Pectus excavatum and carinatum should only be operated in case of functional impact…

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General considerations

A condition known since Hippocrates, the first medical description of pectus excavatum dates back to 1594 and is attributed to Johannes Bauhinus [1]. It is the most common form of anterior chest well deformity, making up 90% of chest wall deformities in Europe. Pectus excavatum is a deformity of the sternum with anterior concavity. In addition, the sternum is translated posteriorly by the 3rd to 7th pairs of costal cartilages. This deformity is commonly known by the mass public as “funnel chest” (Fig. 1)

Fig1: Pectus excavatum

Its prevalence varies relative to the population. It is exceptionally rare in African populations and has an incidence of 1 per 400 to 1000 births according to the authors, with a sex ratio of 4:5.

Pectus excavatum is usually first noticed from birth in a relatively moderate form. It remains stable during infancy and progressively worsens during peak growth velocity at puberty stabilizing in adulthood [2]. A small number of publications have studied the genetic transmission of this disease on a large scale. There is a positive family history in 40% of cases. The most commonly accepted hypothesis is that of a recessive autosomic transmission, which may be related to the X chromosome (which would explain the higher incidence of this anomaly in XXY Klinefelter's syndrome). To this day, no monoallelic mutation has been identified.

The pathophysiology of pectus excavatum has not been well elucidated [3]. For some authors, it may be secondary to exogenous factors such as intrauterine or intrapartum hyperpressure. However, for most authors endogenous or intrinsic factors would be related, especially factors such as a longer costo-cartilagenous complex, or even a dysregulation of the substernal or intercostal musculature [4].

Patients typically have a lanky and thin morphotype and appear to be "wrapping in around themselves", with anterior translation of the shoulder and thoracic hyperkyphosis, which further worsens the unpleasant appearance of this pathology. Most patients consult for severe aesthetic disturbances, many of whom are bullied at school, avoiding topless situations such as wearing swimwear. Some patients have reported discomfort especially during exertion with a loss of endurance and an inability to produce maximal exertion compared to their peers. Finally, some patients report pain at the level of the deformed cartilages and might even suffer sensations similar to a panic attack due to an inability to properly inhale. These findings are not always immediately reported by younger patients and must be systematically ruled out during clinical assessment. To this day, it is not uniformly admitted that corrective surgery in adolescents would objectively improve pulmonary or cardiac parameters and data in the literature are contradictory. Conversely, patients constantly report a subjective improvement in exercise tolerance in the 6-to-12-month postoperative period [5].

Contrary to pectus excavatum, pectus Carinatum has been of interest for only a limited period. Its incidence is much lower at around 1 per 1000 births and patients are often asymptomatic. Pectus carinatum is a deformity of the sternum with anterior convexity. The sternum is anteriorly translated by the 3rd to 7th pairs of costal cartilages. This deformity is sometimes known as pigeon chest (Fig.2)

Fig2: Severe pectus carinatum

Similar to the previous pathology, boys are 3-4 times more commonly affected than girls. The deformity is particularly encountered in South America, the reason for which remains unknown [6]. Pectus carinatum is very rarely noticed at birth and seems acquired rather than congenital. The deformity is noticed by the children themselves at a pre-pubertal age and is accentuated during peak growth velocity during puberty.

The pathophysiology of pectus carinatum has, to this day, not been well elucidated. A portion of these patients present iatrogenic subtypes after sternotomy during childhood, due to injury of the ossification centers. In patients who have not undergone sternotomy during their childhood, a congenital etiology of ossification center injury would seem plausible. For some authors, similar to pectus excavatum, this could be due to an excessively long costo-cartilagenous complex, especially in patients who undergo a staged cartilage resection, in which case it would be possible to suture the ribs almost directly to the sternum, which would lead to said excessive length of the complex.

