# Progress in pathogenesis of pectus excavatum

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#### Abstract:

Congenital pectus excavatum is a common skeletal deformity of the chest wall in children. The progressive aggravation of pectus excavatum can oppress the heart and lungs, even affect growth and development, and affect the physical and mental health of patients. Therefore, it is of great significance to choose the appropriate surgical evaluation index for the formulation of a surgical plan. This article reviews the diagnostic indicators and surgical procedures of pectus excavatum from the perspective of chest wall surgery. The correction effects and postoperative complications of various modified and new surgical procedures for pectus excavatum need to be further observed and evaluated. Postoperative complications should be considered in the design of surgical procedures for pectus excavatum combined with other diseases and the choice of surgical timing. To discuss and review the controversial points in the surgical treatment of pectus excavatum, and to provide a theoretical basis for the improvement of the surgical plan.

**Keywords:** Pectus excavatum, Nuss procedure; Ravitch procedure. diagnostic index.pathogenesis

# **1.** Pathogenesis of pectus excavatum

Clinically, pectus excavatum can be classified into symmetric and asymmetric types. Symmetric type is common in young children, and the proportion of asymmetric type increases gradually with age, and the rotation<sup>[1]</sup> of the sternum can occur. Treatment of asymmetric type is common in older children, some of them are complicated or secondary to scoliosis and other secondary deformities, and most of them show more obvious depression of the right chest wall, and the proportion of scoliosis increases<sup>[2]</sup>. The pectus excavatum has a profound impact<sup>[1]</sup> on patients' physical function. When the posterior edge of the depressed sternum is not compressing the heart, the damage to cardiopulmonary function is less severe in patients with mild pectus excavatum. When pectus excavatum is severe, the depressed chest wall reduces the overall volume of the chest cavity, and the expansion of the lung is inhibited, especially during inspiration, the expansion of the lung is limited, and the resistance is increased, which makes the patient prone to recurrent upper respiratory tract infection<sup>[2]</sup> After the chest cavity is reduced, cardiac activity is limited, stroke volume, palpitation, shortness of

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breath, and dyspnea are other symptoms<sup>[1]</sup>.

The pathogenesis of pectus excavatum mainly includes thoracocostal dysplasia and diaphragmatic dys<sup>[3]</sup>plasia. The ribs form the skeleton of the thorax by connecting with the thoracic vertebrae and sternum. During breathing, the ribs work in concert with the muscles in the gap to expand and contract<sup>[3]</sup> the rib cage. Patients with a pectus excavatum have their sternum recessed inward, creating one or more funnel-shaped depressions that make the chest appear depressed or collapsed<sup>[4]</sup>. The overlong ribs squeeze the sternum backward, and the central tendon of the diaphragm attached to the lower end of the sternum is too short, making the sternum and xiphoid process depressed by backward traction, forming a funnel shape. The deformity of the chest cavity will affect the patient's respiratory movement<sup>[4]</sup>. In addition, the diaphragmatic muscle fibers are attached to the inner surface of the ribs and the lumbar spine<sup>[5]</sup>. They are the most important respiratory muscles and play an important role in maintaining abdominal pressure. In addition, the diaphragm helps the anterior abdominal muscles contract to promote intra-abdominal urination and defecation and helps the anterior abdominal muscles contract to raise abdominal pressure<sup>[5]</sup> when holding a heavyweight in deep inspiration. When the diaphragm is abnormal or incomplete in patients with pectus excavatum, the above functions will be disordered<sup>[5]</sup>.

Most of the literature has confirmed that the embryo is affected by certain factors during development and may develop into a pectus excavatum<sup>[6]</sup>. During embryonic development, the diaphragm is hypertensive, and the abnormal position of the embryo exerts intrauterine pressure on the sternum, resulting in mal<sup>[6]</sup>formation of the thoracic cavity. In addition, the compression of the fetal limb on the thoracic cage and the permanent mechanical stress caused by its extreme position may cause acquired injury and affect the normal development<sup>[6]</sup> of the thoracic cavity.

# **2. Diagnostic techniques for pectus ex-cavatum**

In recent years, the clinical application of three-dimensional reconstruction has provided a new way<sup>[7]</sup> 醿 to evaluate pectus excavatum deformity. The severity of pectus excavatum can be assessed by several indicators. Haller index(HI) is the most widely used deformity index in the world to determine the pectus excavatum, which measures the maximum transverse diameter of the chest at the most depression of the sternum and the distance ratio<sup>[8]</sup>In normal subjects, the average Haller index is 2.52. A Haller index greater than 3.2 is the diagnostic criterion for pectus

