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Retrospective Study

Congenital Mitral Stenosis: About 26 Cases

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Abstract

Introduction: Congenital mitral stenosis (CMR) is defined as any "obstacle to diastolic filling of the left ventricle" due to abnormal development of the structures that form the mitral apparatus (ring, immediately supravalvular area, valves, cords and pillars).

Objective: Clinical, epidemiological and prognostic aspect of congenital mitral stenosis.

Methods: We conducted a retrospective study collected in the cardiology department of CHU Ibn Rochd of Casablanca, between September 2009 and January 2018, involving 26 patients, especially by analyzing clinical, echocardiographic and evolutionary data

Results: During the study period we diagnosed 2,678 congenital heart diseases. CMS was present in 26 patients, a prevalence of 0.97%.

The study population consisted of 13 boys and 13 girls, a sex ratio of 1 and the average age was 6 years (age range was 46 days to 48 years).

Clinically; the functional symptomatology was dominated by dyspnea (17 cases), recurrent bronchitis (8 cases). The clinical examination objectified cyanosis (4 cases) and heart failure (2 cases).

Echocardiography found an isolated CMS in 3 cases; the etiologies were dominated by the supramitral ring (14 cases) and the mitral valve in parachute (8 cases).

The congenital heart disease most associated with CMS was ventricular septal defect (9 cases). The others associated with RMC were: coarctation of the aorta (6 cases), transposition of the great arteries (4 cases), pulmonary stenosis (4 cases), bicuspid aortic valve(4 cases), the double outlet right ventricle (3 cases), the left superior vena cava (3 cases), atrial septal defect (2 cases), supravalvular aortic stenosis (2 cases), ductus arteriosis (2 cases), Shone's syndrome (1 cases) and the cor triatriatum in 1 cases. Pulmonary arterial hypertension was present in 50% of cases, 2 of which were in Eisenmenger.

On the evolutionary level we excluded from the follow-up the patients carrying complex congenital heart disease where the mitral stenosis was in the second plan (single ventricle, double inlet right ventricle).

Surgery was indicated in 13 children; 4 received curative surgery; 4 are lost to follow-up; 4 patients with mitral stenosis considered to be not severe had no operative indication; they are followed regularly and one last patient awaiting surgery. Five died, one in intraoperative, 3 refusing their parents surgery in a respiratory distress table and one in out-of-hospital.

Conclusion: Congenital mitral stenosis is a rare heart disease. The anatomical lesions are often complex at the level of the ring, the area immediately above the valve, the valves, the ropes and the pillars. The supramitral ring remains a rare variety of RMC which was the predominant in our series with a good prognosis after surgery.

Keywords: *Congenital Mitral Stenosis (CMR); Congenital Malformation*

Introduction and Objectives

Congenital mitral stenosis is defined as any "obstacle to the diastolic filling of the left ventricle" due to anomalies in the development of the structures that form the mitral apparatus (ring, immediately supravalvular area, valves, ropes and pillars).

It is a rare congenital malformation, its prevalence is estimated at 4 in 1000 children diagnosed with congenital heart disease [1] and it constitutes 0.6 to 1.2% of autopsied congenital heart diseases [2]; it is rarely isolated, the associated forms were noted in 60 to 90% of cases [3], in particular obstacles on the ejection path of the left ventricle [4].

It is a serious heart disease difficult to identify especially in the associated forms and difficult to treat surgically.

Aim of the Study

The aim of this work was to evaluate the clinical epidemiological echocardiographic aspect and prognosis of RMC at Chu Ibn Rochd in Casablanca.

Methods

We conducted a retrospective study collected in the cardiology department of the Ibn Rochd University Hospital in Casablanca, between September 2009 and January 2018, involving 26 patients.

All children referred to our institution for cardiac evaluation and diagnosed with congenital mitral stricture on echocardiography were included in the study.

The parameters analyzed were as follows: epidemiological data, clinical presentation, electrocardiographic and echocardiographic anomalies, therapeutic modalities and evolution.

Data analysis was made possible using SPSS statistical software (SPSS Inc, version 2014).

Results

During the study period we diagnosed 2,678 congenital heart diseases. CMS was present in 26 patients, a prevalence of 0.97%..

The study population consisted of 13 boys and 13 girls, a sex ratio of 1 and the average age was 6 years (age range was 46 days to 48 years).

Clinically; the functional symptomatology was dominated by dyspnea (17 cases), recurrent bronchitis (8 cases). The clinical examination objectified cyanosis (4 cases), and heart failure (2 cases).

Echocardiography found an isolated CMS in 3 cases; the etiologies were dominated by the supramitral ring (14 cases) and the mitral valve in parachute (8 cases).

