European Journal of Cardio-Thoracic Surgery 2024, 66(1), ezae166
https://doi.org/10.1093/ejcts/ezae166

Cite this article as: Dunning J, Burdett C, Child A, Davies C, Eastwood D, Goodacre T *et al*. The pectus care guidelines: best practice consensus guidelines from the joint specialist societies SCTS/MF/CWIG/BOA/BAPS for the treatment of patients with pectus abnormalities. Eur J Cardiothorac Surg 2024; doi:10.1093/ejcts/ezae166.

The pectus care guidelines: best practice consensus guidelines from the joint specialist societies SCTS/MF/CWIG/BOA/BAPS for the treatment of patients with pectus abnormalities

Joel Dunning [a,]* (UK), Clare Burdett[a] (UK), Anne Child[b] (UK), Carl Davies[c] (UK), Deborah Eastwood[d] (UK), Tim Goodacre [e] (UK), Frank-Martin Haecker [f,g] (Switzerland), Simon Kendall [h] (UK), Shyam Kolvekar[i] (UK), Lisa MacMahon[j,k] (USA), Sean Marven[l] (UK), Sarah Murray[m] (UK), Babu Naidu [n] (UK), Bejal Pandya[i] (UK), Karen Redmond[o] (UK) and Aman Coonar [p] (UK)

[a] Department of Cardiothoracic surgery, James Cook University Hospital, Middlesbrough, UK
[b] Marfan Trust, Bromley, UK
[c] Royal College of Physicians and Surgeons of Glasgow, UK
[d] British Orthopaedic Association, London, UK
[e] Royal College of Surgeons of England, London, UK
[f] Department of Pediatric Surgery, Children's Hospital of Eastern Switzerland, St Gallen, Switzerland
[g] Department of Paediatric Surgery, Faculty of Medicine, University of Basel, Basel, Switzerland
[h] Society for Cardiothoracic Surgery in Great Britain and Ireland, London, UK
[i] National Pectus Centre, Department of Cardiothoracic Surgery, St Bartholomew's Hospital, London, UK
[j] Department of Thoracic Surgery, Phoenix Children's Hospital, Phoenix, USA
[k] Chest Wall International Group (CWIG), Switzerland
[l] British Association of Paediatric Surgeons Thoracic and Airway Group, London, UK
[m] Clinical Research Collaborative BHF and Leicester University, National PPI Group, Leicester, UK
[n] Department of Thoracic Surgery, Birmingham Heartlands Hospital, Birmingham, UK
[o] Department of Thoracic Surgery, School of Medicine, University College Dublin, National Thoracic Subcommittee Lead SCTS, The Mater Hospital, Dublin, Dublin, Ireland, UK
[p] Thoracic Lead at NHS England, President of the Society for Cardiothoracic Surgery in Great Britain and Ireland, London, UK

* Corresponding author. Department of Cardiothoracic Surgery, James Cook University Hospital, Middlesbrough, TS4 3BW, UK. Tel: +44-7801548122; fax: +44-1642854613; e-mail: joeldunning@doctors.org.uk (J. Dunning).

Received 11 September 2023; received in revised form 26 January 2024; accepted 30 April 2024

Executive Summary

Pectus defects are a group of congenital conditions found in approximately 1 in 250 people, where the sternum is depressed back towards the spine (excavatum), protrudes forwards (carinatum) or more rarely is a mixture of both (arcuatum or mixed defects). For the majority of patients, it is well tolerated, but some patients are affected psychologically, physiologically or both.

The deformity becomes apparent at a young age due to the growth of the ribs and the cartilage that links them to the sternum. The majority of defects are mild and are well tolerated, i.e. they do not affect activity and do not cause psychological harm. However, some young people develop lower self-esteem and depression, causing them to withdraw from activities (such as swimming, dancing) and from interactions that might 'expose' them (such as sleepovers, dating, going to the beach and wearing fashionable clothes). This psychological harm occurs at a crucial time during their physical and social development. A small number of patients have more extreme depression of their sternum that impedes their physiological reserve, which can occur when engaging in strenuous exercise (such as running) but can also limit moderate activity such as walking and climbing stairs. The effects can be so extreme that symptoms occur at rest or cause life-threatening compression of the major blood vessels and organs. The group of patients with physiological impairment usually also suffer from low self-esteem and depression.

This paper summarizes the current evidence for the different treatment strategies for this condition, including supportive care, psychological support and non-surgical techniques including bracing and vacuum bell therapy. We also consider surgical techniques including the Ravitch procedure, the Nuss procedure (minimally invasive repair of pectus excavatum), pectus implants and other rare procedures such as Pectus Up.

For the majority of patients, supportive care is sufficient, but for a minority, a combination of the other techniques may be considered.

This paper also outlines best practice guidance for the delivery of such therapies, including standardized assessment, consent to treatment, audit, quality assurance and long-term support.

All the interventions have risks and benefits that the patient, parents and clinicians need to carefully consider and discuss when deciding on the most appropriate course. We hope this evidence review of 'Best Practice for Pectus' will make a significant contribution to those considerations and help all involved, from patients to national policy makers, to deliver the best possible care.

Keywords: Pectus excavatum • Nuss procedure • Pectus carinatum • Ravitch procedure • Pectus brace • Vacuum bell

ABBREVIATIONS

CPET	cardiopulmonary exercise test
CT	computed tomography
ECG	electrocardiogram
MRI	magnetic resonance imaging
NICE	National Institute for Health and Care Excellence
O_2	oxygen
PC	pectus carinatum
PE	pectus excavatum
UK	United Kingdom
VO_2	ventilatory uptake of oxygen

INTRODUCTION

The sternum lies directly over the heart. In patients with pectus excavatum, generally during their teenage rapid growth phase, the sternum is pushed towards the spine, narrowing this space. Whereas the prevalence of severe pectus excavatum in the general population is approximately 1 in 250 [1], most have no symptoms at all. In a subset of patients with severe pectus excavatum and cardiac compression, however, right ventricular filling is impaired by the sternum, which in turn limits the patient's maximum cardiac output because the patient is unable to increase their stroke volume with exercise. They may also suffer an exaggerated compensatory tachycardia and limitations of their exercise ability.

Although it is accepted that surgical procedures can improve cardiopulmonary physiology in these patients, it is not universally accepted that they can have a clinically significant benefit for patients [2].

Currently, 12 cohort studies worldwide including 499 patients demonstrate the benefit of pectus surgery to improve a patient's exercise intolerance in terms of cardiopulmonary exercise testing (CPET). In the largest study, published in 2022 by Jaroszewski *et al.* [3], 130 patients underwent CPET preoperatively and 3 years postoperatively just before removal of the bar. Significant improvement in cardiopulmonary outcomes ($P < 0.001$ for all the comparisons) were seen in the post-repair evaluations, including an increase in maximum and predicted rate of oxygen consumption, oxygen pulse, oxygen consumption at the anaerobic threshold and maximal ventilation. In a subanalysis of 39 patients who also underwent intraoperative transoesophageal echocardiography at repair and at bar removal, a significant increase in right ventricular stroke volume was found ($P < 0.001$). The weakness in this and other studies has been that improvements in CPET have not been further correlated to provide evidence that the patients also benefitted from improvement in quality-of-life scores. As such, while there are excellent data on psychological outcomes, there is a lack of research evidence as to whether physiological improvement leads to a better quality of life for the patient. In addition, a minimum clinically important difference has not been determined for CPET testing in patients with pectus [2].

A large group of patients with pectus abnormalities is also severely affected psychologically by this condition. These patients may also have pectus carinatum or arcuatum; the severity is determined by the impact on their life rather than by measurements on a computed tomography (CT) scan [2].

Evidence of psychological benefit has been documented in multiple studies [2] that have measured quality-of-life scores, depression index, measures of social isolation, self-esteem and body image as well as cosmetic satisfaction scores. Validated psychological tools specific to pectus surgical procedures do exist [4]. However, in contrast to physiological impairment (where the Haller index is the widely accepted measure of severity, and CPET is the most accepted primary outcome measure), no single psychological outcome score has been adopted as the gold standard, thereby weakening comparisons. We do not have an agreed upon threshold for what is an unacceptable level of psychological impairment or what constitutes a meaningful improvement in psychological well-being.

In terms of the safety of pectus surgery, The National Institute for Health and Care Excellence (NICE) performed a technology appraisal in this area and made recommendations for the conduct of safe operations across the United Kingdom (UK) [5] based on their systematic review and also on a contemporary analysis of the UK national data set.

This joint statement seeks to unify the indications for and the treatment of patients with pectus from the clinician's perspective. We hope and envisage that this statement will be utilized to guide the provision of services in the future.

METHODS

A joint societies' group was formed to determine best practice in this area, comprising individuals from a broad range of specialties with diverse but relevant backgrounds and experience (Appendix 1). The method we applied when creating this guideline is based on the methods manual of the European Association for Cardio-Thoracic Surgery for clinical guidelines, which was co-authored by our first author [6]. This method is fully described in the paper but began with the appointment of a task force, which for this guideline, was a multispecialty group. We determined our scope and timelines, took our search

Table 1: Classes of recommendations

Class of recommendations	Definition	Suggested wording
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful and effective	Is recommended/is indicated
Class II	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure	
Class IIa	Weight of evidence/opinion is in favour of usefulness/efficacy	Should be considered
Class IIb	Usefulness/efficacy is less well established by evidence/opinion	May be considered
Class III	Evidence/general agreement that the given treatment/procedure is not useful/effective and may sometimes be harmful	Is not recommended

Table 2: Levels of evidence

Level of evidence A	Data derived from multiple randomized clinical trials or meta-analyses
Level of evidence B	Data derived from a single randomized clinical trial or from large non-randomized studies
Level of evidence C	The consensus of expert opinion and/or small studies, retrospective studies and registries

method from that used by Solutions for Public Health performed for the National Health Service (NHS) England commissioning document on pectus surgery for pectus deformities causing severe physiological impairment and for pectus deformities causing severe psychological impairment [2, 7]. The keywords in this search strategy include 'pectus Excavatum' 'Funnel chest', 'Pectus Carinatum', 'Pigeon thorax', Pigeon chest', 'abnormalities of pectus or sternum or chest wall or thorax', 'MIRPE', 'NUSS', and 'Ravitch'. The full search strategy is given in Appendix 2.

We performed the search for the period October 1966 to October 2022 and included both English and non-English papers and any size of study if relevant to the question. We performed double-blind abstracting with 2 co-authors (JD and CB); any conflicts were resolved by the senior author. We searched Medline, Embase and the Cochrane library, and we accepted guidelines or reviews as well as original papers. We also searched the reference lists of all selected papers. Solutions for Public Health identified 1046 references in their search strategy, and we identified 1753 papers with our extended search.

Results from the literature searches were screened using their titles and abstracts for relevance to answer our PICO (patient, intervention, comparison, outcome) questions (Appendix 3). Full text references of potentially relevant evidence were obtained and reviewed to determine whether they met the inclusion criteria for this evidence review.

We used the recommendations of the European Society of Cardiology 2018 practice guidelines for grading the strength of recommendations and for assessing the levels of evidence in support of them. We present these grades and levels in Tables 1 and 2 [8].

We performed a Delphi consensus process of all co-authors in order to attain consensus for all major recommendations. We had full participation in the Delphi process by all co-authors. Consensus of 80% or more was reached for all issues after the second round except for the single issue of numbers of cases required to determine the competence of a surgeon. For this issue, the majority answer was accepted.

We performed a public consultation at an open forum meeting on 2 February 2023 at the Royal College of Surgeons of England attended by more than 100 patients and their representatives, and the SCTS patient representative is a co-author of the guideline.

INVESTIGATION OF PATIENTS WITH PECTUS EXCAVATUM AND PHYSIOLOGICAL SYMPTOMS

When investigating a patient with pectus excavatum with suspected cardiac compression, we believe that cross-sectional imaging is mandatory to measure the Haller index. We could use low-dose CT scans or magnetic resonance imaging (MRI) scans according to local availability. Using a postero-anterior and lateral chest X-ray is not as reliable and cannot demonstrate cardiac compression. Of note, a CT scan should be performed in end-expiration not end-inspiration, because the Haller index can increase by 30% in end-expiration compared to end-inspiration, and it is important to assess the degree of compression at its most severe [9, 10]. If a cardiac MRI scan is used, the Haller index should similarly be measured when the patient is in the expiratory phase of their breathing pattern. Of note, 3-dimensional CT scans are now routinely available. They allow better elucidation of the craniocaudal extent of the defect or even provide volume measurements and make it easier to better demonstrate the defect to patients.

An echocardiogram should be performed to exclude any other causes of shortness of breath in the patient such as an atrial septal defect or other congenital cardiac abnormality. It should also be used to make sure the aortic root is not enlarged to reduce the likelihood that the patient has a significant connective tissue disorder. Patients are often despondent that the echocardiogram results are normal, but it must be highlighted that this is not a good test for finding an anatomical aetiology for the exercise intolerance. Lung function testing should also be performed, but this is again to exclude asthma or other respiratory conditions and benchmark the patient's function. Malek *et al.* performed a meta-analysis [11] on respiratory function in patients undergoing a surgical procedure. They found 12 studies representing 313 patients with pectus excavatum. Although some patients with pectus did have some reduction in their preoperative forced ventilatory uptake in 1 s and forced vital capacity, he found that these values were not improved postoperatively.

The most reliable and sensitive test to investigate exercise intolerance is a cardiopulmonary exercise test. This test can investigate accurately a patient's exercise intolerance when it is most troublesome, which is during maximal exertion in a vertical position. It is also able to give an accurate estimation as to how abnormal the patient is compared to an age- and gender-matched control and may be repeated postoperatively to assess the results of the operation. All centres seeing patients should have access to CPET services.

Cardiac MRI has been extensively investigated as a tool to measure the physiological impairment due to pectus excavatum. It has the advantage of being able to visualize the right ventricle and observe its filling and ejection. It does, however, have the considerable disadvantage that it is performed supine, when the heart will naturally move into the left chest away from the sternal defect and the patient will be at rest and therefore asymptomatic. It is also more difficult to measure right ventricular function than left ventricular function due to its non-symmetrical anatomy.

However, many studies have reported abnormalities on cardiac MRI scanning. Rodriguez-Granillo *et al.* in 2020 [12] looked at cardiac MRI scans and also stress echocardiography scans. Fifty-nine patients underwent multimodality imaging; 25 of these patients had effort-related symptoms. Twenty controls were also used. The mean Haller index was 5.2. Compared with a sex- and age-matched control group, peak exercise capacity was lower in patients with pectus excavatum (8.4 ± 2.0 METs vs 15.1 ± 4.6 METs, $P < 0.0001$). At stress, significant differences were found between groups regarding left ventricular E/A ($P = 0.004$) and E/A ratio ($P = 0.005$), right ventricular E/A ratio ($P = 0.03$) and trans-tricuspid gradient ($P = 0.001$). With cardiac MRI, only 9 (15%) patients with pectus excavatum had normal septal motion, whereas 17 (29%) had septal flattening during inspiration. Patients were classified according to the site of cardiac compression as type 0 (without cardiac compression); type 1 (right ventricle); and type 2 (right ventricle and atrioventricular groove). Septal motion abnormalities were significantly related to this cardiac compression classification ($P < 0.0001$).

Topper *et al.* [13] in 2016 looked at cardiac MRI in 38 patients pre- and postoperatively. The mean Haller index preoperatively was 9.6, reducing to 3.0 postoperatively. The right ventricular ejection fraction was reduced preoperatively and improved significantly at the 1-year follow-up [45.7% (43.9–47.4%) vs 48.3% (46.9–49.5%), $P = 0.0004$]. Left ventricular function was preserved pre- and postoperatively.

In an interesting study by Monti *et al.* in 2019 [14], a total of 20 patients with pectus excavatum had cardiac MRI; then a vacuum bell was placed on their chest and the study was repeated; it was also done with 10 healthy individuals. Following the vacuum bell application, the patients with pectus showed a 10% increase in the biventricular systolic index. Furthermore, the left ventricular end-diastolic volume index improved by 8%, and the right ventricular ejection fraction increased by 7%. These findings were not mirrored in the healthy individuals when a vacuum bell was applied.

