, Acta 2 mutations in 4, aortitis in 2, and Ehlers–Danlos syndrome in 1. There were 2 patients with a type I TAAA, 56 with a type II, 33 with a type III, and 9 with a type IV.

Results There were 3 hospital deaths (3%), of which 2 were emergent cases. Spinal cord injury occurred in 1 patient (1%). Two patients (2%) had wound complications. Three patients suffered cerebral hemorrhage and 1 had an intramedullary infection, of which all were associated with cerebrospinal fluid drainage. The 3-year survival rate after the operation was 94%, that of 5 years was 94%, and that of 10 years was 91%.

Conclusions Results of thoracoabdominal aortic replacement in patients aged 50 and younger were favorable. While spinal cord complications were rare, cerebrospinal drainage revealed several complications in this series. Evolving endovascular repair needs to be compared with these results, especially in patients aged 50 and younger.

Keywords Connective tissue disease · Thoracoabdominal aortic replacement

Introduction

Open repair of a thoracoabdominal aortic aneurysm (TAAA) has been regarded as one of the most invasive procedures in the area of cardiovascular surgery. In spite of recently improved outcomes from experienced centers [1, 2], it is still considered as an extremely challenging procedure due to the invasiveness of the surgery. Thus, younger patients may survive the grueling operation, with minimal risk of complications.

On another front, thoracic endovascular aortic repair (TEVAR) is undergoing a variety of novel technological advances. TEVAR is currently one of the established treatment strategies to combat aortic diseases. This endovascular technology currently enables the repair of the thoracoabdominal aorta. However, the long-term durability of the TEVAR remains unknown. This technology could most probably be safely applied to the patients, at least for short-term periods.

This investigation analyzed the open surgical repair of thoracoabdominal aorta in patients aged 50 and younger, serving as a benchmark for repair of TAAA using TEVAR.

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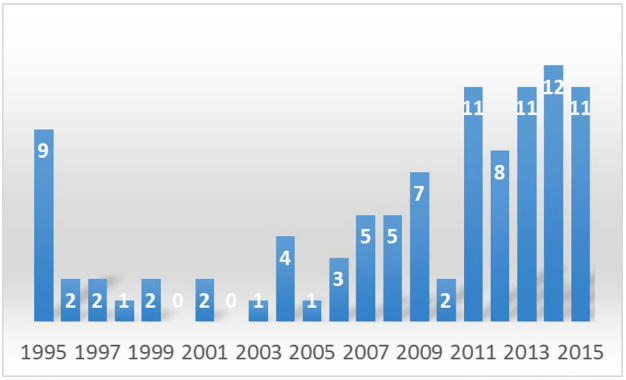

Fig. 1 Annual number of the cases

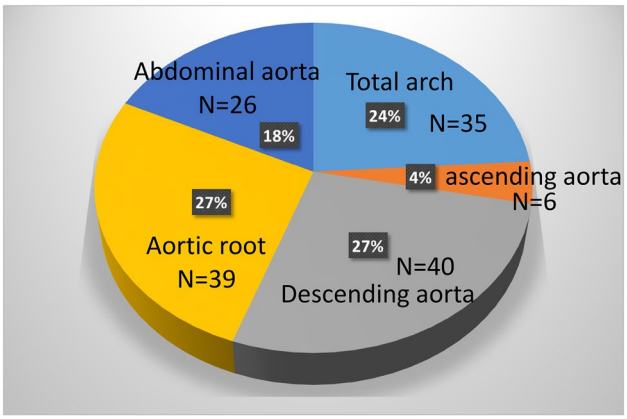

Fig. 2 Previous aortic operations

Patients and methods

Since 1995, 100 patients aged 50 and younger underwent thoracoabdominal aortic replacement in our institution (Fig. 1). There were 63 males, with an average age of 38 ± 7 (range 9–50 years of age). Ninety-six patients had aortic dissection as an aortic pathology, and 6 cases were emergency patients. Marfan syndrome was found in 61 patients, Loeys–Dietz in 10, Acta 2 mutations in 4, aortitis in 2, and Ehlers–Danlos syndrome in 1. Seventy-six patients had 146 aortic operations in total previously (Fig. 2).

