CHCMU
Children's Hospital of Chongqing Medical University
重庆医科大学附属儿童医院

教师简介

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先天性漏斗胸

(Pectus Excavatum)

Objectives： To master the clinical manifestations, diagnosis and treatment principles of PE, be familiar with the surgical treatment of PE, and to understand the causes of PE in children.

Key points： Clinical manifestations, diagnosis and treatment principles of PE.

Difficulties： Surgeries

一、 概述(Overview)

1. 概念(Definition)

Pectus excavatum is a congenital deformity of the anterior thoracic wall in which the sternum and rib cage grow abnormally. This produces a caved-in or sunken appearance of the chest. It is sometimes referred to as cobbler's chest or funnel chest.

2. 流行病学 (Epidemiology)：

Funnel chest is the most common congenital chest wall deformity, accounting for 90% of congenital chest wall deformity, with an incidence rate of 0.1% to 0.4%. The ratio between men and women is about 4-5:1.

3. 自然病程 (Natural history)：

The deformity often aggravates gradually, rarely subsides by itself.

4. 治疗(Treatment)：

Operation is the “only” effective way to treat funnel chest.

二、病因 (Causes)

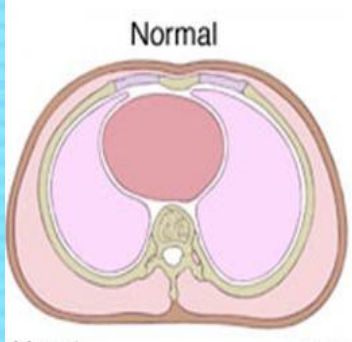
1. Funnel chest is a congenital developmental abnormality, the cause of which is unsure but assume that there is a genetic component for at least some of the cases. Approximately 10%-20% of individuals with PE have a first degree family member with the condition.

2. Relevant theories of pathogenesis：

- Overgrowth of Lower thoracic costal cartilage.
- Shortage of central tendon of diaphragm.
- Long-term inspiratory dyspnea.

3.PE is irrelevant to calcium deficiency.

三、病理（Pathology）



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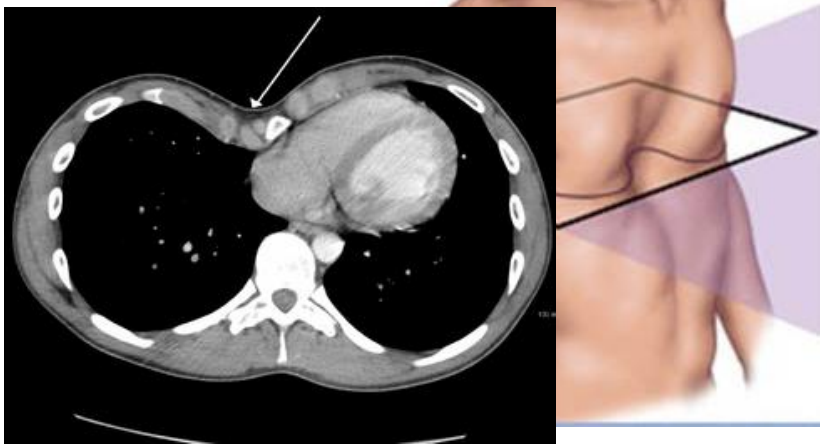
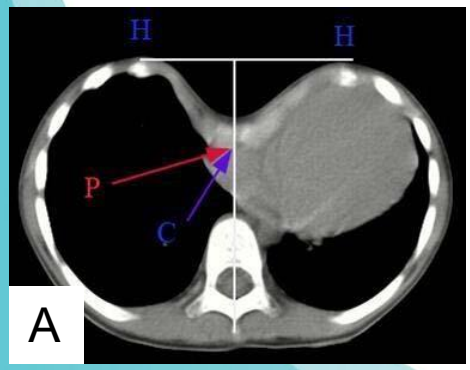


- **Chest wall deformity: Invagination of he lower end of the sternum and the connected ribs, rib cartilages.The anteroposterior diameter of the thorax becomes shorter.**
- **Displacement and deformation of the thoracic viscera.**

➢ 病理分型（Pathological type)：

（1）是否对称:① 对称型（Symmetric type） ②非对称型(Asymmetric type) （2）根据凹陷范围:① 广泛型(Extensive type) ② 局限型(Localized type)

Each type can be accompanied by sternal rotation and scoliosis.



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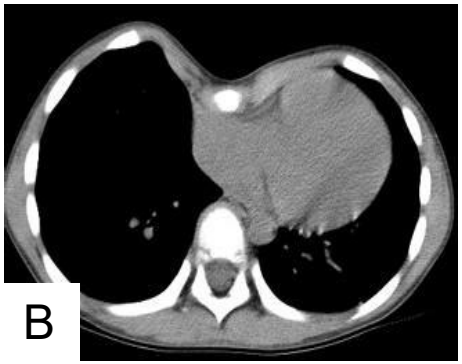

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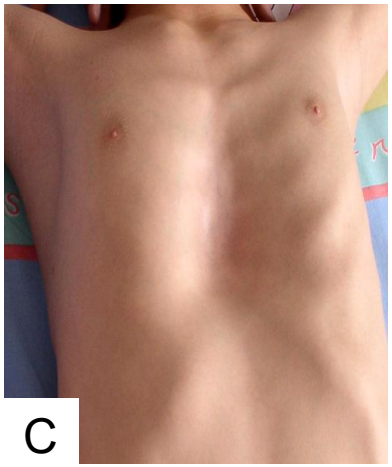


C

D

Symmetric type　　Asymmetric type　　广泛型　　局限型

四、病理生理（Pathophysiology）

1. 肺功能损害(Lung function damage)

- 通气功能损害（Ventilatory impairment）
- 通气储备功能降低（Decreased ventilatory reserve function）

2. 心功能损害(Cardiac dysfunction)

- 舒张功能受损（Impaired diastolic function）
- 瓣膜及传导功能异常（Valve and conduction dysfunction）
- 心脏储备功能降低（Decreased cardiac reserve function）

五、临床表现(Clinical manifestation)

（一）症状（Symptoms）

1. The hallmark of the condition is a sunken appearance of the sternum. The most common form is a cup-shaped concavity, involving the lower end of the sternum and costal cartilages. It can either be present at birth or not develop until puberty.

Patients may also experience chest and back pain, which is usually of musculoskeletal origin.

