

Interim Clinical Commissioning Urgent Policy Statement: Pectus surgery for pectus excavatum deformities resulting in very severe physiological symptoms (all ages) [URN 2256]

Summary

Pectus surgery is recommended to be available as a routine commissioning treatment option for patients of all ages with pectus excavatum resulting in very severe physiological symptoms within the criteria set out in this document.

Committee discussion

The Clinical Panel considered the Preliminary Policy Proposal which was submitted as an urgent policy request. It was agreed that this would proceed as a three-paper summary. It was recommended this proceed as an urgent policy statement.

The condition

Pectus abnormalities cover a range of deformities affecting the anterior chest wall, specifically the sternum and adjacent rib cartilages. Pectus excavatum (PE) and pectus carinatum (PC) are the most common of these. PE, or 'funnel chest', appears as a depression of the sternum; it may be asymmetrical, with the right side deeper than the left (de Oliveira Carvalho, 2014). It can be present at birth or may develop during childhood and adolescence. PC, also known as 'pigeon chest', is caused by the sternum pushing out so the middle part of the chest is more pronounced. It usually manifests at the time of a growth spurt in the early teenage years (Hebra et al 2016). If not corrected, the deformity is permanent.

Both deformities may be inherited, often being present in several members of the same family and associated with other congenital diseases including scoliosis of the spine, Marfan's syndrome and Ehlers-Danlos syndrome (Cobben et al 2014).

Pectus deformities vary from mild and asymptomatic to a more severe form which can alter measures of pulmonary and cardiac function. Diagnosis is by physical examination (Lain et al 2017). Patients with PE may experience severe pain, dysphagia, breathlessness, palpitations and fainting upon certain positions or during exercise. These symptoms cause significant impact to an individual's life (Lomholt et al 2016). In addition, patients with severe pectus deformity may experience life-threatening arrhythmias, Inferior Vena Cava (IVC) obstruction and be of a very high anaesthetic risk, causing necessary procedures (e.g., spinal surgery) unable to be performed.

Treatment choice is determined by assessment of the type of pectus deformity, degree of deformity, simple versus mixed deformity, and determination of whether the deformity is isolated or part of a syndrome. The severity of the deformity can be assessed radiologically using the Haller index, defined as the lateral external

distance divided by the distance between the external deepest point of the chest and external spinous process. Surgery is typically considered when an individual's Haller index is greater than or equal to 3.2 (Abdullah and Harris, 2016).

Current treatments

There are a number of non-surgical management options to support people diagnosed with a pectus deformity, including posture and exercise programmes, bracing and psychological support. One non-surgical treatment of pectus deformity involves the use of vacuum bell therapy. This treatment elevates the "funnel" in PE, through the use of a suction cup placed on the anterior chest wall to create a vacuum using a patient-activated vacuum. This treatment is suitable for patients with *mild* PE or in those in who surgery may not be suitable (Haecker and Sesia, 2016).

Proposed treatments

There are two types of surgical procedure available for the correction of pectus deformity:

- Nuss procedure, which is a minimally invasive procedure generally only used to treat cases of PE; and
- Ravitch procedure, which is an open surgical procedure that can be used to treat both cases of PE and PC.

The Nuss procedure involves placing one or two steel bars under the breastbone with the aim of raising it and correcting the abnormal shape. Each bar, bent into a curve to fit the patient's chest, is inserted through small openings in the chest. The bar (or bars) is usually removed within a few years of placement.

In the modified Ravitch procedure, the rib cartilages are cut away on each side and the sternum is flattened so that it will lie flat. One or more permanent bars or struts are inserted to ensure the sternum keeps its shape.

What we have decided

NHS England has carefully reviewed the evidence to treat PE in patients with very severe physiological symptoms with pectus surgery. We have concluded that there is enough evidence to make the treatment available at this time.

The three-paper summary review which informs this commissioning position can be accessed [here \(Appendix A\)](#).

Links and updates to other policies

This document is in addition to the NHS Clinical Commissioning Policy: Surgery for pectus deformity (all ages) (February 2019):

[1675-Policy_Surgery-for-pectus-deformity.pdf \(england.nhs.uk\)](#)

Plain language summary

About pectus deformities

Pectus deformity is a term used to describe a number of conditions which are related to the shape of the person's chest wall. These are usually present from birth and are

termed a congenital defect, but usually only become visible during early adolescence when the person undergoes growth.