As was the case for pectus excavatum, patients typically have a lanky and thin morphotype and appear to be “wrapping in around themselves”, with anterior translation of the shoulder and thoracic hyperkyphosis, which further worsens the unpleasant appearance of this pathology. Most patients consult for severe aesthetic disturbances, many of whom are bullied at school, avoiding topless situations such as wearing swimwear. In contrast to pectus excavatum, many patients report cartilage pain, and any reported breathlessness is secondary to said pain. These findings are not always immediately reported by younger patients and must be systematically ruled out during clinical assessment. Symptom improvement is usually seen in the 6-to-12-month postoperative period, probably due to improvement in the aesthetic aspect and patient’s self-esteem.

The diagnosis of a pectus deformity is clinical. Physicians should systematically rule out clinical signs or a family history of Marfan’s disease [7].

In pectus excavatum in order to measure the degree of the depth of the deformity, a thoracic CT-scan should be obtained. This allows the measurement of Haller’s index which represents the ratio between the thoracic width and thickness at lowest level of the defect (Fig.3).

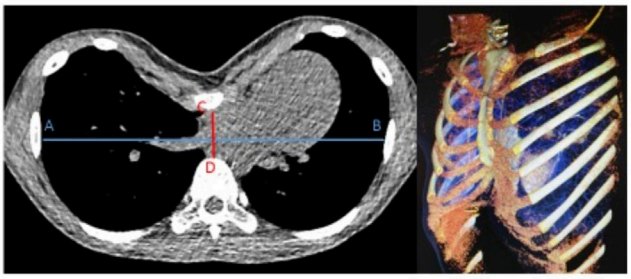

Fig3: Haller’s index and preoperative 3D reconstruction. The cartilaginous deformity of the 3rd to 7th pairs can be seen. Haller’s index = [AB]/[CD]

A normal Haller's index should be situated around 2.56 (i.e., an individual is 2.56 times larger than they are deep). In case of pectus excavatum, this index would be increased, with a surgical cutoff value of 3.2. A CT-scan also allows the measurement of rotation of the sternum and underlying intrathoracic pathologies or scar tissues which could contraindicate any intrathoracic surgeries may be ruled out.

A CT-scan, in accordance with physical exam, through 3D reconstructions of the ribcage, allows the physician to determine the morphology of the pectus. The more extended and asymmetrical forms would render surgical correction more difficult with more inconsistent outcomes.

All of these evaluations lead to a complete anatomical workup of pectus excavatum, after which the functional impact of the deformity must be assessed. Practically, this can be achieved with four tools:

- Symptoms journal: Between consultations, patients are asked to keep a journal tracking the occurrence of different symptoms: chest pain, breathlessness, anxiety. This allows an evaluation of the daily subjective impact of the deformity.
- Echocardiography in order to evaluate any eventual impact of pectus excavatum on the right heart (filling defects, hemodynamic disturbances). The purpose of this exam is to rule out any cardiac pathology which could contra-indicate the surgical management of pectus excavatum. These surgeries require the placement of a retrosternal metallic bar which could render cardio-pulmonary resuscitation and chest compression difficult or even impossible.
- Pulmonary function testing: many adolescents presenting with feelings of breathlessness due to pectus excavatum are actually suffering from exercise-induced asthma and are generally mild.
- Psychological intervention: a psychological intervention in these patients, who are often suffering from a psycho-social standpoint, may be warranted. In fact, one should avoid surgical intervention in subjects presenting with dysmophophobic symptoms and who would not be hesitant to undergo a brutal change in their body image. Furthermore, patients who cite pectus excavatum as the reason for the psychological distress, but which are found to be actually rooted elsewhere, should not be treated surgically. In this subset of patients, surgery would not solve the distress, and the patient would undergo progressively escalating beauty procedures with the final results never being satisfactory.

Concerning pectus carinatum, CT-scanning with 3D reconstructions is also recommended for preoperative planification. Consultation with a psychologist is equally recommended as in pectus excavatum.

Furthermore, some forms which are rather extensive and asymmetrical have been described. An extended or asymmetrical deformity predicts a more reserved aesthetic prognosis after surgical correction. Treatment essentially consists of deformity correction. There are two major types of treatments, operative and non-operative.