The symptoms also include recurrent respiratory infections, wheezing or coughing.

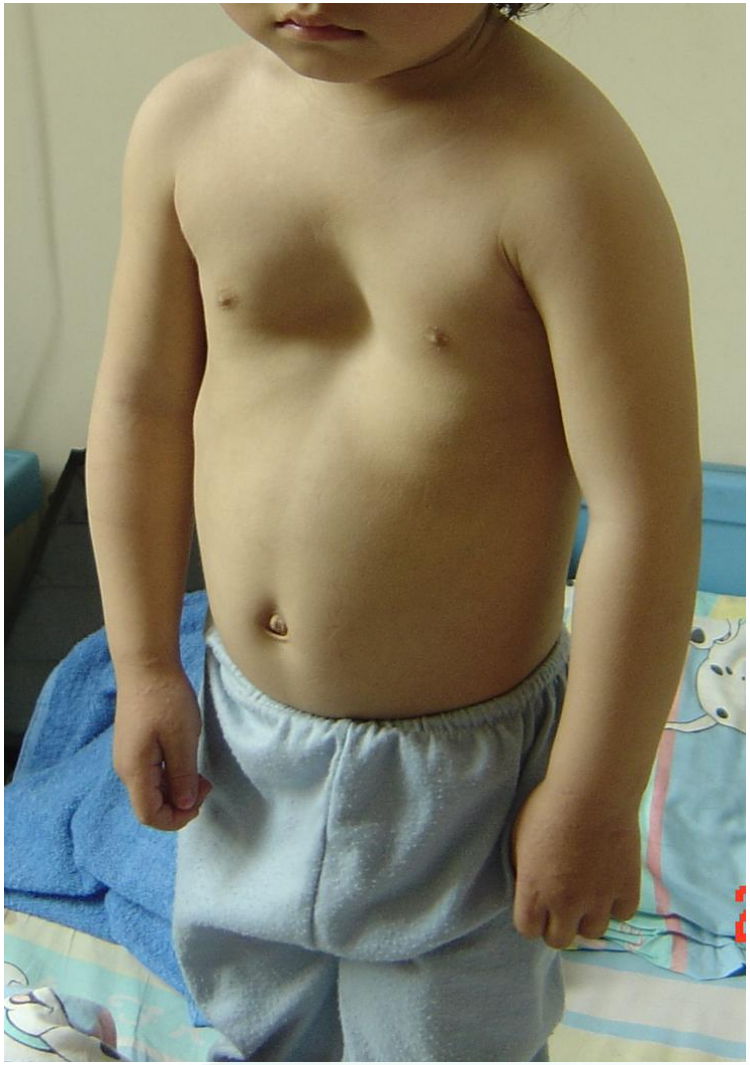

Pectus excavatum is a progressive deformity. This patient progressed from mild to severe & asymmetric before puberty. At the time of puberty, the chest becomes more rigid and patients start to exhibit cardiopulmonary symptoms.（漏斗胸畸形常随年龄的增长而逐渐加重。）

2. In mild cases, cardiorespiratory function is normal, although the heart can be displaced and/or rotated. In severe cases, mitral valve prolapse may be present and physical capability may be limited due to base lung capacity being decreased.

3. Psychological symptoms manifest with feelings of embarrassment, social anxiety, shame, limited capacity for activities and communication, negativity, intolerance, frustration, and even depression.

（二）体征（Signs）

1. Visual examination： Sunken appearance of the sternum,hunched back,anteverted shoulders,bulging abdomen.（漏斗胸体征：前胸凹，背后弓，肩前倾，腹膨隆）



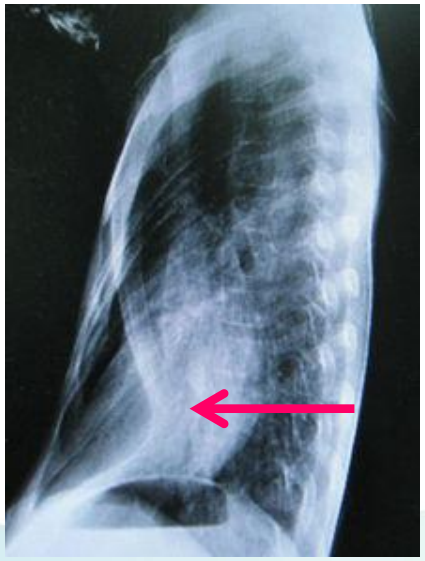
2. Auscultation and palpation：

Auscultation of the chest can reveal displaced heart beat and valve prolapse. There can be a heart murmur occurring during systole caused by proximity between the sternum and the pulmonary artery.Lung sounds are usually clear yet diminished due to decreased base lung capacity.

六、 辅助检查（Auxiliary examina）

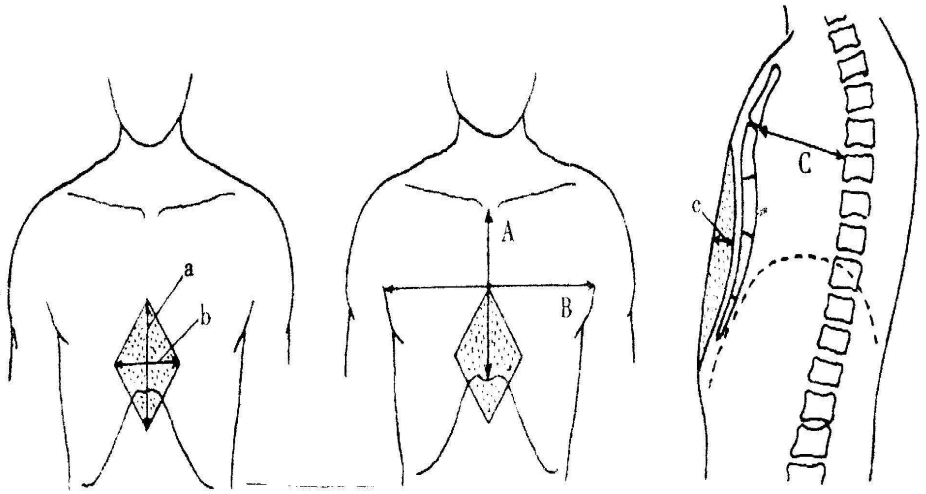
1. Chest x-rays：

This test can visualize the dip in the breastbone and often shows an apparent increased density overlying the right anteromedial aspect of the right middle lobe . The heart and mediastinum were displaced to the left.