There are two main types of pectus deformities:

- *Pectus excavatum* or 'funnel chest', which results from several ribs and the breastbone being pressed inwards, creating a dip between the ribs.
- *Pectus carinatum* or 'pigeon chest', which results several ribs and the breastbone growing outwards, making the chest appear pushed out

These defects may be symmetrical (the same on both sides) or may be prominent on one side of the chest. Some individuals with pectus excavatum or pectus carinatum will lead normal lives, however for patients with very severe pectus excavatum, the deformity can cause pressure on the heart and lungs, leading to life-threatening symptoms. These patients may require surgery to alleviate these symptoms.

Epidemiology and needs assessment

The number of patients likely to meet the clinical criteria for corrective pectus surgery for extremely severe PE is therefore estimated at 5-10 per annum.

Implementation

Inclusion criteria

Patients **must meet the following two** eligibility criteria:

- Have severe pectus excavatum on CT scanning with a Haller index of >3.25 .
- The national multidisciplinary team (MDT) confirms that the patient has potentially life-threatening complications of the greatest severity necessitating the need for urgent surgery.

AND EITHER

- Have an arrhythmia caused by PE

OR

- Have cardiac symptoms (syncope or pre-syncope) caused by PE

OR

- Have evidence of cardiac compression on Cardiac MRI (or equivalent imaging) with symptoms of cardiac or circulatory impairment

OR

- Have dysphagia deemed to be caused by PE

OR

- Have symptomatic cardiac or circulatory complications which would impede surgery for other conditions¹.

¹ This may include surgery for other conditions that require the patient to be in the prone position (e.g., spinal surgery) or cardiac surgery.

