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Original Article

Surgical Outcome of Abdominal Aortic Aneurysm Replacement in Patients with Connective Tissue Disorders under 30 Years of Age

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Objectives: Abdominal aortic aneurysm (AAA) in patients <30 years old is relatively rare. We retrospectively analyzed patients <30 years who received an AAA replacement.
Materials: Among 3,003 patients who received an AAA replacement during the last 40 years, 10 patients <30 years old were retrospectively reviewed. All patients suffered from a connective tissue disease: eight from Marfan syndrome and two from Loeys–Dietz syndrome. Five patients had a history of cardiovascular surgery. Aortic pathologies were a dissection type in eight patients and a non-dissection type in two. All patients received a graft replacement of infrarenal AAA, with a bifurcated graft in six patients and a straight graft in four.
Results: Except for cases that were urgent and emergent, rapid aneurysm expansion was noted in all cases. Mean AAA diameter at surgery was 46.7±9.2 mm. No hospital mortality was recorded. Eight patients required 10 additional cardiovascular surgeries: two root replacements, two total arch replacements, two descending aortic replacements, and four thoracoabdominal replacements.
Conclusion: AAA replacement in patients <30 years is safe. In younger patients with a connective tissue disease, AAA should be included in the routine medical check-up, and earlier surgical indication should be considered for its rapid expansion.

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Introduction

Abdominal aortic aneurysm (AAA) is uncommon in people at an age of ≤60 years. Besides age, smoking habit, the male gender, and arteriosclerosis were found to be risk factors of developing AAA.[1] The incidence of AAA was reported to be 2%–13% in men and 6% in women at an age of ≥65 years.[2] As compared with elderly patients, the etiology and clinical course of AAA in younger patients has not been well recognized. To discuss the optimal surgical timing and surgical outcomes, we retrospectively analyzed 10 patients under the age of 30 years old who received an AAA replacement.

Materials and Methods

Patients

From August 1977 to December 2016, a total of 3,003 patients (2,390 men and 613 women) received an AAA replacement at the National Cerebral and Cardiovascular Center. The age-wise grouping of the 3,003 patients at the time of the AAA operation is as follows: <30 years, 10 patients (0.33%); 31–40 years, 28 patients (0.93%); 41–50 years, 48 patients (1.6%); 51–60 years, 213 patients (7.1%); 61–70 years, 1,037 patients (34.5%); 71–80 years, 1,237 patients (41.1%); 81–90 years, 305 patients (10.2%); and >90 years, 125 patients (4.2%).

The characteristics of the 10 patients under the age of 30 years are listed in **Table 1**. All patients suffered from a connective tissue disease: eight from Marfan syndrome and two from Loeys–Dietz syndrome. Marfan syndrome was diagnosed according to the Ghent criteria, revised Ghent criteria, or through genetic analysis. Loeys–Dietz syndrome was confirmed by genetic analysis.

Two children aged two and 12 years (patients #1 and #2, respectively) had aneurysms without a dissection of

the diameter that were 15 mm and 32 mm, respectively. Another eight patients of age >20 years had the aortic pathology of dissection. In patient #3, localized dissection on aneurysm was complicated while waiting for the aneurysm surgery, which was 38 mm in diameter. In patient #10, AAA with localized dissection on the aneurysm was noted in the initial computed tomography (CT) imaging. In the other six patients (patients #4–9), AAA was observed as a part of chronic aortic dissection. In seven patients (patients #4–10) of age >20 years, the diameter of the AAA was ≥45 mm, with one patient (patient #5) showing a ruptured AAA of 80 mm in diameter.

Before replacement of the AAA, eight patients showed aortic dissection: type A in one and type B in seven patients. Patient #2 suffered from acute aortic dissection type at six months before the AAA replacement, for which total arch replacement was performed.

Indication of surgery

Surgical indication was decided on the basis of the diameter of AAA, as measured by CT, except for in a 2-year-old patient whose AAA was followed by abdominal ultrasonography.

In two children (patients #1 and #2), surgeries were indicated for the rapid expansion of the aneurysm of a diameter of 3 mm in two months and 4 mm in a year, respectively. Among the two patients who were symptomatic, emergent surgery was indicated for a rupture in patient #5. In patient #3, surgery was decided for rapid expansion (10 mm in a year), but urgent surgery was indicated for dissection complicated on the aneurysm.

In the remaining six patients of age >20 years with an aortic dissection, surgical indication was decided when the aneurysm diameter was ≥45 mm; however, in all cases, rapid expansion of the aneurysm was also observed at the rate of 3–6 mm in six months or 4 mm in a year.