漏斗指数（Funnel index，FI）

$$FI = \frac{a \times b \times c}{A \times B \times C}$$



a：漏斗口纵径长度

b：漏斗口横径长度

c：漏斗口水平线至凹陷最深处的长度（深度）

A：胸骨的长度

B：胸部的横径

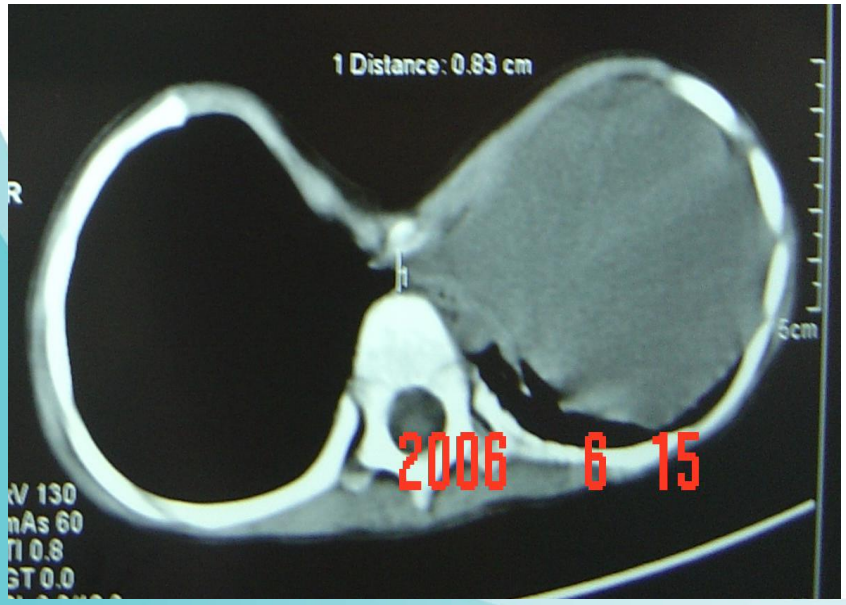
C：胸骨角后缘至椎体前缘的最短距离

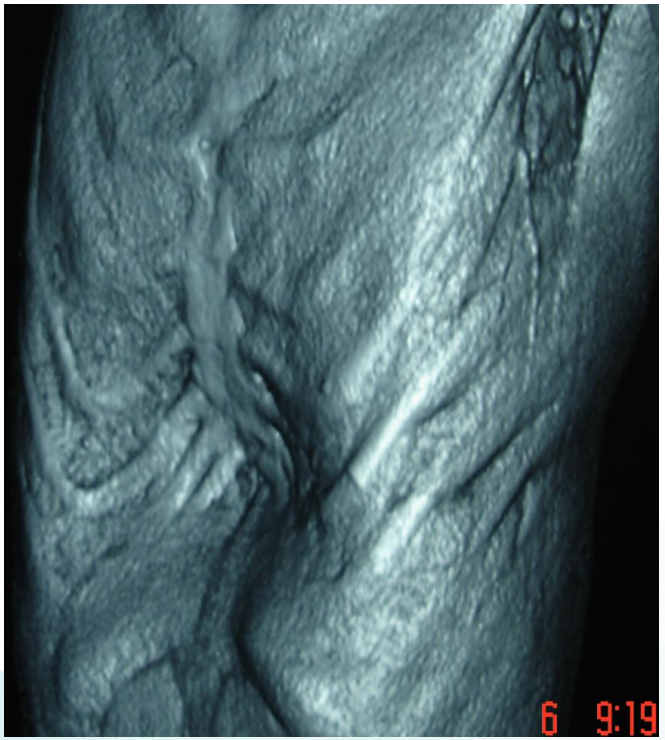
Grading: Mild degree: < 0.2

Moderate degree: 0.2~0.3

Severe degree: >0.3

2. CT： **CT scanning may be used to help determine the severity of the pectus excavatum and whether the heart or lungs are being compressed. CT scans take many X-rays from a variety of angles to produce cross-sectional images of the body's internal structure.**





The Haller index is the ratio between the horizontal distance of the inside of the ribcage and the shortest distance between the vertebrae and sternum.

Haller指数=AB/CD

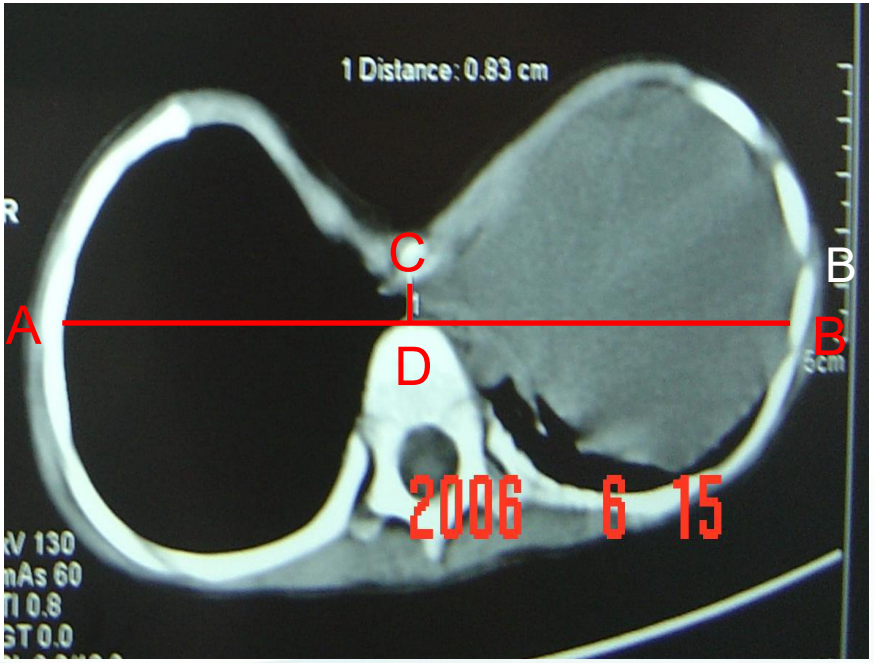
Grading:

Normal value: 2.52 (Average)

