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INNOMINATE ARTERY COMPRESSION SYNDROME WITH PECTUS EXCAVATUM: NUSS REPAIR AND AORTOPEXY AS A RESCUE PROCEDURE.

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A case of pectus excavatum in a 7 years old child with innominate artery compression syndrome is presented.A simultaneous Nuss operation and anterior aortopexy are described.

Key words: pectus excavatum, innominate artery compression syndrome, tracheal compression, nuss operation, aortopexy.

We report a case illustrating pectus deformity excavatum which caused significant tracheal compression and was successfully managed with a combined surgical approach and through the Iinternational co-operation. This index case highlights that patients with airways compression syndrome can be present a major diagnostic and therapeutic challenge. This is the first report of a simultaneous Nuss procedure and anterior aortopexy. A thorough and systematic assessment with individualized decision-making to deal with this rare group of patients can achieve satisfying satisfactory results.

Introduction. Pectus excavatum is one of the more common chest wall anomalies seen in children1, and, although rare, t. Tracheobronchial compression (TBC) is well described concomitant problem in patients with chest such deformities. Though more often it is reported to be associatedin association with scoliosis, the association withrelation of TBC to pectus excavatum should not be underestimated.

The usual cause of airways compression in these patients is a narrow anteroposterior chest diameter (between the manubrium and spine) at is a direct compression at the thoracic inlet, due to a narrow anteroposterior chest diameter (between the manubrium and spine) as well asand an altered anatomic relationship resulting in abnormal 'stacking' of structures in the confined space of the superior mediastinum. This may result in respiratory distress and can become life-threatening2.

Case report. A 7 year old male with a history of clinically asymptomatic severe pectus excavatum was admitted to the ICU of the major pediatric cardiac centers in Kiev after adenoidectomy that resulted in inability to ventilate in one of local hospitals. A 7 year old child, who was known to have pectus excavatum, had an elective adeno-tonsillectomy at a peripheral hospital and could not be weaned from ventilation. He was transferred to the ICU of the paediatric cardiac centre in Kiev for further treatment. The patient was not known to have any other major medical history.

He was had been seen by thoracic surgeons earlier in his life for the pectus excavatum, at which at that time it was judged a purely cosmetic problem and the family was advised to postpone a surgical repair until the teenageteenage years.

The adenoido-tonsillectomy that preceded the below described problem was elective without major expected risks. After planned extubation immediately postsurgically patient developed severe stridor with respiratory distress that required intubation and after failure to extubate he was referred for further investigation and treatment to the Ukrainian Children's Cardiac Center in Kiev. He was admitted to the ICU. As a part of investigation As part of his work-up on arrival in Kiev, a rigid bronchoscopy was performed which revealed a pulsating compression of the lower third of trachea, suggesting a vascular compression. An angio CT performed subsequently had shownrevealed a severe form of pectus excavatum with flattening of the upper flat chest. There was also a leftward displacement of the heart and the manubrium was compressing the aorta and innominate vessels. The trachea was thus compressed by the bracheocephalic trunk at its lower third.

The patient was then referred for the second opinion to The Tracheal Team at The

Great Ormond Street Hospital for Children in London. His clinical data were reviewed there, and the following were suggested;- and a Nuss procedure was employed to push his flat upper sternum forward with to permit a concomitnant aortopexy to lift the aorta and innominate artery away from his trachea was suggested as the best way to treat the condition. On the basis of this advice the cChild was then transferred to the Great Ormond Street Hospital for surgery.

The operation was performed as follows.

1 The *Nuss* procedure was done using minimally invasive convex bar technique (Nuss) under thoracoscopic guidance. Lateral 1" incisions were made opposite the chosen place for lifting the sternum. Tunnels were made to the highest rib anteriorly anterior to the muscle layer. The interior of the chest was inspected using thoracoscopy and entry points created for the pectus bar, which was shaped to the size identifed by a copper template. An 11" bar was chosen. The pectus elevator was introduced without difficulty under direct vision and the pectus bar introduced following it. The bar was flipped into shape, elevating the sternum. The bar was fixed to the rib on either side with figure of 8 stainless steel wire sutures. Stabiliser bars were not used, to protect the axillary structures. The bar was placed more superiorly than usual to lift the sternum and manubrium.

2 *Bronchoscopy*

A fibreoptic flexible bronchoscopy was performed intraoperatively showing severely compressed trachea by the innominate artery, and inflammed internal wall of the trachea with erosion at that site. The anterior wall of the trachea was distorted and will probably be permanently deformedphysically flattened. Although considerably improved compared to the preoperative appearance, there was still room for improvement.

