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Echocardiographic Findings in Congenital Mitral Ring: A Report of Three Cases

Hakim Kaouthar*, Ben Othmen Rihab, Hentati Mouna, Msaad Hela and Ouarda Fatma

Pediatric Cardiology Department, La Rabta Hospital, Faculty of Medecine, Tunis Manar University, Tunisia

***Corresponding Author**: Hakim Kaouthar, Pediatric Cardiology Department, La Rabta Hospital, Faculty of Medecine, Tunis Manar University, Tunisia.

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Abstract

Mitral ring, also known as membranous supravalvular mitral stenosis is a rare congenital heart defect of surgical importance. It may be encountered in isolation or more often in association with other cardiac anomalies. Transthoracic echocardiography is the gold standard to define the anatomy of the ring and mitral valve, to assess the severity of obstruction, and to identify any associated defect. The purpose of this article is to present 3 pediatric cases with Supramitral ring (SMR) and to focus on the value of transthoracic echocardiography in diagnosis and evaluation of this cardiac anomaly and eventual associated lesions. Recognition is important because surgical results are better than for other forms of congenital mitral stenosis.

Keywords: *Congenital Mitral Stenosis; Supramitral Ring; Transthoracic Echocardiography*

Introduction

Congenital mitral ring is a rare congenital cause of mitral stenosis. It was described for the first time by Fisher in 1902 [1]. Its morphology is variable from a thin membrane to a thick fibrous ridge that encircles partially or completely the mitral ring. It can be isolated [2] but it is more often associated to other cardiac lesions that should be identified.

The diagnosis of this type of congenital mitral stenosis is crucial because of the relatively good outcome with appropriate management. Transthoracic echocardiography is the most important imaging tool for the delineation of the anatomy of the ring and mitral valve, and detailed assessment of associated lesions. We report 3 cases of mitral ring with different clinical presentations and varied degrees of obstruction encountered in isolation or associated with other cardiac lesions.

Clinical Presentations

First case

A two-year-old male child, previously healthy, presented two years ago to our department for cardiac murmur with repetitive bronchiolitis. His physical examination revealed a diastolic murmur at the apex. The Electrocardiogram (ECG) showed a left atrial hypertrophy. The Chest X Ray revealed a cardiomegaly (cardiothoracic Index = 0.66) and standard biological exams were normal.

Transthoracic echocardiography examination (TTE) showed a circumferential mitral ring attached to normal mitral valve leaflets with a normal subvalvular anatomy (Figure 1). The ring caused a moderate mitral stenosis (Figure 2) with a mean transmitral gradient of 9 mm Hg (Figure 3). There were no associated lesions and the pulmonary artery was normal

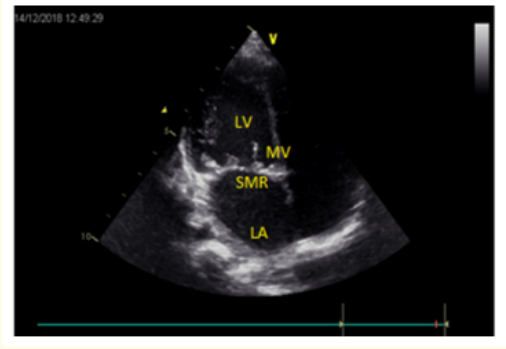

Figure 1: *2-D Apical view showing the Supramitral ring. LA: Left Atrium, LV: Left Ventricular, SMR: Supramitral ring.*

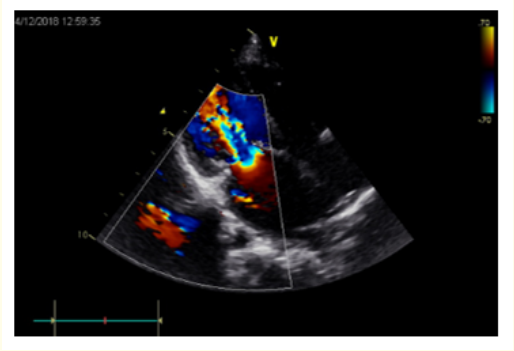

Figure 2: *2-D Color Doppler Apical view TTE view in diastole showing the obstruction caused by the SMR. MV: Mitral Valve.*

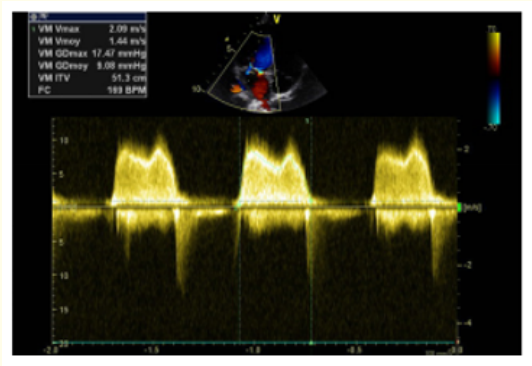

Figure 3: *Continuous Doppler quantifying the peak and mean transmitral gradients.*

We decided a close clinic and TTE follow up for the patient. On the last follow up, so 2 years later he developed pulmonary hypertension of 50 mm Hg of systolic pressure and he started to have dyspnea on exertion. The mean gradient across the mitral valve was measured at 15 mmHg. He is planned for surgical removal of the membranous mitral ring.