Mild degree: < 3.2

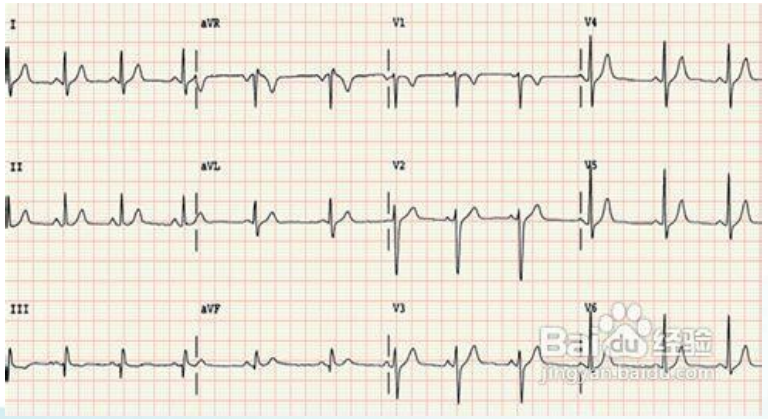
Moderate degree: 3.2～3.5

Severe degree: >3.5



3.心电图(Electrocardiogram)：

An electrocardiogram can show whether the heart's rhythm is normal or irregular. This test is painless and involves the placement of more than a dozen electrical leads, which are attached to the body with a sticky adhesive. 心电图可以显示心律不齐、T波改变、右束支传导阻滞，电轴左偏等异常。



4.超声心动图(Echocardiogram):

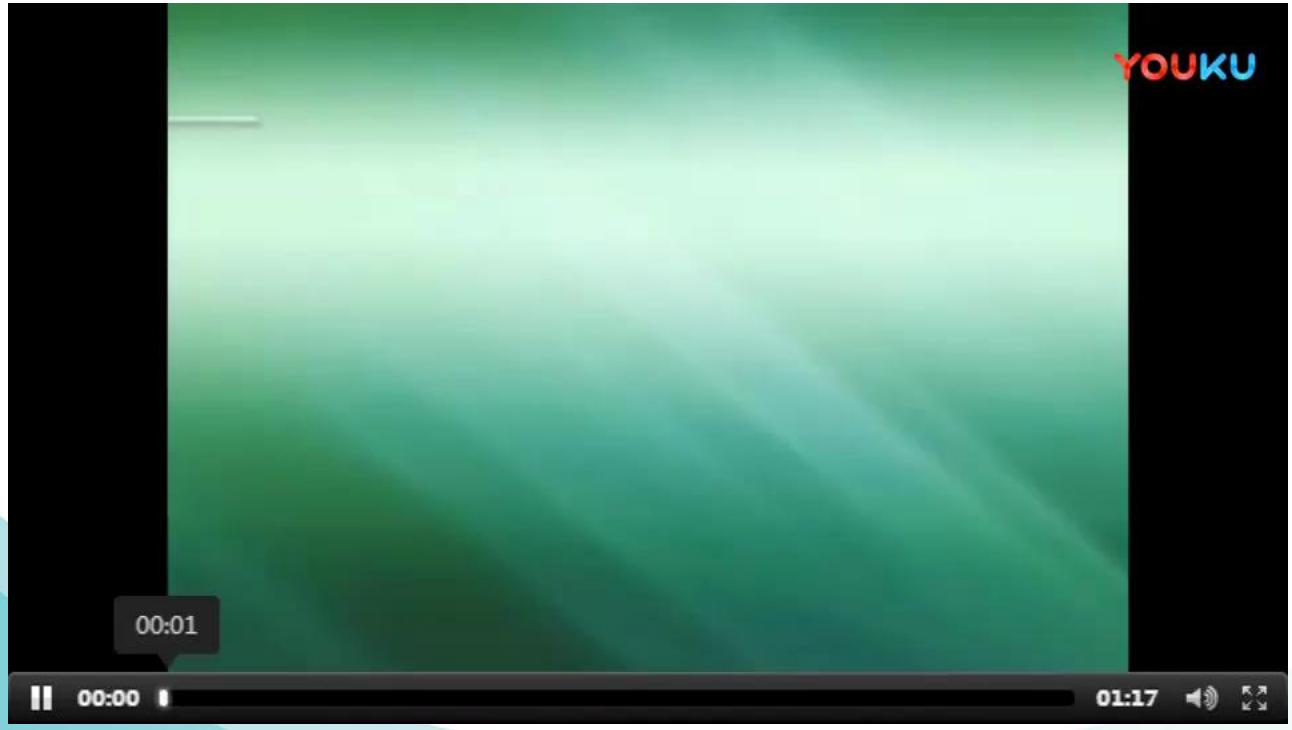
An echocardiogram is a sonogram of the heart. It can show real-time images of how well the heart and its valves are working. The images are produced by transmitting sound waves via a wand pressed against the chest.漏斗胸患者可以出现心腔受压变形、心脏移位、瓣膜反流甚至心功能降低。

5. 肺功能检查(Pulmonary functions):

These types of tests measure the amount of air the lungs can hold and how quickly the testee can empty his/her lungs. 漏斗胸患者可出现小气道阻塞、通气储备功能降低和其他肺功能损害。

