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Surgical and conservative treatment of pectus excavatum

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Abstract

Pectus excavatum is one of the most common chest defect in children. It is characterized by a collapsed sternum, reduced thoracic volume and cardiovascular and respiratory symptoms. In order to determine the degree of deformation and its type, diagnostic management is necessary, including imaging tests such as CT scans, from which the degree and nature of the defect can be determined, as well as other tests to determine the impact of the defect on the patient's functioning. Treatment options include non-invasive methods and invasive methods such as the minimally invasive Nuss procedure or Ravitch procedure. The gold standard of treatment is the Nuss procedure, but in some cases, conservative treatment may be an alternative. Each method has its advantages and disadvantages, and the treatment method is selected after talking to the patient, determining his expectations, age, severity of the defect and its nature.

Purpose

This study reviews anatomy, diagnostic methods and treating procedures to propose the most efficient way of treating this kind of chest deformation.

Materials and Methods

A review of current literature was conducted, focusing on diagnostic tools such as physical exams and imaging. Criteria for conservative versus surgical management are discussed. Most common and gold standard in surgical techniques were evaluated. Literature available in the PubMed and Google Scholar databases was searched using keywords.

Results

The most common surgical procedure in pectus excavatum treatment is Nuss procedure. Currently it's the most efficient and least invasive method of treatment with the highest success rate. That's why it's proposed as gold standard of treating pectus excavatum.

Keywords

Pectus excavatum, Funnel chest, treatment options, outcome

1. Introduction

Pectus excavatum is one of the most common chest deformities in children and accounts for as many as 90% [1] of all chest defects. It occurs 4 to 5 times more frequently in boys than girls [1,2]. Its characteristic anatomical structure with a collapsed sternum, if left untreated, may cause many ailments for the patient in the future.

The clinical picture and complaints presented by patients can be divided into four categories. The first group comprises asymptomatic patients, second group patients with pain, third patients with reduced exercise tolerance and fourth patients complaining of aesthetic defects [2]. The etiology of the disease remains obscure. This results in many known hypotheses for the development of the disease. Starting with histological causes, including abnormal collagen structure, genetic and anatomical diseases [3]. Multiple genetic diseases have scientifically proven predisposition to develop pectus excavatum, such as Marfan syndrome, Ehlers Danlos syndrome, Noonan syndrome or Poland syndrome [4]. What's more, this defect often coexists with mitral valve prolapse or scoliosis [5,6].

Most often, this defect manifests during a period of faster growth. The decision to treat the disease with one of available methods is most often made in adolescence. It can happen that treatment is not carried out at this time for various reasons. There is still a chance for surgical treatment in older age but it is much less common and is associated with more frequent postoperative complications [7]. Despite the evolution of surgical techniques, the currently used minimally invasive methods are still burdened with complications [8]. Therefore, despite the generally accepted gold standard, which is the Nuss method (MIRPe), the non-invasive techniques are also worth considering. The aim of the study is to compare the effects of surgical and conservative treatment, which may allow a better selection of the treatment method for patients.

2. Diagnostic methods

One of the most common and widely used diagnostic tools used to assess and potentially classify a patient for treatment is the Haller index (HI), calculated by taking the ratio of the largest internal width of the chest and the distance between the spine and the sternum[9]. The gold standard is to perform the measurement based on a thorax CT image, but is also possible on the basis of MRI or X-ray. A result below 2.5 is considered the norm and a result >3.25 is assumed to be one of the qualifying factors for surgery. An independent HI assessment cannot be used to decide about surgical treatment, without taking into account other factors such as the patient's symptoms, well-being, age, gender, circulatory and respiratory problems. The Haller Index does not include many variables, causing limitations in its use.

In addition to HI, a modified PCI (Pectus Correction Index) is often used, as an additional element to assess the severity of thoracic deformity, and correlates better, especially with an asymmetric defect, compared to HI [5,10]. For PCI, the width is not measured, but two distances from the tangent line to the anterior surface of the vertebrae. One distance is the same as in HI, i.e. the distance measured from the tangent line with the spine to the sternum, and the other is the distance between the line and the ribs at the widest point. We use the following formula to obtain PCI.

