Valve-Sparing Root Replacement in a Turner Syndrome Patient with Bicuspid Aortic Valve and Juxtacommissural Origin of the Right Coronary Artery: A Case Report

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Case report

A 32-year-old woman with Turner syndrome visited the hospital for an evaluation of cardiovascular complications. Progression of aortic root and ascending aortic dilatation was observed during follow-up. We planned surgery because of the increasing risk of aortic rupture. Preoperative computed tomography (CT) showed ascending aorta dilatation (45.04 mm), aortic root dilatation (48.24 mm), and a high aortic size index (ASI) (36 mm/m²) (Fig. 1A, B). Echocardiography revealed a normal aortic annulus (23 mm), bicuspid aortic valve, and no evidence of aortic regurgitation. We planned aortic valve reimplantation and ascending aorta replacement.

Surgery was performed via median sternotomy. Cardiopulmonary bypass was established as usual, with cannulation of the proximal aortic arch and the bicaval technique. An aortic cross-clamp was performed below the arterial cannulation site. After the aortic cross-clamp, the ascending aorta was transected above the sinotubular junction. A type 0 bicuspid aortic valve was observed, and there was no evidence of leaflet thickening and calcification. Incidentally, we found that the right coronary artery (RCA) was located very close to the anterior side commissure of the bicuspid aortic valve (Fig. 2A, B).

In this case, we designed a graft using a modified Florida sleeve technique [1]. After aortic root dissection below the nadir, we resected the aneurysmal aortic root tissue with left coronary artery (LCA) button formation and saved the anterior commissure with the juxtacommissural-origin RCA (Fig. 2C). A small incision was made on the 28-mm Gelweave Valsalva graft to make a keyhole for encircling the RCA. After the 28-mm Gelweave Valsalva graft was implanted with 4 anchoring sutures below the annulus, a hemostat suture was used for commissural fixation and sinus fixation of the graft. The RCA was encircled by the graft, and the small incision previously made on the Gelweave Valsalva graft was closed using interrupted sutures,
followed by LCA reimplantation (Fig. 3A).

We evaluated aortic valve function using a saline test. No aortic regurgitation was observed. Thereafter, a 26-mm Hemashield graft was sequentially anastomosed to the distal ascending aorta and the Gelweave Valsalva graft (Figs. 2D, 3B). Deairing and cardiopulmonary bypass weaning were also performed. Sternum closure was performed layer by layer using wiring and wound closure.

Postoperative CT showed that the aortic root and ascending aorta were replaced by the graft, and there was no evidence of coronary kinking or stenosis (Fig. 1C, D). Postoperative echocardiography showed no evidence of aortic regurgitation. Intraoperatively, we found hemorrhagic changes in the aortic wall. The resected aortic wall was sent for a pathology examination, which revealed fiber disruption, cystic medial degeneration, and intramural hematoma (Fig. 4A, B). The patient was discharged from the hospital on postoperative day 7 without any complications.

This study was approved by the Institutional Review Board of Seoul National University Hospital (IRB approval no., 2207-179-1344). The requirement for informed consent was waived.

**Discussion**

The association of anomalous anatomy of the coronary arteries and bicuspid aortic valve has been sporadically reported. Previous research indicated that coronary artery anomalies were observed in 7% of patients with bicuspid aortic valve undergoing aortic valve surgery [1]. In this case, the juxtaommissural origin of the coronary artery was detected incidentally, causing difficulties in making a coronary button during aortic valve reimplantation. We therefore suggest that a preoperative evaluation of coronary anomalies in patients with bicuspid aortic valve must strictly be reviewed by a surgeon to decide the appropriate surgical plan, especially for aortic valve reimplantation, and the relationship and distance between the coronary artery and the commissure.

In 2005, the Florida sleeve technique was introduced as a new technique for aortic valve-sparing aortic root replacement [2,3]. The Florida sleeve technique does not require coronary reimplantation, which decreases the risk of surgical bleeding and provides time-saving technical advantages. Studies of the early and long-term outcomes of the Florida sleeve technique have proven it to be safe, effective, and durable [4,5]. However, there have been concerns regarding
the use of this technique for large aneurysms. It is difficult to encase the native aortic root tissue in the graft with a good fit to enable laminar flow. In addition, the coronary geometry can be distorted, resulting in coronary flow problems [6]. In this case, the authors initially decided to use the Florida sleeve technique as an easier procedure; however, we further modified it for coronary securement during aortic valve-sparing aortic root replacement.

In the present case, we encountered an unexpected origin of the coronary artery very close to the commissure to a bicuspid aortic valve in a patient with a large root aneurysm, and we were able to perform aortic valve reimplantation combined with the modified Florida sleeve technique. We resected the aneurysmal aortic root tissue as far as possible and preserved the native juxtacommisural-origin coronary artery with commissure to obtain good aortic valve function and coronary patency. We expect that this operative technique will have all the advantages of conventional valve-sparing aortic root replacement and the Florida sleeve technique, particularly for patients with a juxta-commisural coronary artery and bicuspid aortic valve.

The operative indications for aortic root aneurysms are different in patients with Turner syndrome. The European Society of Cardiology guidelines suggest an ASI of more than 27.5 mm/m² as an indication for surgery in patients with Turner syndrome [7]. Considering current guidelines and pathology results, precautions should be taken when determining the appropriate timing of surgical correction.

**Fig. 2.** Operative findings. (A) A juxtacommisural right coronary artery (RCA) os can be seen (yellow arrow); (B) a coronary probe located at the RCA os (yellow arrow); (C) a blue vessel sling encircled the RCA and left coronary artery, and button formation was performed (yellow arrow); (D) aortic root replacement was performed.

**Fig. 3.** Modified Florida sleeve repair. (A) Four anchoring sutures were made at the lowest level of the annulus of the commissure and mid-point of both leaflets; (B) a hemostat suture was applied with commissure and sinus fixation. LCA, left coronary artery; RCA, right coronary artery.

**Fig. 4.** Aortic wall pathology. (A) Fiber disruption and cystic medial degeneration; (B) intramural hematoma.
in patients with Turner syndrome in order to prevent severe complications.

In summary, we describe a new technique that could achieve appropriate commissural alignment and coronary patency in a patient with bicuspid aortic valve and juxta-commissural origin of the coronary artery who underwent aortic valve-sparing aortic root replacement.

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Conceptualization: SC. Data curation: DHS. Formal analysis: SC. Methodology: SC. Project administration: SC. Visualization: SC, DHS. Writing--original draft: SC. Writing--review & editing: MKS, SC. Final approval of the manuscript: MKS, SC.

**Conflict of interest**

No potential conflict of interest relevant to this article was reported.

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