Transcatheter Aortic Valve Replacement for Patients With Aortic Valve Stenosis Complicated With Moyamoya Disease

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Moyamoya disease (MMD) is a rare disease characterized by occlusive intracranial arteriopathy with formation of abnormal cerebrovascular collateral networks. Conventional cardiovascular surgical procedures using cardiopulmonary bypass for patients with MMD is challenging because low cerebral perfusion pressure and nonpulsatile (continuous) flow during cardiopulmonary bypass can cause severe cerebral ischemia. We successfully performed transcatheter aortic valve replacement in 3 women with severe aortic valve stenosis complicated with MMD. Transcatheter aortic valve replacement may be useful for patients with severe aortic valve stenosis complicated with severe cerebral ischemia, including MMD.


Moyamoya disease (MMD) is a rare disease characterized by bilateral intracranial occlusion of the internal carotid arteries, and anterior and middle cerebral arteries with formation of cerebrovascular collateral networks. It has been reported that cardiovascular surgical procedures using cardiopulmonary bypass (CPB) for patients with MMD is challenging because of decreased cerebral perfusion pressure and nonpulsatile (continuous) flow. We successfully performed transcatheter aortic valve replacement (TAVR) in 3 women with severe aortic valve stenosis complicated with MMD. Here we describe one of the cases because the other 2 patients are enrolled in a clinical trial and the related data are unavailable.

An 84-year-old woman was referred to our institution for the surgical treatment of a severe aortic valve stenosis with a mean pressure gradient of 53 mm Hg and an aortic valve area of 0.52 cm² (aortic valve area index, 0.37 cm²/m²). At the age of 62, she received a diagnosis of MMD during a checkup for recurrent headaches at another institution. Magnetic resonance angiography showed many fine collateral vessels, the so-called moyamoya vessels (Fig 1A, B, arrows) and bilateral occluded internal carotid arteries in the supraclinoid portion (Fig 1A, arrowheads). Positron emission tomography-computed tomography using diamox revealed a severe decrease in cerebral blood flow at the bilateral anterior and middle cerebral arteries and a significant high risk of hemodynamic cerebral ischemia in these areas (Fig 2). Moreover, preoperative electrocardiogram-gated, multislice computed tomography showed that the right coronary artery might be occluded by the native cusp after valve implantation (Fig 3) and atherosclerosis obliterans at the bilateral lower limbs. The patient consented to undergo transapical TAVR after close consideration of the increased risk of neurologic complications in conventional aortic valve replacement with CPB.

With the patient under general anesthesia, we performed transapical TAVR. Arterial and venous sheaths were inserted percutaneously into the right femoral artery and vein, respectively. A primed cardiopulmonary system was made available for an immediate conversion to CPB. Additionally, an arterial sheath was inserted into the left femoral artery for an intraaortic balloon pump (IABP) in case nonpulsatile flow were to occur because of intraoperative complications. First, we placed a microcatheter and a microwire on the right coronary artery. Therefore, we implanted a 23-mm Sapien stent-valve (Edwards Life-sciences, Inc, Irvine, CA) in a precise position under rapid ventricular pacing. Intravascular ultrasonography revealed that the right coronary cusp obstructed the orifice of the right coronary artery. Therefore, we implanted

Fig 1. Magnetic resonance angiograms showing (A, B) moyamoya vessels (arrows) and (A) bilateral occluded internal carotid arteries in the supraclinoid portion (arrowheads).
two bare-metal stents on the orifice of the right coronary artery (Fig 4). After the removal of CPB (pump time, 47 minutes) with stable hemodynamics, a transesophageal echocardiogram showed that there was no paravalvular leakage and that the mean transvalvular gradient was 8 mm Hg. After the operation, neurologic dysfunction was not observed, and the patient was extubated in the operation room. The postoperative course was uneventful, and the patient was discharged 29 days postoperatively after rehabilitation.

Comment

MMD is a rare disease first described by Suzuki and Takaku in 1969 [1], which mainly affects individuals of Asian descent. It is characterized by bilateral intracranial occlusion of the internal carotid arteries, and the anterior and middle cerebral arteries. MMD is a relatively common cause of recurrent, transient neurologic ischemic events, small strokes, and intracerebral hemorrhage. There are some reports of patients with MMD complicated with cardiac disease [2–6].
Cardiovascular surgical procedures with CPB have been reported to constitute an especially high risk for patients with MMD because of decreasing cerebral perfusion pressure owing to the perfusion pressure variability in the initial stages of CPB and nonpulsatile (continuous) flow [2–5]. Therefore, it is a challenging operation and requires strict intraoperative management, including the maintenance of blood pressure, hematocrit, mean arterial pressure, and body temperature in patients with MMD [2–5]. Furthermore, to maintain pulse pressure, the use of an intraaortic balloon pump in coronary artery bypass grafting was reported [6].

TAVR is a therapy developed for severe aortic stenosis in high-surgical risk patients [7, 8]. Surgeons must keep in mind that major complications can occur at any step during TAVR, and we should prepare for fast cannulation to perform CPB. Particularly in patients with MMD, we routinely secure not only one artery and one vein for immediate CPB but also one additional artery for IABP if nonpulsatile (continuous) flow occurs because of intraoperative complications. We performed TAVR in 3 patients with aortic valve stenosis complicated with MMD and did not observe additional neurologic events. Although CPB was not required in the other 2 patients, in this case, unfortunately, the hemodynamics collapsed after balloon valvuloplasty, and CPB was required immediately. Still, IABP was not necessary because the hemodynamics rapidly improved as soon as CPB was started, and the patient’s pulse pressure was well preserved during the CPB support.

The use of CPB and cardiac arrest for TAVR is needed a significant minority of cases, and therefore, this procedure may be a useful treatment for patients with aortic valve stenosis complicated with severe cerebral ischemia, including Moyamoya disease.

Conclusion

We successfully performed TAVR in patients who had severe aortic valve stenosis complicated with Moyamoya disease. TAVR is a very useful treatment for patients with aortic valve stenosis complicated with Moyamoya disease.

References


Primary Modified Bentall’s Procedure in a Case of Laubry-Pezzi Syndrome

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Modified Bentall’s procedure done as part of the primary repair in Laubry-Pezzi syndrome is very rarely described in the literature. We present a case of a 33-year-old man with a subpulmonic ventricular septal defect, aneurysmal dilatation of the aortic root and ascending aorta, with an associated patent ductus arteriosus, corrected by the incorporation of Yacoub’s technique for ventricular septal defect closure with a modified Bentall’s procedure and transpulmonary patent ductus arteriosus ligation. The postoperative course was unremarkable. Early follow-up reports show good biventricular function without residual ventricular septal defect or iatrogenic ventricular outflow tract obstructions.


Charles Laubry and Cesare Pezzi described the clinical features of Laubry-Pezzi syndrome in 1921 in a patient with ventricular septal defect (VSD) with aortic regurgitation caused by aortic valve prolapse [1]. The syndrome may eventually lead to dilatation of the aortic sinus related to the involved leaflet, resulting from a lack of continuity of the aortic media and annulus [2, 3].

It may also be possible that, as in the case of various other pathologic conditions like Marfan’s syndrome and bicuspid aortic valve, the ascending aorta may also be involved structurally in the natural history of the disease [2]. However, it is very rare for these patients to progress to aneurysmal dilatation of the aortic root and ascending aorta at the time of a first operation.

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