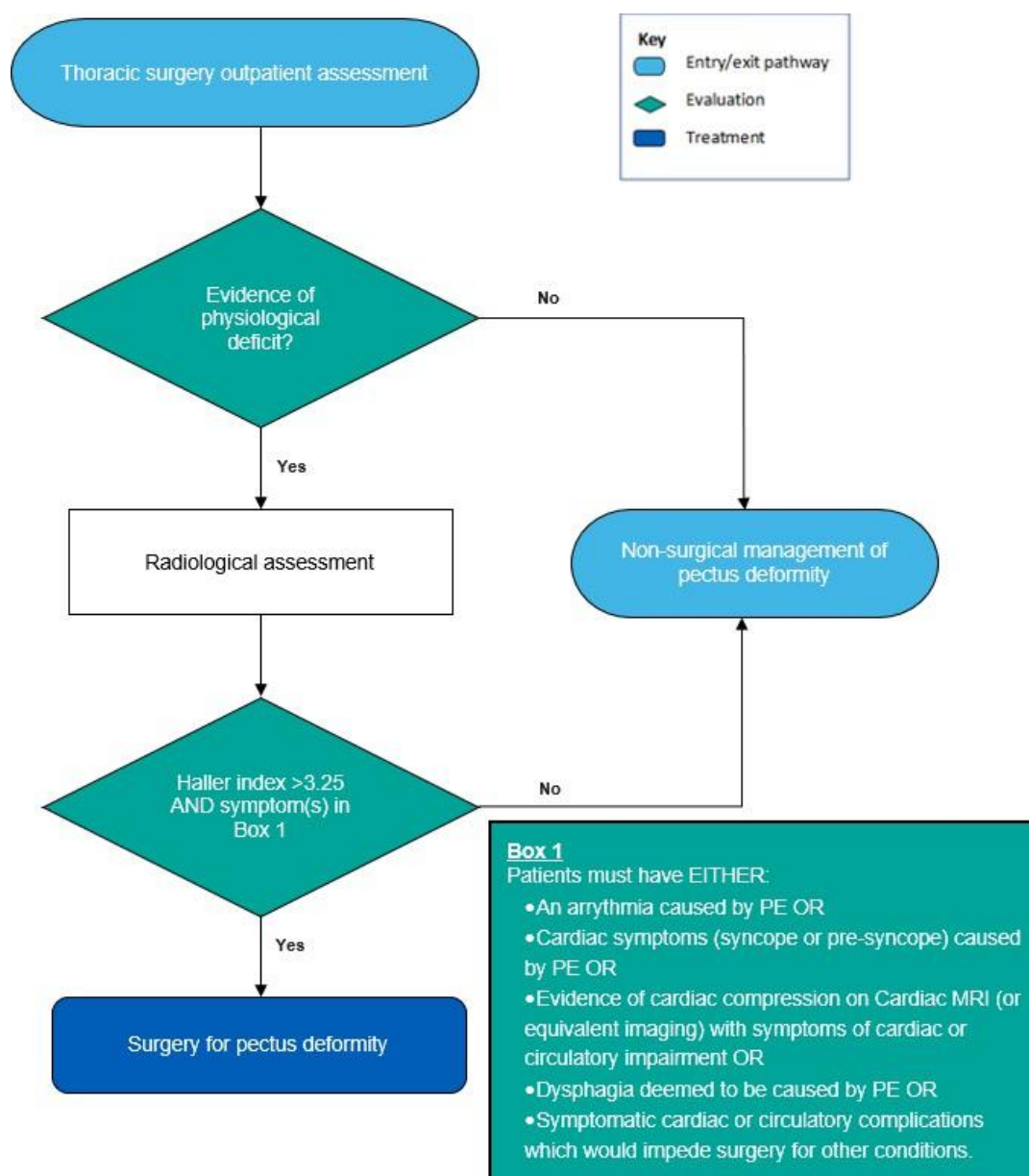
Exclusion criteria

Patients with PC are not eligible for treatment.

Starting Criteria

The patient must be discussed in a **MDT** and a consensus reached that the patient meets the eligibility criteria. The MDT will comprise of a cardiologist, a cardiac surgeon and a thoracic surgeon from each of the two/three specialist centres (including paediatric input as required). The MDT will be chaired by one of the cardiologists, with the chair rotating on an annual basis. The new NHS England National Clinical Lead relating to this subject area will be responsible for establishing the national MDT in association with the Society for Cardiothoracic Surgery in Great Britain and Ireland and British Association of Paediatric Surgeons. The MDT decision will be recorded and is auditable.

Patient Pathway



Governance arrangements

A commissioning plan will identify two adult specialist surgical centres and one paediatric centre, ideally co-located with one of the adult specialist centres that will be commissioned to perform pectus surgery. Centre selection will be considered with the support of the Society for Cardiothoracic Surgery in Great Britain and Ireland.

This policy proposition should be used in conjunction with the NHS England Specialised Services Thoracic Surgery Service Specification and the Paediatric Surgery Service and Congenital Heart Disease specifications which may be found at:

<https://www.england.nhs.uk/publication/thoracic-surgery-adults/>

<https://www.england.nhs.uk/wp-content/uploads/2018/08/Paediatric-congenital-heart-disease-specification.pdf>

<https://www.england.nhs.uk/wp-content/uploads/2013/06/e02-paed-surg-surgi-path-anaes.pdf>

Mechanism for funding

Pectus surgery for PE deformities resulting in very severe physiological symptoms within the criteria in this document will be commissioned and funded by NHS England Specialised Commissioning under existing arrangements for the provision of specialised cancer services and specialised paediatric surgery/congenital heart disease services. All associated activity should be recorded to the following lines:

Adults:

[NCBPS29B COMPLEX THORACIC SURGERY \(inpatient activity\)](#)

[NCBPS29Z THORACIC SURGERY \(outpatient activity\)](#)

Paediatrics:

[NCBPS23B CHILDRENS SERVICES - CARDIAC](#)

[NCBPS23X CHILDRENS SERVICES - SURGERY](#)

Audit requirements

A national registry will be established to collate data on surgical outcomes from pectus surgery, with support from the Society for Cardiothoracic Surgery in Great Britain and Ireland and the British Association of Paediatric Surgeons. It is essential that the national registry collects clinical treatment and clinical outcome data on patients treated via this policy in a timely, regular, and accurate manner. Such data should, at minimum, include demographic patient information, nature of urgency, reasons for treatment (i.e. which eligibility criteria apply), measurements of criteria where such apply, as well as the clinical outcome(s) of surgery at 3 months [whether symptoms and/or criteria have been alleviated] and whether any change is sustained at 6 months. The national dataset establishment will be proposed by the Society for Cardiothoracic Surgery in Great Britain and Ireland and British Association of Paediatric Surgeons. It is to be agreed with the Specialised Services Clinical Panel to ensure it can inform future commissioning decisions. All clinicians performing surgery for pectus deformity must ensure that any additional data collection requirements are met for the purpose of relevant audit and evaluation.