PCI scores in healthy individuals will be close to 0, while those with a pedicle defect above 0. PCI greater than 10 will already be visually apparent. HI and PCI have a close correlation and it is assumed that an HI of 3.25 may correspond to 28 PCI[9]. The PCI itself is more accurate and describes the size of the pectus excavatum, but does not indicate when the procedure should be performed.

In the case of patients with an asymmetric defect, an asymmetry index (AI) can additionally be used to assess this asymmetry. Based on the CT image, we measure between the anterior and posterior surfaces of the ribs on the left and right side of the sternum. Then we divide the score on the right side by the score on the left side and multiply by 100 $(R/L) \times 100$. The more the result exceeds 100, the greater the asymmetry.

In addition to tests and indicators determining the defect itself, tests that determine the impact of the defect on the patient's life and function can be also used.

If cardiovascular or respiratory symptoms occur, the diagnosis should be supplemented with spirometry, echocardiography, electrocardiography or blood tests[11]. In an echocardiographic examination, we look for critical changes in the heart or vessels, especially in high-risk people such as those with Marfan syndrome. [5,12,13] Problems that may arise include changes in

mitral valve or right ventricle or compression of the right ventricle and right atrium resulting in reduced blood flow.

The 12-lead ECG should be performed and may show right bundle branch block, right atrial and right ventricular hypertrophy, and supraventricular arrhythmias.

Spirometry results may indicate a restrictive defect.

Patients may show elevated LDH levels in blood tests.

3 Methods of treatment

In the case of a pectus excavatum, we can distinguish between two therapeutic approaches to the treatment of the defect: surgical and conservative. In the case of surgical treatment, a distinction is made between the minimally invasive Nuss method, which is the gold standard of treatment, and the Ravitch method. For conservative treatment, there is the Eckart Klobe method, which can be combined with rehabilitation. Each method has its own characteristics, a different technique and is applied after individual examination and selection according to the defect and the patient's approach.

3.1 Conservative methods - Eckart Klobe and rehabilitation

This method was initiated in 1910 by Lange and then improved over many years by Haecker and Klobe [14]. VBT (Vacuum bell therapy) is a non-invasive method using a specially designed vacuum pump and sternum lift applicator in a protocol of use adapted to a given patient.

The applicator itself is available in 3 sizes (16, 19 and 26 cm) which are selected individually for each patient. In the initial stage of treatment, the applicator should be used twice a day for 30 minutes over a period of 4-6 weeks.

Then this time can be extended to a maximum of a few hours. Patients with moderate pectus excavatum and those who wish to avoid surgery for various reasons are mainly eligible for this procedure [15]. The main contraindications to this type of procedure are cardiological, hematological contraindications and coagulopathies [16]. The most common complications when using this method are hematomas, petechiae, back pain and upper limbs paresthesia [17]. While wearing the brace, the patient may experience discomfort, pressure or low-intensity pain that persists for the first few days of use. The age of patients qualified for this method has not been strictly defined, the main group are patients under 10 years of age, but due to the small amount of data, far-reaching conclusions cannot be drawn about the use of the device in adults. It was possible to distinguish three main groups of patients in whom the above-mentioned conservative treatment method has been used. Group I are young patients with a flexible thorax and the distance of the sternum from the spine of less than 3 cm, with a treatment duration 12-15 months, group II, adults with a less flexible thorax and the distance of above 3 cm with a treatment duration from 24 to 36 months. Group III, patients with an asymmetric pectus excavatum with high stiffness. This group of patients is at the highest risk of failure of conservative treatment. The results of conservative treatment were not significantly different from those of surgical treatment [17]. Treatment using the Eckart Klobe method can be supplemented with physical, respiratory and cardiac rehabilitation. A suitably prepared personalized rehabilitation protocol can result in 50% greater sternal lift than the use of the device alone [18]. In the case of a pectus excavatum, rehabilitation alone is not recommended as the main treatment method, although results can also be satisfactory and the deformation can be reduced by up to 60% after 3 months [19]. However, due to the lack of research and data,

further research in this area is needed. On average, it can be assumed that correction of the defect using VBT is achieved after 22 month, and it has been shown that no regression of the defect was observed at the end of treatment and in follow-up after an average of 27 months [20]. Long-term studies are needed to see how long-lasting the effects of treatment are.