Thus cardiac MRI scanning can identify abnormalities, but they are more subtle than the gold standard of a cardiopulmonary test and less reliably predict the symptomatic status of a patient with pectus.

Recommendation: Patients with suspected exercise intolerance

We recommend that all patients undergo cross-sectional imaging of the thorax to determine the Haller index and a review of the anatomical compression of the right ventricle.
(Class I, Level of Evidence B)

All patients should undergo echocardiography to exclude other causes of shortness of breath and to measure the size of the aortic root. This test is not sensitive in excluding patients from having cardiac compression.
(Class IIa, Level of Evidence C)

All patients should undergo lung function testing to exclude other causes of shortness of breath. A normal test result does not exclude the patient from having cardiac compression.
(Class IIa, Level of Evidence C)

We recommend that all patients with severe pectus excavatum and exercise intolerance undergo a cardiopulmonary exercise test because it is the best test to identify exercise intolerance that is caused by pectus excavatum. A ventilatory update of oxygen (VO_2) max below 85% of predicted is regarded as an abnormality.
(Class I, Level of Evidence B)

Cardiac MRI scanning may be performed, and it may show abnormalities that recover after an operation or following the application of a vacuum bell, but the test is not reliable enough to exclude a patient from having exercise intolerance as a result of severe pectus excavatum.
(Class IIb, Level of evidence B)

THE EVIDENCE FOR THE BENEFIT OF A SURGICAL PROCEDURE IN PATIENTS WITH PHYSIOLOGICAL IMPAIRMENT DUE TO SEVERE PECTUS EXCAVATUM

Our literature review identified 17 papers that looked at cardiopulmonary exercise tolerance in patients undergoing surgery for pectus excavatum. Twelve studies (499 patients) demonstrated a benefit, and 5 studies (149 patients) demonstrated no benefit (Table 3). Eight of the studies showed statistically significant improvements in all parameters of cardiopulmonary testing. Bawazir *et al.* [15] did not demonstrate an improvement, but of note, this same group from France published the study by Sigalet *et al.* 2 years later [16], which demonstrated a significant improvement in cardiopulmonary exercise postoperatively.

There were a wide range of ages across these studies from an average age of 30 in the Jaroszewski *et al.* study [3] to an average age of just 13 in the paediatric studies. The Haller index was generally above 3.25 in all of the studies; the average was around 4.5 in the patients with exercise intolerance.

The improvements were similar in the studies looking at the Nuss procedure and the Ravitch operation, with the maximal improvement tending to be after bar removal in the patients having the Nuss procedure.

Of note, only 2 of the papers [17, 18] we identified in this literature review were included in the NHS England literature review [2]. In addition, Jaroszewski *et al.* [3] was reviewed by the clinical panel after publication but determined not to provide adequate evidence to support commissioning for patients with cardiac compression.

The view of this guideline group is that, overall, the 17 papers documented in Table 3 do demonstrate a significant benefit following surgical procedures in patients with severe pectus excavatum with cardiac compression and impaired cardiopulmonary exercise ability. A Haller index of 3.25 is consistently used to determine the severe category, and a VO_2 max of <85% predicted is a reasonable physiological measure of exercise intolerance.

It should be noted that some patients who have a strong history of athletic ability prior to their rapid growth phase report marked deterioration as their pectus deformity becomes severe but do not show a VO_2 max below 85%. In this situation, other assessments of cardiac compression could be used to assess the likelihood of improvement such as cardiac MRI scanning or exercise echocardiography to look for objective evidence of right ventricular compression or right atrial compression.

Recommendation

Patients with severe pectus excavatum with a Haller index above 3.25 and objective evidence of cardiac compression benefit physiologically from surgical intervention.
(Class I, Level of Evidence B)

Some patients who experience shortness of breath and exercise intolerance but do not fit the preceding criteria may also benefit from surgery, but other objective parameters should be used to determine the likelihood of improvement in cardiovascular performance.
(Class IIb, Level of Evidence C)

EVIDENCE FOR THE BENEFIT OF SURGICAL PROCEDURES IN PATIENTS WITH EXTREMELY SEVERE PECTUS EXCAVATUM

There are case reports of patients with extremely severe pectus excavatum with symptoms not solely including shortness of breath. In the most severe cases, patients may suffer malignant ventricular arrhythmia, including ventricular fibrillation. We also identified cases of patients with compression of the inferior vena cava including collapse and cases of dysphagia.

In addition there are patients with severe pectus excavatum who require scoliosis surgery. They may be put at significant operative risk because centralizing the spine may cause significant cardiac compression during the procedure.

We performed a literature review of the most severe case reports [19–36] (Appendix 4). We identified patients aged 34 months to 34 years requiring urgent operations. The gender split was 12 males and 7 females. We grouped these patients into 4 categories. The first category includes patients with symptomatic or life-threatening arrhythmias due to severe pectus excavatum. The second category includes patients with symptomatic structural compression of the heart or the inferior vena

Table 3: Summary of papers investigating an improvement in physiological parameters after surgery for pectus excavatum

Study	Population	Intervention and comparison	Outcomes reported
Jaroszewski *et al.*, 2022, retrospective cohort study [3] **USA**	*n* = 130, over 18 and Haller index over 3.25. Age 31.0 ± 9.8 Haller index 4.7 ± 2.3	**Intervention** Modified minimally invasive Nuss procedure **Comparison** No comparator Postoperative CPET performed just before removal at 3 years	VO_2 max 25.9 ± 6.0 preoperatively VO_2 max 28.5 ± 7.0 3 years postoperatively ($P < 0.001$) VO_2 max/predicted 72.8 ± 15.4% preoperatively VO_2 max 84 ± 20.6% postoperatively ($P < 0.001$) O_2 pulse 11.7 ± 3.6 (84.5 ± 16.9%) preoperatively O_2 pulse 12.9 ± 3.7 (94.3 ± 21.4%) ($P < 0.001$)
Del Frari *et al.*, 2021, prospective, open-label, single-arm, single centre, non-randomized study [77] **Austria**	*n* = 19 Age range 13.9–19.6 years Mean Haller index 4.2 ± 1.0 Range 3.1–6.3	**Intervention** Minor open video- assisted repair with horizontal sternal osteotomy, chondrotomies and Nuss type bar inserted **Comparison** No comparator Postoperative follow-up was 6 months after initial surgery [with bar(s) in situ] and 5.7 ± 7.9 months following bar removal at 1–2 years postoperatively	No significant difference in VO_2 max between preoperatively, 6 months postoperatively or after bar removal. Other variables: Peak power (watts) significantly improved post bar removal compared to preoperative sitting ($P = 0.03$) and 6 months postoperative supine ($P = 0.005$) Majority of lung function tests statistically better post bar removal compared to preoperatively or 6 months postoperatively
Castellani *et al.*, 2020, prospective cohort study [78] **Austria**	*N* = 59 Mean age 15.7 ± 4.5 years	**Intervention** Nuss procedure **Comparison** No comparator First test within 12 months prior to surgery. Second test 6–12 months after operation. Third test 6 months to 3 years after implant removal	Mean VO_2 max (ml/kgbw/min) was similar preoperatively and while pectus bar was in situ ($P = 0.368$). After bar removal, comparison to preoperatively showed a significant drop in V02 max ($P = 0.0027$).
Das *et al.*, 2019, prospective cohort study [79] **USA**	*n* = 24 children with Haller index > 3.25. Age 9–18 (mean 12.9 years) Haller index 4.3 ± 0.9	**Intervention** Nuss procedure **Comparison** None	VO_2 max 32 ± 13 preoperatively VO_2 max 45 ± 10 postoperatively ($P = 0.0001$) VO_2 max/percentage 62% ± 25% preoperatively VO_2 max 88% ± 16% postoperatively ($P = 0.0005$) O_2 pulse 9 ± 4 (76% ± 24%) preoperatively O_2 pulse 13+/-5 (96% ± 16%) postoperatively $P = 0.03$ ($P = 0.001$)
Udholm *et al.*, 2016, prospective cohort study [80] **Denmark**	*n* = 15 Designed to look at adult patients > 21 years old Mean age 32	**Intervention** Modified minimally invasive Nuss procedure **Comparison** No comparator Postoperative CPET was at 1 year	VO_2 max 30.4 ± 6.5 preoperatively 33.3 ± 5.4 postoperatively ($P = 0.09$)

Continued

Table 3: Continued

Study	Population	Intervention and comparison	Outcomes reported
Kelly *et al.*, 2013, multicentre prospective cohort study [81] **USA & Canada**	$n = 20$ Subset of study population where: Age range 3–21, mean 13.6 ± 3.26 years CT scan index > 3.2, mean 4.73 ± 1.38	**Intervention** 19 patients had an open procedure and 1 had a Nuss procedure **Comparison** No comparator	VO_2 max values were 10.1% higher postoperatively. VO_2 max (l/min) 3.18 ± 3.33 preoperatively 3.50 ± 3.39 postoperatively after bar or strut removal ($P = 0.002$) O_2 pulse values were 19% higher postoperatively 13.58 ± 3.53 preoperatively and 16.16 ± 4.99 postoperatively ($P = 0.009$) Other measures: Work (watts) was 11% higher postoperatively. 178.38 ± 59.39 preoperatively and 198.01 ± 51.83 postoperatively ($P = 0.012$)
O'Keefe *et al.*, 2013, prospective case series [18] **Canada**	$n = 67$ Age: 13.9 ± 2.3 years Haller index: 4.4 ± 1.3	**Intervention** Nuss procedure with bar removal when the patients had reached skeletal maturity or after 3 years, whichever came first **Comparison** No comparator	VO_2 max 70.1 ± 15.0 preoperatively VO_2 max 1 year postoperatively 72.0 ± 17.9 ($P = 0.14$) VO_2 max 33.2 ± 7.5 preoperatively VO_2 max 34.2 ± 7.5 postoperatively ($P = 0.09$) O_2 pulse 75.8 ± 14.4% preoperatively O_2 pulse 80.5 ± 18.3% postoperatively ($P = 0.01$) Other outcome measures: Ability to exercise (3.3 ± 0.7 vs 4.3 ± 0.6, scale 1–5) increased significantly
Maagaard *et al.*, 2013, prospective cohort study [82] **Denmark**	$n = 49$ patients and 26 controls Age 15.5 ± 1.7 for patients Haller index 4.9 ± 1.4	**Intervention** Nuss procedure **Comparison** 26 healthy age-matched controls also performed tests **CPET** Preoperatively and at 1 year and 3 years (after bar removal)	VO_2 max 26 ± 7.1 preoperatively for patients VO_2 max 29 ± 5.9 postoperatively for patients ($P = 0.0001$) VO_2 max 30 ± 7.7 on first test, 35 ± 6.7 on 1-year test and 31 ± 8.0 at 3 years for controls
Tang *et al.*, 2012, prospective cohort study [83] **Denmark** **[Early results of same cohort as Maagaard *et al.*, 2013]**	49 Patients 26 Controls Mean age 15.5 ± 1.7 Haller index 5.3 ± 2.3	**Intervention** Nuss procedure **Comparison** Healthy control group	Left ventricular diastolic diameter was significantly increased. ($P < 0.05$) at 12 months of follow-up in the patients with excavatum • Right ventricular diastolic diameter and VO_2 max were significantly increased in the control group at 12 months of follow-up. • maximum cardiac index was significantly improved at 1 year postoperatively ($P = 0.0054$) but was still significantly lower than the control group ($P = 0.0008$).
Cooper *et al.*, 2010, prospective cohort study (abstract only) [84] **USA**	$n = 28$ Age range 13–65 years. Mean 24 ± 14 Haller index mean 8.89 ± 5.16	**Intervention** Surgical correction—method not stated **Comparison** No comparator	VO_2 max preoperatively 2.37 ± 0.68 l/min (81% predicted) VO_2 max postoperatively increased by 0.12 ± 0.06 ($P = 0.002$)

Continued

Table 3: Continued

Study	Population	Intervention and comparison	Outcomes reported
Neviere *et al.*, 2011, prospective case series [17] **France**	$n = 70$ Age: 27 ± 11 years (range 18–62 years) Haller index: 4.5 ± 1.1.	**Intervention** Simplified Ravitch-type repair with removal of strut 6 months after initial procedure **Comparison** No comparator	VO_2 max 77 ± 2% preoperatively VO_2 max 1 year postoperatively 87% ± 2% ($P = 0.0005$) VO_2 max 34.9 ± 7.5 preoperatively VO_2 max 37.6 ± 7.1 postoperatively ($P = 0.0001$) O_2 pulse 13.2 ± 3.1 (91% ± 12) preoperatively O_2 pulse 14.8 ± 3.2 (106% ± 8) postoperatively
Sigalet *et al.*, 2007, prospective cohort study [16] **France**	26 patients Age 13.2 ± 2.1 years Haller index 4.5 ± 1.1	**Intervention** Nuss procedure **Comparison** None	VO_2 max, O_2 pulse, respiratory quotient, stroke volume, and cardiac output all showed significant increase ($P = 0.05$) post-bar removal
Bawazir *et al.*, 2005, prospective cohort study [15] **France**	48 patients Age 13.5 ± 1.7 Haller index 3.9 ± 0.8	**Intervention** Nuss procedure **Comparison** None	At preoperative assessment, VO_2 max and anaerobic threshold were significantly below normal for patients of this age and size. • There was a significant decrease in VO_2 MAX and anaerobic threshold at 3 months post-repair ($P < 0.05$), and they had returned to the pre-operative values by 21 months post-repair, while maximal heart rate and the minute volume of ventilation showed no significant changes.
Haller *et al.*, 2000, prospective cohort study [85] **USA**	36 patients 10 non-athletic controls Mean age 16 ± 3 years with age matched controls Pectus index >3 (range 3.2–6)	**Intervention** Ravitch-type repair with removal of strut at 6 months **Comparison** Healthy control group Postoperative studies at 6 months (at time of strut removal)	$V0_2$ max not recorded postoperatively for patients or controls. Preoperatively no significant difference between the 2 groups In the surgical group, O_2 pulse was 11.5 ± 3.7, preoperatively rising to 12.9 ± 3.6 postoperatively. ($P < 0.005$).
Morshuis *et al.*, 1994, prospective cohort study [86] **Netherlands**	35 patients Mean age 17.9 ± 5.6 Haller index NR	**Intervention** Open technique **Comparison** None	There was a significant increase in VO_2 max and oxygen pulse ($P = 0.0333$) during exercise with no change in the maximal work performed or in the heart rate
Wynn *et al.*, 1990, prospective cohort study [87] **USA**	8 PE patients with surgical repair 4 PE patients without repair Age range 10-16 years Operative group mean age 13.6 years Nonoperative group mean age 14 years Thoracic index (depth/width) calculated with calipers was 0.47 ± 0.07 in the operated group and 0.49 ± 0.05 in the non-operated group in the first study. In the second study, there was a significant change in the surgical group of 0.59 ± 0.05 ($P \leq 0.05$)	**Intervention** Open—modified Ravitch **Comparison** PE patients that did not have surgical correction 1st study preoperatively < 3 months preoperatively. Postoperative timing evaluation 6 to 21 months postoperatively (mean 11 months). Interval stated to be similar for non-operated group.	VO_2 max—no significant change between the 2 studies for the non-operated group 41.2 ± 7.3 and 43.0 ± 6.9 Also no significant change in the operated group: preoperatively 36.1 ± 4.4 and postoperatively 38.1 ± 8.1 Other measures: Work performed (% predicted) was significantly higher in the surgical group post-operatively—50.1 ± 19.7 to 69.5 ± 23.2 ($P \leq 0.02$).

Continued

Table 3: Continued

Study	Population	Intervention and comparison	Outcomes reported
Cahill *et al.*, 1984, prospective selective series of patients. [88] **USA**	14 patients with PE 5 patients with PC PE age range 6–17 years with an average age of 10.9. PC age range 13–25 with an average age of 13.4. Severity of PE not commented on Selection criteria for series not expanded on	**Intervention** Surgery—type not stated **Comparison** 5 patients with PC who also had surgery were analysed separately Postoperative studies 3–9 months after surgery	VO_2 max preoperatively was 1.26 ± 0.44 for the PE group but higher in the PC group (2.22 ± 0.13). Postoperatively the PC group had no significant difference in their VO_2 max, but the PE group rose to 1.46 ± 0.42 ($P < 0.01$). Total exercise time (min) was lower in the PE group preoperatively: 6.01 ± 2.12 compared to PC 10.30 ± 1.19. Postoperatively the PC group had no significant difference in their exercise time, but the PE group rose to 7.10 ± 2.58 ($P < 0.01$).

