

〈Case Report〉

TAVI for severe AS in a patient with dextrocardia, situs inversus, horizontal aorta, bicuspid valve, and ascending aortic aneurysm

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ABSTRACT The patient was a 70-year-old male diagnosed with dextrocardia and situs inversus totalis. He had undergone pacemaker (PM) implantation for sick sinus syndrome (SSS) 20 years ago and percutaneous coronary intervention for left anterior descending coronary artery stenosis 15 years ago. Several years ago, he developed dyspnea on exertion, and a detailed examination revealed severe aortic valve stenosis (AS). The patient's symptoms gradually worsened, leading to the decision to proceed with surgery. Although he was relatively young, his activity levels were reduced due to the aftereffects of a traffic accident. Therefore, we opted for transfemoral transcatheter aortic valve implantation (TAVI) using a balloon-expandable aortic valve. Given the patient's complex anatomy, including dextrocardia, situs inversus, an ascending aortic aneurysm, and a Type 0 bicuspid valve with a horizontal aorta, TAVI was performed with particular caution. Owing to the dextrocardia in the patient, the monitor display was inverted during surgery to facilitate prosthetic valve deployment, and the procedure was completed without any complications. The patient had a good postoperative course and was discharged on postoperative day 9. This is a rare case of TAVI performed for AS in a patient with dextrocardia and situs inversus totalis. By inverting the monitor display during prosthetic valve deployment, the procedure was conducted using the same visual orientation as in a typical case, simplifying the operation. doi:10.11482/KMJ-E202551059 (Accepted on February 10, 2025)

Key words : Case report, Dextrocardia, Transcatheter aortic valve implantation (TAVI),

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BACKGROUND

TAVI is a treatment option for symptomatic severe AS, and its range of indications has rapidly expanded in recent years. Prosthetic valves and delivery catheter systems continue to evolve in order to reduce all associated risks, including those related to vascular access complications. TAVI appears to be safe, even in patients with more complex anatomy.

Dextrocardia with situs inversus totalis is a rare congenital heart condition, with an incidence of approximately 1 in 12,000^{1, 2)}. This report describes a complex anatomical case involving severe AS, dextrocardia, situs inversus, an ascending aortic aneurysm, and a Type 0 bicuspid valve with a horizontal aorta. We report this case because TAVI was successfully performed.

CASE REPORT

Patient: 76-year-old male.

Present Illness: The patient had a PM implanted for SSS and was a regular outpatient at the Department of Cardiology in this hospital for post-percutaneous coronary intervention (PCI) follow-up. He was monitored in our outpatient clinic, but his dyspnea on exertion gradually worsened. As a result, the decision was made to perform surgery.

Past History: Complete visceral inversus and dextrocardia, chronic obstructive pulmonary disease.

Two years ago: PM implantation for SSS.

Fifteen years ago: PCI for angina pectoris (#7 TAXUS 3.5 × 28 mm (Boston Scientific, Natick, MA, USA)).

One year ago: Traffic accident; currently walking with a cane due to the aftereffects.

Smoking/Drinking: Smokes 20 cigarettes per day for 40 years. Social drinker.

Physical examination: Height 168.0 cm, weight 71.2 kg, body surface area 1.8 m², pulse 63/min, blood pressure 148/88 mmHg, body temperature 36.5°C, SpO2 97% (room air).

Blood tests: BNP was not elevated, and no abnormal findings were noted in the complete blood count, biochemistry, or coagulation/fibrinolytic system.

Chest X-ray: Cardio thoracic ratio 52%, Costophrenic angle sharp, dextrocardia and post-PM implantation.

EKG (right precordial leads): Heart rate 63/min, PM rhythm (atrial pacing).

Transthoracic echocardiography (TTE): Left ventricular ejection function (LVEF) 63%, severe AS (peak velocity 4.6 m/s, mean pressure gradient

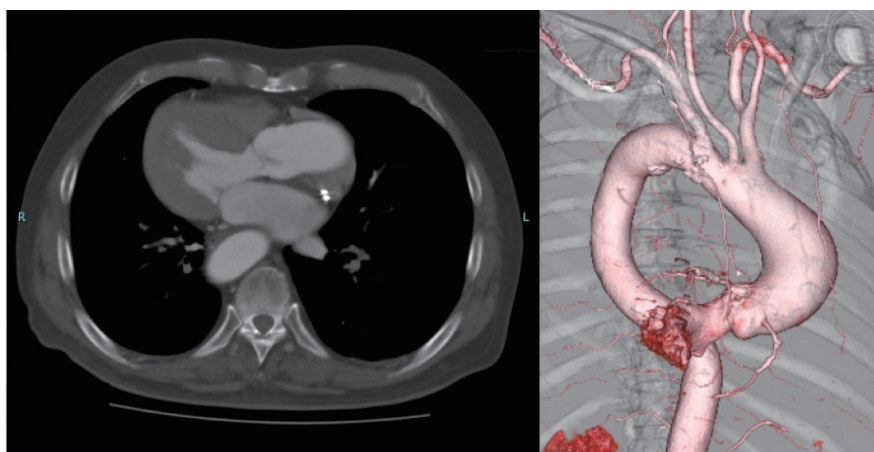


Fig. 1. Contrasted-enhanced CT with 3D reconstruction.