Policy review date

The Clinical Panel will review the policy along with the relevant audit information at one year from publication or if the number of patients treated exceed 20 per annum or on establishment of a clinical trial to review eligible populations. The policy will also be reviewed when information is received which indicates that the policy requires revision. If a review is needed due to a new evidence base, then a new Preliminary Policy Proposal needs to be submitted by contacting england.CET@nhs.net.

Equality statement

Promoting equality and addressing health inequalities are at the heart of NHS England's values. Throughout the development of the policies and processes cited in this document, we have:

- Given due regard to the need to eliminate discrimination, harassment, and victimisation, to advance equality of opportunity, and to foster good relations between people who share a relevant protected characteristic (as cited under the Equality Act 2010) and those who do not share it; and
- Given regard to the need to reduce inequalities between patients in access to, and outcomes from healthcare services and to ensure services are provided in an integrated way where this might reduce health inequalities.

Definitions

Pectus excavatum	A chest wall deformity that results from the inward growth of several ribs and the breastbone, making the chest appear hollow.
Pectus carinatum	A chest wall deformity that results from the outward growth of several ribs and the breastbone, making the chest appear pushed out.
Bracing	The use of a back brace to stop the deformity from getting worse. It is used in children, often until they stop growing.
Ehlers-Danlos syndrome	A group of inherited disorders that affect the connective tissues, primarily skin, joints and blood vessel walls.
Marfan's syndrome	A genetic disorder that affects the body's connective tissue
Nuss procedure	A minimally invasive procedure to repair pectus excavatum
Ravitch procedure	Is an open surgical procedure for the treatment of pectus excavatum and pectus carinatum
Scoliosis	A condition in which the spine can be curved or twisted

References

1. de Oliveira Carvalho, P.E., da Silva, M.V.M., Rodrigues, O.R. & Cataneo, A.J.M. Surgical interventions for treating pectus excavatum. *Cochrane Database of Systematic Reviews* (2014).

<https://doi.org/10.1002/14651858.cd008889.pub2>

2. Hebra, A., Calder, B.W. & Leshner, A. Minimally invasive repair of pectus excavatum. *Journal of Visualized Surgery* **2**, 73. (2016).

<https://doi.org/10.21037/jovs.2016.03.21>

3. Cobben, J.M., Oostra, R.J. & Dijk, F.S. Chest wall anomalies. Pectus excavatum and pectus carinatum. *European Journal of Medical Genetics* **3**, 414-417. (2014).

<https://doi.org/10.1016/j.ejmg.2014.04.017>

4. Haecker, F.M., Sesia, S. Non-surgical treatment of pectus excavatum. *Journal of Visualized Surgery* **2**, 63 (2016).

<https://doi.org/10.21037/jovs.2016.03.14>

5. Lain, A., Garcia, L., Gine, C., Tiffet, O. & Lopez, M. New Methods for Imaging Evaluation of Chest Wall Deformities. *Frontiers in Pediatrics* **5**, 257. (2017).

<https://doi.org/10.3389/fped.2017.00257>

6. Lomholt, J.J., Jacobsen, E.B., Thastum, M. & Pilegaard, H. A prospective study on quality of life in youths after pectus excavatum correction. *Annals of Cardiothoracic Surgery* **5**, 456-65. (2017).

<https://doi.org/10.21037/acs.2016.08.02>

Appendix A

NHS England

Paper summary:

Pectus surgery for very severe pectus excavatum with symptoms above shortness of breath

Drafted: September 2022

Prepared by NHS England Public Health Network on behalf of NHS England Specialised Commissioning

Pectus surgery for very severe pectus excavatum with symptoms above shortness of breath

Summary of papers presented for review

Three papers were presented for review. Paper 1 is a case report of a 14 year old male. Paper 2 is a case report of a 19 year old male. Paper 3 is a case report of a 22 year old female.

