

Cardiovascular Topics

Mitral valve prolapse: a study of 45 children

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Summary

The knowledge pertaining to mitral valve prolapse is mainly based on studies in adults. In this study, the clinical profile as described in adults was compared with that found in children up to the age of 13 years. Forty-five children with echocardiographic-proven mitral valve prolapse and who met the inclusion criteria were included in the study.

The male:female ratio in this study was 1:1.37 and was not statistically significantly different from reported ratios. Most of the children were asymptomatic. Twenty-one of the 31 patients referred from outside the hospital had an incidentally found murmur. The symptoms found in this study were not similar to those described in adults. The most commonly found symptoms were shortness of breath and fatigue, in contrast to those of chest pain and palpitations described in adults. Comparing males to females in this study, significantly lower weight ($p = 0.005$) and body mass index ($p = 0.003$) were found in girls, and a significantly lower pulse rate ($p = 0.002$) in boys. Left-sided cardiac enlargement was diagnosed in 11 patients on chest X-ray and in six patients on electrocardiogram. One patient had Marfan syndrome and four others had a Marfanoid appearance.

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In conclusion, most children with mitral valve prolapse are asymptomatic. Mitral valve prolapse is not an uncommon finding in children younger than 13 years of age. Patients with mitral valve regurgitation were advised to take infective endocarditis prophylaxis prior to invasive procedures.

Cardiovasc J South Afr 2003; 14: 191–194.

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Mitral valve prolapse is defined as the abnormal bulging of one or both of the mitral valve leaflets into the left atrium during systole (Fig. 1).¹ It occurs more often in females,^{2,3} but the exact aetiology of mitral valve prolapse is unknown.⁴

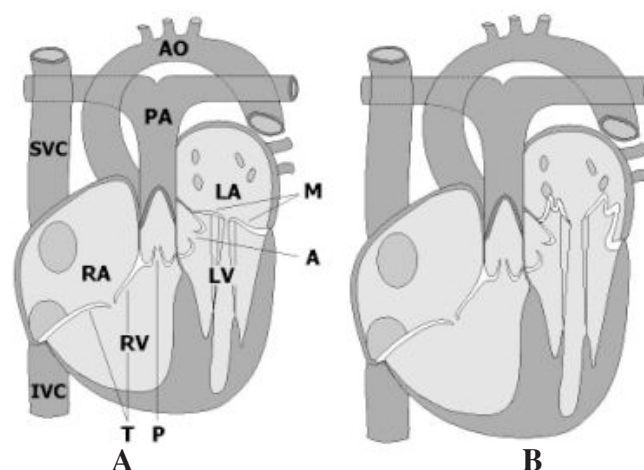


Fig. 1. Cross-section of the heart. Adapted from Devereux.²⁴

A. The normal anatomy of the heart. Labeled are aorta (AO), pulmonary artery (PA), superior vena cava (SVC), inferior vena cava (IVC), right atrium (RA), right ventricle (RV), left atrium (LA), left ventricle (LV), mitral valve (M), aortic valve (A), pulmonary valve (P), tricuspid valve (T). B. A myxomatous degeneration and prolapse of the mitral valve.

The condition was characterised by Barlow *et al.*,⁵ hence, in clinical settings mitral valve prolapse is often called Barlow syndrome. With the development of echocardiography during the 1970s (first M-mode and later 2D-echocardiography) the clinical diagnosis of mitral valve prolapse could be more easily confirmed (Figs 2, 3).¹

The majority of mitral valve prolapse patients are asymptomatic⁶ and, as previously stated, the prolapse is an incidental find.¹ Those who do have complaints suffer from chest pain, palpitations, dyspnoea, and fatigue.^{1,7} Typically, mitral valve prolapse patients have a slender body habitus.^{8,9} Thoracic skeletal abnormalities are common^{1,6} and mitral valve prolapse is often associated with connective tissue diseases.^{6,10} Although most ECGs are normal, ST-segment depression in the inferior leads may occasionally be found.^{1,11} Several complications, such as infective endocarditis, mitral valve regurgitation, rupture of the chordae tendinae, supra-ventricular arrhythmias, atrioventricular conduction disturbances, congestive heart failure, thrombo-embolic episodes and sudden death have been associated with mitral valve prolapse.¹

The knowledge pertaining to mitral valve prolapse is mainly based on studies in adults.¹² Little could be found in the literature on mitral valve prolapse in children under the age of 13 years.¹⁴

Aim

The aim of this study was to compare the clinical presentation in children under 13 years of age with those described in adults.