3 *Aortopexy*

Thus a T-shaped incision was created at the upper sternum and the pre- tracheal fascia was opened, creating space between it and the innominate artery. The pericardium was then opened longitudinally and Teflon- buttressed aortopexy sutures were inserted into the aortic wall at either side of the aorta, at the base of the innominate artery. These

sutures were then passed through the pericardium and then via the sternum to be tied later over pledgets. The pericardium was approximated with Vicryl, the sternum was closed and the aortopexy sutures tied under bronchoscopic control. There was a good physiological result from the Nuss, (not cosmetically) and the aortopexy. The pPatient was extubated at next few hours postoperatively and the early postoperative period was mostly uneventful. After few weeks the patient developed mild reactive pleuritis most likely as a reaction to nickel-containing foreign body (the bar). which This was successfully treated with steroids and was resolved within 6 months period. The A postoperative CT scan showed improved upper chest configuration and absence of tracheal compression by vascular structures.

After having his pectus bar in place for 18 months according to the protocol he was then scheduled for the removal of the bar. The bar was removed via reopening of old lateral incisions by simply pulling out after some mobilization. The upper part of the sternum remained elevated after the bar wasr removed. After operation patient was extubated uneventfully on the operating table. He had uneventful recovery and was discharged home at 1st postoperative day. At 1 month follow-up he is a symptoms-free, active child with good exercise tolerance.

Discussion. Although the correlation between abnormal thoracic configuration and restrictive lung disease is well documented, thoracic deformity that causes obstructive lung disease, such as compression of the large airways (trachea and bronchi), is not well documentedreported. Tracheal compression may be caused by abnormalities of the bony thorax whose previously unrecognized significance can result in unexpected difficulties when extubation is attempted following a routine intubation3. Though not very common, tracheobronchial compression remains a significant morbidity associated with severe pectus excavatum. Patients with this condition should be investigated carefully especially prior to undergoing any other surgical intervention or general anaesthetic as vascular anomalies such as innominate artery compression can co-exist which can remain untreated if the therapy is directed at the correction of thoracic bony abnormalities only2.

There are various surgical treatment options described in literature for treatment of

the tracheobronchial compression by brachiocephalic vessels. Innominate artery reimplantation (which is reported to be a safe and effective treatment for innominate artery compression syndrome with excellent long-term results 4,5) was discussed as a treatment option but was considered to be potentially ineffective due to presence of pectus deformity. As wellFurther, while generally successful, arterial transfer carries the risk of early bleeding and stroke, and the potential for late stenosis at the anastomotic site6. Other large published series strongly support the belief that anterior suspension of the innominate artery is a successful and reliable operation with minimal morbidity and mortality6. These studies however report patients *without* major thoracic deformities.

As patients with airways compression syndrome and pectus deformities are form a rare entity hencesubgroup, there is no unified approach to treat this condition. In this case, tThe combination of minimally invasive convex bar insertion with aortopexy showed to be effective in this particular case and promising though further experience will need to be gained to show the effectiveness of this treatment in larger series.

Patients with airways compression syndrome and concomitant pectus excavatum are a diagnostic challenge. Comprehensive evaluation of these patients prior to undertaking surgical intervention or general anaesthesia avoids potential problems in the immediate postoperative period. Treatment of these patients must be individualised taking into account the nature of the pathology leading to potential or actual airways compression.

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СИНДРОМ КОМПРЕСІЇ БРАХІОЦЕФАЛЬНИМ СТОВБУРОМ ПРИ PECTUS EXCAVATUM:ОДНОЧАСНА ОПЕРАЦІЯ НУССА І АОРТОПЕКСІЯ ЯК ПРОЦЕДУРА ПОРЯТУНКУ

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Представлено випадок pectus excavatum у 7 річної дитини з синдромом трахеальної компресії брахіоцефальним стовбуром.Описано одночасну операцію Нусса і передню аортопексію.

Ключові слова: впалі груди, компресія трахеї, операція Нусса, аортопексія.

СИНДРОМ КОМПРЕССИИ БРАХИОЦЕФАЛЬНЫМ СТВОЛОМ ПРИ PECTUS EXCAVATUM: ОПЕРАЦИЯ НУССА И АОРТОПЕКСИЯ КАК ПРОЦЕДУРА СПАСЕНИЯ

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Представлен случай впалой груди у 7 летнего ребенка с синдромом трахеальной компрессии брахиоцефальным стволом.Описано одновременную операцию Нусса и переднюю аортопексию как процедуру спасения.

Ключевые слова: впалая грудь, компрессия трахеи, операция Нусса, аортопексия.