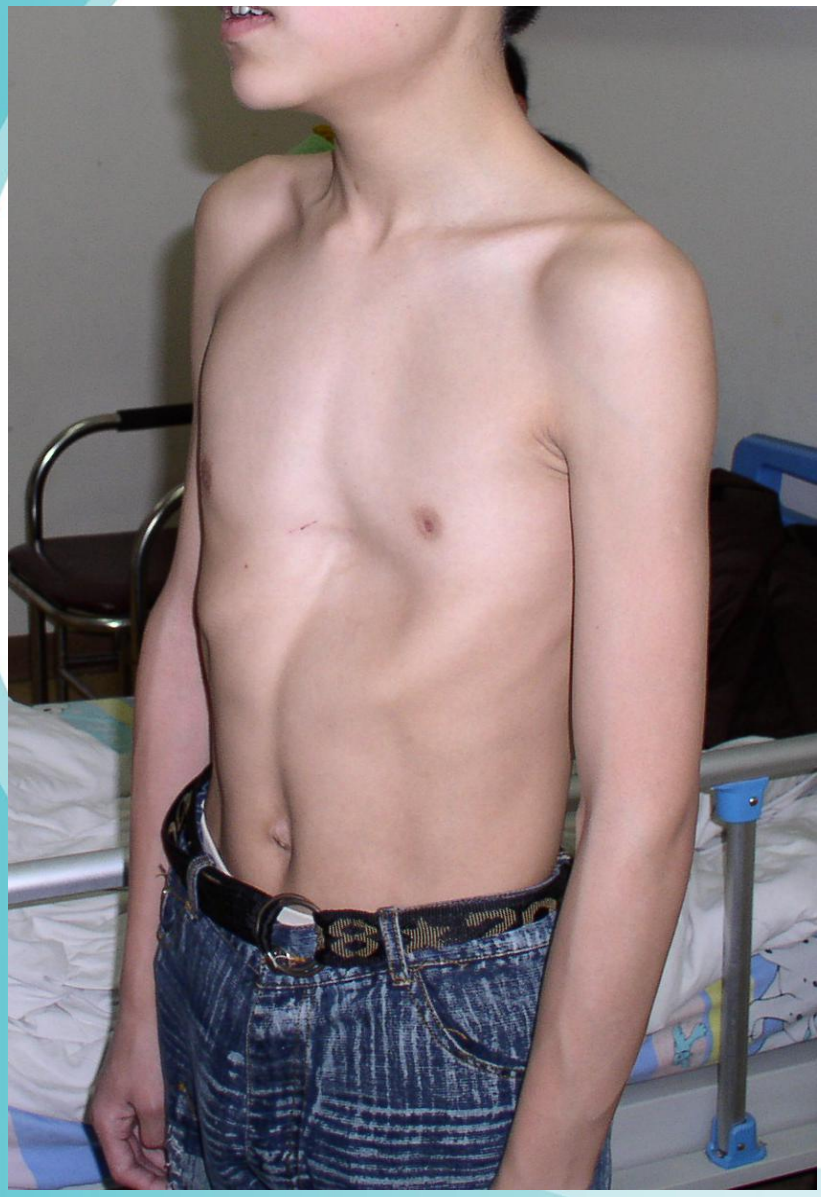
6.运动实验（Exercise test）

This test monitors how well the heart and lungs function while the patient exercises, usually on a bike or treadmill.漏斗胸患者的运动功能常存在不同程度的下降。

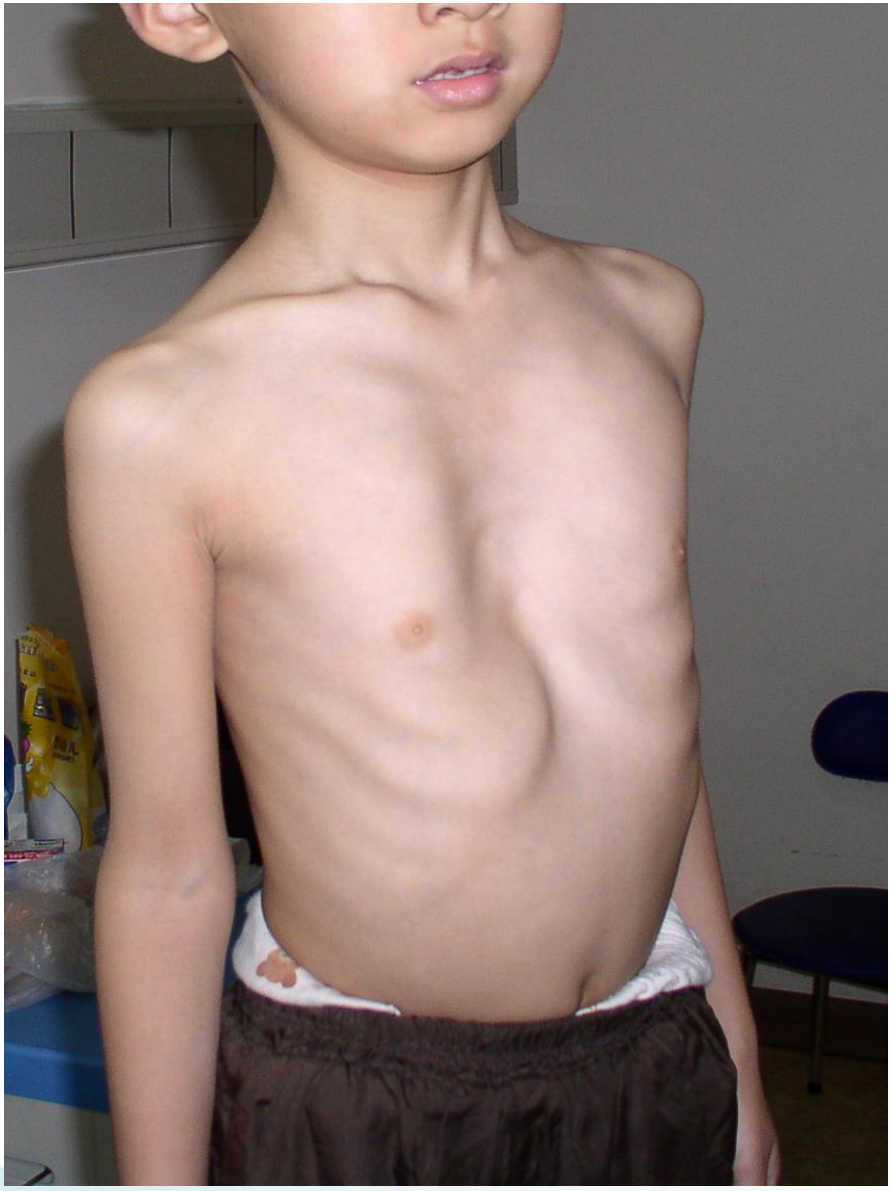


七、诊断(Diagnosis)

Pectus excavatum can usually be diagnosed simply by examining the chest without differential diagnosis. The diagnosis should include the degree of deformity and the associated abnormalities, such as Noonan syndrome, Marfan syndrome and Scoliosis.漏斗胸可以通过简单的体格检查获得诊断，且无鉴别诊断。但详尽的诊断需结合相关的辅助检查，明确严重程度的分级，以及合并的其他畸形，比如Noonan综合征、Marfan综合征、脊柱侧弯。



局限型、不对称型



广泛型、对称型

八、治疗(Treatment)

Pectus excavatum requires no corrective procedures in mild cases. Treatment of severe cases can involve either invasive or non-invasive techniques or a combination of both.

（一） 手术治疗（Surgical treatment）

1. 手术目的（Objevtives）

(1) To correct thoracic deformity, relieve cardiopulmonary compression, and improve cardiopulmonary function.

(2) To prevent the development of funnel chest signs.

(3) To remove the mental disorder caused by the deformity.