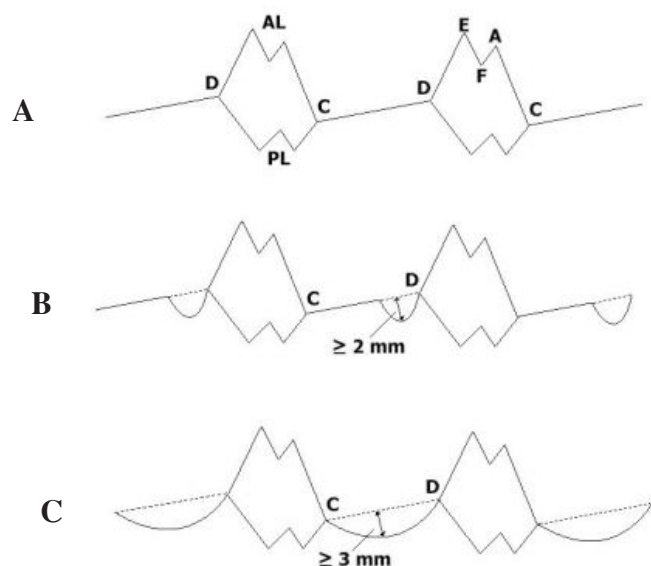


Fig. 2. M-mode echocardiography. Adapted from Prabhu *et al.*²⁵ A. A normal mitral valve M-mode tracing. The points labeled are anterior leaflet (AL), posterior leaflet (PL), end-systole just prior to valve opening (D), peak initial opening of the anterior leaflet (A), nadir of the initial closing of the anterior leaflet (F), peak opening of the anterior leaflet with atrial systole (A), complete valve closure with the onset of ventricular systole (C). B. Late systolic mitral valve prolapse. C. Holosystolic mitral valve prolapse.

Patients and methods

This is a retrospective study of patients suspected of having mitral valve prolapse who presented at our paediatric cardiology department in the last 25 years. One hundred and one patient folders were examined. Patients were excluded if there was no echocardiographic confirmation of mitral valve prolapse, if mitral valve prolapse was due to chronic rheumatic heart disease, or if more than three of the following data modalities were missing: age, sex, reason for referral, symptoms and signs, clinical diagnosis, ECG findings, chest X-ray findings, and treatment modalities. The significance of variables was tested by means of a chi-square test, 2 × 2 contingency table, Wilcoxon signed ranks test or Mann-Whitney U test, where appropriate.

The criteria used for diagnosing mitral valve prolapse were:

- (1) M-mode: prolapse of 2 mm or more for partial late systolic prolapse and 3 mm or more for pansystolic prolapse (Fig. 2).²⁵
- (2) 2D-mode: Visualising prolapse of 3 mm or more measured in the parasternal long axis systolic frame and confirmed in the apical 4-chamber systolic frame.²⁵

NCHS (National Centre for Health Statistics) percentile charts were used for length and weight, and BMI was calculated as follows: weight (kg)/length² (cm). Permission to do the study was obtained from the Ethics Committee of the Faculty of Health Sciences of the Stellenbosch University.

