

## BRIEF REPORT

# Complex morphology of the abnormal muscle bundle in double-chambered right ventricle

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### ABSTRACT

Double-chambered right ventricle is a rare congenital heart disorder involving two different right ventricle pressure compartments that is often associated with malalignment ventricular septal defect. Usually, the obstruction is caused by an anomalous muscle bundle crossing the right ventricle from the interventricular septum to the right ventricle free wall.

A 18-year-old man underwent cardiac surgery procedure for right ventricular outflow tract obstruction caused by anomalous muscle bundles. After the surgical procedure, patient remains free of symptoms and medications.

We report our surgical experience in a case of complex morphology of the abnormal muscle bundle in double-chambered right ventricle.

**Key words:** right ventricle, ventricular septal defect, anomalous muscle band.

Double chambered right ventricle is a rare form of congenital heart disease in which the right ventricle is divided into a high pressure inlet portion and a low pressure outlet portion by an anomalous muscle bundle.

The anomalous muscle band divides the right ventricle in two cavities causing variable degree of obstruction. The anomalous muscle band may be located either high, adjacent to the pulmonary valve, or low, near the apex.<sup>1</sup>

Double-chambered right ventricle is described to be associated with different congenital disorders, most commonly with a membranous or malalignment type ventricular septal defect. The flow abnormalities related to these disorders are considered to be involved in the postnatal development of the proliferation of the muscle bundle.

So a progressive obstruction of the right ventricular outflow tract probably is influenced by the hemodynamic effects of the ventricular septal defect.<sup>2</sup>

A 18-year-old man was referred complaining of mild exertional dyspnea.

Chest X-ray showed significant cardiomegaly. There was not pulmonary congestion (Fig. 1).

A harsh cardiac murmur was heard at the left external border and an echocardiographic examination have shown a 17 millimeter ventricular septal defect associated to a slow velocity flow from left to right ventricle. Aortic root was slightly deviated to the right. In addition there was right ventricular hypertrophy and a muscular septation inside this cavity causing

obstruction with a peak gradient of 60 mmHg. The anomalous muscular bundle divided the right ventricle into two different compartments, causing right ventricular outflow tract obstruction, extremely severe in the systole.



Figure 1, Chest X-ray revealed cardiomegaly.

Surgery was undertaken using standard cardiopulmonary bypass with moderate hypothermia.

The external aspect of the heart revealed an hypertrophied hypertrophied right ventricle with a well-defined accessory chamber (Fig. 2).

A right atrial approach was not enough for the complete resection of the hypertrophied muscle bundles that obstructed the mid ventricle. The obstructions were severe and too bulky to be excised via a right atriotomy.

A small right ventriculotomy revealed a fibromuscular band below the infundibulum that obstructed the right ventricle and a well-formed chamber connected to the right ventricle cavity with a small opening, just below the normal pulmonary valve.

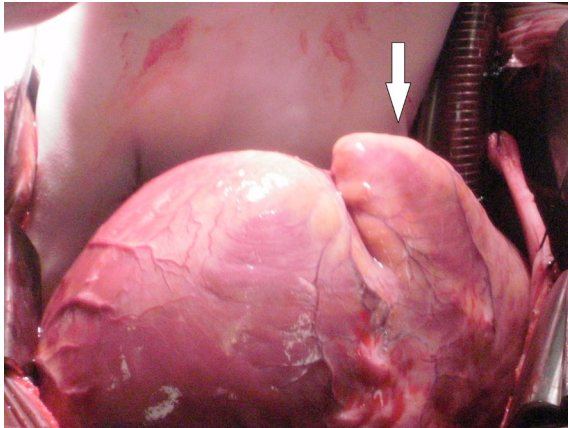
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The abnormal muscle bundle was fibromuscular, discrete, high, and horizontal. The muscular shelf originates from the body of the septomarginal trabeculation.



**Figure 2**, External aspect of the hypertrophied right ventricle with a well-formed accessory chamber.

The obstruction between the right ventricle inlet and the infundibulum had been caused by hypertrophied trabeculae.

The fibrous membrane and the surrounding hypertrophied muscle were excised. The ventricular septal defect was closed with heterologous pericardial patch. The ventriculotomy was closed by patch.

After the procedure, patient remains free of symptoms and medications.

Right ventricular outflow tract obstruction caused by anomalous muscle bundles may be an acquired phenomenon in some patients with ventricular septal defect and valvular pulmonary stenosis and that obstruction may progress with time.<sup>3</sup>

Localisation of the obstructive muscle varies and may be either high (or horizontal), adjacent to the pulmonary valve, or low (or oblique), close to the apex. It may also develop over time as an acquired lesion in patients with an abnormally short distance between the moderator band and the pulmonary valve.

The abnormal muscle bundle is more often local-

ised low than high within the right ventricle. High localisation of the abnormal bundle is invariably associated with discrete morphology, whereas a low obstruction may be either discrete or diffuse, the latter being less common.

The increased blood flow and pressure within the right ventricular outflow tract may act as an initial stimulus for hypertrophy of the crista supraventricularis in patients with ventricular septal defect

and increased pulmonary blood flow. The resulting obstruction usually deteriorates, with subsequent hypertrophy of the muscle and further narrowing of the right ventricular cavity.

So the obstruction is the result of an acquired process whereby increased blood flow among patients with ventricular septal defect causes hypertrophy of the supraventricular crest.

All symptomatic adult patients should undergo surgery to resect the obstructive muscle bundles and to repair any associated lesions. Because the rapid progression of right ventricle outflow tract also asymptomatic patients whose gradients exceed 40 mmHg should undergo surgery.<sup>4</sup>

Surgical treatment of a double-chambered right ventricle carries a low risk and provides excellent hemodynamic and functional results over both the short and long term.

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