2. 手术适应证（Indications for surgery）

(1) Moderate to severe chest deformity.

（Haller Index > 3.2 ）

(2) Evidence of cardiopulmonary dysfunction.

(3) Parents or patients have strong desire for surgical treatment.

3. 手术时机(Operative timing):

(1) 1-3 years：Surgery at this age is generally not recommended unless severe symptoms and signs.

(2) 3-12 years：Most surgeons recommend this age group as the best time for surgery.

(3) >12 years: Surgery can be performed, often with more serious postoperative pain because of the stiffer chest wall.

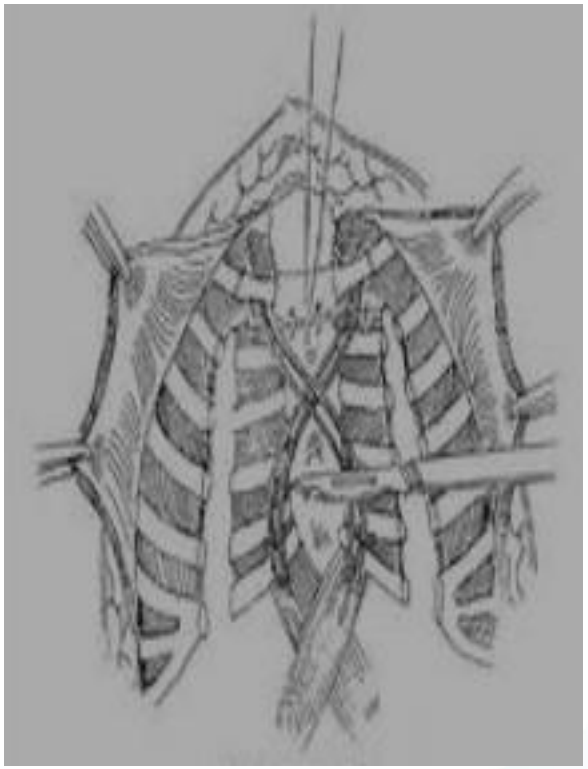
4. 手术方式（Operative methods）：

The surgical treatment of funnel chest has a history of more than 100 years with several techniques applied.

- **Sternal turnover（胸骨翻转术）**
- **Ravitch technique（胸骨上举术）**
- **Minimally invasive repair of pectus excavatum (MIRPE) or Nuss procedure（微创矫正手术（Nuss手术））**

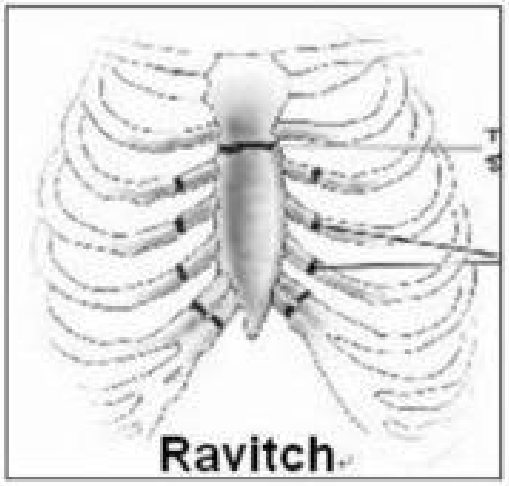
(1) 胸骨翻转术（Sternoturnover，Wada，1944）

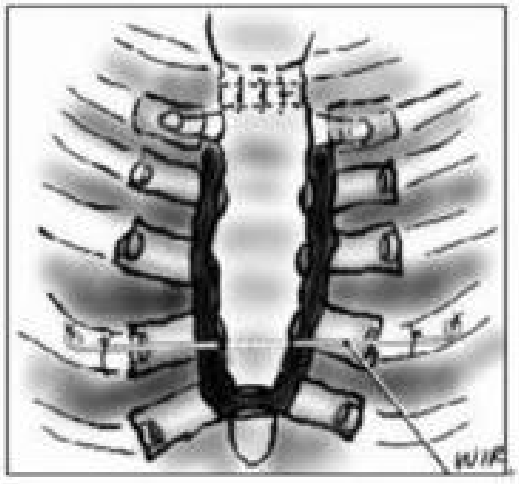
Disadvantages: Relatively more trauma and bleeding. Complications include abnormal respiration, sternal infection, necrosis, and dysplasia of costal cartilage can occur after operation. Recent years, it has been basically eliminated.



(2) 胸骨上举术（sternal elevation， Ravitch，1949）

The Ravitch technique is an invasive surgery that was introduced in 1949 and developed in the 1950s to treat the condition.

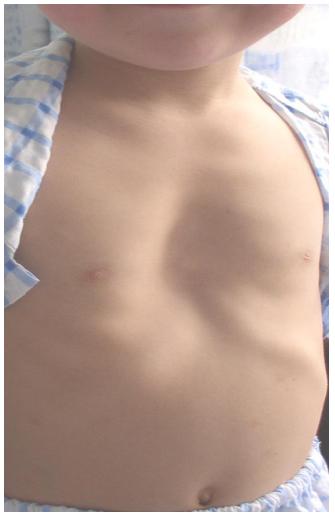


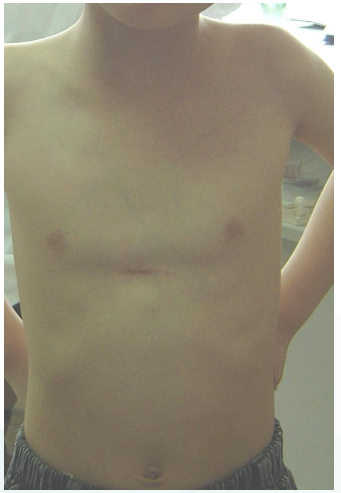


Advantages: Compared with sternoturnover, Ravitch technique is simpler and less traumatic.

Disadvantages: Compared with the minimally invasive Nuss procedure, the trauma is more invasive, and the cosmetic effect is less ideal with the longer incision.

改良Ravitch手术（Modified Ravitch technique）





In recent years, our department has adopted self-designed steel plate for anterior sternal suspension with a small incision with or without severing sternum. Compared with classical Ravitch technique, the modifed procedure has less trauma with less(0.5-1.5cm) or not removal of costal cartilage, and the outcome is excellent.

