

Ostial Coronary Stenosis and Severe Aortic Stenosis in a Patient With Familial Hypercholesterolemia

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A 33-year-old female presented with the typical angina symptoms of dyspnea on exertion. Five years prior, the patient had been diagnosed with familial hypercholesterolemia, as had her living siblings (a brother and a sister). The patient's family history included 2 brothers and 2 sisters that died in childhood due to unknown etiology. Although she was being treated with 80 mg atorvastatin and 10 mg ezetimibe, her total cholesterol level was 607 mg/dL, her low density lipoprotein (LDL) level was 555 mg/dL, her high density lipoprotein (HDL) level was 26 mg/dL, and her triglyceride level was 126 mg/dL. Monthly LDL apheresis was started as an adjunct to drug therapy.¹⁾ On physical examination, she had xanthelasma and plantar xanthomas. Cardiac examination revealed sinus tachycardia of 120 bpm, blood pressure of 110/75 mm Hg, and grade 3/6 systolic ejection murmur that was most prominent over the second intercostal space on the right sternal border. Electrocardiography showed ST depressions in the inferolateral derivations. Transthoracic echocardiography revealed normal left ventricular systolic function, mild hypertrophy of the left ventricle, and calcified aortic stenosis with a peak gradient of 95 mm Hg and a mean gradient of 62 mm Hg (Figs. 1 and 2). Fluoroscopic examination revealed severe calcification of the aortic root (Fig. 3). Coronary angiography demonstrated a 90% narrowing in the ostial part of the left main coronary artery (Fig. 4,

arrows) and 80% in the proximal right coronary artery (Fig. 5, arrow). The patient was referred for surgery.²⁾ Intraoperatively, extensive calcifications of the whole aortic root (porcelain aorta) required a modified Bentall procedure with a mechanical valve conduit.³⁾ The left anterior descending artery was bypassed using the left internal mammary artery, the right coronary artery using the right internal mammary artery, and both diagonal and OM using sequential saphenous veins. Despite inotropic and intraaortic balloon pump support, the patient died during the early postoperative period due to prolonged cardiopulmonary bypass and heart failure.

References

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• The authors have no financial conflicts of interest.

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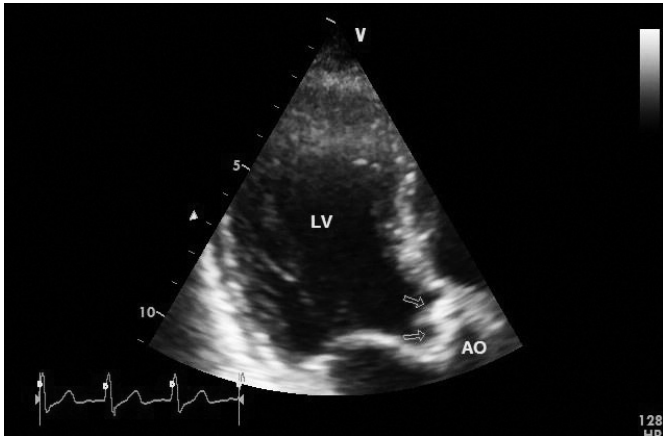


Fig. 1.

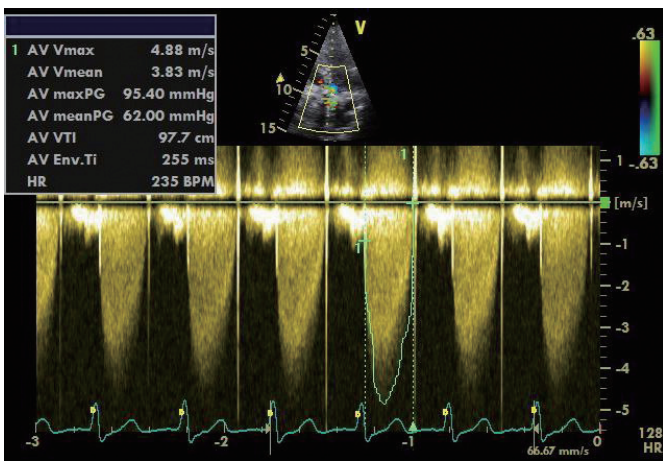


Fig. 2.