Surgical strategy and procedures

All patients received graft replacement of infrarenal AAA using an artificial Dacron graft. The size of the graft was 10 mm in patient #1 and in other patients, 16 to 22 mm for proximal anastomosis and 9 to 11 mm for distal anastomosis when the bifurcated type was used. In patient #5 with a ruptured AAA, emergency graft replacement was performed with partial cardiopulmonary bypass for massive bleeding by means of cannulation of the common femoral artery and vein, followed by the exposure of the infrarenal aorta through the left pararectal skin incision and extraperitoneal approach. In the other nine patients, a graft replacement was performed through the transabdominal approach after a midline skin incision without cardiopulmonary bypass.

The type of reconstruction after aneurysm resection included the bifurcated iliac reconstruction in six patients and straight reconstruction in four. The reconstruction of aortic branches was performed for the inferior mesenteric artery in three bilateral accessory renal arteries in one patient and the right internal iliac artery in another patient.

In four patients (patients #4, #7, #8, and #9), an AAA was observed as a part of chronic type B aortic dissection whose entries were located at the descending aorta and the false lumen was patent, but the sizes of the descending aorta were under surgical indication as follows: patient #4, 32 mm; patient #7, 40 mm; patient #8, 42 mm; and patient #9, 38 mm.

In these indicated patients, proximal anastomosis after aneurysm resection was performed by a "double-barrel" technique, wherein resection of the adjacent dissected intima and graft anastomosis to both true and false lumen was performed, to avoid the elevation of the false lumen pressure by the closure of the outflow of false lumen.

Statistical analysis was performed using JMP (SAS Institute Inc., Cary, NC, USA) to estimate probability of survival using the Kaplan–Meier method.

This retrospective observational study was approved

Table 1 Patients' characteristics

No.	Age (y.o.)	Sex	CTD	Pathology	Rupture	Symptomatic	Diameter	Dilatation
1	2	Male	MFS	Aneurysm	No	Asymptomatic	15 mm	3 mm/2 month
2	12	Female	LDS	Aneurysm	No	Asymptomatic	32 mm	4 mm/year
3	22	Male	MFS	Aneurysm and dissection (localized)	No	Symptomatic	38 mm	10 mm/year
4	22	Female	LDS	Dissection (ST-B)	No	Asymptomatic	47 mm	5 mm/6 month
5	27	Female	MFS	Dissection (ST-B)	Rupture	Symptomatic	80 mm	—
6	27	Male	MFS	Dissection (ST-B)	No	Asymptomatic	53 mm	5 mm/6 month
7	28	Male	MFS	Dissection (ST-B)	No	Asymptomatic	51 mm	4 mm/6 month
8	28	Male	MFS	Dissection (ST-B)	No	Asymptomatic	45 mm	3 mm/6 month
9	29	Male	MFS	Dissection (ST-B)	No	Asymptomatic	60 mm	6 mm/6 month
10	29	Male	MFS	Dissection (localized)	No	Asymptomatic	46 mm	4 mm/year

y.o.: years old; CTD: connective tissue disease; MFS: Marfan syndrome; LDS: Loyes–Dietz syndrome; ST-B: Stanford type B

by the Institutional Review Board, and individual oral and written informed consents were obtained from each patient.

Results

Early results

No hospital mortality and operative morbidity, such as ileus, abdominal wall hernia, and graft infection, was encountered. The mean length of hospital stay was 18 ± 7 days.

Late results

All patients had a follow-up at our institute after a median period of 94 (range: 0–382 months) months. One patient died because of suicide 382 months after surgery for AAA.

Table 2 presents the timing of surgery for the aortic segment other than AAA before or after surgery for AAA is described for each patient. All patients received multiple aortic surgeries: two surgeries in three patients, three surgeries in three, four surgeries in two, and five surgeries in two.

The number of patients who received surgeries for each segment other than the AAA included the aortic root in nine patients, aortic arch in five, descending aorta in four, and thoracoabdominal aorta in five. Among these surgeries, 13 surgeries were performed before AAA and 10 after AAA.

Eight patients received additional aortic surgeries after the operation of AAA: aortic root in two patients, aortic arch in two, descending aorta in two, and thoracoabdominal aorta in four. During the same period, six other patients under the age of 30 years, who were not included in this study, received the repair of the thoracoabdominal aortic aneurysm extending to the infrarenal abdominal aorta.

In four patients (patients #4, #7, #8, and #9), the diameter of the thoracoabdominal aorta enlarged from 32 to 44 mm in five months, 40 to 51 mm in 30 months, 42 to 51 mm in 23 months, and 38 to 42 mm in 10 months, respectively.