Second case

Another case of SMR was diagnosed in a 9 months year old female infant symptomatic of failure to thrive with severe hypotrophy.

Physical examination revealed a moderate systolic cardiac murmur with loud S2 on cardiac auscultation. The TTE showed an intermediate atrioventricular septal defect with a small ventricular septal defect but severe pulmonary hypertension. Assessment of the atrioventricular valve showed a thick mitral ring attached to the left side of the atrioventricular valve and to the superior rim od the atrial septal defect causing a significant obstruction with a mean gradient of 20 mmHg (Figure 4 and 5). There was also an abnormal subvalvular apparatus with direct insertion of a papillary muscle on the anterior bridging leaflet. The patient was recently proposed to surgery.

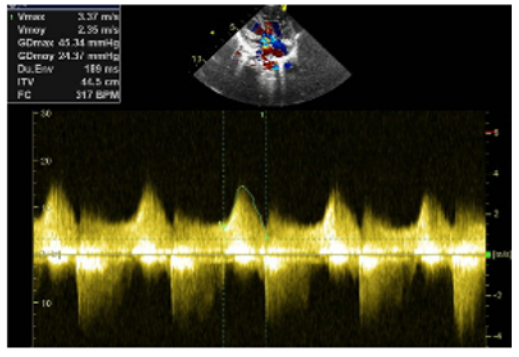

Figure 4: *Continuous doppler quantifying the peak and mean trans atrioventricular valve gradients.*

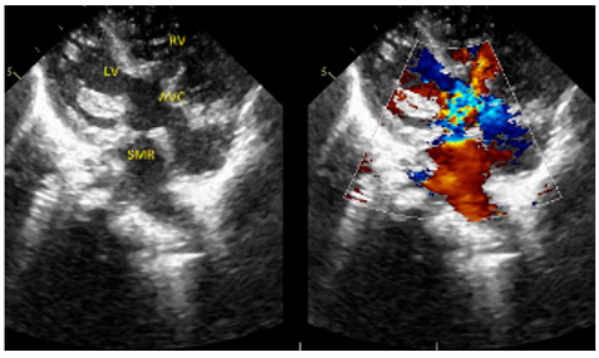

Figure 5: *2-D color doppler apical TTE view in diastole showing the obstruction caused by the SMR.*

Third case

The third case is a about a 14 years male patient who presented with dyspnea on exertion. Clinical examination revealed absent femoral pulses with a blood pressure gradient of 60mmHg between the upper and lower limbs. The TTE showed Shone's complex with

a severe congenital mitral stenosis due to a thin mitral ring (transmitral mean gradient of 36 mmHg) (Figure 6 and 7), bicuspid aortic valve and severe coarctation of the aorta(peak velocity over the isthmus of 4 m/s) (Figure 8). The patient is scheduled for aortic dilation followed by surgical relief of mitral stenosis.

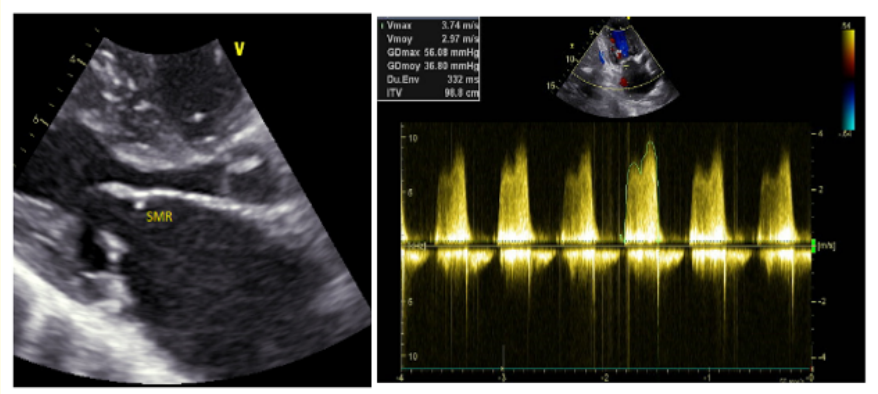

Figure 6 and 7: *2-D Long-axis view in diastole showing the SMR and Continuous Doppler quantifying the peak and mean trans mitral valve gradients.*

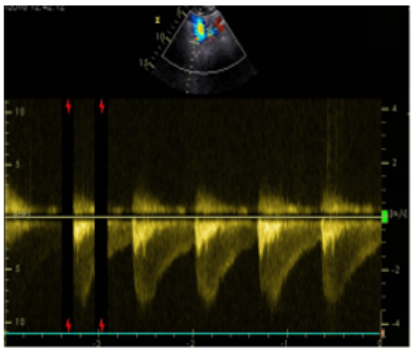

Figure 8: *Supra sternal view continuous doppler quantifying the isthmic aortic gradient.*

Discussion

In Carpentier's classification of congenital mitral stenosis [3], supramitral ring is categorized under congenital mitral stenosis associated with normal papillary muscles. It is a rare malformation: fewer than 100 cases have been reported in literature according to a review published in 2009. Another study indicated that SMR may be present in 8% of all children with congenital mitral valve disease.