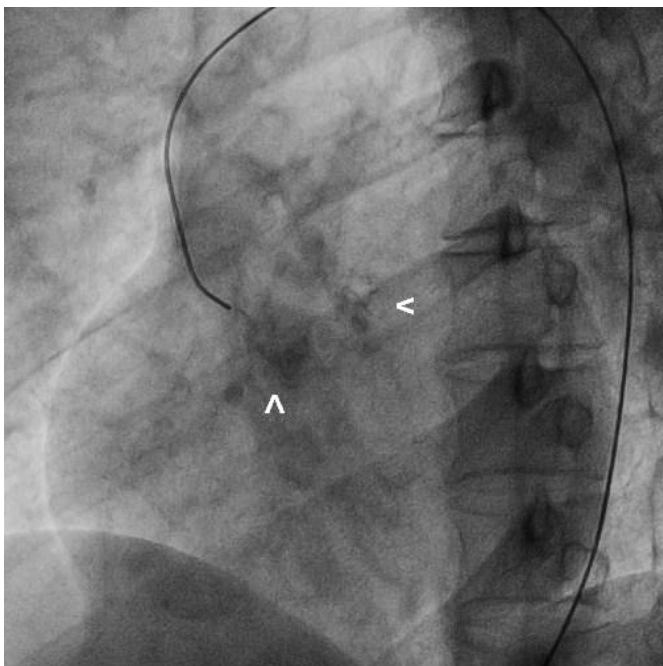


Fig. 3.

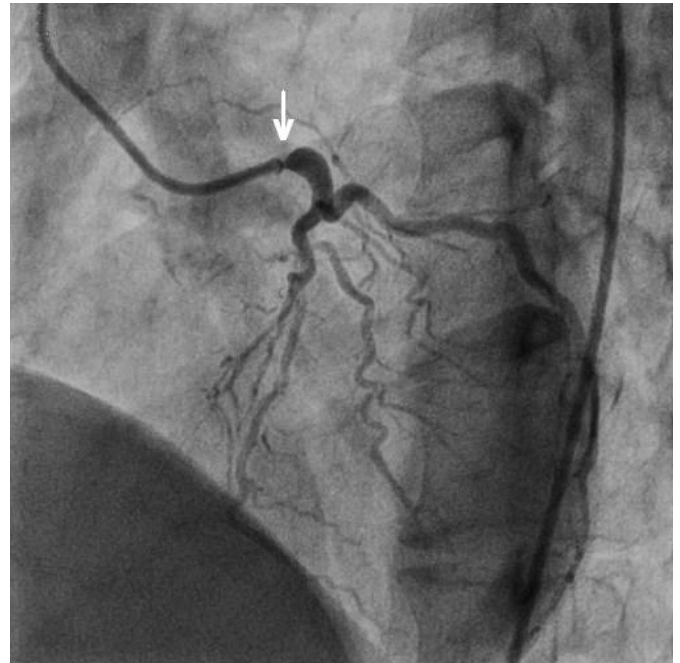


Fig. 4.

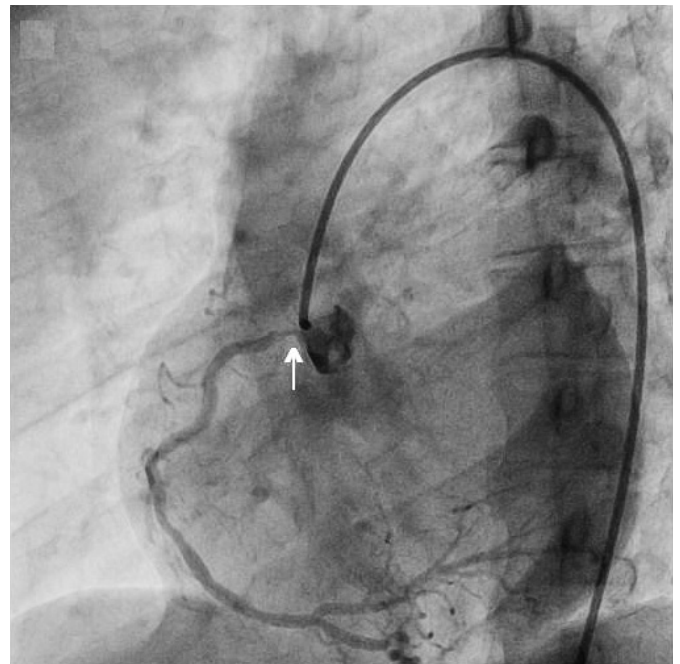


Fig. 5.