

# Aortic root reconstruction by aortic valve-sparing operation (David type I reimplantation) in Marfan syndrome accompanied by annuloaortic ectasia and acute type-A aortic dissection

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To reconstruct the aortic root for aneurysm of the ascending aorta accompanied by aortic regurgitation, annuloaortic ectasia (AAE) and acute type-A dissection with root destruction, the Bentall operation using a prosthetic valve still is the standard procedure today. Valve-sparing procedures have actively been used for aortic root lesions, and have also been attempted in aortic root reconstruction for Marfan syndrome which may have abnormalities in the valve leaflets. We conducted a valve-sparing procedure in a female patient with Marfan syndrome who had AAE accompanied by type-A acute aortic dissection. The patient was a 37-year-old woman complaining of severe pain from the chest to the back. The limbs were long, and funnel breast was observed. Diastolic murmurs were heard. On chest computed tomography, a dissection cavity was present from the ascending aorta to the left common iliac artery, and the root dilated to 55 mm. Grade II aortic regurgitation was observed on ultrasound cardiography. Regarding her family history, her father had died suddenly at 54 years of age. She was diagnosed with type-A acute dissection concurrent with Marfan syndrome and AAE. The structure of the aortic valve was normal, and root reconstruction by a valve-sparing operation and total replacement of the aortic arch was conducted. On postoperative ultrasound cardiography, the aortic regurgitation was within the allowable range, and the short-term postoperative results were good.

**Key words:** Marfan syndrome, acute type-A aortic dissection, aortic valve-sparing operation, aortic root surgery

## INTRODUCTION

Marfan syndrome is a genetic abnormality of the connective tissue of the highest incidence. While main disorders are observed in the eyes, skeleton and cardiovascular systems, the factor determining the prognosis is the cardiovascular events. Thus, it is desirable to reconstruct the aortic root early in those patients with aortic root dilation. For the treatment of dilated ascending aorta and type-A dissection in Marfan syndrome, the Bentall procedure is widely accepted, because this procedure produces good outcomes by replacing the aortic root and the aortic valve with a composite valve graft in parallel with coronary reconstruction [1, 2]. Since the patients with Marfan syndrome are young and often choose artificial valves, they face the issue of remote-phase complications associated with anticoagulation therapies. The aortic root reconstruction by a valve-sparing operation appears reasonable because anticoagulation therapies could be avoided by sparing the native valve without using artificial valves. A report described long-term results of aortic valve-sparing procedures [3]. We conducted root reconstruction by an aortic valve-sparing procedure in a 37-year-old woman with Marfan syndrome and annuloaortic ectasia (AAE) accompanied by acute type-A aortic dissection.

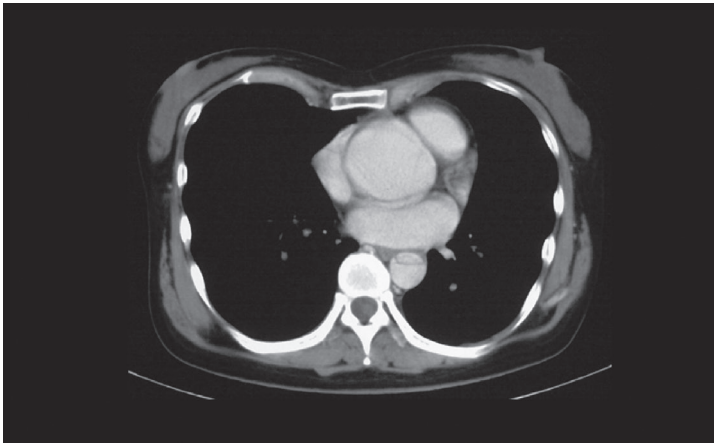
## CASE

The patient was a 37-year old woman. On October 20, 2005, she felt pain from the chest to the back. She was immediately brought to the critical care center of our hospital. She was awake, blood pressure was 148/50 mmHg, and diastolic murmurs were heard. Her height was 167 cm, the limbs were long, and she showed funnel breast. The blood pressure in the right lower extremity could not be determined. Based on the symptoms and physical findings, acute aortic dissection was suspected and chest computed tomography (CT) was conducted. The aortic root dilated to 55 mm, and a dissection was present from the ascending aorta to the left common iliac artery. No retention of pericardial fluid was observed (Fig. 1). Ultrasound cardiography (UCG) also showed the dilated aortic root, and grade II aortic regurgitation was observed. Regarding family history, her father had died suddenly at 54 years of age.