Operative technique

Surgical techniques have been described previously. The strategy of optimal body temperature setting during cardiopulmonary bypass changed during the duration of this study. Until 2009, most patients with type III and IV lesions, and some patients with type I and II lesions, underwent surgery under mild hypothermia (30–33 °C) with a beating heart in this study population. However, since 2010, a deep hypothermia (18–20 °C) setting has been utilized, especially in patients with type I and II lesions. As a result, deep hypothermia was utilized in 62 patients and mild hypothermia in 38 patients in this series. Since 2012, a straight incision with the rib cross (SIRC) thoracotomy had been applied in major cases [3, 4]. Identification of the artery of Adamkiewicz (AKA) was made preoperatively using computed tomography (CT) or magnetic resonance imaging (MRI). We reconstructed or preserved a target intercostal artery (ICA) during the operation as a donor artery to the AKA. Transcranial motor-evoked potentials (tcMEPs) were monitored intraoperatively to identify spinal cord ischemia. A spinal fluid drainage tube was inserted preoperatively and drained continuously to maintain spinal fluid pressure at < 10–15 mmHg if tcMEPs significantly changed after the patient was rewarmed.

Under general anesthesia with separate endotracheal intubation, the patient was positioned in the right lateral decubitus position with the shoulders at 60°–80° and the hips flexed to 30°–40° from the horizontal plane. The left-arm was then extended over the head. An incision was made along the division of the costal arch. The chest cavity was opened through the 6th intercostal space and exposed along the skin incision combined with transection of the 4th, 5th, and 6th ribs when the exposition was not sufficient for the operation planned [3, 4]. The diaphragm was incised circumferentially keeping about 1.0–2.0 cm muscle edge at its costal insertion. The abdominal aorta was then exposed to the retroperitoneal cavity. Using a wishbone retractor, the whole aorta from ascending aorta to the bilateral common iliac arteries was exposed.

Cardiopulmonary bypass (CPB) was established using cannulation of the right common femoral artery and right common femoral vein in major cases. When a graft replacement is planned down to bi-iliac arteries, a bifurcated polyester graft was placed initially at the infra-renal abdominal aorta. Then, the aortic return of the CPB was placed at the bifurcated graft. Cannulation of the ascending aorta is on a gradual upward trend for patients with type I and II lesions. A left heart venting tube was inserted through the left atrium appendage, and in some cases, a pulmonary artery vent was placed. We did not apply the clamp in the aortic arch as a standardized procedure. When both the tympanic and bladder temperatures reached 20 °C, for those patients who required an open proximal aortic anastomosis, a clamp was applied to the mid-descending aorta to maintain femoral artery perfusion after injecting 40 mEq of potassium to obtain cardiac arrest during upper body circulatory arrest. A Dacron four-branched graft was used for thoracoabdominal

replacement. For patients not requiring open proximal anastomosis, the graft anastomosis was performed with proximal and distal clamping.

The entire diseased aorta was then opened, and the patient's ICAs were over-sewn except for the critical ICA and 2 adjacent ICAs. ICA reattachment was performed with a short graft interposition. Visceral arteries including the celiac, superior mesenteric, and bilateral renal arteries were cannulated and perfused at 800–1000 ml/min. Subsequently, a distal aortic anastomosis was performed, and the patient was rewarmed. During rewarming, visceral arteries were reattached to the side arms of the graft. After weaning the patient from CPB and minute hemostasis, cryoablation of the intercostal nerve was used for pain control.

Results

The total operation time was 621 ± 189 min and cardiopulmonary bypass time was 263 ± 85 min. Extension of the replacement is shown in Fig. 3. There were 3 hospital deaths (3%), two of which were emergency cases. A 37-year-old female underwent an emergency operation for an impending rupture of a TAAA and had an acute Stanford type-A aortic dissection on postoperative day 5. She then underwent another emergent operation but could not survive. Another 30-year-old female also underwent an emergent operation for impending rupture of a TAAA and suffered an infection at the wound. This resulted in multi-organ failure (MOF) followed by mortality. A 30-year-old male had an elective operation for a Type II TAAA but then suffered postoperative ischemia of the intestine, which led to MOF.

One patient who had a spinal cord injury was paraplegic (1%). He had an acute Stanford type-A aortic dissection and underwent a total arch replacement with extra-anatomical bypass between the ascending aorta and bilateral external iliac arteries to treat malperfusion of the legs. He required hemodialysis after the surgery due to glomerular nephritis. His dissected thoracoabdominal aorta expanded over 6 years and, therefore, he eventually underwent replacement of the type II TAAA under deep hypothermic circulatory arrest. Initially, the extra-anatomical bypass was planned for the aortic return, but the bypass had a pseudoaneurysm due to physical degradation of the graft. The repair of the pseudoaneurysm required temporal interruption of the bypass. We considered this interruption causative of the postoperative paraplegia. His cerebrospinal fluid drainage (CSFD) was not working due to an occlusion of the tube caused by clots.

