



## Surgical Management of Complex Hypertrophic Cardiomyopathy Causing Severe Mitral Regurgitation in a Child

Authors

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### Abstract

*Mitral leaflet elongation and subvalvular apparatus deformation can be seen in obstructive septal hypertrophy. In such hypertrophic cardiomyopathy (HCM) mitral malformations, the Mitral valve repair will not suffice to reverse the pathology unless surgical myomectomy is done. Outcome is more gratifying when left ventricular outflow tract obstruction (LVOTO) is eliminated along with abolition of systolic anterior motion (SAM) of mitral leaflet. Trans-esophageal echocardiography (TEE) play a major role in making surgical decision on operating table.*

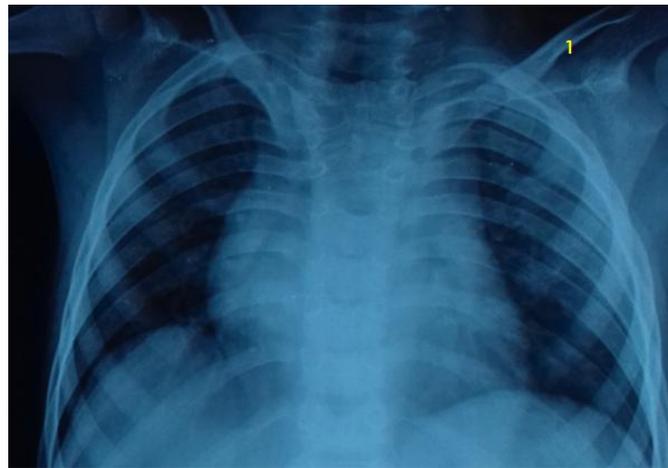
### Introduction

Mitral leaflet elongation and subvalvular apparatus deformation can be seen in obstructive septal hypertrophy. In such hypertrophic cardiomyopathy (HCM) mitral malformations, the Mitral valve repair will not suffice to reverse the pathology unless surgical myomectomy is done. Outcome is more gratifying when left ventricular outflow tract obstruction (LVOTO) is eliminated along with abolition of systolic anterior motion (SAM) of mitral leaflet. Trans-esophageal echocardiography (TEE) play a major role in making surgical decision on operating table

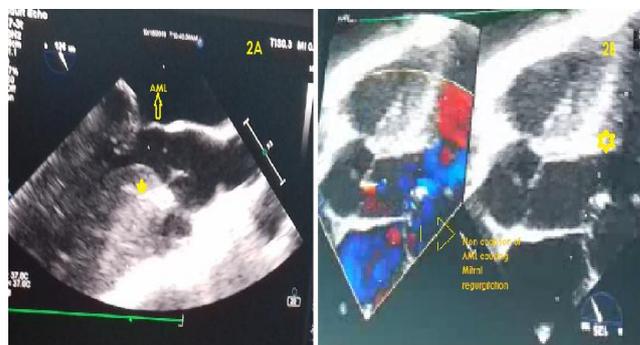
### Case Report

A 4 yr male child, wt-17kg, BSA-0.66m<sup>2</sup>, presented with reduced physical activity with rapid precordial movement for six months. Examination revealed presence of pansystolic murmur in the apical area with radiation to left axilla. Chest X-ray showed Cardiomegaly, dilated left ventricular cavity with no abnormal pulmonary shadow (**Figure-1**). Transthoracic echocardiography revealed prolapse of anterior mitral leaflet (AML), where prolapse of A2segment seen more than A1 and A3, AML length was 21mm. Accessory mitral tissue in relation to AML was getting into left ventricular

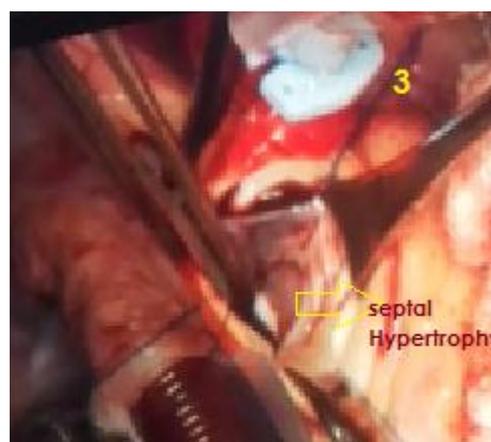
outflow tract (LVOT). Exaggerated movement of mitral valve (MV) leaflet causing non-coaptation in systole, mild left ventricular hypertrophy and dilated left ventricular chamber with normal LV function. TEE before commencing CPB revealed septal hypertrophy causing significant LVOTO (mean gradient =80mmHg)with SAM of AML causing severe mitral regurgitation (**Figure 2A and 2B**).Patient was operated on cardiopulmonary bypass (CPB). Under cold cardioplegic arrest, oblique aortotomy incision given 1cm above the origin of right coronary ostium. Exposure could be enhanced by the retraction of aortic leaflet and retracting the inferior edges of aorta with aortic stay sutures. Septal hypertrophy could be visualised along with a white endocardial scar, which is the contact lesion produced by apposition of septum and anterior mitral leaflet. (**Figure 3**). This lesion serves as a guide to the length of myomectomy which must be extended apically beyond this contact lesion. We used No-11 knife blade on a long handle to begin the myomectomy with an upward incision in the septum 3 to 4mm to the right of the nadir of the right aortic sinus. The incision was then carried upward towards the aortic annulus and then counter clockwise over to the attachment of the anterior leaflet of mitral valve (**Figure 4**). The Transverseplication of redundant leaflet was done with interrupted 5-0 prolensutures (**Figure 5**). Aortotomy was closed in two layers. Patient was weaned from CPB. Post surgical correction TEE showed AML was thick and coapting well with posterior mitral leaflet (PML). LVOT gradient (Peak =26mmHg/mean =10mmhg), mild mitral regurgitation, thickness of interventricular septum (IVS) =12mm, Normal LV function. Post operative course was uneventful and patient was discharged on 5<sup>th</sup> post operative day.