CPET: cardiopulmonary exercise text; O_2: oxygen; PC: pectus carinatum; PE: pectus excavatum; USA: United States of America; VO_2: ventilatory uptake of oxygen.

cava. The third category is that of compression of the oesophagus causing dysphagia and finally there is a category of patients undergoing scoliosis surgery for whom centralizing the spine may cause significant cardiovascular collapse.

In the majority of these case reports, surgical procedures resulted in complete or near complete resolution of all symptoms.

Recommendation

For patients with very severe pectus excavatum and evidence of symptomatic malignant arrhythmias with no other cause identified, surgical treatment is indicated to mitigate the risk of death from that arrhythmia.
(Class I, Level of Evidence C)

For patients with very severe pectus excavatum with strong evidence of significant compression of the right heart or the inferior vena cava, surgery is indicated to relieve cardiac compression.
(Class I, Level of Evidence C)

Surgery is indicated in patients with syncope or presyncope due to severe pectus excavatum.
(Class I, Level of evidence C)

Patients with very severe pectus excavatum and dysphagia (not otherwise explained by oesophageal pathology) should benefit from surgery to relieve these symptoms.
(Class IIa, Level of Evidence C)

We recommend that patients with severe pectus excavatum undergoing scoliosis surgery be carefully assessed by an experienced spinal surgeon and an anaesthetist. If it is felt that scoliosis surgery may reduce the space enough to compress the heart, then pectus surgery must be performed prior to scoliosis surgery.
(Class I, Level of Evidence C)

EVIDENCE FOR BENEFIT OF SURGERY IN PATIENTS WITH PSYCHOLOGICAL IMPAIRMENT DUE TO SEVERE PECTUS EXCAVATUM, CARINATUM OR ARCUATUM DEFORMITY

There is a group of patients who suffer from severe psychological harm due to their pectus deformity. This deformity may be pectus excavatum, but in addition severe psychological harm can be caused by carinatum and arcuatum deformities. The definition of severity of psychological impairment is therefore not assessed according to the physical shape of the deformity but the impact that it has on the patient.

More treatments are available for this group. In addition to the Nuss procedure for pectus excavatum and the Ravitch operation for all deformities, there is the newer Pectus Up procedure and pectus implants that can minimize the psychological impact of the defect. Conservative measures include the vacuum bell for patients with excavatum and bracing for patients with carinatum. Psychological therapy is an option for all types of pectus to help the patients cope with the impact of the deformity.

This topic was reviewed by NHS England in 2021 together with their literature review summarizing the psychological benefits of surgery [2, 7]. In their major literature review summary, they selected a non-systematic review, 5 prospective cohort studies and 2 retrospective papers on the same patient group from Poland [17,18, 37–42] (Table 4).

When summarizing the papers for quality of life, they found that Kuru *et al.* [39] reported median (IQR) patient-assessed scores ($n = 88$) of psychosocial functioning rising from 22.5 (19–25) preoperatively to 33 (30–35) postoperatively ($P < 0.0001$), physical functioning from 9 (9–10) preoperatively and 10 (9 to 12) postoperatively ($P < 0.001$) and total functional scoring from 31 (31 to 35) preoperatively to 43 (43 to 46) postoperatively ($P < 0.0001$). The same case series reported median (IQR) parent-assessed scores of psycho-social scoring from 20 (17 to 23) preoperatively to 24 (21 to 26) postoperatively ($P = 0.001$). All these findings were highly significant.

Table 4: Summary of the studies included in the National Health Service England policy review

Study	Population	Intervention and comparison	Outcomes reported
Kelly *et al.*, 2020, prospective study [38] USA	$n = 996$ children and adults with PE Haller index: 5.46 ± 8.26 No subgroups reported	**Intervention** Nuss procedure **Comparison** No comparator	**Important outcomes** Postoperative cosmetic satisfaction Safety: early (< 30 day) and late (timescale not stated) postoperative complications
Pawlak *et al.*, 2018, retrospective study [41] Poland	$n = 1006$ with excavatum, including $n = 44$ with recurrence after previous surgery. Age: 18.6 ± 5.7 years (range 7–62 years) Haller index: 3.7 ± 1.4 (2.6–17.3) No subgroups reported	**Intervention** Nuss procedure using video-assisted thoracoscopy **Comparison** No comparator	**Important outcomes** Early (up to 30-day) thoracoscopy-dependent postoperative complications
Pawlak *et al.*, 2016, retrospective study [42] Poland	$n = 680$ with PE who had surgery. The authors confirm that this is a subset of their more recent paper [39].	**Intervention** Nuss procedure **Comparison** No Comparator	**Critical outcomes** Cardiopulmonary outcomes—imaging
Luo *et al.*, 2017, prospective study [40] China	$n = 266$ with PE Age: 19.02 ± 4.42 years Haller index: 4.18 ± 1.20 No subgroups reported	**Intervention** Nuss procedure **Comparison** No comparator	**Critical outcomes** Psychological outcomes: Symptom Checklist-90 and Self-rating Depression Scale preoperatively and 1 year post-operatively **Important outcomes** None reported
Kuru *et al.*, 2015, prospective study [39] Turkey	$n = 88$ with PE Age: 18.44 ± 3.93 years (range 14–29 years) Haller index: 4.04 ± 1.34 (range 2.2–9.5) Haller index >3.25: 64 (78%)	**Intervention** MIRPE **Comparison** No comparator	**Critical outcomes** Quality of life: Pectus Excavatum Evaluation Questionnaire (Nuss questionnaire) preoperatively and 6 months postoperatively
Johnson *et al.*, 2014, systematic review [37] USA	Subjects with PE Paediatric (age 0–21 years): 1500 Nuss procedures and 1186 modified Ravitch procedures Adult (age 17–67 years): 262 Nuss procedures and 498 modified Ravitch procedures Average Haller or equivalent index >3.2 Subgroups: outcomes reported for children and adults undergoing the Nuss or Ravitch procedure	**Intervention** Nuss or modified Ravitch procedure **Comparison** No comparator	**Important outcomes** Safety: non-displacement complications, bar/strut displacement, need for reoperation. Mean follow-up between 2.0–3.9 years in different groups (range 0.08–21.5 years)
O'Keefe *et al.*, 2013, prospective study [18] Canada	$n = 67$ with PE Age: 13.9 ± 2.3 years Haller index: 4.4 ± 1.3 No subgroups reported	**Intervention** Nuss procedure with bar removal when the patients had reached skeletal maturity or after 3 years, whichever came first **Comparison** No comparator	**Critical outcomes** Cardiopulmonary outcomes—exercise testing preoperatively and 3 to 6 months after bar removal Cardiopulmonary outcomes—imaging preoperatively and 3 to 6 months after bar removal **Important outcomes** Lung function tests preoperatively and 3 to 6 months after bar removal Postoperative cosmetic satisfaction
Neviere *et al.*, 2011, Prospective study [17] France	$n = 70$ with PE Age: 27 ± 11 years (range 18–62 years) Haller index: 4.5 ± 1.1. No subgroups reported	**Intervention** Simplified Ravitch-type repair with removal of strut 6 months after initial procedure **Comparison** No comparator	**Critical outcomes** Cardiopulmonary outcomes—exercise testing preoperatively and 1 year postoperatively Cardiopulmonary outcomes—imaging pre- and postoperatively: Haller index **Important outcomes** Lung function tests

MIRPE: minimally invasive repair of pectus excavatum; PE: pectus excavatum.

They described the findings of Luo *et al.* [40], who reported the results of 266 adult and paediatric patients. These authors used the Symptom Checklist-90-Revised [43]. The total average score was 152.02 ± 55.04 preoperatively and 141.75 ± 19.56 postoperatively, which was statistically significant ($P = 0.037$). The somatization score was 1.57 ± 0.59 preoperatively and 1.23 ± 0.62 postoperatively ($P = 0.001$). The interpersonal sensitivity score was 1.92 ± 0.72 preoperatively and 1.77 ± 0.64 postoperatively ($P = 0.025$). The depression score was 1.74 ± 0.78 preoperatively and 1.57 ± 0.69 postoperatively ($P = 0.003$). The anxiety score was 1.73 ± 0.71 preoperatively and 1.58 ± 0.73 postoperatively ($P = 0.013$). The score for other aspects of the symptoms was 1.67 ± 0.66 preoperatively and 1.57 ± 0.49 postoperatively ($P = 0.104$). The numbers whose scores were considered to indicate mental health problems were 161 (60.53%) preoperatively and 79 (29.70%) postoperatively; both were highly statistically significant with a $P < 0.001$.

NHS England summarized 3 papers that looked at cosmetic satisfaction (Kelly *et al.* in 431 patients, O'Keefe *et al.* in 87 patients and 1 of the papers by Pawalak *et al.* with 680 patients) [18, 38, 41].

O'Keefe *et al.* ($n = 67$) reported patients' self-rated appearance on a scale from 0 = looks terrible to 5 = looks normal [18]. The mean (± standard deviation) score preoperatively was 2.51 ± 0.82 and 4.43 ± 0.54 ($P < 0.001$) postoperatively. The findings of a satisfaction survey completed at the postoperative visit found that 96% of respondents said they were 'very happy' or 'happy' with their operation. Pawlak *et al.* reported satisfaction with the results of the operations. In children with a mean age of 12.2 ± 2 years (range 7–14 years) ($n = 156$), they reported a satisfactory outcome in 96.8% and an unsatisfactory outcome in 3.2%. They documented the paper by Steinmann *et al.* that described significant psychological distress leading to concerns about appearance, withdrawal and social isolation in a group of 90 patients case matched with 80 controls [44].

The NICE technology appraisal of 2009 [5] also reviewed the psychological benefits of the procedure in 10 papers [45–55] and in the UK national data set. In their analysis of the UK national data set, the patient's score of the cosmetic appearance of their chest increased from a mean of 3.1 out of 10 to 8.4 postoperatively; this result was repeated in 3 case series that they reviewed containing 1441 patients in which the patients reported an excellent result in 82% of cases.

In addition, we identified Kelly *et al.* from 2008 [56], who reported on 264 child patients and 291 parents from multiple centres using a validated Pectus Excavatum Evaluation Questionnaire. The children noted a dramatic improvement in their body image and physical difficulties postoperatively. Parents also noticed an improvement in the children's emotional and physical difficulties and social self-consciousness. Patient satisfaction with the chest appearance was found to be very good, with excellent to good results reported in more than 95% of patients at the time of bar removal. We also identified Lomholt *et al.* [57], who reported 107 patients and 106 parents who completed the generic health-related quality-of-life measure. The Child Health Questionnaire was assessed preoperatively and at 3 and 6 months following pectus repair. A control group of 183 school children completed the same measure on 1 occasion. In the postoperative study, patients and parents reported improved emotional well-being and self-esteem. Additionally, patients at both 3 and 6 months postoperatively reported increased physical and social activities.

The Nuss questionnaire (otherwise known as the Pectus Excavatum Evaluation Questionnaire or PEEQ) has been extensively used to assess the psychological success of the operation [4]. It was originally designed for paediatrics and has been successfully modified for adults [58]. The use of this questionnaire has been shown to demonstrate good improvements in patient satisfaction with 90% chest appearance satisfaction reported [59–62] and quality of life improved in 90% of patients.

The literature highlights many improvements in psychological well-being. but there are no studies that are able to determine a threshold of psychological harm that justifies an operation, and a level below which an operation is not beneficial. This result is likely due to the fact that all the patients in the cohort studies have sought out a surgical procedure as a solution to their pectus abnormality. They have consented to have the operation, which in itself is likely to define the patient as being significantly psychologically impacted because they are willing to undergo a significant operation with a lengthy recovery period with associated pain. In addition, we found no studies that investigated psychological therapy as an alternative to an operation, and no comparative or randomized trials on this subject. We note that surgeons are not trained in psychological assessment and in the delineation of body dysmorphia or somatization of other issues in a patient's life. Therefore we recommend using professional psychological services in the selection of patients for an operation, in the recovery to support patients postoperatively and to independently assess the success of the operation when it is performed for psychological benefit.

Recommendation

There is good evidence that patients who are psychologically impacted by their pectus abnormality benefit from surgery in terms of improved quality of life, reduced depression and anxiety scores and that the operation has good patient satisfaction.
(Class IIa, Level of Evidence B)

The assessment of the psychological impact of a pectus abnormality on a patient should be assessed by a psychologist prior to the operation for psychological benefit.
(Class IIa, Level of Evidence C)

The NUSS questionnaire or the NUSS questionnaire modified for adults (also known as the Pectus Excavatum Evaluation Questionnaire–PEEQ) should be used to demonstrate an improvement in quality of life in patients undergoing pectus surgery.
(Class IIa, Level of Evidence B)

OPERATIVE TECHNIQUE

Several potential operations are available for patients. The Nuss procedure has been performed more than 50 000 times worldwide [63] and is the most popular operative choice for pectus excavatum. The Ravitch operation was the original operation performed for pectus excavatum in the past, prior to the Nuss procedure, and is still an option for patients who do not wish to have a second operation for bar removal or for patients with carinatum not amendable to bracing or asymmetrical deformities. In studies comparing the outcomes of both in patients with excavatum deformities, a 2016 systematic review on this subject by Kanagaratnam *et al.* [64] found across 13 studies that in paediatric patients there were no differences between the Nuss and the Ravitch operation including wound infection, haemothorax, pneumothorax or pneumonia. In adults, the Ravitch operation had a slightly lower complication rate, driven mainly by reoperation for bar displacement, but the patients must have reached full bone maturity to avoid the severe complication of acquired asphyxiating thoracic dystrophy [65].

The choice should therefore be made after a discussion with the surgeon and the patient, balancing the risks with the perceived benefits both physiologically and psychologically.

Of note, various modifications to the Ravitch procedure have been published with regard to the size of the incision and the methods of elevating the sternum and of resecting ribs, but these technical variations should all be regarded as essentially similar in risk and benefit.

A new option for patients is the Pectus Up procedure, also called a taulinoplasty. It involves a 4-cm incision over the front of the chest and then a plate across the front of the chest to pull up the sternum via a screw device into the sternum. This plate is permanent, and this procedure is only for patients with pectus excavatum [66]. In contrast to the operations described previously, only a few more than 100 of these procedures have been performed so far; thus no long-term follow-up data or large cohort data are available to guide safety. It should therefore be regarded as a novel procedure currently under development and evaluation. It is hoped that it will significantly reduce pain in the postoperative period and avoid a second operation at the expense of a 4- to 5-cm incision in the midline of the chest.

For patients with pectus excavatum undergoing surgery for the psychological benefit, it is also possible to insert a pectus implant. This procedure is designed from the patient's CT scan; the implant is inserted below the pectoral muscles through a 4- to 5-cm incision in the midline of the chest. It comprises a silicone material that has a firmness similar to that of human cartilage. Because there is no surgical or pulling force on the bones of the chest wall, it is a much less painful operation and patients invariably leave the hospital on days 1–2. There is also no risk of cardiac, thoracic or other organ injury. The major complication is seroma formation, and patients often require seroma aspiration 1 to 5 times postoperatively. This procedure may be an alternative to the other more invasive surgical procedures for those with normal exercise tolerance who are seeking a less invasive solution.

Recommendation

For patients with asymmetrical pectus excavatum and pectus carinatum or arcuatum, the Ravitch procedure and its variations are recommended.
(Class I, Level of Evidence B)

For patients with pectus excavatum, the Nuss procedure and the Ravitch procedure have similar surgical risks and efficacy. The decision as to which operation should be offered should be based on the preference of the patient after a detailed discussion with their surgeon.
(Class IIa, Level of Evidence B)

The Pectus Up procedure, also known as a taulinoplasty, is currently a novel operation under development and evaluation and has currently been performed in only small numbers of patients. Therefore, patients interested in this operation should be aware of the comparative lack of safety data and long-term efficacy data prior to selecting this operation.
(Class IIa, Level of Evidence C)

In carefully selected patients with pectus excavatum and normal exercise tolerance, pectus implants may represent an alternative to more invasive procedures. The same criteria used for the more invasive methods should be used for patient suitability for implant surgery.
(Class IIa, Level of Evidence C).

