

# Accessory mitral valve tissue causing severe left ventricular outflow tract obstruction in a post-Senning patient with transposition of the great arteries

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## Abstract

Accessory mitral valve tissue is a rare congenital anomaly associated with congenital cardiac defects and is usually detected in the first decade of life. We describe the case of an 18-year old post-Senning asymptomatic patient who was found to have accessory mitral valve tissue on transthoracic echocardiography producing severe left ventricular outflow tract obstruction.

## Introduction

Accessory mitral valve tissue (AMVT) has an incidence of 1 per 26,000 echocardiograms in adults.<sup>1</sup> It is generally found in association with congenital anomalies of the heart or great vessels, such as ventricular septal defect, patent ductus arteriosus, or transposi-

tion of the great arteries (TGA).<sup>1-4</sup> Patients with isolated AMVT usually present with symptoms during the first decade of life depending on the severity of left ventricular outflow tract (LVOT) obstruction.<sup>2</sup> Embryologically, AMVT is due to incomplete separation of the mitral valve from the endocardial cushion.<sup>1,5</sup> The accessory tissue is usually attached to the ventricular aspect of anterior mitral valve leaflet, the chordae of the mitral valve or to an accessory papillary muscle.<sup>1-3</sup> We describe the case of an 18-year old post-Senning asymptomatic patient who was found to have AMVT on transthoracic echocardiography producing severe LVOT obstruction.

## Case Report

An 18-year old male patient was referred to our clinic for murmur evaluation. The patient was known to suffer from TGA and had undergone a Senning operation in the UK in 1992 when he was six months old. Since then he had been asymptomatic, in good health with no medical follow up. Cardiovascular examination revealed a harsh 4/6 ejection systolic murmur radiating widely over the precordium as well as to the patient's neck and back. ECG demonstrated sinus tachycardia with biventricular hypertrophy. The transthoracic echocardiography showed situs solitus, D-TGA with pulmonary vein baffle to right atrium, and the systemic vein baffle to left atrium demonstrating normal flow and no stenosis or leak. The left ventricle was small with an intact interventricular septum bulging into the left ventricle, but with good systolic function. The right ventricle was dilated with moderate right ventricular systolic dysfunction. There was no asymmetric septal hypertrophy or systolic anterior motion of mitral valve or presence of any subaortic membrane. There was a well defined mobile leaflet-like structure measuring 1.3×0.5 cm, prolapsing in systole into the LVOT (Figure 1A and B, arrowheads). On color Doppler examination, there was

severe turbulence in the LVOT (Figure 2A) with LVOT peak gradient of 79 mmHg and a mean gradient of 49 mmHg (Figure 2B). On careful examination, the mobile mass represented an AMVT which was seen to adhere to the ventricular side of the anterior mitral valve leaflet (Figure 1C, arrowheads, Movie 1) and prolapsing into the LVOT during systole. There was no aortic regurgitation. There was grade I left ventricular diastolic dysfunction. There had been no previous history of prolonged fever or hospitalization since his surgery. In view of severe LVOT obstruction, he was advised surgery but he and his family preferred a medical follow up. He is being followed up on aspirin prophylaxis.

## Discussion

Accessory mitral valve tissue can be asymptomatic with presence of a murmur or it can present with mild to severe LVOT obstruction causing exercise intolerance, chest pain, or syncope on exertion, as well as recurrent transient ischemic attack or stroke.<sup>1-3</sup> In our case there was severe LVOT obstruction but with no symptoms. AMVT is classified as: Type I (fixed: A = nodular, B = membranous), and type II (mobile: A = pedunculated, B = leaflet-like). Type IIB was further subdivided as rudimentary chordae and developed chordae.<sup>2,3</sup> In this patient, AMVT was of type IIB with rudimentary chordae. The mobile type is a parachute-like leaflet floating in the LVOT with or without obstruction and is usually thickened or rarely dysplastic. The fixed type is attached to the interventricular septum by short chordae and may reduce the size of a ventricular septal defect.<sup>4</sup> LVOT obstruction may be due to mass effect of the accessory tissue or due to progressive deposition of fibrous tissues within the LVOT, secondary to the turbulent flow leading to fibro-muscular hypertrophy.<sup>5</sup> In the majority of cases there is severe LVOT obstruction, with a median LVOT gradient of more than 50 mmHg.<sup>2,6</sup> Other types of left ventricu-

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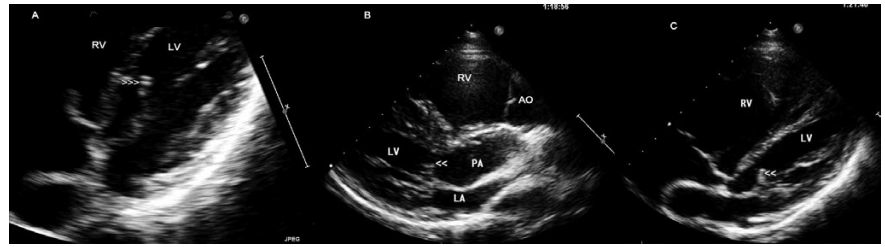
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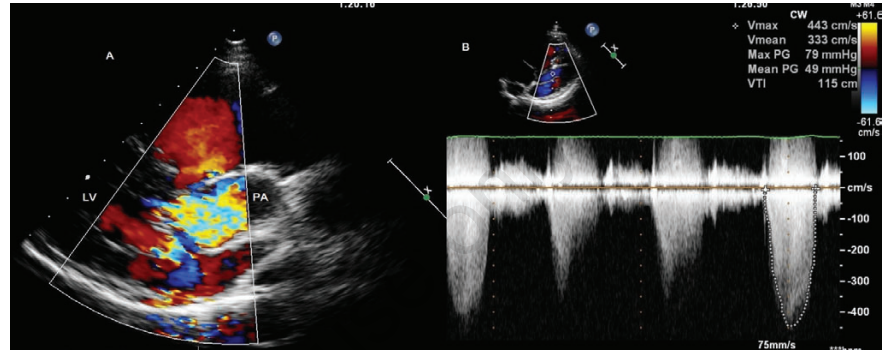
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lar mass, like tumors or vegetations, can produce similar findings on echocardiography. However, these lesions more often originate from cardiac muscle (tumors), or build-up on the low-pressure side of a heart valve (vegetations),<sup>3</sup> and do not have a leaflet-like appearance. Subaortic membrane has a different appearance of a single, linear membrane in the LVOT.