Clinical presentation and age at presentation depend on the degree of obstruction, associated mitral regurgitation and associated cardiac defects including other subtypes of congenital mitral stenosis.

Transthoracic echocardiography detects the mitral ring in up to 70% of cases [4]. It is considered the best method to identify the anatomy of the congenital left ventricular inflow obstruction [5]. The limitation of this non-invasive tool is either the difficult visualization

of a thin membrane that can be smaller than 1 mm or its attachment to the mitral valve [6]. Different complementary views were required in our patients to visualize the ring, to assess its attachment to the mitral leaflets and to detect any other lesion of the mitral leaflets or subvalvular structures. Both pulsed-wave and continuous-wave Doppler ultrasound were used to assess transmitral flow signals and peak velocities of flow. The addition of Doppler color flow techniques has proved essential for defining the proximal isovelocity surface area at the point where the ring becomes obstructive. Associated defects were methodically sought.

Two echocardiographic types of mitral rings were described according to the relation with the mitral annulus [7]. The first type, called "Supravalvar ring", shows a membrane located in close proximity to the mitral valve, above the valve annulus and beneath the opening of the left atrial appendage. This type is usually associated with a normal mitral subvalvular apparatus and must be differentiated from the cortriatriatum [8]. In the latter condition, the membrane is generally proximal to the left atrial appendage, well separated from the mitral valve, and divides the left atrium into distinct proximal and distal chambers.

The second type is the "Intravalvar ring". The diagnosis of this type is more difficult because the membrane is closely adherent to the mitral leaflets [9]. It is usually associated with an abnormal subvalvular apparatus.

The three cases we reported were those of this second type but in the first one, no associated abnormalities of the mitral subvalvular apparatus were noticed.

There is no cases described in literature of mitral ring in fetus and only rare cases have been diagnosed at birth. This finding could suggest postnatal development of the ring or difficult diagnosis of a thin membrane in the fetus and neonate. The mitral ring can be complete, circumferential or partial. It creates a stenosis that is generally progressive with a median age at diagnosis of 36 months in the largest published series [9].

Increase in venocapillary pressure as well as pulmonary arterial pressure that result from left atrium obstruction explain congestive heart failure and failure to thrive in some children with severe obstruction or associated lesions, as described in the second case in this article.

Supramitral ring is commonly associated with other cardiac defects [2]. Rarely, it may be encountered in isolation, such as in the first case reported here. Lesions frequently associated to SMR are ventricular septal defects and left-sided obstructive lesions [10]. This association is illustrated by Shone's syndrome [11] that is composed of SMR, parachute mitral valve, subaortic stenosis, and coarctation of the aorta, as described in our third patient.

The addition of three-dimensional echocardiography to 2D echography improves the diagnosis and characterization of SMR. It provides better anatomic analysis of the entire mitral valve apparatus from the supra-annular region to the insertion of the papillary muscles to the myocardium [12]. This diagnostic accuracy provided by real time three-dimensional transesophageal echocardiography (RT3DTEE) aids the surgeon in successful resection of the mitral ring and repair of associated mitral valve abnormalities [13].

SMR is a congenital heart defect of surgical importance. The first surgical correction was described by Lynch and colleagues [14] in 1962, the study of 14 patients showed excellent results after the operation. In case of "supravalvular" ring, the excision of the membrane is straight forward and definitive. Whereas, it is more difficult and not always sufficient in case of "intravalvular" ring because the anatomy of subvalvular apparatus also contributes significantly to mitral stenosis. In Toscano., *et al*. study [9], the membrane did not recur after resection in supravavular type. Whereas, recurrence of intravalvular ring was documented in half of the cases. Furthermore, associated lesions can make the surgery more difficult, especially in the newborn. A key example is shone's complex, in which associated obstructive lesions at different levels and unfavorable mitral valve anatomy amplify the surgery risk. In addition, the degree of mitral stenosis can be underestimated due to the left ventricular outflow tract obstruction, which may mask the necessity of surgical intervention.

Conclusion

Congenital mitral ring can be supravalvar or intravalvar. Mitral ring develops generally in infancy and can cause rapidly progressing mitral stenosis. Transthoracic echocardiography is the gold standard for diagnosis, severity assessment and exploration of associated lesions. Accurate diagnosis is important because postoperative outcome is good and this defect may be considered as a treatable form of congenital heart disease, particularly when associated with normal underlying mitral valve anatomy.

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