THE SAFETY OF PECTUS OPERATIONS

This topic has been comprehensively addressed by the NICE 2009 technology appraisal for the placement of a pectus bar for pectus excavatum [5]. The NICE technology appraisal collaborated with the Association of Paediatric Anaesthetists of Great Britain and Ireland, the British Association of Paediatric Surgeons and the Society for Cardiothoracic Surgery in Great Britain and Ireland to create this document.

In their systematic review of the literature, they looked at 10 case series containing more than 3000 patients, and they reviewed the UK national register of 260 patients. They found that in the National Register 13 events were classed as major perioperative adverse events. Of these 13 events, 3 were pneumothoraxes; 3 were pleural effusions; 1 was a bar migration; 1, an infection; 1, pain; 1, lower lobe collapse; 1, an air leak; 1, a pericardial effusion; and 1, hypotension on bar insertion due to adhesions (resolving on repositioning of the bar). There were 49 postoperative adverse events; 23 were categorized as 'major': infection ($n = 13$), bar migration ($n = 4$) and 1 case each of pleural effusion, haematoma, pain, granulomas over stabilizer plates, a broken retaining wire causing discomfort (removed as a day case) and a protruding wire (removed surgically).

In their literature review, they found that across 4 case series, pneumothorax occurred in 55% (369/668), 9% (15/167), 7% (24/322) and 3% (5/172) of patients, most of which resolved spontaneously [45–48]. Bar or stabilizer displacement occurred in 10% (70/668), 5% (9/172), 3% (11/322) and 2% (3/167) of patients in 3 case series [45, 47, 48]. In the first case series, 50 bar displacements required surgical revision; in the second case series, 4 bar displacements were flipped bars that were classified as major complications; in the last case series, all 3 bar displacements required reoperation [45–47]. Wound infection, pneumonia, pleural effusion, pericardial effusion, and pericarditis were reported in 1–3% of patients in 3 case series. The case series of 668 patients reported transient Horner's syndrome secondary to epidural analgesia in 24% (162/668) of patients [45]. In both case series of 167 and 172 patients, there was 1 case of liver perforation in each series. In both case series of 167 and 322 patients, there was 1 case each of intraoperative cardiac perforation. One case report reported 4 cases of cardiac injury during surgery including 1 death of the injury [49].

The case series of 167 patients also reported 15 cases of intraoperative rupture of the intercostal muscles (in older patients), 10 cases of haemothorax or haemopneumothorax and 7 cases of minor pericardial tears [46].

NICE made a series of recommendations. Firstly, they stated that the current evidence on the safety and efficacy of placement of a pectus bar for pectus excavatum is adequate to support its use provided that normal arrangements are in place for clinical governance, consent and audit.

Secondly, they stated that placement of pectus bars for pectus excavatum should be carried out only by surgeons with cardiac and thoracic training and experience who are capable of managing cardiac or liver injury, and where there are facilities for handling these issues.

Finally, they stated that this procedure should be carried out only by surgeons with specific training in inserting the device and that these surgeons should perform their initial procedures with an experienced mentor. No figures were given for guidance in the NICE Technology appraisal. In personal correspondence, Donald Nuss advised a minimum of 20 cases per annum. He also highlighted an observed drop in the number of complications over time as the procedure has become established as a widely practiced surgical technique. There are also advances in surgical equipment that can assist with safety, such as devices for sternal elevation [67] that lift the chest wall off of the mediastinal structures before the bars are inserted. Some centres utilizing this technique such as Professor Park's practice (described in Kim *et al*. 2023 [68]) have large case series (1526 in the last 10 years) with no deaths and very low complication rates. High-volume centres in the United States use bilateral thoracoscopy with high-pressure CO_2 at around 10 mmHg and also demonstrate excellent safety.

We considered very carefully as a guideline group the parameters by which surgeons and programmes might be assessed and what might be acceptable thresholds. The Delphi process found a majority in favour of the view that there was no single figure above which one could determine that a surgeon or programme was safe. But surgeons should perform pectus operations frequently and regularly in their practice; 2 or more surgeons should be encouraged to be active in each programme and potentially to operate together; practices should be audited regularly; and results should be shared regularly between units. Furthermore, national governing bodies should consider pectus operations as a subspecialty for accreditation and mandate entry of results in national registries.

Recommendation

The Nuss procedure is a safe operation supported by the National Institute of Health and Clinical Excellence when carried out by surgeons with cardiac and thoracic experience, with the knowledge and facilities to deal with any complications that may arise and who perform the operation frequently and regularly.
(Class I, Level of Evidence B)

All patients should have their data entered in a national registry with the ability to follow the patient to assess the safety and efficacy of the procedure.
(Class IIa, Level of Evidence C)

ANALGESIA IN PERIOPERATIVE CARE

An operation for pectus excavatum is painful, and the pain is similar between the 2 most established operations—the Nuss and the Ravitch procedures. Many hospitals run enhanced recovery programmes that advocate multimodal analgesia and pre-emptive analgesia preoperatively; this approach should be regarded as best practice. In addition to paracetamol, a non-steroidal anti-inflammatory and opiate analgesia, many centres use gabapentin or similar agents for nerve-based pain relief. Gabapentin currently is used as an off license indication in this setting, although a UK multicentre randomized trial called the GAP trial is looking at its use in abdominal, cardiac and thoracic surgery and is near completion. Also, epidural analgesia is used in a large number of UK centres but internationally in pectus surgery this practice has largely been superseded by administration of cryoanalgesia. Epidurals can only remain in place for 3–5 days, and there is a risk of spinal cord injury with every epidural. Cryoanalgesia comprises the placement of a probe at -70 degrees Celsius onto the intercostal nerves prior to or at the start of the operation. Approximately 5 nerves treated on either side from around the second to the third to the sixth or the seventh axon successfully grow back over the next 2–6 months. It is a low-risk procedure, and nerve regrowth has been found to be highly successful. In the recent Frost trial [69] in cardiac surgery patients, cryoanalgesia reduced opiate usage to control pain and also increased the forced ventilatory uptake in 1 s in the cryoanalgesia group. Of note, an intercostal blockade can still be used with cryoanalgesia to provide pain relief.

Recommendation

Cryoanalgesia is a promising alternative to epidural anaesthesia in pectus patients and should be considered for patients having pectus surgery.
(Class IIa, Level of Evidence B)

Neuromodulating medications such as gabapentin may be beneficial in the postoperative period but, until further evidence is published on the effectiveness and safety, we make no recommendation either way for its use. Administration should be guided by local practice and clinical experience.
(Class III (lack of benefit), Level of Evidence C)

Enhanced recovery protocols should be used for patients undergoing pectus surgery in a manner similar to that of patients undergoing other types of thoracic surgical procedures.
(Class IIa, Level of Evidence B)

FOLLOW-UP AFTER AN OPERATION

All patients should be followed up within the first 6 weeks postoperatively to check for any early postoperative complications. For the Nuss procedure, it is mandatory to follow up patients while the bars are in situ and to plan for timely removal. Patients should be monitored for any evidence of bar rotation or displacement, and lateral plain imaging as well as postero-anterior chest X-rays or CT scanning should be performed to identify movement of the bar. If the procedure is performed to improve exercise ability, then postoperative CT scanning should be performed to demonstrate elimination of cardiac compression and reduction of the Haller index to normal levels. Ideally, a postoperative CPET test should be performed at least 1 year postoperatively or after bar removal. For open (Ravitch) and Nuss bar (post removal) procedures, it is good practice to continue to follow up to check that the procedure has been effective and to check that there is no recurrence. Surgeons may want to follow up their patients for 5 years after the initial operation. This amount of time provides quality control and ensures that any issues can be resolved for the individual patient but also feeds back into modifications of the base technique where lessons can be learned.

PRINCIPLES AND PRACTICE OF CONSENT

It is important to provide a detailed and documented consent process with the patient and family. If the patient has cardiovascular compromise, then it is important to discuss with the patient the expectations for improvement. Our literature review demonstrates consistent improvement in exercise ability. However, the studies show that the patient does not return to the cardiopulmonary exercise ability of a normal person of their age and gender, and it is important to ensure that the patient is given this information. In addition, the studies document that maximal exercise ability is not attained until the bar is removed following the Nuss procedure or after 6 months to a year for the Ravitch procedure.

For patients with severe psychological impairment due to their pectus deformity, it is imperative that a full psychological assessment be made prior to surgery, and in addition it is preferable if psychological support is available postoperatively. The expectations of the patient should be discussed, and a detailed account of the likelihood of normality for the patient's chest should be documented. In particular, rib flaring and upper chest deformities should be mentioned because there can often be some residual abnormality. We highly recommend preoperative and postoperative photography because it is helpful in the postoperative period if the result can then be compared to the preoperative images. These should ideally be taken by the hospital photography department and are also important for documentation purposes.

Although it may be stated that this is a very safe operation, deaths can occur both during the operation and on bar removal. In a review and survey in 2018 from Hebra *et al*. [63] of the 50,000 operations performed since 1998, there have been 11 deaths during the primary operation and 2 on bar removal. This result represents a mortality risk of 0.03%. In addition, 27 published and 32 unpublished life-threatening events were identified for patients who eventually survived, giving a risk of 0.1% of a major complication. Minor complications are much more common and include pneumothorax, which is the most common complication (up to 20%), wound or chest infection (<5%) or bar infection (1%), bar displacement, which is around 1%, or pleural effusion.

Pain is present in all patients to varying degrees, and the expectations around pain and its management should be discussed.

Recommendations concerning time off from activity vary, but most patients are encouraged to walk immediately, minimize upper body activity for around 6 weeks and return to studies or work at around 4–6 weeks. Full upper body activity should only be resumed after 3 months. For the Nuss procedures, some centres warn against significant upper body trauma such as kick-boxing or high-risk activities such as downhill mountain biking for the full 3-year period that the bar(s) is in place.

Nuss bars must be removed after 3 years (patients with Marfan's syndrome often have their bars left in place for 4 years), and it is not acceptable to leave the bars in situ longer-term. The surgeon should bear significant responsibility to ensure that the patient attends again at the indicated time for bar removal, but ultimately it is the patient's responsibility to reattend or seek removal if they change location. There are virtually no long-term data on leaving Nuss bars in situ for the rest of a patient's life, and there is a significant risk that it will naturally migrate into the chest with time because the sternum continues to remodel over the lifetime of the patient.

Recommendation

The perceived benefits of the operation must be documented, including the perceived physiological benefit and the psychological benefit.
(Class I, Level of Evidence C)

Preoperative medical photography should always be performed if the patient is seeking psychological benefit.
(Class IIa, Level of Evidence C)

Risks should be documented and recorded according to internationally published data supplemented with local experience.
(Class IIa, Level of Evidence B)

A full pathway to complete recovery should be documented in advance, including time to return to work or study and time to full activity.
(Class IIa, Level of Evidence C)

It is not recommended that a Nuss bar be left in place for the lifetime of the patient; although the surgeon bears significant responsibility to ensure the patient returns for removal, ultimately it is the patient or their legal guardian(s) who must agree to reattend for removal
(Class III (harm), Level of Evidence B)

BRACING FOR PECTUS CARINATUM

Bracing is a non-surgical option for patients with pectus carinatum. The principle is that a brace is placed over the highest point of the chest and used to compress the sternum and position it into a more normal position. This brace is then worn for a period of time during which the bones of the chest wall remodel so that the sternum eventually stays in this position (or at least a less prominent position) once the brace is no longer worn. The success of bracing is dependent on the severity of the pectus carinatum abnormality and also on the bone density of the patient (which becomes denser and therefore less malleable with age). Complications can occur if excessive pressure is placed on the skin, and some devices measure the pressure applied to the skin to help regulate this. There is a lot of variation in 'wear regimes', multiple manufacturers of braces and differing levels of support given during the time the brace is worn. De Beer *et al.* [70] reported their findings in 740 patients using a dynamic compression brace with a pressure-measuring device from 2013 to 2020 to evaluate factors that may predict final success. Full treatment took between 1 and 2 years. Each additional centimetre in height of the carinatum deformity increased the treatment time by 2 months. An initial correction pressure of more than 7.6 psi increased treatment time by 3.5 months. Patients with an asymmetrical chest were more likely to abandon treatment (74 patients in total abandoned the treatment—10%). The mean age of the successfully treated patients was 13.8 years and was a year lower than the average of those who abandoned treatment. Aesthetic scoring of the chest rose from 4.0 before treatment to 7.8 out of 10 after treatment in the cohort. In a systematic review by the same authors [71], they found 8 other studies comprising 1185 patients using a brace similar to theirs and found similar results with a low complication rate (mostly chest discomfort 12% or skin lesions 5%) and a 6% dropout rate. In a systematic review by Hunt *et al.* in 2019 [72], 16 articles were identified. The brace was found to be a highly effective conservative solution for pectus carinatum with a low complication rate and high success rate. The bracing protocol significantly influenced success with the 24-h wear time being the most effective protocol and low wear times that gradually increased being least effective. Our guidelines group recommends that bracing for pectus carinatum is a highly effective non-surgical alternative treatment of this condition. With good patient selection, high wear protocols and support, it will effectively avoid surgery in a large proportion of patients if offered to them in their early teenage years. It is likely to be highly cost-effective with a much better patient experience.

Recommendation

Bracing is a highly effective and safe conservative treatment for patients with significant pectus carinatum and must be available to all suitable patients who wish to be treated for their condition. It has fewer risks and is cheaper than an operation and still provides good outcomes.
(Class I, Level of Evidence B)

We recommend treating patients with regular professional support to supervise the wearing time; documented high-wear protocols should be used.
(Class IIa, Level of Evidence B)

All patients with a pectus carinatum should be encouraged to visit a specialist clinic as early as the condition is identified: Treatment becomes significantly less effective the older a patient is when starting treatment.
(Class IIa, Level of Evidence B)

Many types of devices are available, including ones that measure the correction pressure. We make no recommendations as to the optimal device.

VACUUM BELL THERAPY FOR PECTUS EXCAVATUM

Treatment with a vacuum bell is a conservative treatment for patients with pectus excavatum. The vacuum bell is placed over the abnormality, and a vacuum is induced that brings not only the skin and soft tissue forwards but also the sternum. The principle is then similar to that of bracing for pectus carinatum in that the body will remodel in this new position if the sternum is held forwards for a long period of time. The success of this treatment is again dependent on the depth of the abnormality and the age of the patient (compliance of the chest).

In a systematic review published in 2019 [73], Patel and Hunt identified 7 papers on this subject. They reported that an initial chest wall depth of $<$ 1.5 cm, chest wall flexibility and age $<$11 when starting treatment were all predictive of a better result. The largest study they identified was by Obermeyer *et al.* [74] in 180 patients. An excellent result was reported in only 20% of patients and wear time had to be more than a year.

Toselli *et al.* [75] in 2022 reported their results in 186 patients. One third of their patients achieved an excellent or good result, but more than half had a fair or poor result. Success was dependent on a smaller initial depth of $<$1.8 cm, and treatment time was again more than a year.

Thus vacuum bell therapy is much less successful overall than bracing is for pectus carinatum. A major factor may be that the vacuum bell cannot be worn for long continuous periods of time, and many patients are able to wear it only in the evenings and cannot sleep with it. Close support is mandatory for patients undergoing vacuum bell therapy. Children need to be old enough to want to comply with treatment, but it is imperative to treat these patients as early as possible because results become much less effective because the chest becomes less compliant with age.

However, it is a safe conservative treatment for pectus excavatum and in selected patients may also avoid the requirement for an operation in later years.

Recommendation

Vacuum bell therapy is a safe conservative treatment for patients with pectus excavatum and should be available to all suitable patients who wish to have treatment for their condition, although results are best in less marked defects and in younger patients.
(Class IIa, Level of Evidence B)

We recommend treating patients with regular professional support to supervise the wearing of the device.
(Class IIa, Level of Evidence B)

Patients with pectus excavatum should be assessed early by specialists who will be able to assess whether vacuum bell therapy could be a suitable treatment for young patients.
(Class IIa, Level of Evidence B)

PECTUS AND ITS ASSOCIATION WITH CONNECTIVE TISSUE DISORDERS

It should be noted that the incidence of pectus in the general population is around 1/1000 births and the incidence of Marfan syndrome is 1/3000 births in European countries; thus both are relatively common conditions in our society. About half of patients known to have Marfan syndrome have a documented pectus abnormality, but when considering the reverse, whereby a patient presents with an isolated pectus abnormality, it is found that around 5% of patients will subsequently be found to have Marfan syndrome [76]. Thus, 95% of patients with an isolated pectus abnormality will not have Marfan syndrome, but it is important to identify which patients require testing.

