



Total aortic replacement in an adolescent case

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Abstract: Total aortic replacement in adolescents is rare. Herein, we present a case of a 13-year-old girl who underwent thoracoabdominal aortic replacement following valve-sparing aortic root replacement and total arch replacement 5 months ago.

Keywords: Adolescence; Loeys-Dietz syndrome; aortic replacement

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Introduction

Successful thoracoabdominal aortic replacement is still a major challenge; additionally, this procedure in adolescents has rarely been reported. We report a successful thoracoabdominal aortic aneurysm repair (TAAAR) following aortic valve-sparing root replacement (AVSRR) and total arch replacement in a 13-year-old girl. TAAAR was performed with the patient under deep hypothermic circulatory arrest (20 °C), and the whole dissected aorta was replaced with a 20-mm, four-branched Gelweave Coselli thoracoabdominal graft (Vascutek-Terumo, Tokyo, Japan).

Case presentation

A 13-year-old girl was admitted to our hospital for intermittent massive hemoptysis. Five months ago, she had undergone valve-sparing aortic root replacement and total arch replacement for acute aortic dissection. Although no definitive diagnosis was established, her pediatrician highly suspected Ehlers-Danlos syndrome. Emergent computed tomographic angiography (CTA) demonstrated a dilated proximal descending aorta and fistula between the aorta and bronchus. Other parts of the aorta were not dilated but were small. Moreover, iliac and femoral arteries were small in size. The chest cavity was deformed and showed pectus excavatum. Ultrasound cardiography demonstrated no aortic regurgitation but showed low ejection fraction, which was identified after the previous operation performed

for acute aortic dissection. Therefore, an emergent thoracoabdominal aortic replacement was performed via a straight incision using rib-cross thoracotomy (*Figure 1*) (2). In patients with flat chest, this incision provides better exposure of the distal aortic arch than the standard incision. A previously implanted side branch of the aortic arch was used owing to the small femoral arteries. Cardiopulmonary bypass was established by placing the venous return in the left femoral vein. Moreover, a vent tube was placed in the left atrium from the left appendage. When the nasopharyngeal temperature reached 20 °C, circulatory arrest was commenced. To avoid cerebral embolism, an aortic clamp was placed at the arch graft between the second and third branches.

The fistula between the aorta and lung was examined. Proximal anastomosis was made at the elephant trunk implanted in the previous operation. Perfusion was re-established with the original aortic return from the side branch of the arch graft. Because of the severe deformity of the chest cavity, the descending aorta was located behind the heart, as observed through the surgical view. The descending to abdominal aorta was completely opened, and the dissected flap was resected. Celiac, superior mesenteric, and bilateral renal arteries were started to enable perfusion with balloon-tip cannulas. Origins of intercostal arteries were sealed with z-suture using 3-0 polypropylene suture, except for the targeted intercostal artery that was supposed to connect the Adamkiewicz artery (AKA). A feeder of the



Figure 1 Total aortic replacement in an adolescent case (1). Available online: <http://www.asvide.com/article/view/26963>

AKA was preoperatively detected using CTA. In this case, the critical segmental artery was the left eighth intercostal artery, which was reconstructed to maintain perfusion to the spinal cord. A 10-mm diameter graft was anastomosed to the critical segmental artery using inclusion technique and to the main graft in an end-to-side fashion. The ninth intercostal artery was additionally reconstructed to provide collateral circulation into the critical artery to the AKA (3,4). Because the abdominal aorta was only 10 mm in diameter where the distal anastomosis was performed, the incision was made longitudinally and the distal anastomosis was made in an end-to-side fashion. The celiac, superior mesenteric, and bi-renal arteries were reconstructed using side branches of the graft individually. Despite the cardiac dysfunction, the patient could be weaned from cardiopulmonary bypass safely. Additionally, the motor-evoked potential was monitored during the operation. It had disappeared during the hypothermia once but reappeared during rewarming.

The patient required few months to recover from the low cardiac output syndrome. Overall, the postoperative course was uneventful, and the patient was discharged without complications. She currently attends school without any difficulties.

Discussion

There were several issues regarding this operation. Firstly, thoracoabdominal aortic replacement was performed in an adolescent patient during growth period. It is generally assumed that a vascular prosthesis should be placed at an anatomic position, and its length should be as short as possible to avoid kinks and bents. However, the influence

of total aortic replacement on growth spurt has not been evaluated. Therefore, in this case, the main body of the graft at the thoracoabdominal aorta was kept slightly long and bent outward. Because the main body of the graft was bent, the length of the side branches was kept longer than usual. Careful monitoring is required for both the graft length during the growth period and for other native aorta and aortic valve competencies following AVSRR. Secondly, the strategy of deep hypothermia could have posed a threat to the patient's cardiac dysfunction. Deep hypothermia is our routine strategy for types I and II thoracoabdominal aortic replacements. This patient had severe chest deformity with the aorta located behind the heart. Considering this, performing aortic replacement with the heart beating was regarded difficult. She was administered catecholamine for a few months postoperatively; however, her cardiac function completely recovered in the long run. Careful monitoring is mandatory not only for the cardiac function after total aortic replacement but also for the aorta during the growth period (5).

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were

in accordance with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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