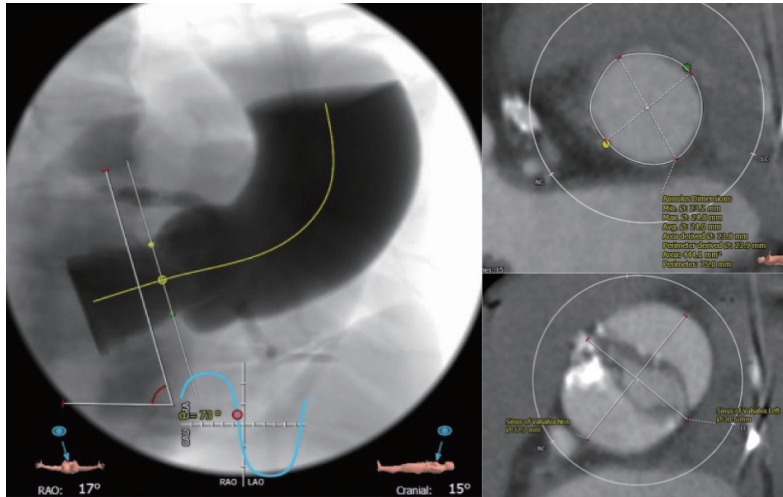


Fig. 2. Contrast-enhanced CT showing analysis of the aortic valve.

47 mmHg, aortic valve area (AVA) 0.69 cm^2 , AVA index $0.39 \text{ cm}^2/\text{m}^2$).

Contrast-enhanced chest computed tomography (CT) scan (Fig. 1): Revealed dextrocardia with situs inversus totalis, ascending aortic aneurysm (50 mm), and a horizontal aorta.

Analysis of the aortic valve (Fig. 2): Aortic valve annulus: $23.2 \text{ mm} \times 24.8 \text{ mm}$, area: 444.1 mm^2 , perimeter: 75.0 mm , sinuses of Valsalva: $30.5 \times 37.7 \text{ mm}$, intercommisural distance: 29-31 mm, bicuspid valve (Sievers Type 0), ascending aorta: $45.2 \times 46.4 \text{ mm}$, aortic root angulation: 73° .

Therefore, the patient was diagnosed with symptomatic severe AS and an ascending aortic aneurysm. The patient presented with a complex anatomical structure, including dextrocardia, situs inversus totalis, an ascending aortic aneurysm, and a Type 0 bicuspid valve with a horizontal aorta. Although the patient was relatively young, his activities of daily living had declined because of the aftereffects of a traffic accident. After discussions at a multidisciplinary conference involving cardiology, cardiovascular surgery, and anesthesiology, it was decided that TAVI was appropriate for the patient. Based on the valve analysis, a 26 mm Sapien3 balloon-expandable prosthetic valve (Edwards

Lifesciences, Irvine, CA, USA) was selected.

SURGERY

Transfemoral-TAVI Sapien3 26 mm 21 mL (2 mL underfill)

The patient was placed in the supine position under general anesthesia. A 14 Fr arterial dilatation sheath (Edwards Lifesciences) was inserted into the right femoral artery. A pigtail catheter was inserted into the right coronary cusp through the left femoral artery. We established a perpendicular view (Right anterior oblique, 17° ; Cranial, 15°), and the aortic valve was passed through a straight Radifocus wire (TERUMO, Tokyo, JAPAN). After crossing the aortic valve with a straight Radifocus wire (TERUMO, Tokyo, Japan), a Safari wire (Boston Scientific, Natick, MA, USA) was positioned in the left ventricle. Aortic balloon angioplasty (BAV) was performed using a 23 mm balloon (Edwards Lifesciences) under rapid pacing, with the fluoroscopic display monitor inverted to the left and right sides. Rapid pacing was performed after inserting a temporary pacing lead and changing the permanent pacemaker setting to DDD 60/130. Because of the presence of a horizontal aorta and valve stiffness, the balloon slipped; therefore,