excavatum, a Haller index less than 3.25 is mild, and a Haller index of  $3.25^{[8]}$  funnel chest index(FI) is a method proposed by domestic scholars to evaluate the degree of funnel chest deformity. Mild: FI < 0.2, moderate: 0.3 > FI > 0.2, severe: FI >  $0.3^{[8]}$ The lower vertebral index (LVI) was measured by the lateral chest X-ray film of the patient and was calculated as the ratio<sup>[8]</sup>The average LVI of normal subjects was 0. 21, while the LVI of patients with pectus excavatum was > 0. 22, and the maximum was  $0^{[8]}$  anthropometric index(AI) The ratio of sternal depression depth during deep inspiration to thoracic transverse diameter during deep inspiration at the lower 1/3 of the sternum plane. The AI value ranges from 0 to 1, and AI≥ 0.12 can be judged as pectus excavatum. The higher the AI value, the higher<sup>[8]</sup>

# **3.** Treatment techniques for pectus excavatum

It is generally considered that the age of patients who are suitable for pectus excavatum surgery is 3-12 years old, and the optimal age is 6-8 years<sup>[9]</sup>Before surgery, it is generally necessary to check the following two indicators, including the pectus excavatum index >3 on CT; Pulmonary function test results: small airway ventilation impairment and decreased ventilatory reserve function, suggesting the existence of restrictive or obstructive airway disease; Cardiac function tests showed incomplete right bundle branch block and cardiac valve prolapse by echocardiography. The pectus excavatum was progressively aggravated with definite complications<sup>[9]</sup>. The existing surgical methods of pectus excavatum mainly include invasive, minimally invasive, and non-invasive surgery.

#### 3.1 Invasive surgery

Invasive surgery is mainly divided into sternal lifting and sternal inversion.

In the Ravitch procedure, a transverse incision was made at the xiphoid process. After the chest wall tissue was separated, the depressed 4-5 pairs of costal cartilage were removed<sup>[10]</sup>A transverse wedge incision was made at the beginning of the sternal depression, and then a "V" shaped incision was made at the lower part of the sternum. The depression of the lower part of the sternum was corrected<sup>[11]</sup>Finally, a metal strut was placed across the sternum for internal fixation<sup>[10]</sup>

The whole length of the costal cartilage was removed under the costal cartilage periosteum. The xiphoid process was removed first, and the intercostal muscles attached to the two sides of the sternum and the depressed coastal perichondrium were separated along the sternum. Then

the retrosternal connective tissue was separated, and the pleura was pushed to the side until it reached the level<sup>[12]</sup>Then, a transverse osteotomy was performed on the posterior wall of the bend of the depressed sternum, and an autologous cartilage piece was embedded for suture fixation. The dissociated sternum was lifted and kept in the lifted position<sup>[12]</sup>. Finally, the second costal cartilage was obversely cut from the anteromedial to posterolateral direction, and the broken end of the inner costal cartilage was transferred and fixed on the broken end of the posterolateral costal cartilage by overlapping suture. The contralateral second costal cartilage was also treated to form a three-<sup>[12]</sup>Sternal lifting is suitable for children aged 5-10 years with soft bones. For asymmetric pectus excavatum, a wedge incision of half sternum on the severely depressed side can be used for correction. The advantages of this operation are more like to control the scope of surgical trauma and fewer complications<sup>[13]</sup>. The disadvantage is that the effect of the operation is not as good as that of the sternoplasty, and the patients with stent implantation need a second operation to remove the stent. There was a certain probability of recurrence after the operation. The most serious condition was the recurrence of pectus excavatum (5.7%).

The periosteum of the deformed costal cartilage on both sides of the sternum was first separated, the deformed costal cartilage was dissociated, the 3rd to 7th costal cartilage was cut off, and the xi<sup>[14]</sup>Then the rib cartilage was pulled forward and above with the forceps, and the rib cartilage was moved up according to the shape of the rib care, close to the normal rib. Finally, the excessive bending deformity of the costal cartilage was removed, and the corresponding broken ends of the costal cartilage on both sides were sutured and fixed. Because of the upward force on the costal cartilaginous sides, the depressed sternum can be lifted to maintain the position<sup>[14]</sup> of the anterior elevator.

In the sternum inversion, the entire length of the 2-7 costal cartilage periosteum and the costal cartilage arch were dissected from top to bottom, and the costal cartilage and rib junction were cut off and flipped<sup>[15]</sup>The broken end of the sternum was sutured with stainless steel wire end-toend, and the excess costal cartilage<sup>[15]</sup> was trimmed under the condition of maintaining the normal thoracic shape. Finally, all the intact costal perichondrium and intercostal muscles were sutured back to the two edges of the sternum and the costal cartilage and rib junction of<sup>[16]</sup> the inverted bone flap. After this operation, attention should be paid to the distortion of bilateral internal thoracic arteries and other blood vessels after turning over the sternum, which may cause lumen stenosis and thrombosis, which may affect<sup>[16]</sup> the blood supply of the bone plate.