1) Non-operative treatment.

The first-line management of pectus excavatum includes physiotherapy. Although this intervention does not improve the pectus deformity, it allows an improvement in the child's posture. Some authors recommend pre-operative respiratory exercises in order to facilitate post-operative management. Additionally, sessions with aspiration bells, such as the "Vacuum-Bell®", are also indicated in an attempt to depress the ribcage. However, duration of treatment and frequency of the sessions are not well established. Eckart Klobe, the designer of these bells, recommends two daily sessions of 30 minutes each, until correction of the pectus deformity is achieved [8]. In our experience, adolescents should not be treated expectantly. In young children, the outcomes are more encouraging with a reversal of at least 50% of the deformity, although there is rapid recurrence as soon as this rate of two sessions per day is abandoned. Owing to its high price-range, the strict nature of its use, and the generally partial and disappointing outcomes of such devices, we do not recommend their use. A literature review is quite disappointing from this point of view. St-Louis et al. [9] studied 31 patients over 4 years and did not find any improvements in the depth of the pectus deformity with the use of a Vacuum-Bell, but they did report a modification of Haller's index. Since their studied population had a mean age of 14 years old, the modification in Haller's index could just as well be simply due to the modification of ribcage volume due to rapid growth during puberty, thus rendering the Vacuum-Bell's effectiveness uncertain. Alaca et al. [10] studied 26 patients treated with an aspiration bell, categorized into two groups, one of whom also received physiotherapy. The group that had also received physiotherapy seemed more motivated, with an improved posture and global quality of life. Similar findings have also been reported in scoliotic patients treated with bracing with or without physiotherapy. There is not enough evidence to suggest that the pectus deformity would improve in the long term, both in terms of anatomical correction and functional symptoms. Finally, Haecker et al. [11] followed 450 patients aged 2 to 61 years old who were treated with such devices since 2003 and reported encouraging results, although post-treatment middle-and long-term data are still lacking.

To this day, isolated conservative management of these deformity has not yielded satisfactory results, whether in terms of aesthetic or functional demands. As such, these findings are reason enough to provide patients with surgical options.

However, in pectus carinatum, conservative treatment is much more commonly used. It consists of compressing the deformity with a brace, ideally at the time of peak height velocity during puberty when the thorax gains 50% of its definitive volume. Numerous studies have reported high success rates in terms of the aesthetic outcome [12-15]. In a cohort of 460 patients, Dekonenko et al. [16] found a 94% satisfaction rate. Poola et al. [17] reported 47% complete correction within 7.5 months in a series of 340 patients. In our experience, a brace should be worn for 12 to 16 hours a day, with at least 8 hours a day provided for the patient during sleep or at school. This allows a better acceptance and observance of the brace. Most patients wear the brace for a period of 12 to 18 months. Very few complications have been reported, primarily erythema in 10% of cases [17].

2) Operative treatment

Different treatment options exist in order to better control the risk-to-benefit ratio in patients with pectus deformity. Largely, three different techniques have shown satisfactory results:

- **Prosthetic packing:** This is a palliative technique where the pectus deformity is packed by a silicone prosthesis that is custom made for each patient. This technique has the advantage of being simple and allowing a very simple post-operative management. Nevertheless, certain problems have been raised, such as the longevity of the implant, which should be indefinitely, and the fact that it does not allow correction of the underlying chest-wall defect [18]. In patients with respiratory discomfort or compression of the right heart, this technique would not resolve the primum movens and is therefore not recommended.
- **Correction by sternochondroplasty [19]:** Different techniques have been described which have in common the resection of the deformed cartilages. This entails a curative solution for both pectus excavatum and pectus carinatum deformities. Ravitch has suggested since 1949, that all of the costal cartilages be resected along with the perichondrium. These days, the most frequently used technique is the simplified subperichondrial sternochondroplasty of Wurtz. The intervention consists of subperichondrially resecting the pairs of cartilages from the 3rd to the 7th arc while performing an osteotomy the sternum and elevating it, which is then fixated by a metallic splint. This intervention is undertaking by a transverse bi-sub-mammary approach. In case of cardiac anomalies, a vertical and median approach is preferred in order to prepare for future cardiac surgery. The first part of the intervention consists of detaching and reclining the pectoralis major, followed by the rectus abdominis from the 7th arc while respecting their posterior aponeuroses. The second part consists of isolating the cartilages from the perichondral envelope with an Obwegeser periosteal elevator and resecting them in totality except for the 7th and eventually the 8th arc (if it is inserted on the sternum). A 2-cm resection would suffice for these last two arcs in order to reattach them at the end of the intervention. The third part includes a sternal osteotomy at the level of the manubrio-sternal junction with the aid of an oscillating saw and an osteotome. An anterior fragment is removed, and the osteotomy is closed with large diameter absorbable sutures, such as Vicryl® 2, in pectus excavatum, while an anterior opening osteotomy is realized in pectus carinatum. In pectus excavatum, the osteotomized sternocostal unit is stabilized from anterior to posterior with the help of a Wurtz metallic splint (MedicalexTM, Bagneux, France) which is passed with a dissector and surgical loop overlying the insertion of the 6th costal arcs (Fig.4). The splint is sutured to the sternum with the same suture, and the perichondrium is minutely repaired with Vicryl® 0, and the different layers are repaired over a drain with negative pressure. In extended and flexible forms such as in patients with Marfan syndrome, STRATOS (Strasbourg Titanium Osteosynthesis System) type rib bridges are used and are hooked directly onto the ribs. This intervention allows correction of all subtypes of the deformity, and more particularly extended and asymmetrical forms.

Fig.4: A) Partial subperichondrial resection of costal cartilage. B) Intra-operative photograph, with a high sternal osteotomy and the perichondral envelopes emptied of their cartilage. C) Stabilization of the sternum with a metallic splint. D: Example of correction with vertical surgical scar in a 13-year-old patient with a severe type of pectus excavatum with Marfan syndrome.

- Minimally invasive correction with cartilage growth guidance [5]: This includes the technique developed by Donald Nuss at the end of the 1990s. The technique includes the biomechanical principle of "3-point flexure" with a metallic bar introduced within the ribcage. This bar leans on the two hemi-ribcages over which the sternum is supported. The first stage of the procedure consists in bending a metallic bar to the shape of the ribcage based on a flexible metallic phantom which serves as a mold. Afterwards, a dermographic pen is used to locate the entry and exit points from the thorax, overlying the point at which the sternal deformity is most shallow. The incisions are made on the axillary line in boys and within the submamillary groove in girls. Digital subcutaneous dissection is carried out at the contact with the ribs. A thoracoscope is introduced through the incision into the lowest intercostal space with slight insufflation of CO2 to detach the lung. A 30° thoracoscope is used to properly explore the ribcage. While under thoracoscopic control, a saber-shaped dissector is introduced to release the pericardium from the posterior aspect of the sternum and is exited from the ribcage. A surgical loop is then attached to the saber, which is then removed. It serves as a distance line for the plate that is then introduced into the thorax and exited while tracking over the surgical loop. With the help of a bender, the extremities are molded such that it properly covets the thorax, then the bar. As such, it should be concave in the front. The plate is then turned such that the convex part is anterior, which pushes the sternum outwards. The bar is attached to stabilizers which, in our experience, are bulky and uncomfortable for children and adolescents. Fixation is achieved with Vicryl® 2 sutures onto the ribs [20] (Fig.5 and 6).