There is a genetic test for mutations in the fibrillin-1 gene. However, contrary to the expectations of many patients that a simple test can be done to exclude Marfan syndrome, the results of genetic testing are not always straightforward. Sometimes in a limited group of people, they may require more than the usual testing to identify the alteration. For example, gene sequencing may not find mutations if part or all of one copy of a gene is missing. Mutations in the fibrillin-1 gene can also cause conditions other than Marfan syndrome. Therefore, it can be difficult to predict what condition to expect when a mutation is found. In addition, a full Marfan screen in a patient without a family history can cost more than £1000. Therefore, working with a medical geneticist or a genetic counsellor is often necessary after clinical assessment.

Therefore the diagnosis of Marfan syndrome should be a clinical diagnosis based mainly on a clinical examination; the gene test can then be used to help back this up.

A child must have 2 out of 3 major systems seriously affected to be suspected of having Marfan syndrome. If these systems are not present, then meeting with a geneticist is not required. In addition, 75% of cases are suspected from a family history because the child of a parent with Marfan syndrome has a 50% chance of inheriting the condition because it is autosomal dominant.

The 3 major systems that are affected are as follows:

1. Eyes: patients will have dislocated lenses or retinal detachments on examination with a slit lamp or may already have been noted to have this on eye examination. Importantly, just having myopia is not a sign of Marfan syndrome.
2. Cardiac: Patients with Marfan syndrome have aortic root dilatation or aortic dissection or aneurysm. This can be assessed on an echocardiogram but is not otherwise assessable on clinical examination.
3. Skeletal: Many patients with Marfan syndrome have scoliosis, and it is often severe in nature.

As part of a full clinical examination of a patient with an isolated pectus abnormality, in addition to assessing the eyes, heart and skeletal system, a family history should be sought, including for eye, heart or skeletal abnormalities or sudden cardiac death under 50 years of age. The physical examination should document height, arm span, lower segment joint score, look for a high palate, arachnodactyly, test the thumb and wrist for hypermobility, look at the spine for kyphosis or scoliosis, assess for pes planus (flat feet) and look for lumbar striae. Modified Ghent scoring, the Marfan calculator risk score or Ehlers-Danlos score may identify less obvious phenotypes.

Forty percent of patients with Marfan syndrome have arrhythmias; thus, an electrocardiogram (ECG) in all patients with an isolated pectus abnormality is recommended.

If you do not uncover 2 out of the 3 major systems as having a significant abnormality, then the patient can be reassured and the findings documented (including a record of any minor factors observed), but it should be noted that for those who end up with a diagnosis of Marfan syndrome, there are increased risk factors for surgery (in addition to the dilated aortic anatomy). Thus, surgery should typically be delayed until the patient has been fully investigated by a geneticist.

Twenty percent of children with Marfan syndrome have asthma. A smaller proportion are prone to pneumothorax (~5%), and others may have fibrosis, bronchiectasis, lung cysts or fungal infections. A CT scan should be considered to check the lung fields in addition to looking at the aorta and the Haller index.

Recommendation

Five percent of patients presenting with a pectus abnormality will have an underlying connective tissue disorder such as Marfan syndrome. Thus an initial evaluation must be performed in every patient to see if further specialist assessment is required.
(Class I, Level of Evidence B)

The initial assessment must include evaluation of the ocular, cardiac and musculoskeletal systems and whether there is a family history suggestive of a connective tissue disorder.
(Class I, Level of Evidence B)

Any patient thought to be at risk of heritable disorders of connective tissue must be referred to their local genetics service.
(Class I, Level of Evidence B)

AUDIT AND BEST PRACTICE IN PECTUS SURGERY

It is mandatory for all clinicians seeing and treating these patients to keep records of their assessments and outcomes and to perform local audits on a regular basis. We recommend that these data be collected to create national registries of patients with pectus abnormalities and that this information be used to monitor safety and efficacy nationally and internationally. Because pectus treatment is performed by paediatric surgeons with an interest in thoracic surgery, collaboration among adult thoracic surgeons and congenital heart surgeons is required across professional and other societies.

We would also recommend that each centre has the following minimum standards in order to be a commissioned pectus centre and make the following recommendations:

1. A centre must be able to offer a range of services including the Nuss procedure and the Ravitch procedure but also the conservative treatments of bracing and vacuum bell therapy in order to make sure that patients have all options available to them when they are offered treatment.

2. A centre must have an audit process and at least annually review the number of patients seen and the number of patients who receive conservative or surgical treatments together with the number of major complications.
3. Medical photography must be available at each treatment centre.
4. Access to psychological services must be available at each treatment centre.
5. Access to genetic services must be available if a connective tissue disorder is suspected.
6. A centre should see a minimum of 50 patients per year for assessment of their pectus abnormality.
7. A centre should be performing at least 20 pectus operations annually to maintain the minimum perioperative skills to perform safe surgical procedures.
8. A centre must have the ability to deal with cardiac injury, lung injury and liver injury onsite.
9. Depending on jurisdiction, patients classified as paediatric should be seen at a centre accredited for paediatric pectus care and surgery. Older patients should be seen in teenage/young adult or adult pectus care and surgery centres.
10. Transition of care between age-dependent providers must be seamless.

FUTURE RESEARCH IN PECTUS SURGERY

Many questions have not been fully answered in the current published literature. Therefore, the need exists to perform high-quality studies in this area. To date, no randomized controlled trials in pectus surgery have been published. Questions remain as to how much benefit the patient experiences with regard to their own quality of life, how closely this benefit is related to the improvements seen in CPET and how long-lasting this benefit is. In addition, the optimal timing of the surgical procedures in terms of a patient's age and the optimal type of surgery, of bar removal and of reliable investigations to identify patients who will benefit from surgery are still not fully elucidated. The National Institute for Health Research in the UK has recently awarded £1.9 million for a 200-patient randomized trial to investigate the cardiovascular benefits of performing the NUSS or the Ravitch procedure compared to a control group (NIHR158749—A randomized trial of surgery versus no treatment to RESTORE cardiopulmonary function in severe pectus excavatum—The RESTORE Trial). This study will go some way in being able to answer these questions and will commence in July 2024, with the one-year results being reported 3 years later and the final 3- year results in 2029. We propose to update these guidelines based on the findings of that study at that stage.

CONCLUSION

We provide a comprehensive best practice guideline that has been developed across multiple national and international organizations. Our goal is to unify provision of services for patients with pectus abnormalities and ensure best practice. We also aim to provide a framework that will lead to national auditing of the provision of care for these patients to ensure that safe and equitable care is provided for all patients with pectus abnormalities.

FUNDING

There was no funding.

Conflicts of interest: Joel Dunning—Research grant—I am chief investigator of the RESTORE Trial, which is a randomized trial sponsored by NHS England into pectus surgery in patients with cardiovascular impairment. I have been asked my expert opinion on a case that took NHS England to Judicial Review recently with regard to their decommissioning decision for pectus excavatum in England. This was without reimbursement.

APPENDIX 1: GUIDELINES WORKING GROUP

Author and clinical specialty	Affiliations	Capacity
Joel Dunning (Consultant Thoracic Surgeon)	Member Thoracic Subcommittee Society for Cardiothoracic Surgery in Great Britain and Ireland (SCTS) European Society of Thoracic Surgery Co-Chief Investigator of the RESTORE Trial (a randomized trial of surgery versus no treatment to RESTORE cardiopulmonary function in severe pectus excavatum)	On behalf of the SCTS Thoracic Subcommittee and systematic reviewer
Clare Burdett (Specialty Registrar Thoracic Surgery)	SCTS Member Royal College of Surgeons of England	Systematic reviewer
Anne Child (Clinical Geneticist)	Medical Director of the Marfan Trust, UK Fellow Royal College of Physicians	On behalf of the Marfan Trust
Simon Kendall [Cardiothoracic Surgeon(ret)]	Medical Director National Health Service (NHS) England—North East, Cumbria and Yorkshire (Professional Standards and System Improvement) Past President, SCTS	On behalf of SCTS
Karen Redmond (Professor of Thoracic Surgery)	National Thoracic Subcommittee Lead SCTS Fellow Royal College of Surgeons in Ireland	On behalf of the SCTS Thoracic Subcommittee
Bejal Pandya (Consultant Cardiologist)	National Pectus Multidisciplinary Team Chair Member of the British Cardiovascular Society Fellow Royal College of Physicians	On behalf of the NHS England National Pectus Committee
Babu Naidu (Professor of Thoracic Surgery)	Academic & Research Committee Thoracic Lead SCTS Fellow Royal College of Surgeons of England Fellow Royal College of Surgeons of Edinburgh	On behalf of the SCTS Academic & Research Committee (Thoracic)
Lisa MacMahon (Paediatric thoracic surgeon)	Guideline Chair of the Chest Wall International Group (CWIG)	On behalf of CWIG
Frank-Martin Haecker (Professor of Paediatric Surgery)	Treasurer and Executive Committee Chest Wall International Group (CWIG)	On behalf of CWIG
Sarah Murray (Patient Representative)	Society for Cardiothoraic Surgery in Great Britain and Ireland	On behalf of patients
Carl Davis (Consultant Paediatric and Neonatal Surgeon)	Clinical Lead for Scottish National Chest Wall Deformity Service Royal College of Physicians and Surgeons of Glasgow	On behalf of the Scottish National Chest Wall Deformity Service
Shyam Kolvekar (Consultant Cardiothoracic Surgeon)	Lead Surgeon for the NHS England National Pectus Service at St Batholomew's Hospital, London	On behalf of the NHS England National Pectus Committee
Tim Goodacre (Consultant Plastic Surgeon)	Vice President of the Royal College of Surgeons of England President of the British Association of Plastic Reconstructive and Aesthetic Plastic Surgeons	Personal
Deborah Eastwood (Consultant Paediatric Orthopaedic Surgeon)	President of the British Orthopaedic Association	On behalf of the British Orthopaedic Association
Sean Marven (Consultant Paediatric Surgeon)	British Association of Paediatric Surgeons Thoracic and Airway Group and Northern Representative	On behalf of the British Association of Paediataric Surgeons Thoracic and Airway Group
Aman Coonar (Consultant Thoracic Surgeon)	Thoracic Lead at NHS England SCTS	On behalf of NHS England and SCTS

CWIG: Chest Wall International Group; NHS: National Health Service; SCTS: Society for Cardiothoracic Surgery in Great Britain and Ireland.

APPENDIX 2: FINAL SEARCH STRATEGY

1	Funnel chest/or pectus carinatum/
2	(pectus excavatum or funnel chest or pectus carinatum or pigeon thorax or pigeon chest).ti, ab, kw.
3	((chest wall or thoracic wall or pectus or sternum or chondromanubria) adj2 (deform* or malform* or abnormal* or anomal*)).ti, ab, kw.
4	1 or 2 or 3
5	surgical procedures, operative/or minimally invasive surgical procedures/or reconstructive surgical procedures/or thoracic surgical procedures/
6	Reoperation/
7	(minimal* invasive adj3 (repair* or surg* or procedure* or operat* or correct*)).ti, ab, kw.
8	((pectus excavatum or funnel chest) adj5 (repair* or surg* or procedure* or operat* or correct*)).ti, ab, kw.
9	(postoperat* or post-operat* or postsurg* or post-surg* or postrepair* or post-repair* or postprocedure* or post-procedure* or postcorrect* or post-correct*).ti, ab, kw.
10	(repair* or surg* or procedure* or operat* or correct*).ti.
11	((repair* or correct*) adj2 (surg* or procedure* or operat*)).ti, ab, kw.
12	(mirpe or nuss or ravitch).ti, ab, kw.
13	(reoperat* or re-operat* or revision*).ti, ab, kw.
14	5 or 6 or 7 or 8 or 9 or 10 or 11 or 12 or 13
15	4 and 14
16	Funnel Chest/su or Pectus Carinatum/su
17	15 or 16
18	exp animals/not humans/s
19	17 not 18

APPENDIX 3: PICO QUESTIONS FOR THE SEARCH STRATEGY

For [Patients with severe pectus excavatum] is [Nuss or Ravitch surgery] superior to [no treatment] in terms of improving [physiological cardiovascular ability]

For [Patients with severe psychological impairment due to their pectus abnormality] is [surgery] superior to [no treatment or psychological therapy] in terms of improving [psychological well-being]

For [Patients undergoing treatment for pectus abnormalities] should [specialist areas] versus [general ward areas] be used in order to optimize [patient experience].

For [patients undergoing pectus surgery] what are the [optimal safety conditions] to ensure [safe surgery with low morbidity and mortality]

APPENDIX 4: CASE REPORTS OF THE MOST SEVERE PATIENTS

Studies supporting indication for pectus surgery 'the prevention or treatment of symptomatic cardiac or circulatory (acute or chronic) complications following surgery for other conditions'

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
1	Adachi R, Nishihara T, *et al.*, Hemodynamic deterioration due to increased anterior and posterior cardiac compression during posterior spinal fusion for scoliosis with pectus excavatum. SAGE Open Med Case Rep. 2022; 10: 1-7. (Case report)	13-year old male	Prone position causing hypotension during spinal surgery for scoliosis	Diagnosed with Marfan syndrome at 11 years old with PE. T12-L4 posterior spinal fusion for lumbar scoliosis performed 2 years previously. No problems during peri-operative period. Scoliosis of thoracic spine progressed; patient underwent T3-L4 posterior spinal fusion surgery. The patient had hypotension and tachycardia during surgery in the prone position or when the thoracic spine was corrected to the left front.	Pre-scoliosis surgery: Modified Haller index 9 %VC, 52.3 RV inflow pressure gradient, 15 mmHg Spinal penetration index, 10% Thoracic scoliosis was convex to the right. Sternal depression was on the left. Post-scoliosis surgery: Modified Haller index was 13.4 Spinal penetration index, 16% Imaging showed Cobb angle of the thoracic spine was corrected from 60° (T6–L4) to 32° (T7–L1), and the thoracic	No	N/A

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
				Circulation was maintained by adjusting position, administering several boluses of phenylephrine and loading infusion solution.	spine was moved to the left. Thoracic kyphosis changed from 39° to 27° and the thoracic spine was moved forward. The space in the mediastinum decreased, and the cardiac compression due to the corrected thoracic spine and depressed sternum became stronger.		
3	Alexianu D, Skolnick ET, *et al*., Severe hypotension in the prone position in a child with neurofibromatosis, scoliosis and pectus excavatum presenting for posterior spinal fusion. Anesth Analg. 2004; 98(2): 334-5, table of contents. (Case report)	34-Month-old male	Prone position causing hypotension during spinal surgery (neurofibromatosis)	History of failure to thrive, neurofibromatosis, severe progressive scoliosis, PE and impaired pulmonary function. Previous general anaesthetic for gastric feeding tube 3 months before was uneventful. Attempted posterior spinal fusion but developed severe hypotension when positioned prone and abandoned. Subsequent MRI demonstrated increased size of previously identified abdominal masses. Multidisciplinary team agreed-on surgical removal of masses before fusion not feasible because of haemorrhage risk and possibility of recurrence. Continuous TOE during repeat spinal surgery demonstrated RV compression by sternum, not the fibromas. Used longitudinal bolsters rather than transverse bolsters to manage this.	A thoracic MRI 3.5 months before surgery revealed numerous paraspinal neurofibromas without involvement of the great vessels. Repeat MRI after failed operation revealed large paraspinal, posterior mediastinal and retroperitoneal confluent masses that had increased substantially from the previous MRI. The masses were displacing and encircling the great vessels in the upper abdomen and the aorta at the level of the arch. TTE showed a dilated IVC that was displaced to the right. The typical appearance of mild compression of the RV seen with pectus excavatum was also evident.	No	N/A
5	Borrhomée S, Lenoir M, *et al*. Syncope caused by right ventricular obstruction by pectus excavatum. J Thorac Cardiovasc Surg. 2016; 151(4): e67-9. (Case report)	23-year-old female	Syncope due to adhesions developing after previous cardiac surgery.	Recurrent syncope. 11 months old—repair of ventricular septal defect and infundibular stenosis. 16 years old—redo of infundibular stenosis and also noted to have mild asymptomatic PE. Both procedures were performed through a median sternotomy. Three years after second operation, had a syncopal episode. Frequency rapidly increased until episodes occurring weekly.	Initial investigations only demonstrated premature atrial contractions and did not explain her symptoms. TTE and CT scan showed severe compression of right ventricle by the sternum.	Yes	Ravitch procedure. Released dense adhesions between the sternum and the anterior mediastinum. ECHO—major improvement in RV size and tricuspid valve function. At 6 months—no reported symptoms or syncopal episodes.