Surgical excision of the AMVT is required while repairing other congenital defects or if significant LVOT obstruction is present.<sup>2,7</sup> In patients with TGA, either pre- or post-operative LVOT obstruction commonly occurs due to bicuspid or unicommissural pulmonary valve, fibrous ridge, obstructive muscular conus, malaligned outlet septum, bulging muscular septum, straddling mitral valve, anterior mitral valve rotation and AMVT.<sup>8</sup> Table 1 shows the common types of obstruction with management options in patients with TGA.<sup>9,10</sup> Yoshimura *et al.* reported 2 cases of AMVT associated with TGA. In one of these, release of the LVOT obstruction due to AMVT was necessary to perform the arterial switch operation.<sup>4</sup> In the second patient, it was not possible to perform the arterial switch operation because the AMVT could not be resected completely through the pulmonary artery. It was felt that there is a high chance of overlooking the AMVT intra-operatively, especially if no pre-operative diagnosis is made. In their series of



**Figure 1.** Transthoracic echocardiogram in different views showing accessory mitral valve tissue attached to ventricular aspect of anterior mitral valve leaflet prolapsing into left ventricular outflow tract in systole (A, B, C arrowheads) in a post-Senning surgery patient.



**Figure 2.** (A) Color Doppler echocardiogram showing severe turbulence in the left ventricular outflow tract. (B) Continuous wave Doppler examination showing severe left ventricular outflow tract obstruction with peak gradient of 79 mmHg and a mean gradient of 49 mmHg, in a patient with accessory mitral valve tissue.

**Table 1. Possible causes of LVOT obstruction and their management in TGA.**

Type of LVOT obstruction	Associated lesions	Management
Dynamic (septal shift due to pLV/pRV < 1)	IVS/VSD (high pulmonary blood flow, SAM, tricuspid tissue hernia may contribute to gradient)	ASO usually relieves gradient by reversing pLV/pRV.
Pulmonary valve abnormalities (unequal cusp sizes, bicuspid, dysplasia, annular hypoplasia commissural fusion)	IVS VSD	Rare in isolation, usually part of complex LVOT obstruction in TGA with VSD. Not always obstructive in isolation. ASO may result in neo-aortic leak or root dilatation. Intraventricular rerouting (Rastelli or REV) is the method of choice in the majority of patients.
Subvalvar fibrous membrane	IVS VSD	Similar to subaortic membrane in concordant hearts. Usually resectable. ASO + resection.
Subvalvar fibromuscular tunnel	IVS VSD	Often associated with hypoplastic pulmonary valve. Difficult to relieve by resection. Rastelli/REV, Nikaidoh-Bex (aortic translocation), ASO, all options possible.
Septal malalignment (deviation of outlet septum into LVOT)	VSD	Difficult to relieve by resection. Nikaidoh-Bex or REV good option.
Accessory tricuspid valve tissue	VSD	Prolapses through VSD to cause LVOT obstruction. Usually resectable. ASO+resection.
Accessory mitral valve tissue	IVS VSD	Prolapses into LVOT. Usually resectable. ASO+ Resection.
Anomalous insertion of mitral valve chordae or papillary muscle to outlet septum, straddling tricuspid valve	VSD	Difficult to relieve by resection, even if septation is otherwise possible. Various techniques such as Nikaidoh-Bex may be necessary or single ventricle approach if severe mitral valve abnormalities.
Accessory endocardial cushion tissue relating to a VSD	VSD	Prolapses into LVOT. Usually resectable. ASO+ resection

ASO, arterial switch operation; IVS, intact interventricular septum; SAM, systolic anterior motion of mitral valve; LV, left ventricle; LVOT, left ventricular outflow tract; pLV/ pRV, LV to right ventricle pressure gradient; VSD, ventricular septal defect; REV procedure, RepARATION A L'etage Ventriculaire.

TGA patients, Hazekamp *et al.*<sup>8</sup> observed that mitral valve anomalies prevented LVOT obstruction relief, permitting either Senning or univentricular palliation. The possible reasons in this patient may be either the AMVT may have been overlooked or prevented LVOT obstruction relief, or it did not produce significant obstruction at that time. The reason for such severe LVOT gradient now may be progressive thickening of the tissue over the years or development of fibro-muscular hypertrophy of LVOT, as noted by some authors.<sup>4,5</sup> At the current time, even though transthoracic echocardiography is sufficient to visualize AMVT, addition of transesophageal echocardiography and three-dimensional echocardiography may further delineate the structure precisely. Due to their susceptibility to neurological events, patients with AMVT are advised to take aspirin prophylaxis.<sup>7</sup> In conclusion, AMVT should be considered in the differential diagnosis of LVOT obstruction, specifically in pre- and post-operative congenital heart disease patients.

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