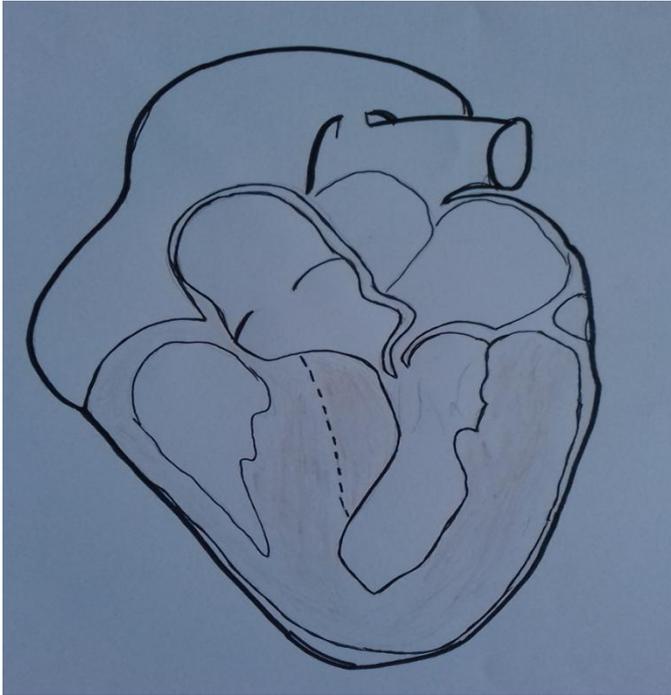
**Figure 1**-Chest X-ray showing Cardiomegaly, dilated left ventricular cavity with no abnormal pulmonary shadow



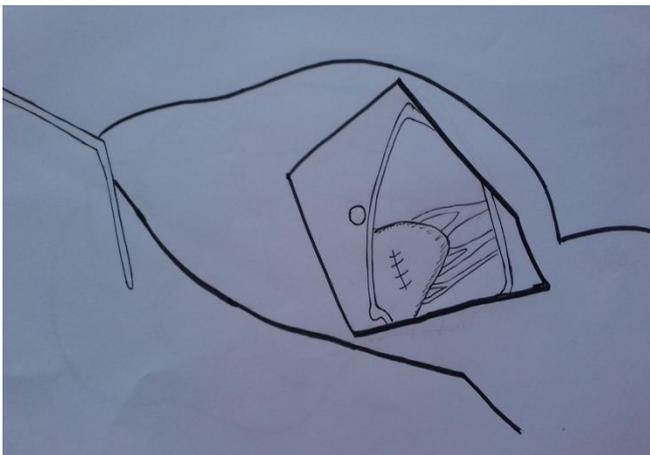
**Figure 2**-Intra operative transesophagealechocardiography (TEE) revealing septal hypertrophy causing significant LVOTO (**Figure 2A**) and SAM of anterior mitral leaflet causing severe mitral regurgitation(**Figure 2B**).



**Figure 3**-Septal hypertrophy with a white endocardial scar, which is the contact lesion produced by apposition of septum and anterior mitral leaflet.



**Figure 4** -septal hypertrophy involving sub aortic and mid ventricular region.Dotted line shows the incision line of septalmyomectomy by trans-aortic approach.



**Figure 5**-Schematic diagram showing transverse plication of redundant anterior mitral leaflet (+++).

### Discussion

In HCM, LVOT obstruction can be caused by SAM of mitral valve<sup>(1,2)</sup>. Investigators have observed mitral slack is a necessary component of dynamic SAM.<sup>(3,4)</sup> Displaced anomalous anterior papillary muscle inserting directly to the mitral leaflet without chordae tendinae, elongated posterior leaflet causing SAM are the other causes of LVOTO<sup>(5,6)</sup>. The most common cause of

mitral regurgitation in HCM is non-coaptation of posterior leaflet which is not long enough to cause coaptation<sup>(7-9)</sup>. Treatment is always surgical for mitral structural abnormalities and hypertrophied septum. Surgical myomectomy along with plication, valvuloplasty and papillary muscle relocation improves the coaptation and reduces mitral regurgitation<sup>(7,9,10-12)</sup>. TEE is the best modality of investigation when preoperative imaging are suboptimal. It is always to be done in operating room before CPB to formulate the final operating plan and after CPB to assess the adequacy of surgical correction. Shortening of leaflet can be achieved by plication technique as described by Swistel et al, stiffen the AML and reduces the billowing, which in turn reduces the LVOTO and increases the coaptation<sup>(12-14)</sup>. Excision is an alternative procedure to plication in shortening of redundant leaflet segment.

### Conclusion:

Trans aortic approach for surgical myomectomy and repair of mitral leaflet is the surgical procedure defined for mitral regurgitation in HCM. TEE is the best modality of imaging before CPB to plan the surgery and after CPB to assess the adequacy of correction.

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