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COMBINED MINIMALLY INVASIVE RESECTION OF SUB AORTIC MEMBRANE AND MODIFIED RAVITCH PROCEDURE

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This, in turn, results in the further obstruction and hyperdynamic function.

Pectus carinatum is characterized by anterior protrusion of chest wall with0.2% incidence. Subvalvular aortic stenosis causes clinically significant left ventricle outflow obstruction resulting in the development of concentric left ventricular hypertrophy, frequently with a septal bulge. This, in turn, results in the further obstruction and hyperdynamic function. We report a very rare case where an adolescent admitted for surgery for symmetrical marked, symptomatic pectus carinatum and incidentally found to have severe sub aortic stenosis giving significant left ventricle outflow gradients in pre-operative evaluation. A successful Minimally invasive resection of Subaortic membrane with concomitant chest wall reconstruction was performed.

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INTRODUCTION

Pectus carinatum is characterized by anterior protrusion of the sternum and adjacent cartilage, and its reported incidence is 0.2%, with higher frequency in men than women [1].

Although it can present in early childhood, it often is not apparent until puberty at which time it can progress in severity during rapid linear growth. Pectus carinatum may present with or without symptoms. The most common symptoms elicited may include exercise intolerance, chest pain, chest wall tenderness, shortness of breath, palpitations, or wheezing. In children whose chest wall significantly deviates from normal, reconstructive and psychosocial concerns are reasons families may seek medical attention.[2]

Surgical and nonsurgical methods are available for treatment of pectus carinatum. The nonsurgical method involves external compression of the sternum using a brace. Bracing is generally the first option for treatment; if it fails, surgical correction can be considered [3]. The classic method for surgical repair of chest deformities was described by Ravitch [4]. Associated Subaortic membrane with Pectus carinatum deformity has not been documented in the literature.

Cardiopulmonary complications resulting from mechanical compression by the deformed chest wall may be an indication for surgical correction, especially in patients with underlying cardiac disease.

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Cardiac compression can contribute to postoperative hemodynamic instability if the pectus deformity is left uncorrected. Subvalvular aortic stenosis is a rare disorder seen in infants and newborns but is the second most common type of aortic stenosis. It is responsible for approximately 1% of all congenital heart defects (8 in 10,000 births) and 15% to 20% of all fixed left ventricular outflow tract obstructive lesions.

10 to 14% of subvalvar aortic stenosis is observed amongst children with congenital aortic stenosis. It is more common in males and is responsible for 65% to 75% of the cases with a male to female ratio of 2:1. The prevalence of SAS is 6.5% of all the adult congenital heart diseases.

In the present study, we describe a combined strategy to correct the pectus carinatum deformity and the resection of underlying Sub-aortic membrane performed through a right anterior mini-thoracotomy and central cannulation.

Case Report

A 19 year old boy from Turkmenistan, case of Pectus carinatum-Chondrogladiolar symmetrical type [Figure 1]. Patient presented with chief complaints of progressive exercise intolerance and chest discomfort since last 3 months which had progressed to angina on routine activities. Past history of any major medical condition or surgical procedure was absent. Chest radiograph showed marked protrusion of chest wall with substantial retrosternal space. On pre-operative evaluation, Transthoracic echocardiography showed incidental finding of turbulence across LVOT with maximum gradient of 35mmHg and mean gradient of 19mmHG. Aortic cusps were mildly thickened, aortic root of 27mm. Normal left ventricular function with ejection fraction of 55%. Computed tomography

of chest revealed Pectus carinatum deformity, dilated aortic arch 3.9 cms and both lungs to be hyperinflated. Transesophageal echocardiography intraoperatively revealed tethering of Subaortic membrane between right and non coronary cusps resulting in moderate Aortic regurgitation.

Post evaluation, it was not conclusive enough whether the symptoms were occurring due to the Subaortic membrane or the Pectus deformity. Since the patient has admitted for Pectus Carinatum correction for psychosocial reasons as well as symptoms; a decision was taken forPectus Carinatum correction with concomitant minimally invasive Subaortic membrane resection.

After inductionskin flap from xiphisternum to 2nd intercostal space was raised and entry to thorax was gained through right 2nd intercostal space. After systemic heparinization (400 IU/kg), aortic cannulation was done and venous drainage was achieved using a 3-stage cannula to Inferior vena cava via right atrial appendage. Aorta was cross clamped, heart was arrested with cold blood cardioplegia and patient cooled to 28°C. Subaortic membrane was resected and aortic valve was assessed was its competency and was found adequate. Aortic cross clamp time was 73 minutes and cardiopulmonary bypass time was 55 minutes. After a smooth weaning from cardiopulmonary bypass, hemostasis was achieved.

After achieving hemostasis, chest wall repair was started. Reconstruction of Pectus cranium deformity with Sternochondroplasty multiple and transverse sternal osteotomies and fixation [Figure 2]. The deformed portion of the sternum was transacted in three points, and the divided sternum was fixed and corrected in elevated position by using a dynamic compression plate (KLS Martin 2.3mm and KLS Martin 2.0mm and AO 8-hole Box Plate) passed horizontally posterior to the ribs to support the lower xiphisternum[Figure 3]. Total duration of the surgery was six hours and post op hemodynamic and cosmetic results were satisfactory.



Figure 1 Patient's pre-operative Pectus carinatum- Chondrogladiolar symmetrical type.



Figure 2-Post heparin reversal and decannulation for Subaortic stenosis resection, reconstruction of Pectus cranium deformity with Sternochondroplasty and multiple transverse sternal osteotomies and fixation.



Figure 3 The deformed portion of the sternum was transacted in three points, and the divided sternum was fixed and corrected in elevated position by using a dynamic compression plate (KLS Martin 2.3mm and KLS Martin 2.0mm and AO 8-hole Box Plate) passed horizontally posterior to the ribs to support the lower xiphisternum.

DISCUSSION

Wearing a compression brace is a valid nonsurgical treatment for pectus carinatum and is generally the first option considered. For patients who have difficulties with brace compliance, surgical correction may be offered [6].

The mean age of the patients of 24.35 ± 13.20 years in the present study was higher than those reported in similar studies: 14.30 years, 14 years [5], and 15.70 years [7]. Nonetheless, our outcomes and results are similar to those of previous studies. After the growth spurt in adolescents, the chest wall cannot be easily modified via nonsurgical procedures, and these procedures therefore have a poor outcome in adults, as well as poor patient compliance. According to Yuksel *et al.* [5], the optimum age range for minimally invasive surgery is between 12 and 18 years, because the deformity is more prominent during the rapid growth phase of puberty and the chest wall is still flexible.

In our study, a symptomatic symmetrical pectus carinatum deformity was presented with, incidentally found, significant gradient across LVOT due to sub aortic membrane which warranted urgent intervention. Hence, a decision for minimally invasive SAM resection along with correction of pectus carinatum was taken in view of patient's choice as primary intervention due to psychological reasons.

In our study, there were no plate dislocation or wire problems and post op gradient across LVOT was 1 mmHg, and patient was subsequently discharged on post-operative day 9.

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