(3) 微创漏斗胸矫正术（Minimally invasive repair of pectus excavatum (MIRPE), 1997）

MIRPE, widely known as the Nuss procedure, involves slipping in one or more concave steel bars into the chest, underneath the sternum. The bar is flipped to a convex position so as to push outward on the sternum, correcting the deformity. The bar usually stays in the body for more than two years.The Nuss procedure is a two-stage procedure (two surgeries, 2-5 years apart).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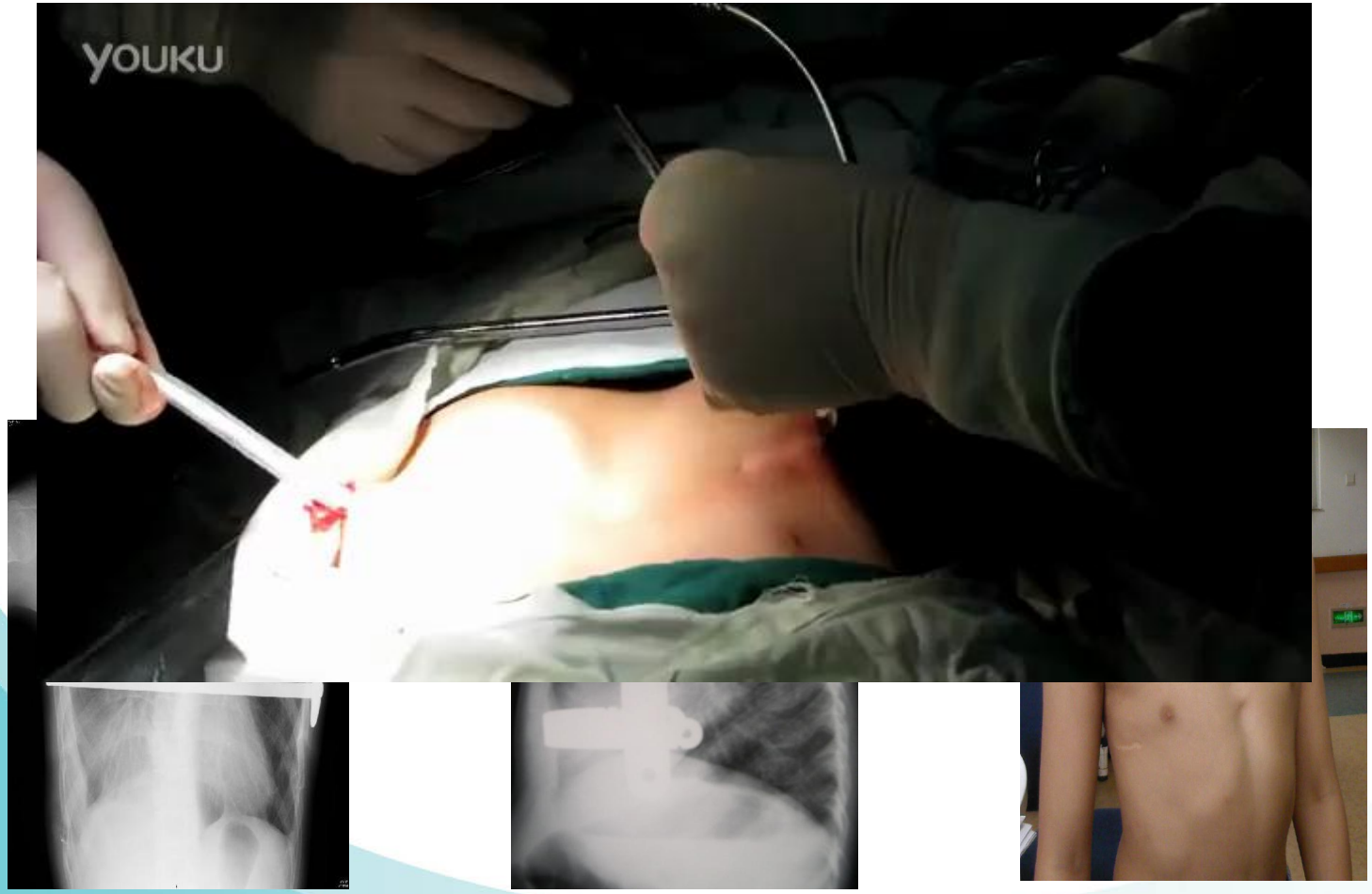
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Nowadays, surgery design assisted by 3D printing technology is applied in the treatment of funnel chest, which has improved the operation outcome.

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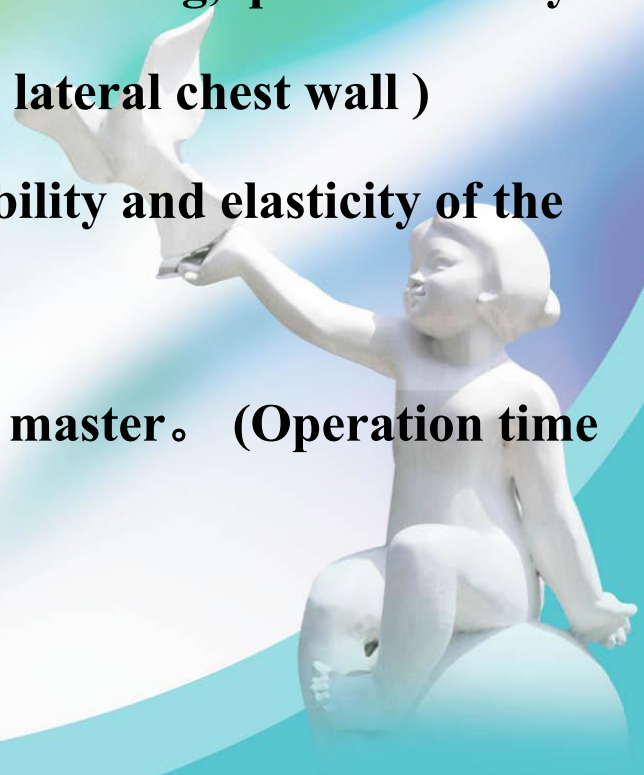
Advantages of Nuss procedure:

- Minimally invasive with no osteotomy, less bleeding, quick recovery.
- Good looking of incision. (Incision is on the lateral chest wall)
- It can maintain the stretch, expansion, flexibility and elasticity of the chest for a long time.
- Nuss procedure is quite simple and easy to master。 (Operation time is about just 20-30 minutes) .