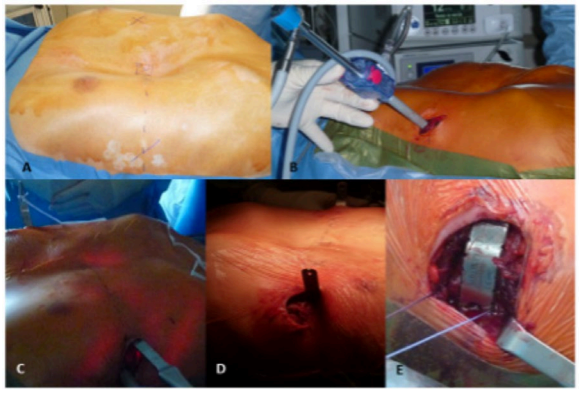

Fig.5: The cutaneous landmarks are drawn (A), and lateral cutaneous flaps are then made. A thoracoscope is introduced on the right side of the thorax (B). It allows control of the dissection by the saber (C), to which a distance line is attached thus facilitating the passing of the pre-contoured plate (D). The latter is thus bended to meet the extremities, turned and fixed to the ribs (E).

Fig.6: Preoperative and 6-month postoperative aspect of severe pectus excavatum operated by the Nuss technique.

Numerous complications have been reported in the above-cited treatment methods:

- The Vacuum Bell is contra-indicated in Marfan's disease since it has an increased hypothetical risk of injuring the aortic arch, although such a complication has never been reported. Cutaneous lesions have also been reported, which are most often just temporary, such as skin bruising and presternal dysesthesias which could present as severe paresthesia of the upper limbs [11].
- Sternochondroplasties have not shown increased cardiopulmonary risks since these techniques are purely extrathoracic. In contrast, they run the risk of scar formation and may sometimes relapse, in which case revision surgery with a Nuss technique would be indicated. Secondary displacement of the Wurtz splint may also occur if the patient does

not follow post-operative instructions, such as avoiding trunk flexion/extension and rotation/inclination for the first month post-operatively.

- The Nuss technique also has its complications, such as a classic of the bar for similar reasons as mentioned above. Furthermore, this is an intrathoracic technique and cases of cardiac lesions, even death by cardiac perforation, have been described. The available literature tends toward avoiding this technique in adult patients with an extremely rigid thorax, in case of sternal rotation >35°, and in case of a sterno-vertebral distance <5cm at its nadir [21].

Contrary to sternochondroplasties, the risks related to the technique are hypercorrection of the deformity, the reason for which plate bending should be undertaken with extreme care. The learning curve and experience of the operator are elements that are more sensitive in minimally invasive techniques than in sternochondroplasty.

Functional impact

Just as Obermeyer et al. [22] had highlighted, many patients complain of limited tolerance during exercise or even activities of daily living. Nevertheless, these complaints make up only 15 to 20% of the reasons for consultation, lagging far behind aesthetic concerns. These subjective findings have not been corroborated by objective physiological observations, except in limited cases. The patient and their family should be made aware that surgical correction may not lead to significant objective improvement in cardiopulmonary function.

Chen et al. [23] published in 2012 a meta-analysis of 23 studies in patients operated with either the Nuss technique or sternochondroplasties, with over 2476 patients included, 1555 of which were operated according to the Nuss technique. The forced expiratory volume (FEV) was statistically increased at 3 years in both techniques, even more so with the Nuss technique. Nevertheless, short term results were better with sternochondroplasties, probably from the reduced thoracic expansion due to the presence of the intercostal bar. Similarly, at 3 years post-operatively, there is increase in vital capacity (VC), forced vital capacity (FVC), and total lung capacity (TLC), with better results in the Nuss technique. Most studies included in this meta-analysis show that the results for the Nuss technique were better on the long term due to a larger gain in thoracic volume and more satisfactory thoracic expansion after removal of the bar.

In another meta-analysis on 456 patients, Wang et al. [24] showed that, within the first year post-operatively with the Nuss technique, a decrease in the FEV may be seen. This has been related to the presence of the bar, especially since a net increase is observed after its removal 2 years post-operatively. As a result, in order to avoid patient disappointment, they should be informed that their performance may paradoxically decrease in the short term after surgical correction.

Moreover, although this study included only a small sample size, Jeon et al. [25] showed that patients with severely asymmetrical types of pectus excavatum regularly had lower preoperative VC, FVC and Peak flow. Post-operatively, these values normalized, with Peak flow showing the

least amount of improvement in patients with asymmetrical deformities compared to those with symmetrical ones.

