

Severe Aortic Stenosis and Left Main Disease in a 20 Years Old Girl: A Case Report

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Abstract

A young 20 years old female suffering from familial hypercholesteremia presented with severe aortic stenosis and left main coronary artery disease. Her complete workup was done which showed an extremely small aortic annulus of 14mm and severe left main disease with ostial disease of obtuse marginal branch of circumflex artery. There is no medical treatment available and complex surgery is required in such a case. Her surgery was planned and labeled as high-risk surgery in the face of limited facilities for dealing with such pathology. CABG with 2 grafts and aortic valve replacement with aortic root extension was taken for the patient. This is one of the rarest case presentations with successful surgical management described in this case report after obtaining informed written consent from patient.

Key words: Aortic stenosis, left main disease, familial hypercholesteremia, CABG, AVR, Aortic root extension.

Introduction

Familial hypercholesteremia is an autosomal dominant disease with a high incidence of coronary artery disease and premature death. The disease is attributed to mutations in low-density lipoprotein gene receptor (LDLR) located on chromosome 19.¹ Xanthomas are visible on the tendons of hands and feet. LDL-C levels of more than 500 mg/dl are associated with an early presentation of coronary artery disease and calcified aortic valve disease.²

Case Report

A 20 years old afghani girl presented with severe shortness of breath NYHA-3. She was a diagnosed case of familial hyperlipidemia (Figure-1a, 1b). Echocardiographic findings showed severe aortic stenosis with an effective aortic orifice area of 0.5cm², MPG of 50 mmHg and, PPG of 85mmHg. Aortic annulus of 14mm, aortic sinus of 15mm, STJ = 16mm, ascending aorta = 15mm, aortic arch = 17mm, and descending aorta of 14mm. Ejection

fraction of 60% and TAPSE of 18mm. Left heart catheterization was done which showed significant left main stenosis and stenosis at the origin of OM 1 artery (Figure-1c). CT scan of the patient was done which showed the annulus of aorta to be 14mm (Figure-1d).



a: Xanthomas on feet.

b: Xanthoma on an elbow.



c: RAO view showing significant LM disease and ostial OM1 disease.

d: A small aortic sinus with a left main arising from left aortic sinus.

Figure 1

The case was discussed in the heart valve team at NICVD Karachi and in the high-risk meeting team comprised of surgeons, anesthetists, cardiologists, and intensivists. It was decided to proceed with the case. The minimum size of aortic valve available was Medtronic 16mm. The surgery

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decided for the patient was CABG + AVR + Aortic Root Extension ± Aortic Root Replacement.

The case was processed through standard sternotomy, LIMA was harvested and vein conduit was harvested from left leg through minimal incision to keep the cosmesis keeping in view the young age of the female patient. A plaque-free zone was identified in aorta for the cannulation. Aortocaval cannulation was done and temperature drifted to 32 degrees. A vent was placed in the left ventricle. Antegrade cardioplegia was given after applying the aortic cross-clamp. The vein conduit was grafted to OM. Aortotomy was done which showed severe aortic stenosis and poor quality of aorta with numerous atherosclerotic plaques deposited in aorta. There was a supra-annular ridge which was excised first then the valve was excised. Different valve sizars were inserted to measure the annulus size, 16mm was the last size available and was not passing through the annulus (Figure-2). The valve-conduit available to proceed with the root replacement was 21mm valve so the option for root replacement was dismissed as 21mm valve could not be seated on a 14mm annulus. Therefore, a decision for root extension was taken. Nick's procedure for aortic root extension was taken and we were able to get 2.5 mm through a bovine pericardial patch used for the extension. A 16 mm mechanical valve was placed. During this time, the cardioplegia was given through coronary Ostia. The Aortotomy was closed. There was a diffuse atherosclerotic disease in the proximal LAD so distal target was found to graft LIMA to the LAD. A hotshot was given and aortic clamp was removed. Per-operative TEE was done which showed normal valve mobility with minimal transvalvular gradient and no paravalvular leak. The patient came off CPB smoothly. Hemostasis was secured and fresh frozen plasma was transfused to achieve hemostasis. Both the pleura were open so we put right and left pleural drains, one mediastinal drain, and one redivac drain. The patient was shifted to ICU.

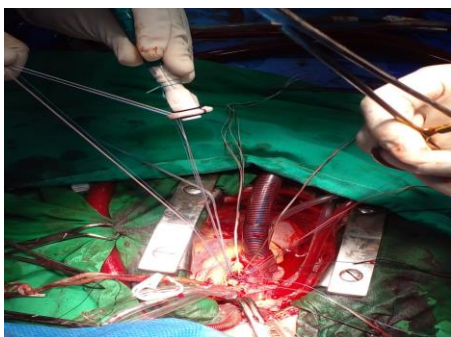


Figure 2: A 16 mm valve is anchored to the aortic annulus with sutures.

The patient got extubated early morning the next day. Post-operative Transthoracic echo was done which showed MPG of 12mmhg and PPG of 25mmhg. The patient had an uneventful recovery and was sent home on 5th POD day.

The ethical approval was obtained from the Ethical Review Committee of National Institute of Cardiovascular Diseases, Karachi vide reference No. ERC-55/2022

Discussion

It was a very challenging case to do in a limited resource where a small valve conduit for root replacement was not available. The operation needed a longer bypass time and cross-clamp time. The case had to be operated urgently as severe LM stenosis and severe aortic valve stenosis was the worst possible combination for a patient to have to get a sudden death. The timely operation has helped the patient to live a symptom-free life.

A similar case of familial hypercholesterolemia in a 17 years old boy who presented with cardiogenic shock and had severe aortic stenosis has been reported but no management has been discussed.³ The management of another case is reported in literature where the age of the patient was 70 years with severe aortic stenosis and left main disease. He was treated by transcatheter aortic valve placement and simultaneous rotablation of the left main stem.⁴

Severe symptomatic aortic stenosis needs aortic valve replacement as class 1 recommendation according to ACC guidelines⁵ and similarly, significant left main stenosis is classified as a class 1 recommendation for CABG.⁶ So, patient underwent both the procedures and post-operative strict medical follow-up is kept for the patient.

Conflict of interest: None declared.

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