The congenital heart disease most associated with CMS was ventricular septal defect (9 cases). The others associated with RMC were: coarctation of the aorta (6 cases), transposition of the great arteries (4 cases), pulmonary stenosis (4 cases), bicuspid aortic valve(4 cases), the double outlet right ventricle (3 cases), the left superior vena cava (3 cases), atrial septal defect (2 cases), supravalvular aortic stenosis (2 cases), ductus arteriosis (2 cases), Shone's syndrome (1 cases) and the cor triatriatum in 1 cases. Pulmonary arterial hypertension was present in 50% of cases, 2 of which were in Eisenmenger.

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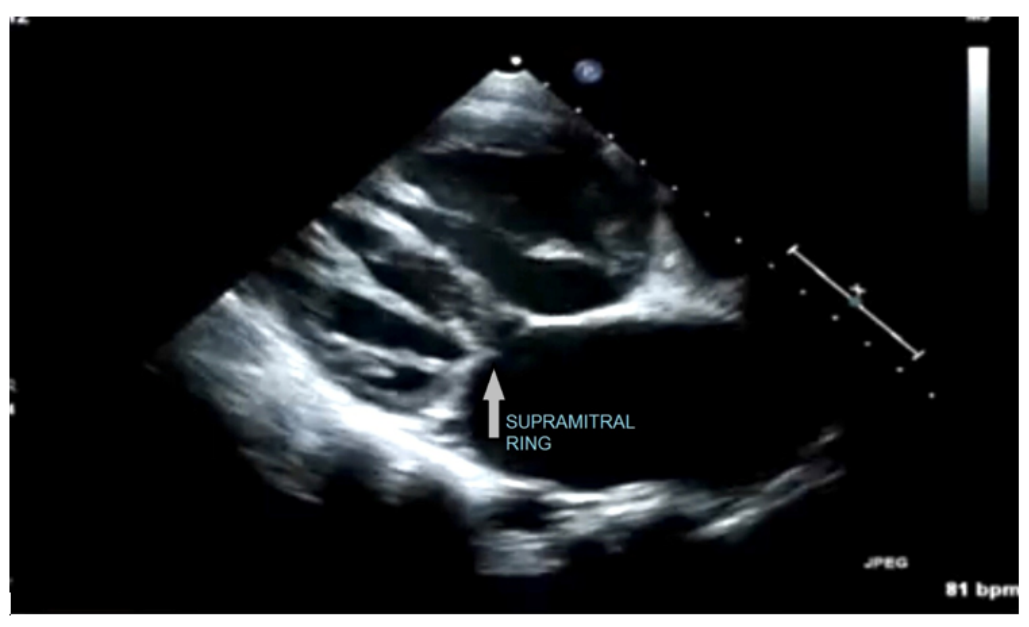
Good operative result of patients who have benefited from curative surgery (all CMS by supramitral ring) with the occurrence in a single patient of a post-operative atrioventricular block (resection of membrane under associated aortic).



Discussion

The retrospective nature means that our study has limits. As a result, data was missing with incomplete or unrecovered files that were excluded.

The prevalence of CMS in our series was 0.97%. A review of the literature analyzed epidemiological studies addressing RMC. Thus, Keith J., *et al*. report an estimated prevalence of 4 in 1000 children diagnosed with congenital heart disease [1] and RL Collins Nakai., et al. report a similar prevalence of 0.6 to 1.2% of autopsied congenital heart disease [2]. The average age of our patients (6 years) remains higher than that noted by RL Collins Nakai., *et al*. (1.5 years) in their study in Boston on 38 patients [2] and by Fekih M (1 year 8 months) in their study in Tunis on 13 patients [5]. The maximum age was 48 years in our study. In terms of symptoms, dyspnea was the most frequent (65%) as described in the literature: Fekih M., *et al*. (69%), RL Collins Nakai., *et al*. (39%) [2,5]. The other circumstances of discovery were recurrent bronchitis, more rarely cyanosis. The etiologies of RMC were dominated by the supramitral ring with a high prevalence (14 cases therefore 53%) but described as being a rare entity in the literature with less than 100 cases which were reported according to Mychaskiw G II., *et al*. [6], Agarwal S., *et al*. indicates that the supramitral ring can be present up to 8% of all children with

congenital mitral valve disease [7] unlike EM Walter., *et al*. who have a prevalence of supramitral ring in children carriers of significant RMC in Germany (48 cases, 35%) [8].

The congenital heart disease most associated with CKD was ventricular septal defect (34% of cases), coarctation of the aorta (23%), transposition of the great arteries (15%), and bicuspid aortic valve (15%). EM Walter., *et al*. [8], SP Collison., *et al*. [9] also noted more ventricular septal defect (30% and 53% respectively), coarctation of the aorta (35%, 43%) but also an association with the sub aortic membrane. (35%, 28%) which was not present in our series of patients.

Conclusion

Congenital mitral stenosis is a rare heart disease. The anatomical lesions are often complex at the level of the ring, the area immediately above the valve, the valves, the ropes and the pillars. The supramitral ring remains a rare variety of RMC which was the predominant in our series with a good prognosis after surgery.

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