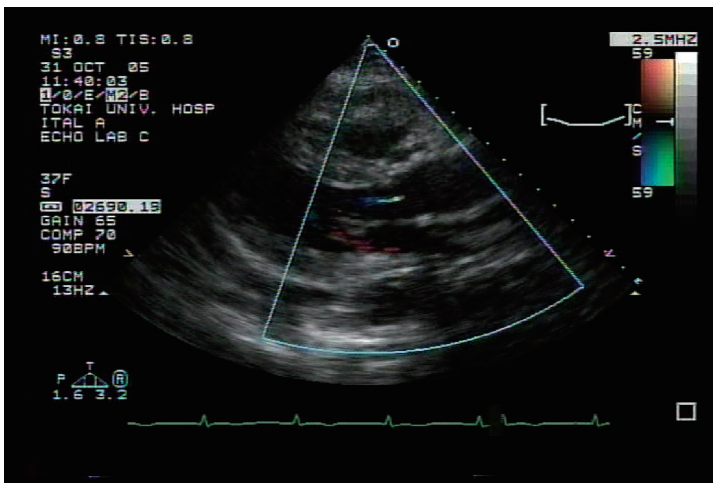
Based on the above examinations, she was diagnosed with complicated type-A acute dissection accompanied by Marfan syndrome and AAE, and an emergency operation was conducted. Diagnostic criteria for Marfan syndrome followed the paper by De Paepe [4].

## OPERATIVE PROCEDURE AND RESULTS

The proximal thoracic aorta was approached by means of a median sternotomy. Both axillary arteries and the right femoral artery were anastomosed



**Fig. 1** Preoperative CT.  
The dissection is present from the ascending aorta to the left common iliac artery, and the root enlarged to 55 mm.



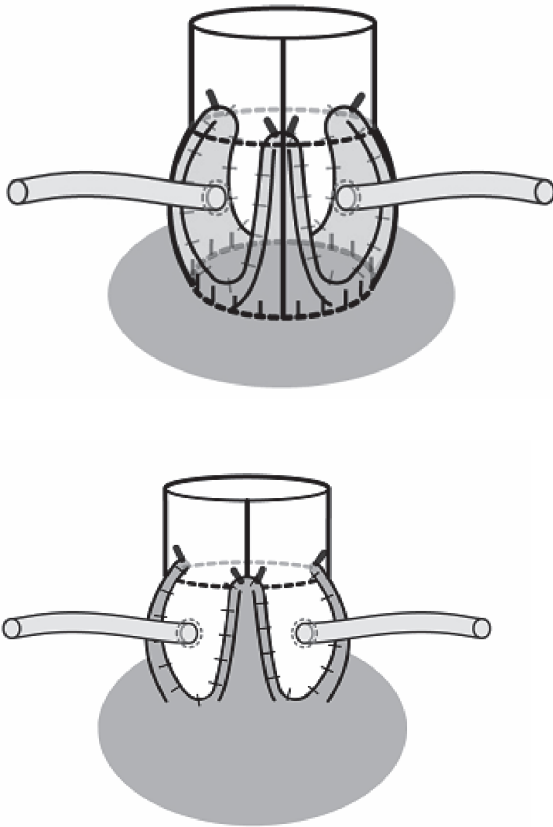
**Fig. 2** Postoperative UCG.  
Trivial aortic regurgitation was observed.

to a 6-mm polytetrafluoroethylene graft for arterial return, and the superior and inferior vena cavae each received a single right-angled cannula for venous access. Cardiopulmonary bypass was established, and core cooling was conducted. A cannula for retrograde cardioplegia was placed in the coronary sinus, and a vent tube was placed through the right superior pulmonary vein. Myocardial protection was achieved by using intermittent retrograde cold blood cardioplegia with direct antegrade coronary ostia infusion once the aorta was opened. The entry was present at the right sino-tubular junction. The dissection did not extend into the sinus of Valsalva, the aortic valve coaptation was good, and no abnormality was observed in its structure.

Since the structure of the valve was normal, root construction by a reimplantation method using a valve-sparing procedure was selected. Both coronary arteries were cut out in the shape of a cuff from the aortic wall (the Carrel patch method). The aortic wall was excised at a distance of 3 mm from the aortic annulus. A mattress suture was placed from within outward with 2-0 polyester sutures reinforced with felt along the entire circumference of the basal ring immediately below the skeletonized aortic annulus. The membranous septum was placed at the aortic annulus. A 4-branched 28-mm graft was selected. The graft was cut to 4 cm and placed on the skeletonized valve, and the mattress suture in the aortic annulus was passed through the stump of the graft from the inside to

outside and ligated. The respective commissures were fixed to the graft with a 4-0 polypropylene suture. The aortic valve was fixed to the graft by continuous suturing with a 4-0 polypropylene suture. The aortic valve was reimplanted in the graft. Water was infused into the graft to confirm that there was no leakage. Both coronary arteries were fixed to the graft by the Carrel patch method using a 4-0 polypropylene suture in a running fashion.