To this day, few studies have assessed the role of postoperative chest physiotherapy in the improvement of surgical outcome. In a series of 34 patients, Noguchi et al. [26] found a net benefit to exercises in force expiration on a spirometer, 4 times a day, for three months. The group that underwent chest physiotherapy presented a VC of 1400mL, compared to 900mL for the group without physiotherapy. As such, management of the pulmonary aspect in these patients should be addressed, in an attempt to increase pulmonary function post-operatively. Patients with pectus carinatum have shown little alteration in cardiopulmonary function [27].

From a cardiac point of view, it is admitted that deformity correction essentially relieves potential compression of the right heart. Sigalet et al. [28] concluded in 2007 on the positive effect of surgery in these patients with a reported increase in performances in terms of endurance, both on the cardiac and pulmonary fronts. Névière et al. and the team at Lille [29] observed, in patients in whom pre-operative pulmonary function tests were within the normal range, a post-operative improvement of Maximal VO2 with exertion within the first year, increasing from 77% of the expected theoretical value to 87%. According to them, this increase in VO2 could be explained by improved right cardiac function allowing for a more adapted gas exchange. A 2016 literature review on this subject by Maagard [30] concluded that, in a similar manner, cardiac function improves after surgery of thoracic deformities.

From an anatomical standpoint, Oezcan et al. [31] conducted ultrasonographic explorations that found either pericardial effusion, tricuspid valve prolapse, or dynamic anomalies of the right ventricle in 1/3 of patients, and in 10% of cases a lower-than-expected right ventricular ejection fraction. The correction of these anomalies may also explain the post-operative improvement in performance.

In addition, ECG studies are commonplace in adolescents. However, in adults, only 5 to 10% of patients would develop, probably due to the irritation of the autonomic cardiac innervation at the level of the bundle of His, reentrant-type anomalies, such as a Wolff-Parkinson-White syndrome [32]. The correction of these thoracic deformities may also prevent these complications, when the patient is operated in their adolescence.

In patients with pectus carinatum, in the absence of cardiac compression, functional cardiac repercussions do not develop [33]. Nevertheless, 30% of these patients complain of chest pain and anxiety or palpitations [34], without any ultrasonographic anomalies, as was reported by Port et al. Since these findings were published, a systematic cardiac ultrasound has been recommended in these patients, if only to rule out any underlying cardiac disease, especially if the patient has a marfanoid habitus [35]. Conversely, a study by Ates et al. [36] shed some light on conservative treatment. Some studies have reported cardiopulmonary complications related to the stiffening of the thorax after sternochondroplasties [37], while contrarily, treatment by compression has shown aesthetic improvements without cardiopulmonary compromise.

Psychological impact

Most patients who consult for pectus excavatum deny functional impact as a primary argument when seeking corrective surgery. Rather, most adolescents present with major aesthetic complaints, with around 80% of demands for correction being solely due to aesthetic considerations. Some might avoid all social interactions, with some even reporting being physically aggressed, being humiliated on a daily basis, and attempting suicide. In our experience, this extreme psychological burden is not related to the severity of the deformity. Some patients undergo a battery of preoperative testing, with PFTs within the normal range, and only a slightly altered Haller's index (<3). As such, surgery seems to be indicated only in patients with functional impact. It is obvious that surgery is not a benign procedure with its own risks attached, and that all procedures should be properly balanced in terms of expected psychological improvement compared to, on the one hand, the potential complications, on the other hand, the demanding pre- and post-operative recovery period.

By force of habit, there is a tendency to recommend a psychological intervention in all patients presenting with a thoracic deformity regardless of the reason for consulting, in order to ensure that the patients have understood the constraints related with the proposed management, and that the patient wishes the correction for the proper reasons. This is also to ensure that there is no tendency toward dysmorphophobia, an entity that is very frequent at an age where the body is transforming drastically. One should also avoid an escalation of demands for beauty procedures, while the original intention was to undergo thoracic reconstructive surgery.