The three intracerebral hemorrhages (3%) and one intramedullary infection (1%) were likely related to the CSFD. These four patients recovered without any adverse after-effects.

Besides these four patients, one patient had a major stroke, and two had convulsions postoperatively. However, following recovery, all three patients were discharged without symptoms. Two had temporal respiratory dysfunction (2%). The 3-year survival rate after the operation was 94%, that of 5-years was 94%, and that of 10-years was 91% (Fig. 4).

Discussion

Thoracoabdominal aortic lesions in the younger generation are usually not caused by an atherosclerotic aneurysm, but rather by dissecting an aneurysm. In this study, 96 patients

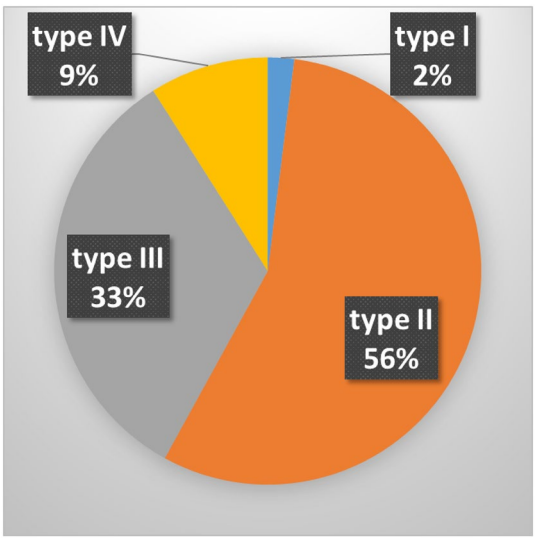

Fig. 3 Extension of the replacement (Crawford classification)

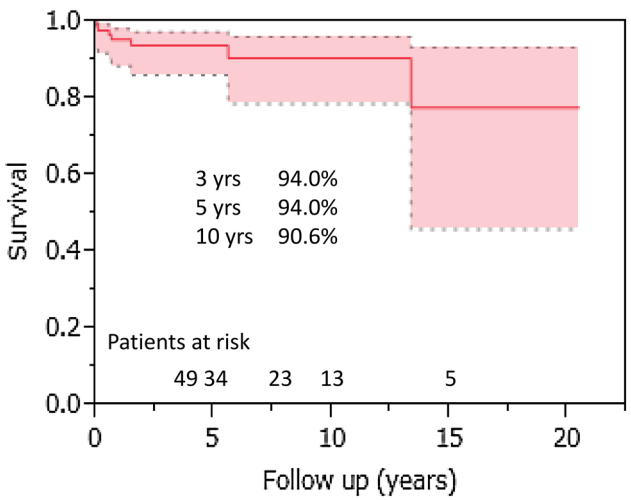

Fig. 4 Survival curve after the operation

(96%) underwent aortic dissection as a pathology of the TAAA, and 78 patients (78%) had connective tissue disorders (CTD). Luozzo et al. reported open repair of descending aorta and TAAA in patients younger than 60 [5]. The patients with CTD account for only 14% of the population in their study. Another report of TAAA open repair in patients aged younger than 60 showed that 18.9% of the enrolled patients had a CTD [6]. The much higher rate of the CTD presented in this investigation may be associated with the relatively younger population studied herein. The CTD described herein has been known to be associated with connective tissue disorders. The CTD are genetic-linked diseases, and the genes have been found to be identified. Over 30 genetic and epigenetic relationships with thoracic aortic aneurysms and dissections have been identified [7], and this number is likely to increase. It seems likely that many patients may have unknown CTD. The reports regarding TAAA repair in patients with CTD have indicated that the patients were abreast around 40 years old [8, 9]. Coselli and his colleagues reported the surgical experiences of TAAA in 127 patients with MFS only with an average age of 43.4 ± 12.5 years, and the TAAA showed all aortic dissection as a pathology [10]. Importantly, their surgical results were excellent as only 4% suffered mortality, there were no strokes, and only 1.6% of patients suffered spinal cord complications. Omura and his colleagues reported their experiences of TAAA repair in 20 patients with MFS [11]. These patients were also young (mean age of 45 years), and they were all dissecting aortic pathology. Their surgical results were also outstanding as there was no in-hospital mortality and no neurological complications including stroke and permanent spinal cord ischemia. Thus, TAAA repair in patients with CTD could be performed with minimal risk, particularly in young patients. In our study, the mean age was 36 ± 8 years, which is much younger than other reports, and the mortality was 3% in total and 1% in elective cases. We speculate that our patients in this series survived the operations due to the excellent functional capacity of the young population.