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
				Episodes usually occurred during physical activity, beginning with dizziness or weakness immediately followed by a sudden loss of consciousness. Usually resolved spontaneously after several seconds.			
6	Crossland D, Auldist A, Davis A. Malignant pectus excavatum. Heart. 2006; 92(10): 1511. (Case report)	7-year-old female	Syncope—patient had previous sternal surgery for Raphe syndrome.	Anterior raphe syndrome At 3 years old—sternal repair of midline defect. Presented with recurrent syncope (6 episodes over 6 months). Most episodes occurred in the morning and were usually associated with showering. An episode occurred while swimming.	Obvious PE but rest of clinical examination normal. ECG, 24 ambulatory ECG and exercise test results were normal. No evidence of long QT syndrome. TTE showed a structurally and functionally normal heart, but the sternum appeared to compress the RV. There was no flow acceleration across the inflow of the RV. Syncope continued despite salt and fluid loading. Chest CT scan showed significant RV compression by the sternal deformity.	Yes	Sternochondroplasty. At 2 years—no further episodes of syncope.
7	Galas J, van der Velde M, *et al*. Echocardiographic diagnosis of right ventricular inflow compression associated with pectus excavatum during spinal fusion in prone position. Congenit Heart Dis. 2009; 4(3): 193-5. (Case report)	15-year-old male	Prone position causing hypotension during spinal surgery	Significant PE and thoracolumbar scoliosis. During elective spinal fusion, the patient immediately developed severe hypotension with systolic blood pressure dropping to 60 mmHg after being placed in the prone position. The hypotension failed to respond to volume and pharmacologic treatment with ephedrine, but promptly improved to 139/67 mmHg after the patient was placed back in the supine position. Scoliosis surgery was postponed while the PE was repaired.	TOE when under anaesthesia and in supine position demonstrated anterior compression of the right heart by the sternum at the level of the right atrial–right ventricular junction, with a mean RV inflow gradient of 3–4 mmHg and a peak gradient of 7 mmHg. Applying external pressure to sternum increased mean inflow gradient to ∼9 mmHg and the peak gradient to ∼16 mmHg. This was reproduced when patient was placed in the prone position on the spine table. Mean inflow gradient increased to 8 mmHg with a peak gradient of 17 mmHg and systolic blood pressure decreased to ∼60 mmHg. There was also associated E:A wave reversal.	Yes	Modified Ravitch procedure with Nuss bar placement. Six weeks later, a follow-up TTE in the supine position revealed no tricuspid inflow gradient, though mild compression of the RV persisted. Ten weeks later, the patient underwent posterior spinal fusion. Prior to this procedure, TOE confirmed no significant tricuspid inflow gradient in the prone position. The patient tolerated the procedure well and was discharged home without further cardiac complications.

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
OEI 9	Jaroszewski D, Eldeib A, *et al*. Cardiac transplantation and consecutive minimally invasive pectus excavatum. Ann Thorac Surg. 2021; 111(1): e11-e14. (2 case reports)	**Case A** 24-year-old female **Case B** 34-year-old female	Acute haemodynamic compromise following a cardiac transplant (both cases).	**Case A** At 10 years old had a cardiac transplant for familial hypertrophic cardiomyopathy. Developed severe PE during adolescence. 14 years post-transplant , developed allograft failure and had a retransplant (bicaval anastomosis). Because her failed graft was not dilated, the anatomic space was extremely limited due to PE deformity. Removal of sternal retractor was not tolerated despite maximal pressor support. The cardiac compression from the PE caused haemodynamic instability with systolic hypotension as low as 30 to 35 mmHg. PE repair undertaken. **Case B** Non-ischaemic cardiomyopathy and PE with recurrent episodes of ventricular tachycardia/ fibrillation and subsequent cardiovascular collapse requiring emergency extracorporeal membrane oxygenation. Transplant with bicaval anastomosis. The sternum was unable to be closed because of haemodynamic instability from pectus compression on the heart graft in addition to bleeding and coagulopathy. Stabilized for 36 hours on ITU and returned to theatre for simultaneous sternal closure, PE repair and removal of breast implants.	**Case A** Prior to retransplant, Haller index was 3.27. **Case B** Prior to transplant, CT showed the failing native heart had shifted and dilated in the left thorax. Haller index, 5.8	Yes—both cases	**Case A** Modified MIRPE using two 11.5-inch bars (Fig. 1B). The chest was subsequently closed without haemodynamic issues after bars were placed to elevate the sternum. At 19-month follow-up, the patient has no pectus-related issues or sternal instability. **Case B** Modified MIRPE with 2 14-inch bars. At 6-month follow-up, the patient has no pectus-related issues or sternal instability.
11	Löhnhardt M, Hättich A, *et al*. Rescue Nuss procedure for inferior vena cava compression syndrome following posterior scoliosis surgery in Marfan syndrome. Eur Spine J. 2019; 28(Suppl 2): 31-36. (Case report)	15-year-old male	Acute IVC compression following scoliosis surgery (in Marfan syndrome with pre-existing stenosis in IVC).	At 8 years old, diagnosed with Marfan syndrome (scoliosis and extreme PE). Following a growth spurt, patient needed posterior spinal fusion for severe back pain and loss of spinal mobility. A dorsal instrumentation from T4–L4 vertebrae was performed. Blood loss, circulatory unstable, required blood, plasma	Preoperative CT scan showed the scoliosis and pectus excavatum with a clinically irrelevant compression of the IVC. Postoperative CT scan—subtotal compression of the IVC in proximity to the RV, between the sternum and spine.	Yes	Nuss procedure using a substernal bar as an internal brace to increase venous flow. Postoperative TTE showed an increased filling of the RV and a regular flow of the IVC. The volume status improved

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
				and platelets. Transferred to ITU following surgery. Complicated with pleural effusions and ascites. Patient developed further symptoms including right pleural effusion, inferior inflow congestion, congested gastritis and a congested liver ascites and swelling of the lower limbs. Consistent with severe cardiovascular depression due to severe vena cava compression syndrome. PE surgery was undertaken.			due to removed compression of the IVC with distinctive inferior inflow congestion. 3 Weeks after the Nuss procedure, the patient was discharged with minimal residual pleural effusions and an oral diuretic therapy.
15	Rouch A, Rabinel P, Accadbled F, Brouchet L. Emergency Ravitch procedure for inferior vena cava compression after surgical scoliosis correction. Ann Thorac Surg. 2020; 110(4): e299-e301. (Case report)	14-year-old female	Acute IVC compression following scoliosis surgery	Patient had painful left lumbar spine idiopathic scoliosis associated with asymptomatic funnel chest. Not Marfan disease. Scoliosis progressed despite conservative management and had posterior spinal instrumentation and fusion T3– L3. Uncomplicated operation but developed abdominal pain and anuria on the first day. Diagnosed acute compression of the IVC with ascitic oedematous decompensation with peritoneal and retroperitoneal fluid collection and acute renal failure.	US scan showed abdominal and perirenal fluid collection suggesting bladder wound. CT scan showed large perirenal, retroperitoneal fluid collection and could not rule out posterior bladder wound. Cystoscopy normal. Injected thoracic CT scan—low volume bilateral pleural effusion associated with compression of IVC between the lower plate of T8 and upper plate of T10 and the distal portion of the sternum, without obvious thrombosis. ECHO—confirmed compression of IVC.	Yes	Ravitch procedure Urine output improved perioperatively. Haemodynamics improved intraoperatively when the sternum was pulled up. Symptoms improved postoperatively. Disappearance of fluid in her abdomen and lungs. At 6 months, the patient was asymptomatic.

CT: computed tomographic; ECG: electrocardiogram; ECHO: echocardiography; ITU: intensive therapy unit; IVC: inferior vena cava; MIRPE: minimally invasive repair of pectus excavatum; MRI: magnetic resonance image; PE: pectus excavatum; RV: right ventricle; TOE: transoesophageal echocardiography; TTE: transthoracic echocardiography; US: ultrasound; VC: vital capacity.

Studies supporting indication for pectus surgery "Symptomatic and/or life threatening arrythmias caused by pectus excavatum"

No.	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
2	Ahn J, Choi J, *et al*. Right ventricular compression mimicking Brugada-like electrocardiogram in a patient with recurrentpectusexcavatum. Case Rep Cardiol. 2017; 2017: 3047937. doi: 10.1155/2017/3047937. Epub 2017 Feb 20. PMID: 28321340 (Case report)	23-year-old male	Arrhythmias—atrial tachycardia (and Brugada-like ECG pattern)	Presented with 10 min of palpitations at rest. Previous PE surgical correction at 3 years of age. No episodes of syncope or family history of sudden cardiac arrest.	Haller index, 5.21. ECG—rSR′ pattern with slight ST-segment elevation and T-wave inversion on the right precordial leads (Brugada-like ECG pattern). Laboratory findings were within normal range. Both signal-averaged ECG and flecainide challenge test showed no significant findings. Treadmill testing—no tachyarrhythmia was noted except an infrequent single ventricular premature beat at stage 4. 24-hour ambulatory Holter monitoring—several episodes of sustained atrial tachycardia with a maximum duration of 5 min. ECHO and cardiac CT demonstrated external compression on the basal-to-mid portion of the RV and dilation of the right atrium and RV apical site with mild hypokinesia.	Yes	Nuss procedure CXR—effective prevention of sternal compression. ECHO—RV was decompressed and contractility was normal. At 6 months, no symptoms and no tachyarrhythmias documented. ST segment morphology appeared to be normalized on lead V2-3.
4	Awad S, Barbosa-Barros R, *et al*. Brugada phenocopy in a patient with pectus excavatum: systematic review of the ECG manifestations associated with pectus excavatum. Ann Noninvasive Electrocardiol. 2013; 18(5): 415-20. (Case report)	24-year-old male	Abnormal ECG—Brugada phenocopy	Presented with chest pain. Otherwise well and not on medication. No history of syncope and denied any family history of sudden cardiac death.	PE but normal results from cardiovascular examination. CXR—narrowing of the anteroposteriordistance between the sternum and the spine with relative compression of the heart. ECHO—no structural or wall-motion abnormalities. Initial standard 12-lead ECG with the precordial electrodes placed in the 4th ICS showed sinus rhythm, heart rate 65 bpm, P-wave duration 80 ms, PR interval 160 ms, QRS duration 80 ms and normal axis. A "saddle-back" ST-segment elevation in lead V2 was consistent with a type 2 Brugada pattern. 12-lead ECG with high-precordial lead placement in 2nd ICS showed sinus rhythm, heart rate 75 bpm, P-wave duration	No	N/A

Continued

Continued

Studies supporting indication for pectus surgery "Symptomatic and/or life threatening arrythmias caused by pectus excavatum"

No.	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
					80 ms, PR interval 120 ms, QRS duration 100 ms, and normal axis. The ECG was consistent with an iRBBB pattern with rsR' complex in leads V1–V2 and a "saddle-back" ST-segment elevation in lead V3. When the ECG was obtained in the 5th ICS, away from the RV outflow tract, it showed disappearance of both the iRBBB and the "saddle-back" ST-segment elevation. Pharmacological challenge test with ajmaline (class Ia antiarrhythmic with potent sodium channel blocking effects) did not induce a type 1 Brugada pattern.		
12	Moossdorff M, Maesen B, *et al*. Case report: ventricular fibrillation and cardiac arrest provoked by forward bending in adolescent with severe pectus excavatum. Eur Heart J Case Rep. 2021; 5 (10): ytab373. (Case report)	19-year-old male	Arrhythmias—syncope, VF and cardiac arrest	Patient with severe PE scheduled for modified Ravitch repair. Previous right-sided video- assisted thoracoscopic surgery bullectomy and pleurectomy for spontaneous pneumothorax. For epidural catheter placement, the patient was sitting and bending forwards. Became hypotensive and light headed. Blood pressure increased after ephedrine but symptoms were progressive and he collapsed. Went in to VF and cardiac arrest. Resuscitated and quickly recovered without any neurological sequelae. Additional history regarding postural symptoms and syncope was taken. The patient reported experiencing light headedness and dyspnoea when bending over, for example when tying shoelaces. He always considered this unremarkable.	Haller index, 4.72 Preoperative ECG showed a sinus rhythm 76 bpm, with subtle RBBB with a small r' in the anterior chest leads. After VF arrest: • ECG directly after return of spontaneous circulation showed sinus tachycardia with broadening of the QRS-complex and ST-elevations most pronounced in the inferior leads that resolved within min • Cardiac US in acute setting showed global hypokinesia without local wall motion abnormalities and no pericardial effusion or other abnormalities. • Electrolytes in arterial blood gas normal. • Coronary angiography showed normal coronaries. • No signs of anaphylaxis. Cine cardiac MRI and chest CT—severe PE and extensive compression of the heart (in particular of the left atrium, and to a lesser degree of the right	Yes	Modified Ravitch procedure. He was well 18 months after surgery. His postural symptoms had decreased.

Continued

Continued

Studies supporting indication for pectus surgery "Symptomatic and/or life threatening arrythmias caused by pectus excavatum"

No.	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
				After excluding other causes, thought to be due to cardiac compression secondary to severe PE. ICD not inserted and listed for pectus surgery.	atrium and ventricle) between the depressed sternum and the thoracic spine. Cardiac MRI further showed normal right and left ventricle contractility and a normal right outflow tract. No signs of arrhythmogenic right ventricular cardiomyopathy and no delayed enhancement. Ajmaline challenge was negative for Brugada syndrome.		
13	Pimenta J, Vieira A, Henriques-Coelho T. Ventricular arrhythmia solved by surgical correction of pectus excavatum. Interact Cardiovasc Thorac Surg. 2018; 26(4): 706-708. (Case report)	14-year-old male	Arrhythmias—single, paired and tripled polymorphic premature ventricular contractions arising from lower part of RV.	Previously healthy. Presented with frequent palpitations lasting 15 min. No other CVS symptoms. Medical history unremarkable.	Severe PE on physical examination. Haller index, 4.4 ECG—mild right axis deviation. TTE—compression of RV with normal tricuspid flow and normal systolic and diastolic function. 24-hour ambulatory ECG—single, paired and tripled polymorphic premature ventricular contractions arising from lower part of RV. Exercise stress test—a triplet of polymorphic premature ventricular contractions was observed with the same origin described in ambulatory ECG monitoring. Chest CT compression of right ventricle.	Yes	Nuss repair. After 2 years patient was asymptomatic with no palpitations. 24 hour ambulatory ECG showed very few ventricular contractions without pairs or triplets. Exercise stress test results completely normal. Chest CT resolution of compression of RV.
14	Rashwan R, Purpura A, Kah wash B. Sudden cardiac arrest in a young patient with severe pectus excavatum. Am J Med Sci. 2018; 356(6): 570-573. (Case report)	19-year-old male	Arrhythmias—VF and SCA	Previously healthy and had SCA. Collapsed at home—VF and shocked out of it.	Physical examination, only PE. Haller index, 22.7 ECG in emergency department—entirely negative P wave in lead V1, inverted T waves in leads V1–V2 and incomplete RBBB. Normal corrected QT interval. Normal complete blood count and electrolytes. Troponin 1.4 ng/ml. Negative urine drug screen. CXR—steep angulation of anterior ribs with loss of right heart border on postero-anterior view and posterior	Yes	Ravitch procedure. ICD interrogation at 1 and 2 months did not record any arrhythmogenic events.