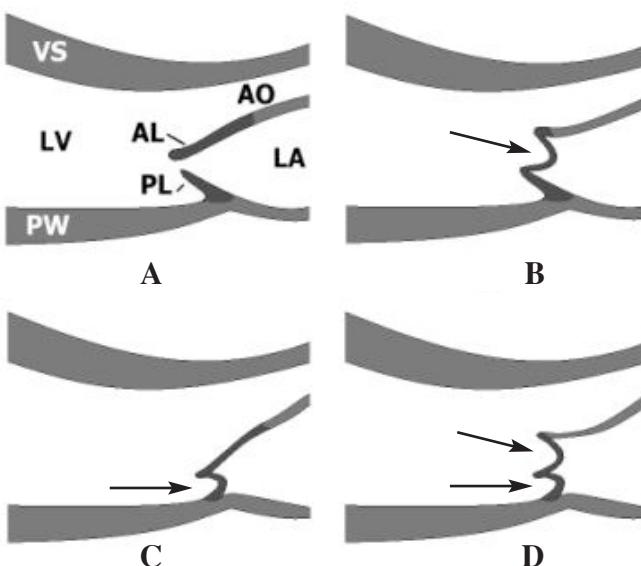


Fig. 3. Two-dimensional echocardiograph in the parasternal long-axis view. Adapted from Prabhu *et al.*²⁵

A. A parasternal long-axis view at end-diastole immediately preceding mitral valve closure. Labeled are anterior leaflet (AL), posterior leaflet (PL), ventricular septum (VS), posterior wall (PW), aorta (AO), left atrium (LA) and left ventricle (LV).

Systolic prolapse may be predominantly anterior leaflet (B, arrow), predominantly posterior (C, arrow), or both (D, arrows).

Results

Forty-five patients met the inclusion criteria for analysis. Of these, 19 were male and 26 female. Thirty-nine of these patients were of mixed racial origin, three were black and three were Caucasian. The mean age at presentation was 7.23 years (range 0–12.4 years).

Reasons for referral are summarised in Table I. Thirty-one patients were referred from outside the Tygerberg Children's Hospital and 14 from the general outpatient department. In 21 of the 31 patients referred from outside and in all 14 of the patients referred from outpatients, a murmur was an incidental finding which led to the patient's referral. In only one patient was there a family history of

TABLE I. REASON FOR REFERRAL			
	Percentage of the number of		
	reasons (n)	reasons (n = 36)	patients (n = 31) ^a
Incidental finding	21	58.3	67.7
Chest pain	4	11.1	12.9
Palpitations	1	2.8	3.2
Dyspnoea	1	2.8	3.2
Fatigue	0	0.0	0.0
Cardiac abnormalities ex murmurs etc	2	5.6	6.5
Joint pains	2	5.6	6.5
Other	5	13.9	16.1
Not referred	14		

^aA number of patients had more than one reason for referral

TABLE II. COMPLAINTS AT PRESENTATION			
	Percentage of the number of		
	complaints (n)	complaints (n = 49)	patients (n = 43) ^a
No complaints	27	55.1	62.8
Dyspnoea	6	12.2	14.0
Fatigue	5	10.2	11.6
Chest pain	2	4.1	4.7
Palpitations	0	0.0	0.0
Mitral valve prolapse- related complaints ex fear and panic attacks etc	9	18.4	20.9
No information available	2		

^aA number of patients had more than one complaint

TABLE III. SKELETAL AND CONNECTIVE TISSUE ABNORMALITIES	
Case number	Diagnoses
1	Marfan's syndrome
2, 3, 4	Marfanoid appearance
5	Marfanoid appearance, scoliosis
6	Scoliosis
7	Barrel chest
8	Straight back syndrome
9	Straight back syndrome

heart disease. Table II lists the patient's complaints on presentation. Sixty-three per cent (27/43) were asymptomatic and only a minority (14% and 12% respectively) complained of dyspnoea or fatigue.

Forty-two of the 45 patients had a systolic murmur on auscultation while only eight patients had an audible systolic click. In 27 patients the murmur was graded between 2/6 and 3/6.

Compared to the 50th percentile scores for length, weight, BMI, systolic and diastolic blood pressure and pulse frequency, the entire group had a significantly lower weight ($p = 0.029$) and BMI ($p = 0.031$). When comparing males to females, in our study, a significantly lower pulse rate ($p = 0.002$) was found in boys, while girls had a significant lower weight ($p = 0.005$) and BMI ($p = 0.003$). Eleven skeletal and connective tissue abnormalities were found in nine patients and are summarised in Table III.