Paper 1: Pimenta et al, 2018

This paper is a case report of one patient (14 year old male) who presented to the paediatric cardiology department due to frequent episodes of palpitations lasting 15 minutes. On physical examination, he presented with severe pectus excavatum (PE). The rest electrocardiogram (ECG) had mild right-axis deviation without any other abnormality, and transthoracic echocardiography revealed compression of the right ventricle (RV), with normal tricuspid valvular flow and normal systolic and diastolic function. Twenty-four-hour ambulatory ECG monitoring showed single, paired and tripled polymorphic premature ventricular contractions arising from the lower part of the RV. During the exercise stress test, a triplet of polymorphic premature ventricular contraction was observed with the same origin described in ambulatory ECG monitoring. A chest computed tomography (CT) scan was performed to evaluate thoracic anatomy and a Haller index of 4.4 was obtained with compression of the RV. Because of this severity, surgical repair of the pectus excavatum was performed using the Nuss technique with no complications.

Paper 2: Moosdorff et al, 2021

This paper is a case report of one patient (19 year old male) who was scheduled for elective surgical correction of severe pectus excavatum. During forward bending for epidural catheter placement, syncope and ventricular fibrillation (VF) occurred resulting in cardiac arrest. After successful cardiopulmonary resuscitation, extensive analysis indicated that cardiac compression due to severe pectus excavatum was the cause for VF. Magnetic Resonance Imaging (MRI) and chest CT identified severe pectus excavatum with a Haller index score of 4.72. Postponed correction by modified Ravitch was performed, without implantable cardioverter-defibrillator (ICD) placement since VF was considered reversible, with no complications.

Paper 3: White et al, 2009

This paper is a case report of one patient (22 year old female) with a long history of recurrent syncope which began in early adolescence. On physical examination, the patient was found to have a marked pectus deformity with a straight back appearance. An ECG showed an incomplete right bundle-branch block, and a transthoracic echocardiogram was performed which was of poor quality owing to the patient's severe PE. Pulmonary function testing also showed vital capacity and total lung capacity were restricted. The patient lost consciousness on tilt-table testing at 80° at 2 minutes and 30 seconds with drops in both blood pressure and pulse wave amplitude. Cardiothoracic MRI demonstrated marked expiratory compression of the inferior vena cava (IVC) with near obliteration of the intrathoracic space between the sternum and thoracic vertebrae. The patient had a Haller index score of 18.8. The patient underwent a minimally invasive surgical repair of her pectus deformity by use of

the Nuss procedure with a pectus bar being inserted via bilateral video-assisted thoracoscopy, with resolution of the inferior vena cava compression.

Effectiveness

Complete response

Pimenta et al, 2018 reported that 2 years after surgical repair, the patient remained asymptomatic, with no palpitations, normal ECG monitoring and no arrhythmia on exercise stress testing. The postoperative chest computed tomography scan showed resolution of the RV compression.

Moosdorff et al, 2021 reported that 18 months after surgery, the patient remained well only reporting unremarkable occasional dizzy spells when tying shoelaces upon specific request.

White et al, 2009 reported that a predischARGE chest CT scan showed no IVC compression during maximal expiration and marked improvement in the minimal expiratory thoracic depth from 1.2 cm to 5.7 cm. Four months after surgery, the patient reported no further syncopal episodes, being able to stand and walk for extended periods of time without difficulty.

- **One paper reported resolution of syncope, one paper reported resolution of arrhythmia and one paper reported resolution of inferior vena cava compression with near resolution of all symptoms.**

Safety

- **All 3 case reports stated that the patients had successfully completed surgery.**

References

- Ventricular arrhythmia solved by surgical correction of pectus excavatum. Pimenta J, Vieira A, Henriques-Coelho T. *Interact Cardiovasc Thorac Surg*. 2018 Apr 1;26(4):706- 708. doi: 10.1093/icvts/ivx397.PMID: 29244154
- Case report: ventricular fibrillation and cardiac arrest provoked by forward bending in adolescent with severe pectus excavatum. Moosdorff M, Maesen B, den Uijl DW, Lenderink T, Franssen FAR, Vissers YLJ, de Loos ER. *Eur Heart J Case Rep*. 2021 Oct 25;5(10):ytab373. doi: 10.1093/ehjcr/ytab373. eCollection 2021 Oct.PMID: 34738057
- Images in cardiovascular medicine. Pectus excavatum with compression of the inferior vena cava: a rare cause of recurrent syncope. White JA, Fine NM, Shargall Y. *Circulation*. 2009 Oct 27;120(17):1722-4. doi: 10.1161/CIRCULATIONAHA.109.866400