3.2 Surgical methods

The main indication for surgical treatment of pectus excavatum is the symptomatic form of the disease [1,21], i.e. respiratory or circulatory symptoms resulting from the compression of the deformed thorax on the mediastinal structures. Characteristic symptoms of funnel chest include [1,5,11,21,22,23,24,25]:

When the defect is asymptomatic, it requires monitoring of progression every 1-2 for rapid detection of changes [11,12]. Objective assessment of respiratory and cardiovascular abnormalities may include reduced spirometry parameters, reduced Vo2max, diagnosed restrictive lung disease, right-sided cardiac compression in ECG or imaging tests, or shift of the heart to the left side of the chest [6,11]. Less common indication is an asymptomatic form of the disease, in order to remove a cosmetic defect, which, however, can cause many mental problems in adolescent children. [11,21,25] According to some sources, the indication for surgery is Haller index >3.25 [6,11] or correction index $>28\%$ [26] or $>20\%$ [27]. However, it is not the established values, but the occurrence of symptoms or impaired organ function on functional and imaging tests that indicate surgical treatment [2,9]. When planning surgical treatment, children are most often targeted during puberty; the surgery is optimally performed at the age of 10-14 [1,18,28,29,30], the average age is 13.5 [1]. This age allows the child to grow and develop properly and reduces the risk of recurrence. [14,20, 22]. The older age of patients does not disqualify them from surgical treatment. Surgical procedures are also the standard of care in this group of patients, but the higher risk of complications and the lower risk of complete recovery compared to the pediatric population must be taken into account. [5,11,22,24,29]. The Nuss procedure with special modifications adapted to this group of patients and reducing the risk of complications in this group [8,24,29] and the Ravitch procedure are used. Treatment methods are developing very quickly and are increasingly being tailored to individual patient needs. In some centers, it is even possible to combine surgical correction of the defect with breast implant surgery [27].

3.3 Ravitch procedure

The first commonly used method of surgical treatment of pectus excavatum was the Ravitch method [11,25]. The first documented procedure took place in 1949 [2,21,25]. Nowadays, it is a less frequently used method due to its invasiveness, giving way to the Nuss method. However, it is still used, especially for treating defects with high asymmetry, in older patients [1,2,11,27,31,32] and in cases of recurrence [1]. The method involves a longitudinal or transverse thoracic incision and bilateral resection of deformed costal cartilages in the parasternal area and the xiphoid process, as well as a transverse osteotomy of the sternum in

the place of the posterior intussusception, thus correcting the shape of the rib cage [25]. Rehbein and Wernicke modified the original method by additionally implanting wires to stabilize the newly positioned sternum.[33] The wires are inserted through a bilateral skin incision under the nipples, or through a central skin incision along the sternum. The wires had to be removed after a year and the patients had to be operated on again. [33] Therefore, another modification replaced the use of rods and replaced them with metal plates and orthopedic screws, allowing for better customization to the individual needs of the patient and not requiring reoperation to remove the material.[18]

3.4 Nuss procedure

Currently, the most commonly used surgical method is the minimally invasive Nuss method. It involves inserting an arched metal or titanium rod into the rib cage so that it is located under the most depressed part of the sternum and the most deformed part of the ribs.[33] Small incisions, about 3 cm, are made in the skin in the anterior axillary line, in front of which, initially under thoracoscopic guidance, a metal guide is inserted in such a way as to cut and create a space for the rod between the anterior pericardium and the posterior surface of the sternum. The guidewire is tied on the opposite side to the rod and then removed in the same way it was inserted. In the cage, the rod is inserted with the bend downwards and then rotated 180 degrees, causing the bend of the rod to push the sternum upwards. The bar is then fixed using stabilizing plates. The current recommended stabilization time is 3 years [5], once the intended shape of the sternum is achieved, after which the plate is removed during a short surgical procedure under general anesthesia. [1,11,27]. The method is constantly being improved, aiming to minimize the number of complications [8]. Additionally, patients should be tested for metal allergy before surgery, in which case a titanium rod is an alternative [1,12,30]. Hospitalization after the procedure is 3-5 days, and return to normal physical activity is expected within 4-6 weeks. [1,11]