ECGs were done in 32 patients with two showing ST-T segment abnormalities in the inferior ECG leads. Chest roentgenograms were done in 21 patients. In 11 of these patients, left ventricular or left atrial enlargement or cardiomegaly was reported.

Thirty-six patients were referred with a clinical diagnosis of a cardiac abnormality, of whom 29 were diagnosed as having mitral valve prolapse, or mitral valve regurgitation, or both. Infective endocarditis prophylaxis was recommended in the 42 patients with mitral valve regurgitation.

Discussion

This study has all the shortcomings of a retrospective study, with special reference to incomplete folder information, which may skew the data and its interpretation. The male:female ratio reported in the literature varies between 1:1⁹ and 1:6.² There was no statistically significant difference between the numbers of males and females ($p = 0.297$) in our study. From the literature, it was obvious that the method by which the diagnosis of mitral valve prolapse was made and the number of patients in the study may influence the reported ratios. For example, in a study by McLaren *et al.*³ (male:female ratio 1:1.9) the diagnosis was made on auscultation alone, while the study population in a study by Hickey *et al.*⁷ (male:female ratio 1:1.3) was small.

The presence of mitral valve prolapse in the different race groups in this study is just a reflection of the composition of our hospital patient population and does not indicate that it is more common in patients of mixed origin. No data of mitral valve prolapse in patients of mixed origin could be found in the literature.

The main reason for referral from outside clinics or physicians was an incidentally found murmur (67.7%). This finding was significantly higher ($p < 0.001$) than the 32.7% in a paediatric population described by Bisset.¹³ At presentation, the majority of patients were asymptomatic. Dyspnoea and fatigue were the most common complaints in our study population. In contrast, adults more often present with palpitations and chest pain.^{1,7} Our findings support the claim in the literature that children have fewer complaints.^{14,15}

According to the literature, mitral valve prolapse patients have a lower weight, a slender body habitus and might be taller when matched for age and gender.^{2,8,9} However, in this study only a significantly lower weight and BMI were found. These findings were more prominent in females. A significantly lower pulse frequency was found in males than in females, which to our knowledge, has not been reported previously in the literature. The mechanism for the slow heart cannot be explained.

The number of skeletal abnormalities found fall within the reported range of 1.9 to 75%.^{15–17} The growth spurt during puberty might explain the higher incidence of skeletal abnormalities, such as scoliosis, in adults.^{18,19} Mitral valve prolapse is often found in patients with Marfan syndrome²⁰ but only one of our patients had Marfan syndrome, while four others were described as having a Marfanoid appearance.

Two (6.5%) of the 32 patients who had had ECGs done, had the previously described ST-segment abnormalities associated with mitral valve prolapse in adults. This was significantly fewer than found in adults²¹ ($p = 0.013$) and in a study in children by Greenwood¹⁵ (age 2 months to 18 years, $p = 0.033$). It, however, correlates well with the findings of McLaren *et al.*³ ($p = 0.916$) in a study of children (age 2–18 years). None of our patients had signs of supraventricular tachyarrhythmias, another well-described finding in adults.^{1,6}

Few chest roentgenogram abnormalities were described in patients with mitral valve prolapse.^{1,14,15,22} In our study, 11 of the 21 patients in whom chest X-rays were done, had reported left ventricular and left atrial enlargement. Possible causes for these findings might be an over reporting of ventricular and/or atrial enlargement, or that the degree of mitral valve regurgitation might have contributed to these findings.

Systolic clicks were found in only eight of our patients. This finding might be skewed, depending on the clinician who examined the patient and noted the clinical findings. Children with mitral valve regurgitation were advised to inform their dentists that they have a heart lesion, so that infective endocarditis prophylaxis, according to the criteria of the American Heart Association,²³ could be given.

Conclusion

Mitral valve prolapse is not uncommon in children under the age of 13 years. The clinical findings differ from those described in adults, as chest pain and palpitations were uncommon complaints in children.

Most of our patients were asymptomatic, had a slender body habitus, and presented with an incidentally found ejection systolic murmur. Systolic clicks were found in only eight of our patients. Skeletal or connective tissue abnormalities were present in 20% of our patients.

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