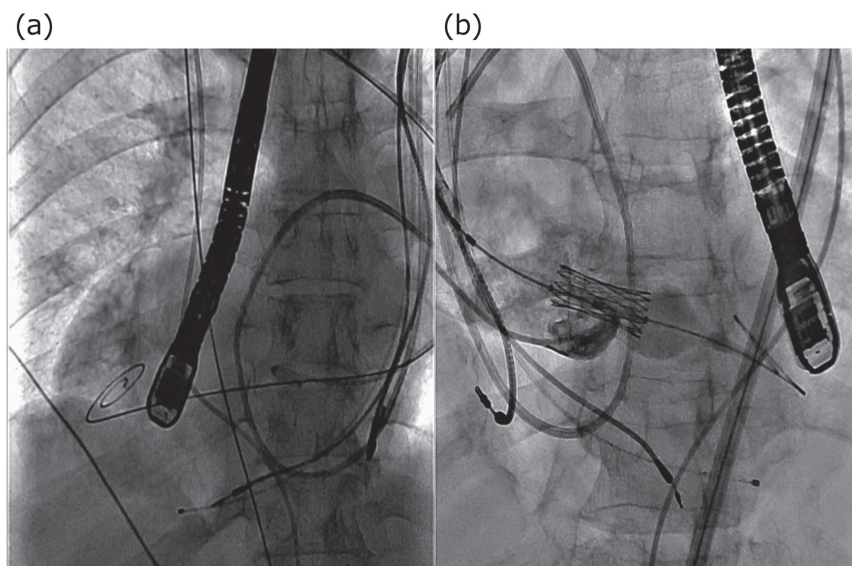


Fig. 3. X-ray images during TAVI.

(a) Normal image.

(b) Left-right reversed image.

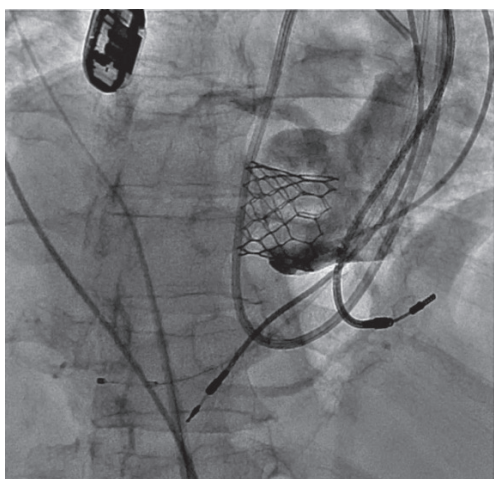


Fig. 4. X-ray image during TAVI showing prosthetic valve deployment.

BAV was performed four times. The Commander delivery system (Edwards Lifesciences) was rotated 180° and a 26 mm Sapien3 valve was deployed (nominal volume minus 2 mL) with inverting the monitor display (Fig. 3). Transesophageal echocardiography revealed paravalvular leak (PVL) from the anterolateral side of the non-coronary cusp. To address this, post-dilatation using delivery

system 26 mm balloon minus 1.5 mL of contrast was performed (Fig. 4). The procedure details were as follows: operation time, 1 h 33 min; rapid pacing time, 146.4 s; contrast volume, 190 mL; and fluoroscopy time, 35.6 min.

POSTOPERATIVE COURSE

The patient was managed in the intensive care unit on the surgery day and was transferred to the general ward on postoperative day (POD) 1.

Postoperative TTE: LVEF 63%; aortic valve: peak velocity 2.3 m/s, mean pressure gradient 12 mmHg, doppler velocity index 0.36, acceleration time 67 ms, no PVL.

The patient had a good clinical course and was discharged POD 9. He has since been under outpatient observation with no reported complications.

DISCUSSION

Dextrocardia with situs inversus totalis is a rare condition, and this is the third reported case in Japan where TAVI was successfully performed^{3, 4)}.

This is the first report involving an ascending aortic aneurysm, a Type 0 bicuspid valve with a horizontal aorta, and complex anatomy.

Of the 13 case reports to date, one-third used self-expanding aortic valve devices, while two-thirds used balloon-expandable devices. Some reports suggest that self-expanding aortic valve devices are advantageous in complex anatomies because of their recapturable and repositionable design^{2, 3)}. However, actual reports show that balloon-expandable devices are commonly used. While there has been one case of right ventricular perforation of a PM lead with a self-expanding aortic valve device and one case of complete atrioventricular block with a balloon-expandable device as postoperative complications, aortic valve implantation itself can be completed regardless of the device type²⁻⁶⁾. An important consideration is preoperative projection. Understanding cardiovascular anatomy through preoperative contrast-enhanced CT 3D construction is crucial. In addition to the fact that we are accustomed to performing TAVI with balloon-expandable aortic valves, we have selected balloon-expandable aortic valves because the aortic root angle is over 70°, which is the upper limit of the angle for self-expandable aortic valves, and therefore, we consider that self-expandable aortic valves are not indicated. And the dextrocardia with situs inversus totalis is visually confusing to catheterization. Furthermore, the patient has a horizontal aorta, a bicuspid aortic valve, and an ascending aortic aneurysm, each of which makes it difficult to predict the behavior of the prosthetic valve during deployment based on past literature reports.

Another important aspect of aortic valve implantation is the inversion of the radiograph monitor display, as shown in this case. This technique has been reported multiple times, and in addition to reversing the monitor display, rotating the delivery system by 180° is also crucial²⁻⁶⁾. In

practice, the procedure can be performed in the same manner as in typical cases, and it appears to be a very effective method.

CONCLUSIONS

TAVI is an important intervention option for cases with complex anatomy, such as severe AS associated with dextrocardia and situs inversus totalis. Moreover when performing TAVI, reversing the monitor display during BAV and prosthetic valve deployment allows the procedure to be conducted with the same visual information as in typical cases. This approach enhances ease of and safety during the procedure, making it a useful technique.

ACKNOWLEDGEMENTS

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