4. Efficacy and complications of both treatments

Both the Ravitch and Nuss methods have comparable effectiveness, perioperative mortality and length of hospitalization after surgery, ranging from 3-5 days to 1 week, depending on the sources [11,21,34]. In both methods, patients show comparable satisfaction rates of up to 95% [1]. The risk of recurrence varies in the literature, with an estimated incidence of 2-10% [1]. In the pediatric population, the same risk of complications was observed, although in the group of adult patients the Nuss procedure had a higher complication rate than the Ravitch method [5,29], which, however, is constantly being improved in the form of the use of special modifications in Nuss procedures in adult patients, reducing the risk of complications [29]. Both methods may cause postoperative complications, with a risk estimated at 15-20% [1], such as:

The Nuss method, compared to the Ravitch method, is characterized by a higher risk of postoperative pneumothorax and bleeding into the chest. [5,21] These risks have been

significantly reduced over time with the use of videothoracoscopy during rod placement [14]. Operation with this method, compared to the Ravitch method, results in a smaller scar, lower risk of infection [1,5], and lower risk of perioperative complications [5]. Surgery time using the Nuss method is shorter and is characterized by lower intraoperative blood loss.

Rare cases of scoliosis after Nuss surgery have been reported [5], but Nuss surgery may have a positive effect on co-occurring mild scoliosis, especially when operated on during adolescence [5].

Complications that occur especially after Ravitch surgery:

- postoperative cases of thoracic constrictive dysplasia have been observed in some patients under 6 years of age undergoing Ravitch surgery [11,28]
- extensive cartilage removal with subsequent damage or removal of growth plates has resulted in inhibition of growth and thoracic development in some patients [11]
- postoperative complications in the form of abnormal bone fusion, osteonecrosis or thoracic instability [11]

5. Operational vs conservative comparison

Surgical treatment is much more effective in correcting the defect, but conservative treatment is also used in the treatment of pectus excavatum.

First of all, conservative treatment can be offered to younger patients, who, due to their age, are not yet eligible for surgery, and the defect causes symptoms.

For treatment with non-invasive methods, the undoubted advantage is the absence of scarring and of exposure to the inconvenience of hospitalization. There is no risk of perioperative complications, hospital stay or pain after the procedure. This is a good alternative for patients who do not want to undergo surgery. The visual effect after completing treatment is comparable to that of surgical patients. However, these methods are not without drawbacks. The biggest disadvantage of these methods is the treatment time and the fact that it requires a lot of commitment and regularity from the patient. Depending on the case, treatment lasts on average several months. During this time, the patient must regularly use the device selected for him, several times a day for several dozen minutes. This is accompanied by discomfort, pain, hematomas or petechiae, and paresthesia in the upper limbs may occur, especially in the initial stage of treatment. There are cases of patients who give up during treatment because they are unable to adapt to regular use of the applicator. Another disadvantage may be that not every patient will achieve a good treatment effect and this risk decreases as the patient's age increases, reduced chest elasticity and severe chest asymmetry. There is too little information on adults treated with this method, and further research is required here.

The main advantage of surgical correction of the defect is its effectiveness, resulting in 95% of patients' satisfaction. The speed of the procedure and recovery is much faster than non-invasive treatment methods. This treatment can also be offered to adult patients. It does not require as much regularity from patients as the non-invasive method. Operative methods also have many disadvantages. First of all, it is an invasive procedure, which poses a much greater risk of serious and life-significant complications. This results in preoperative stress, hospitalization, possible complications and post-operative pain. After surgery, patients have scars in the chest area. The use of the surgical method in children before their development is complete may result in damage to the growth cartilages and abnormal development of the child.

Although research into the safest and most effective surgical treatment of pectus excavatum is ongoing, further research in this direction is still required.

6. Conclusion

The gold standard for treating a funnel chest is the minimally invasive Nuss method due to fewer perioperative complications and faster patient recovery. The conservative method is an alternative for patients who are afraid of the procedure, but it has several significant drawbacks, which may cause the treatment to fail. The patient should be informed about the advantages and disadvantages of both methods. Before deciding on the choice of treatment, it is necessary to carry out diagnostic tests to determine the type and degree of the defect and to select an appropriate treatment method. Further research is necessary to determine the most customized treatment option for each patient.

Disclosure

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