Continued

Continued

Studies supporting indication for pectus surgery "Symptomatic and/or life threatening arrythmias caused by pectus excavatum"

No.	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
					displacement of sternum on lateral view. CT chest—compression of heart by severe sternal deformity resulting in ballooning of RV apex and leftward shift. Cardiac MRI—CT chest findings confirmed with no myocardial scarring or no evidence of arrhythmogenic RV dysplasia. CT angiography—normal. TTE—normal left and right ventricular size and contractility. Procainamide provocation test ruled out Brugada syndrome. Electrophysiology study—inducible VF with triple ventricular extra stimuli at RV apex. Non-specific and no target site could be isolated for catheter ablation. Genetic testing results negative. Scheduled for operative correction of PE and wore defibrillator vest while awaiting ICD.		

CT: computed tomography; CVS: cardiovascular; CXR: chest X-ray; ECG: electrocardiogram; ECHO: echocardiography; ICD: implantable cardioverter defibrillator; ICS: intercostal space; iRBBB: incomplete right bundle branch block; MRI: magnetic resonance imaging; PE: pectus excavatum; RBBB: right bundle branch block; RV: right ventricle; SCA: sudden cardiac arrest; TTE: transthoracic echocardiography; US: ultrasound; VF: ventricular fibrillation.

No	Studies supporting indication for pectus surgery "Symptomatic cardiac or circulatory obstruction caused by pectus excavatum"						
	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
8	Iannucci G, Slesnick T, *et al*. Lower extremity oedema in a child due to pectus excavatum. Ann Thorac Surg. 2015; 99: e29-e30 (Case report)	11-year-old female	Chronic peripheral oedema of lower limbs due to impaired venous return (IVC compression).	2-Year progressive bilateral lower extremity swelling. Previously well. No family history of thrombophilia. Moderate PE in mother.	Moderate bilateral pitting oedema and severe PE. No hepatomegaly or ascites. No other findings to suggest an underlying connective tissue disorder. MRI of limbs—soft tissue swelling. Tests for proteinuria, renal dysfunction, hepatic dysfunction, rheumatic disease, angioedema, hypothyroidism, and deep venous thrombosis were negative. ECHO—mild compression of the IVC and increased flow velocity within. Cardiac MRI—severe reduction in the anterior–posterior diameter of the chest cavity at the level of the IVC–right atrial (RA) junction with resultant compression of the IVC. There was turbulent flow in the IVC at the RA junction, with a peak velocity of 1.2 m/s, predicting a pressure gradient of 5 mmHg while she was supine. The IVC narrowed to $\sim 6 \times 12$ mm.	Yes	Modified Ravitch procedure. At 3 weeks she had complete resolution of oedema and leg pain. ECHO at 6 months demonstrated unobstructed flow in the IVC. At 1 year, no symptoms or oedema.
16	Siniorakis E, Arvanitakis S, *et al*. Pectus excavatum: Right ventricular compromise with orthostatic syndrome and Brugada phenocopy. J Saudi Heart Assoc. 2017 Jul; 29 (3):223-226. (Case report)	23 -year-old male	Orthostatic syndrome	Presented for preoperative check before PE surgery for predominantly cosmetic reasons. He also reported exertional dyspnoea, atypical chest discomfort, reduced endurance, and frequent orthostatic hypotension. Past medical history clear. Family history clear and no record of syncope or sudden cardiac death.	In supine position, clinical examination unremarkable except for PE. Symptomatic orthostatic syndrome noted on standing—blood pressure dropped 50 mmHg, heart rate increased 30%; patient had dizziness and near fainting. Haller index, 3.8 CXR—enlarged cardiac silhouette with left displacement of heart. Electrolytes and arterial blood gases normal. ECG—consistent with Brugada phenocopy. Sinus rhythm with right axis deviation and leads V1–V2 depicted an rSR pattern with down-sloping ST segment and negative T waves. Lead V3 had	Yes	Ravitch procedure At 6 months asymptomatic and no orthostatic syndrome. ECG—findings in leads V3 and V4 were attenuated although negative P waves in leads V1–V2 persisted. RV decompressed with normal tricuspid annular plane systolic excursion. MRI—RV ejection fraction increased to 56% Haller index 2.0

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
	Studies supporting indication for pectus surgery "Symptomatic cardiac or circulatory obstruction caused by pectus excavatum"						
					bifid T waves with a prominent late component. P waves in V1 were negative. Procainamide test negative. TTE showed compression of right atrium and ventricle, with reduced tricuspid annular plane systolic excursion. Short axis M-mode imaging revealed a sudden pulling of the interventricular septum towards the anterior chest wall at the onset of diastole, causing almost total obliteration of RV cavity. Evident in inspiration and expiration resulting in constrictive pathophysiology. Potential intracavitary pressure gradients were not recorded due to positional deformities of the RV. Imaging and function of the great vessels, left heart cavities and valves were normal. MRI—squeezing of RV outflow tract with reduced RV ejection fraction 48%.		
17	White J, Fine N, Shargall Y. Images in cardiovascular medicine. Pectus excavatum with compression of the inferior vena cava: a rare cause of recurrent syncope. Circulation. 2009; 120(17): 1722-4. (Case report)	22-year-old female	Intermittent IVC obstruction with syncope	Long history of recurrent syncope starting in early adolescence. Episodes occurred after prolonged periods of standing or walking. Started with prodrome of diaphoresis and blurred vision culminating in syncope. Patient found she could manage symptoms by twisting her torso or performing rapid, shallow breathing. However, still had frequent syncope and stopped school at 16 years. Brother and paternal uncle had PE.	On examination, marked PE deformity with straight back. Haller index, 18.8 (chest CT) Heart sounds normal. Grade 2 systolic ejection murmur at left sternal border. No jugular venous distension or peripheral oedema. ECG—incomplete right bundle branch block. TTE—poor quality due to PE. Pulmonary function tests—restrictive pattern—vital capacity 2.5 l (66% predicted) and total lung capacity 3.32 l (65% predicted) Exercise test—unable to complete due to rapid onset of light-headedness.	Yes	Nuss procedure. Chest CT predischarge showed no IVC compression during maximal expiration and marked improvement in the minimal expiratory thoracic depth from 1.2 cm to 5.7 cm. At 4 months postoperatively, patient reported no syncopal episodes and was able to stand and walk for extended periods. She returned to school and reported a

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
	Studies supporting indication for pectus surgery "Symptomatic cardiac or circulatory obstruction caused by pectus excavatum"						
					Tilt table test (bed 80 degrees) - after 2 min, the patient insisted on twisting her torso dramatically to the side and in doing so maintained consciousness for 15 min. When instructed to straighten her posture, she lost consciousness at 2 min and 30 seconds, with a documented drop in both blood pressure and pulse wave amplitude. Cardiovascular MRI—marked expiratory compression of IVC with near obliteration of the intrathoracic space between the sternum and thoracic vertebrae. The cross-sectional area and anterior-posterior diameter of the IVC decreased from 6 cm^2 and 21 mm during inspiration to 1.2 cm^2 and 3 mm during expiration, respectively. Significantly increased mean IVC flow velocity at end expiration with marked blunting of the normally biphasic flow pattern, suggesting obstruction to caval flow.		significant improvement in exercise tolerance and overall activity level.
18	Yalamanchili K, Summer W, Valentine V. Pectus excavatum with inspiratory inferior vena cava compression: a new presentation of pulsus paradoxus. Am J Med Sci. 2005; 329(1): 45-7. (Case report)	29-year-old male	Pulsus paradoxus caused by intermittent IVC obstruction on inspiration	Dyspnoea on exertion for 1 year. PE that increased in depth during adolescence but no limitations when playing a variety of sports. Recently described immediate tachycardia and progressive dyspnoea after initiating aerobic activities. Particularly noted after climbing several flights of stairs that previously posed no problems. No chest pain, dizziness, syncope, generalized weakness or peripheral oedema.	Healthy appearing with marked PE with symmetrical chest expansion. No kyphosis or scoliosis. Blood pressure, 110/70 mmHg Heart rate, 82 bpm Respiratory rate, 12 bpm Heart rhythm normal with expiratory split S2 that narrowed with inspiration. No murmurs or jugular venous distension. Pulses were equal in all extremities, but pulse pressure was palpably reduced with normal respiration and absent with deep inspiration. 16 mmHg reduction of systolic blood pressure with inspiration. ECG normal.	No	N/A

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
	Studies supporting indication for pectus surgery "Symptomatic cardiac or circulatory obstruction caused by pectus excavatum"						
					Pulmonary function test results normal. Haller index, 7.18 Chest CT—heart shifted to left. Sternum in close approximation to IVC, superior vena cava and right heart. ECHO—normal cardiac anatomy. During normal breathing while supine, tricuspid valve maximum flow velocity decreased from 0.96 m/s with expiration to 0.69 m/s with inspiration. Left ventricle stroke volume decreased from 0.152 l with expiration to 0.123 l during inspiration. When in the upright position, there was a similar decrease in tricuspid valve flow and stroke volume with inspiration. Cardiac output remained preserved at ~7–8 l/min at all times due to a great increase in heart rate. Left and right heart Doppler studies suggested a venous return limitation during inspiration. Treadmill exercise ECHO showed early tachycardia (180 beats/min) and associated significant dyspnoea. Patient achieved 12.9 MTS and appropriate hypercontractile ventricular response. Cardiopulmonary exercise test—VO_2 max 2.24 l/min (ref 3.26–4.22 l/min) and VO_2 at anaerobic threshold of 1.08/l was 26% to 33% of VO_2 max reference (normal range 50% to 60% of VO_2 max reference). MRI—leftward deviation of heart associated with corresponding deviation of the normally straight IVC into the left chest. The IVC appears constricted at the level of the		

Continued

Continued

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
	Studies supporting indication for pectus surgery "Symptomatic cardiac or circulatory obstruction caused by pectus excavatum"						
					diaphragm during inspiration, with corresponding dilation of the hepatic venous system.		

CT: computed tomography; CXR: chest X-ray; ECG: electrocardiogram; ECHO: echocardiogram; IVC: inferior vena cava; MRI: magnetic resonance imaging; MPS: myocardial perfusion scintigraphy; PE: pectus excavatum; RV: right ventricle; TTE: transthoracic echocardiography

Studies supporting indication for pectus surgery "Oesophageal obstruction caused by pectus excavatum"

No	Reference	Population	Clinical issues	Case summary	Examination and investigations	Pectus surgery performed	Outcomes following pectus surgery
10	Kim S, Jeong J. Pectus excavatum: A rare cause of dysphagia. J Thorac Cardiovasc Surg. 2017; 153(1):217. (Case report—Letter)	23-year-old male	Dysphagia	Presented with chest discomfort and a feeling of fullness after eating.	Chest appeared extremely sunken. Haller index, 37.5 Chest computed tomography scans showed severe depression of the anterior chest wall with indentation on the heart and liver and inadequate space around the oesophagus.	Yes	Modified Nuss procedure. Symptoms resolved. Haller index, 2.76.

REFERENCES

[1] Biavati M, Kozlitina J, Alder AC, Foglia R, McColl RW, Peshock RM *et al.* Prevalence of pectus excavatum in an adult population-based cohort estimated from radiographic indices of chest wall shape. PLoS One 2020;15:e0232575.

[2] NHS England Evidence Review. Pectus surgery for pectus deformities resulting in severe psychological symptoms (with no physiological impairment) NHS England URN: 2102b, Drafted: June 2021 Prepared by: Solutions for Public Health (SPH) on behalf of NHS England Specialised Commissioning. https://www.england.nhs.uk/publication/surgery-for-pectus-deformity-all-ages/

[3] Jaroszewski DE, Farina JM, Gotway MB, Stearns JD, Peterson MA, Pulivarthi VSKK *et al.* Cardiopulmonary outcomes after the Nuss procedure in pectus excavatum. J Am Heart Assoc 2022; 11:e022149.

[4] Lawson ML, Cash TF, Akers R, Vasser E, Burke B, Tabangin M *et al.* A pilot study of the impact of surgical repair on disease-specific quality of life among patients with pectus excavatum. J Pediatr Surg 2003; 38:916–8.

[5] Rachwan R, Purpura A, Kahwash B. Sudden cardiac arrest in a young patient with severe pectus excavatum. Am J Med Sci 2018; 356:570–3.

[6] Sousa-Uva M, Head SJ, Thielmann M, Cardillo G, Benedetto U, Czerny M *et al.* Methodology manual for European Association for Cardio-Thoracic Surgery (EACTS) clinical guidelines. Eur J Cardiothorac Surg 2015;48:809–16.

[7] NHS England Evidence Review: Pectus surgery for pectus deformities causing severe physiological impairment with or without psychological symptoms NHS England URN: 2102a. Drafted: June 2021 Prepared by: Solutions for Public Health (SPH) on behalf of NHS England Specialised Commissioning. https://www.england.nhs.uk/publication/surgery-for-pectus-deformity-all-ages/

[8] Kanagaratnam A, Phan S, Tchantchaleishvili V, Phan K. Ravitch versus Nuss procedure for pectus excavatum: systematic review and meta-analysis. Ann Cardiothorac Surg 2016;5:409–21.

[9] Rodríguez-Granillo GA, Martínez-Ferro M, Capuñay C, Bellia-Munzón G, Deviggiano A, Raggio I *et al.* Preoperative multimodality imaging of pectus excavatum: state of the art review and call for standardization. Eur J Radiol 2019;117:140–8.

[10] Albertal M, Vallejos J, Bellia G, Millan C, Rabinovich F, Buela E *et al.* Changes in chest compression indexes with breathing underestimate surgical candidacy in patients with pectus excavatum: a computed tomography pilot study. J Pediatr Surg 2013;48:2011–6.

[11] Malek MH, Berger DE, Marelich WD, Coburn JW, Beck TW, Housh TJ. Pulmonary function following surgical repair of pectus excavatum: a meta-analysis. Eur J Cardiothorac Surg 2006;30:637–43.

[12] Rodriguez-Granillo GA, Raggio IM, Deviggiano A, Bellia-Munzon G, Capunay C, Nazar M *et al.* Impact of pectus excavatum on cardiac morphology and function according to the site of maximum compression: effect of physical exertion and respiratory cycle. Eur Heart J Cardiovasc Imaging 2020;21:77–84.

[13] Topper A, Polleichtner S, Zagrosek A, Prothmann M, Traber J, Schwenke C *et al.* Impact of surgical correction of pectus excavatum on cardiac function: insights on the right ventricle. A cardiovascular magnetic resonance study. Interact CardioVasc Thorac Surg 2016;22:38–46.

[14] Monti L, Montini O, Voulaz E, Maagaard M, Morenghi E, Pilegaard HK *et al.* Cardiovascular magnetic resonance assessment of biventricular changes during vacuum bell correction of pectus excavatum. J Thorac Dis 2019;11:5398–406.

[15] Bawazir OA, Montgomery M, Harder J, Sigalet DL. Midterm evaluation of cardiopulmonary effects of closed repair for pectus excavatum. J Pediatr Surg 2005;40:863–7.