Next, the aortic arch was treated. The rectal temperature was 25°C. Distal perfusion from the femoral artery was stopped (open distal anastomosis), the innominate artery and the left subclavian artery were clamped, a 12 Fr balloon perfusion cannula (Fuji Systems, Tokyo, Japan) was placed at the left common carotid artery, and antegrade cerebral perfusion was started. The aorta was cut after the branch of the left subclavian artery. Since the diameter of the distal aorta was as small as 19 mm, placement of an elephant trunk was abandoned. A 20-mm straight graft was selected, the stump of the distal aorta was wrapped with a felt strip and anastomosed to the graft by a mattress suture with 2-0 polyester sutures, and continuous suturing with a 3-0 polypropylene suture was added. The 20-mm graft on the distal side was sutured to a 28-mm 4-branched graft, distal perfusion through the lateral branch was restarted, and warming was started. Subsequently, the graft for root reconstruction was sutured to a 4-branched graft, and myocardial ischemia was released. The arch vessels were reconstructed from



**Fig. 3** Schematic drawing of the surgical techniques. Reimplantation procedure (upper) and remodeling procedure (lower).

the distal side. The absence of aortic regurgitation was confirmed by transesophageal echocardiography. Warming was continued until the patient's rectal temperature reached 36°C. The cardiopulmonary bypass time was 358 minutes, myocardial ischemic time was 228 minutes, and systemic circulatory arrest time was 90 minutes.

On postoperative UCG, trivial aortic regurgitation was observed (Fig. 2). The follow-up of the patient was ongoing as of the submission of this report.

### DISCUSSION

The advantage of the valve-sparing procedure is that it provides excellent survival rate and it helps avoid cardiac accidents, because anticoagulant therapy is not necessary and there is no risk of long-term complications resulting from implantation of an artificial valve. There are following two main operative procedures for valve-sparing aortic root reconstruction (Fig. 3): (1) the remodeling method which has been performed since its original description in 1978 by Yacoub [5, 6], and (2) the reimplantation method which has been performed since its original description in 1988 by David *et al.* [7]. However, the indications and selection of procedure, including the sizing of the graft, are controversial. Initially, the decision was made according to the paper by David [7]. However, an increasing number of reports suggested that grafts of 26 to 30 mm were frequently selected [8]. Our standard for the diameter

of the graft is up to 30 mm, with the diameter of annulus +1 to 2 mm. We have so far performed the aortic valve-sparing operation in total of five patients including the present patient: one patient underwent the remodeling method, and four patients were treated by the reimplantation method; the diameter of the graft was 26 mm in one patient, 28 mm in three patients and 30 mm in one patient.

The advantage of the remodeling method is that the hemodynamic properties can be reproduced since the structure of the Valsalva sinus can be reproduced. The disadvantages of this method are that dilation of the aortic annulus cannot be prevented and that the risk of bleeding increases since the anastomosis line becomes long. The advantages and disadvantages of the reimplantation method are precisely opposite to that of the remodeling method. However, a graft with the structure of the Valsalva sinus was recently invented by De Paulis *et al.* [9] and is expected to improve the durability of the composite aortic valve, a disadvantage of the reimplantation method due to the lack of structure of the Valsalva sinus.

In Marfan syndrome, it has been reported that abnormalities of the connective tissue of the valve leaflet are present even if it is macroscopically normal, and the durability after valvuloplasty is controversial [10]. Birks *et al.* [11], however, reported that the aortic valve-sparing procedures were indicated for about 70% of patients with Marfan syndrome and that the survival rate was almost the same as that in patients without Marfan syndrome. Further, Tambour *et al.* [12] reported that the 10-year survival rate was 100% in patients with Marfan syndrome undergoing the remodeling method (23 patients) or reimplantation method (19 patients) and that no patient needed reoperation. De Oliveira *et al.* [3] also reported that the 10-year survival rate was 96% in patients with Marfan syndrome undergoing the reimplantation method (39 patients) or remodeling method (22 patients) and that the rate of avoidance of reoperation in 10 years was 100%. These results indicate that the valve-sparing procedures for Marfan syndrome are producing good long-term outcomes. However, more time is required for the valve-sparing procedures for Marfan syndrome to become recognized widely in the world. Until then, the standard procedure for root reconstruction will still be the Bentall procedure using the Carrel patch method [2]. In order for the valve-sparing procedures to achieve excellent long-term results in the future, we should consider root reconstruction before the aortic root dilates to the present standard of 50 mm [8], select and revise the method so as not to induce aortic regurgitation at an early postoperative period.

### CONCLUSION

We have experienced root reconstruction by the valve-sparing procedure in a patient with Marfan syndrome who had AAE accompanied by acute type-A aortic dissection and confirmed good short-term postoperative results. In Marfan syndrome with AAE, dilation of the Valsalva sinus increases the incidence of dissection and deformation of the valve, making the valve-sparing procedures complex and difficult. Therefore, early surgical treatment is desirable for

patients with Marfan syndrome and AAE.

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