Disadvantages of Nuss procedure:

1.Complications may be serious, such as pneumothorax, lung injury, heart rupture.Use of thoracoscopy can basically avoid accidental injury).

2.Operation age is limited (> 3 years).

3.Orthotic plates are more expensive。

Case 1

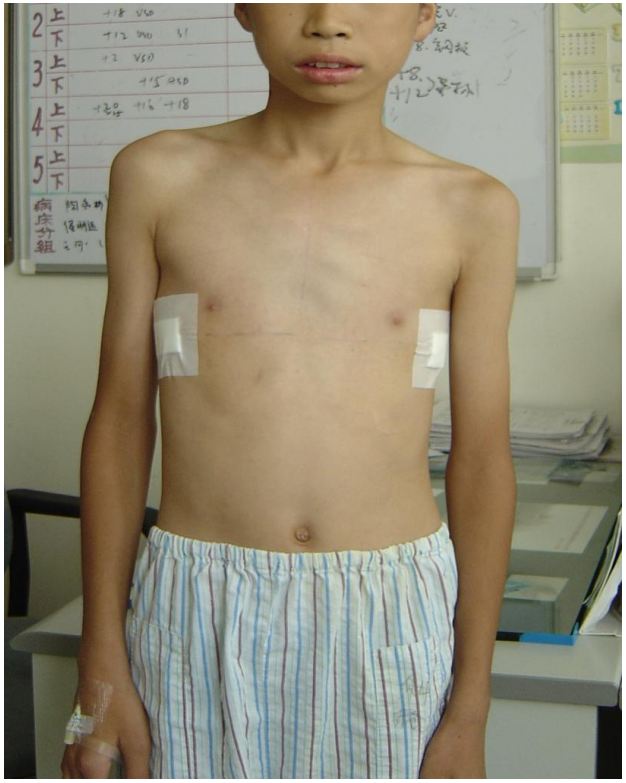
Before operation

1w after operation

1.5y after operation

（Male, 1y and 8m, Modified Ravitch technique）

Case 2



Before operation **1w after operation**

（Male, 13y, Nuss procedure ）

Case 3

Before operation **3y after operation**

（Male, 9.5y, Nuss procedure ）

(4) 整形美容手术（ Plastic surgery，Cosmetic surgery）

- From a purely morphological perspective, implant procedure is simple, reliable, and minimally intrusive, but does not correct existing cardiac and respiratory problems.Plastic surgery is indicated in patients who wish to avoid corrective surgical procedure.

Silicone implant for PE

3D image of custom-made implant to treat PE

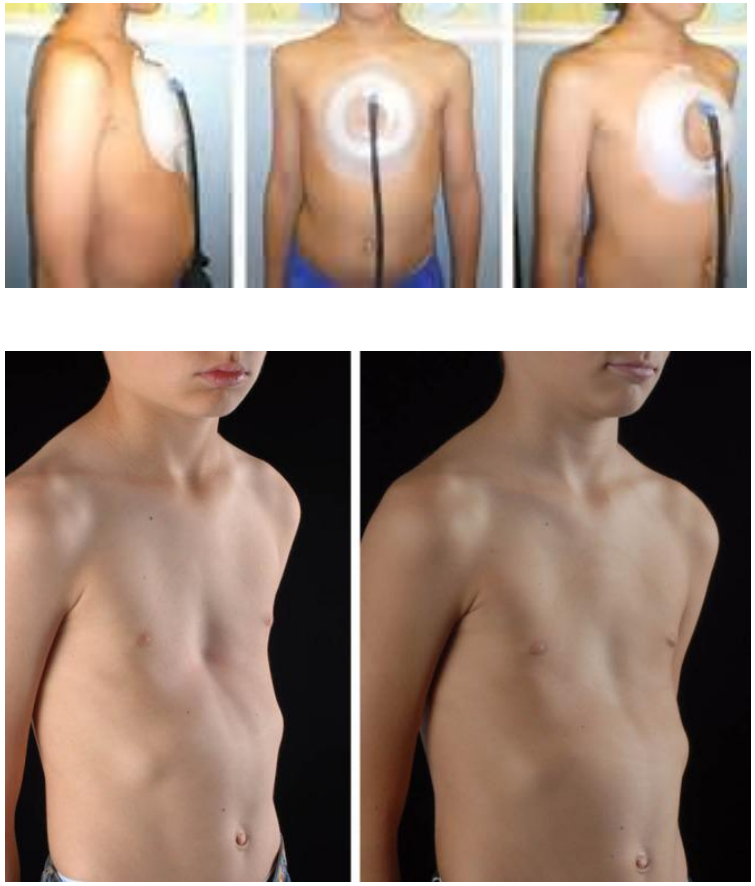
Woman with pectus excavatum with an implant

（二） 非手术治疗（No-surgical treatment）

(1) 真空钟治疗（Vacuum bell）

VB therapy is indicated in patients who present with mild PE and/or wish to avoid surgical procedure.

Vacuum bell for pectus excavatum treatment, with hand pump (left) and measuring rod (right)



Male, aged 10, before (left: depth of PE =2.2 cm) VB therapy, and 12 months after cessation of VB therapy (right: depth of PE =0.5 cm); duration of therapy: 36 months.

（2） 运动锻炼（Exercise）

- Physical exercise is not seen as a means to resolve the condition on its own, but has an important role in PE treatment programs.
- Exercises are aimed at improving posture, strengthening back and chest muscles, and enhancing exercise capacity, ideally also increasing chest expansion. Pectus exercises include deep breathing and breath holding exercises, as well as strength training for the back and chest muscles.
- Aerobic exercises are helpful to improve cardiopulmonary function.

小结（Summary）

- Funnel chest is the most common congenital chest wall deformity。
- The cause of PE is unsure, but it is irrelevant to calcium deficiency.
- The hallmark of PE is a sunken appearance of the sternum, with a degree of impairment in cardiopulmonary function, and Psychological problems.
- Pectus excavatum can usually be diagnosed simply by examining the chest, and it's important to evaluate the degree.
- Surgery is the “only” effective way to treat funnel chest.
- At present, the most mainstream treatment: Nuss procedure.

➢ Questions：

1.What are the clinical manifestations of funnel chest? How to evaluate the severity of deformity?

2.When and how to perform surgery for PE?

➢ Main reference materials：

1. 《小儿外科学》（第五版）；

2.Sabiston and Spencer Surgery of the Chest(Eighth Edition).


谢谢！