#### 3.2 Minimally invasive surgery

In the Nuss procedure, the appropriate steel plate was selected according to the ideal shape and size of the chest before operation. The lowest part of the sternal depression was marked, and a transverse incision<sup>[17, 18]</sup>With the assistance of thoracoscopy, the gap between the lower part of the sternal depression and the front of the pericardial was separated, and the expansion clamp was crossed behind the sternum and threaded through the opposite side. The plate was connected to the expansion clamp and dragged behind the sternum. Then, after the brace was in place, the bow-back was turned 180 degrees upward, so that the sternum and anterior chest wall protruded into the desired state<sup>[17]</sup>. Finally, the two ends of the support frame were set into the fixator and sewn on the rib periosteum. The retrosternal drainage tube was placed and removed<sup>[19]</sup>The average length of hospital stay was 5 to 7 days, and the stent<sup>[20]</sup>The Nuss procedure has the advantages of an 85 excellent and good rate and a 10 recurrence rate<sup>[20, 21]</sup>. The disadvantages of the Nuss procedure include that the plate will be tightly connected with the lung and heart and react<sup>[20, 22, 23]</sup>

The most important difference between the Wang procedure and the Nuss procedure is the placement<sup>[24, 25]</sup>In Wang's procedure, the risk of surgery is eliminated<sup>[25]</sup>In addition, Wang's operation time is shorter, the average operation time is 30 to 50 minutes, no drainage is needed, the patients can be discharged after 5 to 6 days, and the support bar is removed after 1 to 2 years. The postoperative effect is satisfactory, the risk factor is low, and there is no additional pleural damage<sup>[22, 25]</sup>

The fixation site of external thoracic suspension fixation was selected at the lowest point of thoracic depression, and the Kirschner wire with a diameter of 2.5 mm was selected as the external thoracic support rod <sup>[26]</sup>. The skin and subcutaneous tissue were cut, the muscles were separated, the xiphoid process was lifted, and the sternum<sup>[27]</sup> Finally, the thread was lifted and fixed on the chest wall support rod. Postoperative management was strengthened, and the vital<sup>[26]</sup> signs were monitored by an electrocardiogram monitor.

#### 3.3 Non-invasive surgery

Noninvasive surgery for pectus excavatum mainly uses negative pressure suction cups, which can be used as postoperative adjuvant treatment. The suction cup can be selected according to the range of the funnel chest depression (three sizes of suction cups are available for selection). The suction cup is close to the edge of the chest wall depression, and the negative pressure (lower than 15% of the surrounding atmosphere) is generated

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by a manual exhaust pump. When the negative pressure is generated, the sternal elevation can be seen, generally not less than 2 times/day, 30 minutes/time<sup>[28]</sup>The negative pressure suction cup is suitable for patients with mild pectus excavatum, patients with pectus excavatum who have not reached the surgical age, patients with recurrence after surgery, patients who refuse surgical treatment, and patients with stiff chest wall to prepare for surgery<sup>[28]</sup>. However, negative pressure suction cups also have side effects, including skin bleeding or subcutaneous hematoma at the site of use, some patients will have back pain, transient upper limb pathetic, and rib fracture. It can be used by reducing the negative pressure or time to adapt to the suction cup treatment, and if there are very serious side effects, the use of suction cup treatment<sup>[28]</sup> should be suspended.

### 4. Conclusions:

This article summarizes the recent Chinese and English literature on the diagnosis and treatment of pectus excavatum and compares the diagnostic indicators and surgical techniques of pectus excavatum. Based on the analysis of the literature content, we found that there are many controversial points in the surgical treatment of pectus excavatum in clinical practice, such as the Nuss procedure has replaced the traditional open Ravitch procedure and become the most commonly used technique for correction of pectus excavatum. However, the incidence of plate displacement is the highest among the complications of minimally invasive surgery, followed by allergic reactions and infections. However, it is necessary to be aware of rare complications such as cardiac perforation, and large vessel injury. Therefore, it is recommended that patients with heart diseases should be treated with pectus excavatum correction surgery at the same time under the guidance of a multidisciplinary team. However, the modified Ravitch procedure can provide more room for cardiac surgery, and other deformity correction procedures can also be performed at the same time as thoracic correction. In conclusion, this study summarizes the advantages and disadvantages of each surgical procedure to select appropriate evaluation indicators and individualized surgical procedures for patients with pectus excavatum. In the future, the surgical treatment of pectus excavatum will be proactive, and the patients will not be limited by age and condition, and achieve more perfect orthopedic effects in a more minimally invasive and more efficient way.

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