Because of the good risk of the young patients, we recently applied the aggressive extended replacement of the aortic lesion strategy. Thirty-two patients who had undergone a Stanford type-B chronic dissection since 2012 had an extended graft replacement regardless of the aortic size, excluding the dissected area. No mortality and no major complications occurred in this recent series. This strategy is already reported by Omura et al. [9, 11] and the positive results are likely attributed to the younger population incorporated in the study. The residual dissected aorta in young patients has a high possibility to expand to a critical size during their lives, wherein the patients would be obliged to have a follow-up operation. We believe that the extended repair in this young generation could be performed safely. However, we cannot disregard the risk involved in the extended repair. Keschenau and his colleagues also reported their experiences of open thoracic and thoracoabdominal aortic repair in 72 patients with CTD [9]. Although the range of the age was 19–70, the median age was also as low as 41 years. The hospital mortality was 14%, and 31% of the patients required revision surgery for bleeding or hematoma formation. Amazingly, 15% of these patients also developed sepsis. As a result, these authors reported that they preferred a staged procedure which would reduce the operating time. Etz and his colleagues also recommended a staged repair, as a means to reduce the incidence of paraplegia [12]. However, the patients included in their study were much older, and thus the possibility of spinal cord injury might have not been equivalent.

Hicks and his colleagues described the open surgical results of the thoracoabdominal aortic aneurysm and compared these cases with those with CTD and degenerative diseases [8]. The age is much younger in the CTD group (39 ± 1.9 years) compared with the degenerative group (68 ± 1 years). However, the mortality rate was 10% in the CTD group and 6% in the degenerative group, which was not significantly different ($P = 0.40$). From our experiences, TAAA repair in the younger patients who were mainly with CTD is easier than those in the older patients, as they usually have atherosclerosis. There were no technical difficulties associated with a case of Ehlers–Danlos syndrome in this series. However, TAAA in degenerative disease sometimes shows complications associated with the spinal cord. Previously we reported that embolism might be a cause of the spinal complications [13].

Thoracic endovascular aortic repair (TEVAR) has become the treatment of choice for thoracic aortic disease. However, its application in the younger population is controversial due to the uncertainty of its long-term durability. Thoracic endovascular aortic repair (TEVAR) in patients with MFS carries a substantial risk of early and late complications, consisting mainly of endoleaks and surgical conversions and death by the midterm follow-up. Therefore, current guidelines do not recommend the use of TEVAR in MFS [14, 15]. These complications are relatively frequent in patients undergoing TEVAR for chronic dissection [16]. The repair of TAAA using TEVAR requires the preservation of blood flow to the major abdominal branches or reconstruction of the major abdominal branches in major cases. Thoracic endovascular aortic repair (TEVAR) is considered an advantage for the treatment of descending aortic lesions because the descending aorta has no major branches. For the Crawford type I TAAA repair, in order to obtain the landing zone of the distal end of TEVAR, TEVAR is sometimes placed on the celiac artery following the closure of the celiac artery. This scenario is technically simple as a repair of TEVAR for descending aortic lesion. However, we believe the usage

of TEVAR should be limited in this young population of patients with TAAA.

Postoperative spinal cord injury (SCI) is one of the most devastating complications and a decline in spinal cord blood flow is often related to the complications. Younger patients and patients undergoing aortic dissection tend to have more patent intercostal arteries, and management of the bleeding from the patent intercostal arteries is considered to have a significant effect on the rate of the SCI. Our data demonstrate an SCI rate of only 1% among the survivors. The benefit of CSFD has been established, and it had been employed as a standard strategy in this series. However, three patients experienced cerebral hemorrhaging and one suffered an intramedullary infection. These complications were related to the CSFD. Wynn and his colleagues reported that neurologic deficit from spinal fluid drainage occurs in only 0.8% of patients [17]. However, it is also associated with high morbidity and mortality. Therefore, we suspended placing the tube preoperatively in cases that occurred within the last 2 years. Once the MEP has resolved, the spinal fluid drainage is initiated as an alternative strategy.

Conclusions

Surgical results of thoracoabdominal aortic replacement in patients aged 50 years and younger were favorable. While spinal cord complications were rare, cerebrospinal drainage itself showed several complications. Evolving endovascular repair should be compared with these results, especially in patients aged 50 and younger.

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