

# Supravalvular aortic membrane-like stenosis: A case report

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## Abstract

Supravalvular aortic stenosis is a type of congenital heart disease in which membrane-like stenosis rarely occurs. Herein, we present the case of a 52-year-old female patient who was admitted to the hospital because of palpitation. Two-dimensional echocardiography and real-time three-dimensional echocardiography showed supravalvular membrane-like stenosis with aortic insufficiency. The patient underwent surgery, with the ascending aortic fiber stenosis ring removed intraoperatively. She was followed up closely after discharge. [If there is no word count limitation, please provide the implication of this case.]

## Supravalvular aortic membrane-like stenosis: A case report

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## Author contributions:

Drafting article and interpretation of echocardiography by Jie Zhao and Shasha Duan; data collection by YiLu Shi, WuPing Xiao and Min Zhao; interpretation of CT by Zhao Jin; critical revision of article by Yaxi Wang and Xiao shan Zhang.

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## Abstract

Supravalvular aortic stenosis is a type of congenital heart disease in which membrane-like stenosis rarely occurs. Herein, we present the case of a 52-year-old female patient who was admitted to the hospital because of palpitation. Two-dimensional echocardiography and real-time three-dimensional echocardiography showed supravalvular membrane-like stenosis with aortic insufficiency. The patient underwent surgery, with the ascending aortic fiber stenosis ring removed intraoperatively, mechanical aortic valve replacement, and ascending aortic plasty (aortic ring enlargement). She was followed up closely after discharge. She is now in good condition and totally asymptomatic.

## Keywords:

Supravalvular aortic stenosis; two-dimensional echocardiography;;real-time three-dimensional echocardiography

## CASE PRESENTATION

A 52-year-old female patient was admitted to the hospital because of palpitation and discomfort after an activity for 10 years and aggravation for half a year. On physical examination, her blood pressure was 130/83 mmHg, there was no protrusion in the precordial area, there were no palpable tremors and pericardial friction, and the heart rate was 65 beats per minute. Low heart sounds, irregular rhythms, and diastolic sigh-like murmur can be heard over the auscultation area of the aortic valve. Electrocardiography showed occasional premature ventricular contractions and an abnormal ST segment. Echocardiography was performed to determine the source of the murmurs. Two-dimensional transthoracic echocardiography showed that the inner diameter of the aorta was widened to 40 mm, and a linear strong echo protruding from the anterior and posterior walls of the sinus junction into the lumen was seen at 10 mm above the aortic valve. The membranous structure causes narrowing of the lumen. On continuous Doppler measurements, the velocity of the aortic valve increased to 354 cm/s, and the maximum pressure difference was 50 mmHg. Color Doppler flow imaging showed mild aortic regurgitation. Therefore, transthoracic echocardiography primarily diagnosed supravalvular aortic membrane-like stenosis. Then, real-time three-dimensional echocardiography was performed to clearly show the anatomical structure of the membrane-like tissue. Annular patchy tissue was then found above the aortic valve. Moreover, auxiliary examinations were performed to further clarify the anatomy of the aorta. Chest plain computed tomography showed dilation of the ascending aorta, and the inner diameter of the aorta was widened to 37 mm. However, it failed to show the membranous structure above the aortic valve. Coronary angiography was performed to determine the structure and origin of the coronary arteries. The results revealed that the left and right coronary arteries were of normal origin, and no obvious stenosis or dilation was observed. Thus, the patient was diagnosed with supravalvular aortic stenosis, mild aortic regurgitation, and widened ascending aorta. The cardiac surgeon indicated the patient for surgery. With the consent of the patient and her family, she received surgical treatment. During surgery, the doctor found that the fibrous stenosis ring in the aortic wall was located 5-10 mm above the left and right coronary artery opening. In addition, the aortic valve junction was adherent, and the valve leaflet was thickened and partially calcified. The surgical method was expanded to mechanical aortic valve replacement, supra-aortic septectomy, and ascending aortic plasty (aortic ring enlargement). Finally, the supravalvular membranous structure and aortic valve were removed.

Pathological examination was performed. Pathology revealed mucoid, hyaline, fibrosis, and calcification within the aortic valve tissue. One week after the operation, echocardiography showed that the mechanical valve ring of the aortic valve was fixed, the function of the mechanical valve was good, no obvious perivalvular leakage was found, the ascending aorta stenosis disappeared, and the blood flow was unobstructed. Half a year after discharge, the patient was in good health.

The patient signed the consent form, and the report was approved for publication by the ethics committee.

## Discussion

Congenital supra-ventricular aortic stenosis is a rare disease of the left ventricular outflow tract, which is caused by partial loss of chromosomes and abnormal arterial elastin<sup>1</sup>. Because the stenosis is located above the aortic valve and the opening of the coronary artery, the pressure of the proximal vessel is increased; therefore, supra-ventricular stenosis is often accompanied by coronary artery dilatation, stenosis at the opening of the coronary artery, aortic insufficiency, and widened aortic sinus<sup>2</sup>. Once found, surgical treatment is often needed to relieve the obstruction and restore the normal structure and function of the aortic valve<sup>3-5</sup>. According to morphology, supra-ventricular aortic stenosis can be divided into three types: hourglass type, ascending aorta diffuse dysplasia type, and intersaepum type, of which the intersaepum type is the least common<sup>6</sup>.

Echocardiography is widely used in congenital heart disease because of its convenience, non-invasiveness, non-radiation requirement, and real-time display of cardiac cavity structure. Echocardiography can be used not only to diagnose supra-aortic stenosis but also to evaluate the surgical effect and dynamic follow-up changes. The sonographic manifestations of membranous stenosis are linear strong echoes protruding from the anterior and posterior walls of the sinus junction into the lumen. Color Doppler imaging shows colorful turbulence signals in the stenosis area and at its distal end, and the blood flow velocity at the stenosis site measured by spectral Doppler can evaluate the degree of stenosis. Real-time three-dimensional echocardiography can display the overall shape of the intersaepum and clearly display a pore-like structure in the middle of the intersaepum. At the same time, it can display the dynamic changes of the intersaepum with the cardiac cycle and its relationship with the aorta and aortic valve to determine whether there is stenosis and prolapse and to provide accurate information for surgical treatment<sup>7,8</sup>. The ultrasound findings of this patient were consistent with the intraoperative findings. After treatment, the patient's clinical symptoms significantly improved. Half a year after discharge, the patient was in good health.

## CONFLICT OF INTEREST:

All of the authors have no conflict of interest.

## DATA SHARING:

No additional data.

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**Figure 1** Transthoracic echocardiography shows the supra-avalvular aortic membrane-like stenosis (panel A and B, arrow). Continuous wave Doppler (CW) shows the velocity of aortic valve is increased.

LA=left atrium; LV=left ventricle; RA=right atrium; RV=right ventricle; AV=aortic valve.

**Figure 2** Real time three-dimensional echocardiography shows the supra-avalvular aortic membrane-like stenosis (panel D, arrow). Coronary angiography showed coronary artery initiation and morphology were normal (panel E). Operative finds fibrous stenosis ring in the aortic wall (panel F, arrow)