[16] Sigalet DL, Montgomery M, Harder J, Wong V, Kravarusic D, Alassiri A. Long term cardiopulmonary effects of closed repair of pectus excavatum. Pediatr Surg Int 2007;23:493–7.
[17] Neviere R, Montaigne D, Benhamed L, Catto M, Edme JL, Matran R *et al.* Cardiopulmonary response following surgical repair of pectus excavatum in adult patients. Eur J Cardiothorac Surg 2011;40:e77–82.
[18] O'Keefe J, Byrne R, Montgomery M, Harder J, Roberts D, Sigalet DL. Longer term effects of closed repair of pectus excavatum on cardiopulmonary status. J Pediatr Surg 2013;48:1049–54.
[19] Adachi R, Nishihara T, Morino T, Sekiya K, Kitamura S, Konishi A *et al.* Posterior cardiac compression during posterior spinal fusion for scoliosis with pectus excavatum. SAGE Open Med Case Rep 2022;10:1–7.
[20] Ahn J, Choi JI, Shim J, Lee SH, Kim YH. Right ventricular compression mimicking Brugada-like electrocardiogram in a patient with recurrent pectus excavatum. Case Rep Cardiol 2017;2017:3047937.
[21] Alexianu D, Skolnick ET, Pinto AC, Ohkawa S, Roye DP Jr, Solowiejczyk DE *et al.* Severe hypotension in the prone position in a child with neurofibromatosis, scoliosis and pectus excavatum presenting for posterior spinal fusion. Anesth Analg 2004;98:334–5.
[22] Awad SFM, Barbosa-Barros R, Belem LdS, Cavalcante CP, Riera ARP, Garcia-Niebla J *et al.* Brugada phenocopy in a patient with pectus excavatum: systematic review of the ECG manifestations associated with pectus excavatum. Ann Noninvasive Electrocardiol 2013;18:415–20.
[23] Borrhomée S, Lenoir M, Gouton M, Fadel E. Syncope caused by right ventricular obstruction by pectus excavatum. J Thorac Cardiovasc Surg 2016;151:e67–9.
[24] Crossland D, Auldist A, Davis A. Malignant pectus excavatum. Heart 2006;92:1511.
[25] Moossdorff M, Maesen B, den Uijl DW, Lenderink T, Franssen FAR, Vissers YLJ *et al.* Case report: ventricular fibrillation and cardiac arrest provoked by forward bending in adolescent with severe pectus excavatum. Eur Heart J Case Rep 2021;5:ytab373.
[26] Iannucci GJ, Slesnick TC, Kogon B, Samai C. Lower extremity edema in a child due to pectus excavatum. Ann Thorac Surg 2015;99:e29–30.
[27] Jaroszewski D, Eldeib A, Gotimukul A, Lackey J, Gaitan B, DeValeria P *et al.* Cardiac transplantation and consecutive minimally invasive pectus excavatum. Ann Thorac Surg 2021;111:e11–4.
[28] Kim S, Jeong J. Pectus excavatum: a rare cause of dysphagia. J Thorac Cardiovasc Surg 2017;153:217.
[29] Löhnhardt M, Hättich A, Andresen A, Stangenberg M, Mir TS, Reinshagen K *et al.* Rescue Nuss procedure for inferior vena cava compression syndrome following posterior scoliosis surgery in Marfan syndrome. Eur Spine J 2019;28:31–6.
[30] Galas JM, van der Velde ME, Chiravuri SD, Farley F, Parra D, Ensing GJ et al Echocardiographic diagnosis of right ventricular inflow compression associated with pectus excavatum during spinal fusion in prone position. Congenit Heart Dis 2009;4:193–5.
[31] Pimenta J, Vieira A, Henriques-Coelho T. Ventricular arrhythmia solved by surgical correction of pectus excavatum. Interact CardioVasc Thorac Surg 2018;26:706–8.
[32] National Institute For Health and Clinical Excellence Interventional Procedures Programme. Interventional procedure overview of placement of pectus bar for pectus excavatum (also known as MIRPE or the Nuss procedure), 2009, IP 135/2. https://www.nice.org.uk/Guidance/IPG310
[33] Rouch A, Rabinel P, Accadbled F, Brouchet L. Emergency Ravitch procedure for inferior vena cava compression after surgical scoliosis correction. Ann Thorac Surg 2020;110:e299–301.
[34] Siniorakis E, Arvanitakis S, Tzevelekos P, Panta S, Balanis A, Aivalioti F *et al.* Pectus excavatum: right ventricular compromise with orthostatic syndrome and Brugada phenocopy. J Saudi Heart Assoc 2017;29:223–6.
[35] White J, Fine N, Shargall Y. Images in cardiovascular medicine. Pectus excavatum with compression of the inferior vena cava: a rare cause of recurrent syncope. Circulation 2009;120:1722–4.
[36] Yalamanchili K, Summer W, Valentine V. Pectus excavatum with inspiratory inferior vena cava compression: a new presentation of pulsus paradoxus. Am J Med Sci 2005;329:45–7.
[37] Johnson WR, Fedor D, Singhal S. Systematic review of surgical treatment techniques for adult and pediatric patients with pectus excavatum. J Cardiothorac Surg 2014;9:25.
[38] Kelly RE Jr, Obermeyer RJ, Goretsky MJ, Kuhn MA, Frantz FW, McGuire MM *et al.* Recent modifications of the Nuss procedure: the pursuit of safety during the minimally invasive repair of pectus excavatum. Ann Surg 2022;275:e496–e502.
[39] Kuru P, Bostanci K, Ermerak NO, Bahadir AT, Afacan C, Yuksel M. Quality of life improves after minimally invasive repair of pectus excavatum. Asian Cardiovasc Thorac Ann 2015;23:302–7.
[40] Luo L, Xu B, Wang X, Tan B, Zhao J. Intervention of the Nuss procedure on the mental health of pectus excavatum patients. Ann Thorac Cardiovasc Surg 2017;23:175–80.
[41] Pawlak K, Gąsiorowski Ł, Gabryel P, Dyszkiewicz W. Video-assisted-thoracoscopic surgery in left-to-right Nuss procedure for pectus excavatum for prevention of serious complications—technical aspects based on 1006 patients. Wideochir Inne Tech Maloinwazyjne 2018;13:95–101.
[42] Pawlak K, Gąsiorowski Ł, Gabryel P, Gałęcki B, Zieliński P, Dyszkiewicz W. Early and late results of the Nuss procedure in surgical treatment of pectus excavatum in different age groups. Ann Thorac Surg 2016; 102:1711–6.
[43] Derogatis LR. SCL-90-R: Administration, Scoring and Procedures. Manual II for the (Revised) Version and Other Instruments of the Psychopathology Rating Scale Series. Clinical Psychometric Research, 1983.
[44] Steinmann C, Krille S, Mueller A, Weber P, Reingruber B, Martin A. Pectus excavatum and pectus carinatum patients suffer from lower quality of life and impaired body image: a control group comparison of psychological characteristics prior to surgical correction. Eur J Cardiothorac Surg 2011;40:1138–45.
[45] Nuss D. Recent experiences with minimally invasive pectus excavatum repair "Nuss procedure". Jpn J Thorac Cardiovasc Surg 2005;53:338–44.
[46] Castellani C, Schalamon J, Saxena AK, Höellwarth ME. Early complications of the Nuss procedure for pectus excavatum: a prospective study. Pediatr Surg Int 2008;24:659–66.
[47] Hosie S, Sitkiewicz T, Petersen C, Göbel P, Schaarschmidt K, Till H *et al.* Minimally invasive repair of pectus excavatum—the Nuss procedure. A European multicentre experience. Eur J Pediatr Surg 2002;12:235–8.
[48] Park HJ, Lee SY, Lee CS, Youm W, Lee KR. The Nuss procedure for pectus excavatum: evolution of techniques and early results on 322 patients. Ann Thorac Surg 2004;77:289–95.
[49] Bouchard S, Hong AR, Gilchrist BF, Kuenzler KA. Catastrophic cardiac injuries encountered during the minimally invasive repair of pectus excavatum. Semin Pediatr Surg 2009;18:66–72.
[50] Nuss D, Kelly RE Jr. Minimally invasive surgical correction of chest wall deformities in children (Nuss procedure). Adv Pediatr 2008; 55:395–410.
[51] Lam MWC, Klassen AF, Montgomery CJ, LeBlanc JG, Skarsgard ED. Quality-of-life outcomes after surgical correction of pectus excavatum: a comparison of the Ravitch and Nuss procedures. J Pediatr Surg 2008; 43:819–25.
[52] Metzelder ML, Kuebler JF, Leonhardt J, Ure BM, Petersen C. Self and parental assessment after minimally invasive repair of pectus excavatum: lasting satisfaction after bar removal.[see comment]. Ann Thorac Surg 2007;83:1844–9.
[53] Rushing GD, Goretsky MJ, Gustin T, Morales M, Kelly RE, Nuss D *et al.* When it is not an infection: metal allergy after the Nuss procedure for repair of pectus excavatum. J Pediatr Surg 2007;42:93–7.
[54] Shin S, Goretsky MJ, Kelly RE, Gustin T, Nuss D Jr. Infectious complications after the Nuss repair in a series of 863 patients. J Pediatr Surg 2007;42:87–92.
[55] Dzielicki J, Korlacki W, Janicka I, Dzielicka E. Difficulties and limitations in minimally invasive repair of pectus excavatum—6 years experiences with Nuss technique. Eur J Cardiothorac Surg 2006;30:801–4.
[56] Kelly RE, Cash TF, Shamberger RC, Mitchell KK, Mellins RB, Lawson ML *et al.* Surgical repair of pectus excavatum markedly improves body image and perceived ability for physical activity: multicenter study. Pediatrics 2008;122:1218–22.
[57] Lomholt JJ, Jacobsen EB, Thastum M, Pilegaard H. A prospective study on quality of life in youths after pectus excavatum correction. Ann Cardiothorac Surg 2016;5:456–65.
[58] Krasopoulos G, Dusmet M, Ladas G, Goldstraw P. Nuss procedure improves the quality of life in young male adults with pectus excavatum deformity. Eur J Cardiothorac Surg 2006;29:1–5.
[59] Sacco Casamassima MG, Gause C, Goldstein SD, Karim O, Swarup A, McIltrot K *et al.* Patient satisfaction after minimally invasive repair of pectus excavatum in adults: long-term results of Nuss procedure in adults. Ann Thorac Surg 2016;101:1338–45.
[60] Hanna WC, Ko MA, Blitz M, Shargall Y, Compeau CG. Thoracoscopic Nuss procedure for young adults with pectus excavatum: excellent midterm results and patient satisfaction. Ann Thorac Surg 2013;96:1033–6.

[61] Hoksch B, Kocher G, Vollmar P, Praz F, Schmid RA. Nuss procedure for pectus excavatum in adults: long-term results in a prospective observational study. Eur J Cardiothorac Surg 2016;50:934–9.
[62] Patel AJ, Hunt I. Is vacuum bell therapy effective in the correction of pectus excavatum? Interact Cardiovasc Thorac Surg 2019;29:287–90.
[63] Hebra A, Kelly RE, Ferro MM, Yüksel M, Campos JRM, Nuss D. Life-threatening complications and mortality of minimally invasive pectus surgery. J Pediatr Surg 2018;53:728–32.
[64] European Society of Cardiology. Governing policies and procedures for the writing of ESC Clinical Practice guidelines. 2017. https://www.escardio.org/static-file/Escardio/Guidelines/About/Recommendations-Guidelines-Production.pdf (22 May 2021, date last accessed).
[65] Jaroszewski DE, Notrica DM, McMahon LE, Hakim FA, Lackey JJ, Gruden JF *et al.* Operative management of acquired thoracic dystrophy in adults after open pectus excavatum repair. Ann Thorac Surg 2014 ;97:1764–70.
[66] Núñez García B, Álvarez García N, Aquino-Esperanza J, Esteva Miró C, Pérez-Gaspar M, Jiménez Gómez J *et al.* Efficacy and safety of taulinoplasty compared with the minimally invasive repair of pectus excavatum approach to correct pectus excavatum. J Laparoendosc Adv Surg Tech A 2021;31:1402–7. Dec
[67] Haecker FM, Krebs T, Kocher G, Schmid R, Sesia S. Sternal elevation techniques during the minimally invasive repair of pectus excavatum. Interact CardioVasc Thorac Surg 2019;29:497–502.
[68] Kim H, Rim G, Park H. Technical advances in pectus bar stabilisation in chest wall deformity surgery: 10-year trends and an appraisal with 1,500 patients. J Chest Surg 2023;56:229–37.
[69] Lau WC, Shannon FL, Bolling SF, Romano MA, Marc P, Sakwa MP *et al.* Intercostal cryo nerve block in minimally invasive cardiac surgery: the prospective randomized FROST trial. Pain Ther 2021;10:1579–92.
[70] de Beer S, Volcklandt S, de Jong J, Oomen M, Zwaveling S, van Heurn E. Dynamic compression therapy for pectus carinatum in children and adolescents: factors for success. J Pediatr Surg 2022;58:1440–5.
[71] de Beer SA, Blom YE, Lopez M, de Jong JR. Measured dynamic compression for pectus carinatum: a systematic review. Semin Pediatr Surg 2018;27:175–82.
[72] Hunt I, Patel AJ. Effectiveness of compressive external bracing in patients with flexible pectus carinatum deformity: a review. Thorac Cardiovasc Surg 2020;68:72–9.
[73] Viggiano D, Bongiolatti S, Borgianni S, Lo Piccolo R, Voltolini L, Gonfiotti A. Nuss technique for pectus excavatum in adult patients: cosmetic satisfaction and improvement of quality of life in a single-center experience. Front Surg 2022;9:903791.
[74] Obermeyer RJ, Cohen NS, Kelly RE Jr, Ann Kuhn M, Frantz FW, McGuire MM *et al.* Nonoperative management of pectus excavatum with vacuum bell therapy: a single center study. J Pediatr Surg 2018;53:1221–5.
[75] Toselli L, Chinni E, Nazar-Peirano M, Vallee M, Sanjurjo D, Martinez J et al Determinants of success associated with vacuum bell treatment of pectus excavatum. J Pediatr Surg 2022;57:550–4.
[76] Behr CA, Denning NL, Kallis MP, Maloney C, Soffer SZ, Romano-Adesman A *et al.* The incidence of Marfan syndrome and cardiac anomalies in patients presenting with pectus deformities. J Pediatr Surg 2019;54:1926–8.
[77] Del Frari B, Blank C, Sigl S, Schwabegger AH, Gassner E, Morawetz D *et al.* The questionable benefit of pectus excavatum repair on cardiopulmonary function: a prospective study. Eur J Cardiothorac Surg 2021; 61:75–82.
[78] Castellani C, Windhaber J, Schober PH, Hoellwarth ME. Exercise performance testing in patients with pectus excavatum before and after Nuss procedure. Pediatr Surg Int 2010;26:659–63.
[79] Das BB, Recto MR, Yeh T. Improvement of cardiopulmonary function after minimally invasive surgical repair of pectus excavatum (Nuss procedure) in children. Ann Pediatr Cardiol 2019;12:77–82.
[80] Udholm S, Maagaard M, Pilegaard H, Hjortdal V. Cardiac function in adults following minimally invasive repair of pectus excavatum. Interact CardioVasc Thorac Surg 2016;22:525–9.
[81] Haller JA Jr, Loughlin GM. Cardiorespiratory function is significantly improved following corrective surgery for severe pectus excavatum. Proposed treatment guidelines. J Cardiovasc Surg 2000;41:125–30.
[82] Maagaard M, Tang M, Ringgaard S, Nielsen HHM, Frøkiær J, Haubuf M *et al.* Normalized cardiopulmonary exercise function in patients with pectus excavatum three years after operation. Ann Thorac Surg 2013; 96:272–8.
[83] Tang M, Nielsen HHM, Lesbo M, Frøkiær J, Maagaard M, Pilegaard HK *et al.* Improved cardiopulmonary exercise function after modified Nuss operation for pectus excavatum. Eur J Cardiothorac Surg 2012; 41:1063–7.
[84] Cooper CB, Abrazado M, Saleh R, Elashoff R, Finn JP, Fonkalsrud EW. Anatomical and physiological improvements following surgical correction of pectus excavatum: a prospective study. Am J Respir Crit Care Med 2010;181:A5841.
[85] Kelly RE Jr, Mellins RB, Shamberger RC, Mitchell KK, Lawson ML, Oldham KT *et al.* Multicenter study of pectus excavatum, final report: complications, static/exercise pulmonary function, and anatomic outcomes. J Am Coll Surg 2013;217:1080–9.
[86] Morshuis WJ, Folgering HT, Barentsz JO, Cox AL, van Lier HJ, Lacquet LK. Exercise cardiorespiratory function before and one year after operation for pectus excavatum. J Thorac Cardiovasc Surg 1994; 107:1403–9.
[87] Wynn SR, Driscoll DJ, Ostrom NK, Staats BA, O'Connell EJ, Mottram CD *et al.* Exercise cardiorespiratory function in adolescents with pectus excavatum. Observations before and after Operation. J Thorac Cardiovasc Surg 1990;99:41–7.
[88] Cahill JL, Lees GM, Robertson HT. A summary of preoperative and postoperative cardiorespiratory performance in patients undergoing pectus excavatum and carinatum repair. J Pediatr Surg 1984;19:430–3.

European Journal of Cardio-Thoracic Surgery, 2024, 66, 1–34
https://doi.org/10.1093/ejcts/ezae166
Guidelines