Steinmann et al. [38] published the first significant study on the psychological impact of thoracic deformities on adolescent patients. 90 patients were compared to a control group, all of whom had to fill out the NQ-mA modified Nuss questionnaire for adults, the SF-36 quality of life questionnaire, the FKB-20 body image questionnaire and the Dysmorphic Concern Questionnaire. The perception of body image was negatively altered in these patients, without correlation with the functional impact of their deformity. Depression was significantly more prevalent in patients with pectus carinatum since this deformity was more difficult to hide beneath their clothing. Body image was constantly associated with more frequent depressive traits, as well as major alterations in self-esteem, while, paradoxically, quality of life did not seem significantly altered in a multivariate analysis. It should also be noted that, according to the DSM-IV, a psychiatric assessment of these patients did not find a higher prevalence of associated psychiatric disorders compared to the rest of the population. The authors thus concluded that a psychological evaluation was paramount in these patients, since there seemed to be a dissociation between the psychological suffering induced by the deformity and its functional impact. The authors also found it reasonable to consider undertaking corrective surgery based solely on the patient's psychological concerns as to avoid progression toward a psychiatric disorder. Nuss et al. [39] highlighted that, one year after corrective surgery, patients reported an improved perception in the ability to exercise, even if this was mainly due to the improvement in body image. This would also explain the improvement in physical performance in adolescents in whom no significant post-operative changes in cardiac or pulmonary parameters were noted. As such, it would seem reasonable to question whether surgical correction of thoracic deformities

should also respond to aesthetic concerns [40] in order to reinforce self-esteem. Considering all available surgical options, there are two distinct approaches, one of which is purely aesthetic (prosthetic filling), with the other being reconstructive aiming to correct the cause of the deformity (sternochondroplasty and the Nuss technique).

Similar tendencies are found in pectus carinatum [41] after surgery, even though treatment of such a deformity in adolescent patients is rarely surgical owing to the possibility of treatment with a compressive brace while their deformity is still flexible at the level of the ribcage. Contrary to pectus excavatum, even though surgery has shown a constant tendency to improve quality of life on the psychological level, it has shown to sometimes induce dysesthesia during the first 6 months post-operatively without significantly affecting the patients psychologically [42]. Recently, Robert Kelly's team developed a quality-of-life questionnaire related to the perceived body image in pectus carinatum, called the PeCBI-QOL [43] for both adolescents and their parents. The latter has shown a strong correlation between poor body image and poor quality of life, with both parameters showing equivalent results in both the patients and their parents.

Conclusion: should patients with pectus excavatum and carinatum only be operated in case of functional impact?

Thoracic deformities in adolescents may be treated both conservatively and surgically. In contrast to pectus carinatum, conservative treatment in pectus excavatum has constantly shown disappointing outcomes. Conversely, while surgical treatment of pectus excavatum has shown to improve both the aesthetic aspect of the deformity and cardio-pulmonary function, such treatments have only an aesthetic impact in patients with pectus carinatum, in whom conservative strategies should be the first line of treatment.

An analysis of the available scientific literature shows that many studies focalize on cardio-pulmonary function as a means to objectively justify surgical management, such as improvements in endurance, pulmonary function, or the hemodynamic improvements of the right heart. However, the reason for consultation remains primarily the aesthetic disturbances, which sometimes severely alters the quality of life of these patients. Regardless of the legitimate reservations that surgeons might have of undertaking potentially risky surgery for aesthetic concerns, the literature on the psychological impact of these deformities on these patients is increasingly in favor of treating these deformities. What remains is to assess when to treat conservatively, especially in patients where concerns are predominantly aesthetic without any cardio-pulmonary impact, compared to surgically to correct both the aesthetic aspect and cardio-pulmonary function. To this day, it is impossible to differentiate between the two, and there is a lack of objective treatment algorithms. As a result, different schools have different approaches, with much importance being given to the unbiased and enlightened information provided to the adolescent on the different treatment options and their advantages, risks, and limitations on the expected outcome.

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