Discussion

According to a recent nationwide database study conducted in the USA, the average age of patients who received elective surgical intervention for AAA was 73 years, and an AAA is uncommon in people under the age of 60 years.[1,2] In the patient cohort of our institute, patients' distribution, such as the number of patients under 60 years of age, was 10%, with male gender accounting for 80% of the study population; this result is in concordance with those of an American cohort study.[3] In this study, all patients with an age <30 years and received an AAA surgery suffered from connective tissue diseases, and the aortic pathology was dissection in eight-tenths of the patients. These characteristics completely differ from those of usual patients with an AAA of age 60–70 years.

The risk of rupture in degenerative AAA depends on its size and the degree of aneurysmal expansion. The annual risk of rupture based on the aneurysm size is estimated as follows: <0.5% in ≤40 mm diameter, 0.5%–5% in 40–49 mm, and 3%–15% in 50–59 mm.[3,4] In considering the incidence of rupture, the aneurysmal diameter of >50 mm was found reasonable as the surgical indication of degenerative AAA. However, several case reports about AAA in patients with Marfan syndrome report that the rupture of AAA could be complicated in patients with an AAA of a size <40 mm.[5,6] In these past reports, the fragility of the tunica media due to cystic medial necrosis was suspected as the cause of aortic rupture.[7] In this study, all six patients >20 years without symptoms revealed rapid expansion of the AAA within six months to one year. When an AAA is detected in young patients with Marfan syndrome, routine surveillance every six months might be recommended.

Table 2 Surgical timing for each aortic segment

No.	Age (y.o.)	Sex	Root	Arch	Descending	Thoracoabdominal
1	2	Male			30 M after	
2	12	Female	40 M before	6 M before		7 M before
3	22	Male	26 M before			
4	22	Female	70 M before	10 M before	3 M before	5 M after
5	27	Female	22 M after			
6	27	Male	3 M before		10 M after	
7	28	Male	21 M after			30 M after
8	28	Male	8 M before	23 M after		23 M after
9	29	Male	62 M before	50 M before	20 M before	10 M after
10	29	Male	54 M before	7 M after		

y.o.: years old; M: months

Months before or after surgery for abdominal aortic aneurysm shown for surgery of each aortic segment.

Murdoch et al. reported the life expectancy of patients with Marfan syndrome as 32 ± 16.4 years, with the primary life-threatening complication of Marfan syndrome being cardiovascular disease, especially an aortic aneurysm or dissection involving multiple segments of the aorta.[8] The development of surgical techniques to repair aortic aneurysms has been predicted to prolong the life expectancy of patients with Marfan syndrome.[9,10]

In this study, seven patients had a history of aortic surgery under close follow-up, and two patients were under the surveillance of an AAA. These close follow-ups allowed us to detect the rapid expansion of the AAA and perform safe graft replacement without any complications. In seven adult patients without rupture, rapid expansion was detected after the aneurysm enlarged to >40 mm in size. In one patient with extremely rapid enlargement (10 mm in a year), localized dissection was complicated at the size of 38 mm in diameter. The fair surgical outcomes of an AAA in young patients suggest that earlier surgical indication at around a 40 mm diameter size may be justified to reduce the risk of progressive aortic dilatation, further dissection, or rupture.

Patients with connective tissue diseases such as Marfan syndrome and Loeys–Dietz syndrome frequently developed aneurysms or dissections involving multiple segments of the aorta, and they occasionally required a staged replacement of the entire aorta.[11,12] In this study, all 10 patients required multiple aortic surgeries, and three of the 10 patients received a replacement of the entire aorta from the aortic root to aortic bifurcation.

Six patients (patients #4–9) suffered from chronic type B aortic dissection. Two patients had the history of descending replacement prior to AAA surgeries, and one patient received a descending replacement after the AAA surgery. Four patients received a thoracoabdominal replacement after their AAA surgeries, and two of them received the replacement within one year. All these surgeries are indicated according to the usual operative indication based on the routine follow-up protocol.

In this study, four patients received a thoracoabdominal aortic repair after an AAA replacement, and five patients received descending and thoracoabdominal aortic replacement within two years before and after an AAA replacement. Niinami et al. reported a relatively low associated morbidity and mortality with aggressive aortic surgical treatment in patients with Marfan syndrome.[13] If the multiple aortic surgeries are highly expected in the near future, aggressive one-stage repair of the thoracoabdominal aorta with earlier surgical indication should be considered.

Conclusion

Patients under the age of 30 years who received an AAA replacement were all complicated with a connective tissue disease. In patients under the age of 30 years, an AAA replacement was safely performed and early surgical indication could be considered.

Disclosure Statement

All authors have no financial relationship with a biotechnology manufacturer, a pharmaceutical company, or another commercial entity that has an interest in the subject matter or materials discussed in the manuscript.

Author Contributions

Study conception: JM, YI, HM
Data collection: JM, YI, AO, YS, KU
Analysis: JM, YI
Investigation: JM, YI
Writing: JM, YI, HM
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