

Cardiac & Respiratory Implications for Pectus Patients

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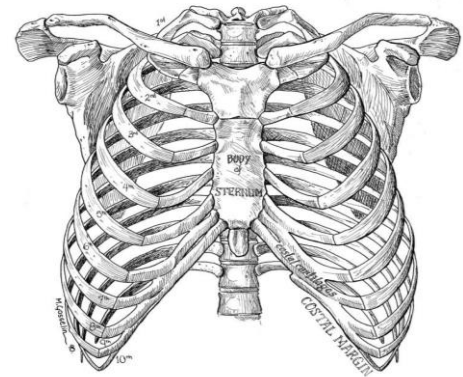
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THIS IS MODERN MEDICINE

Overview

- Pectus Excavatum (PEx)
 - Presentation
 - Mechanism for Cardiorespiratory Limitation
 - Controversy
 - Evidence Base
 - Surgical Considerations
-
- Other conditions: Pectus Carinatum (PC), Pectus Arcuatum (PAr), Poland Syndrome



Pectus Excavatum

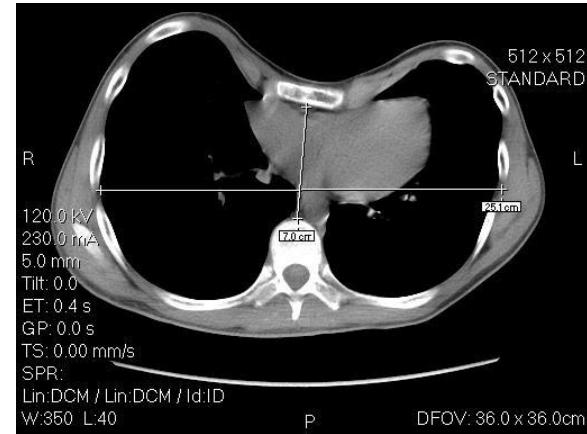
Pectus excavatum is a structural deformity of the anterior thoracic wall in which the sternum and rib cage are shaped abnormally

- This produces a caved-in or sunken appearance of the chest
- Most common chest wall deformity 1/400 – 1/600 births¹
- 90% of anterior chest wall deformities

Most frequently recognized during early childhood, may worsen during adolescent growth spurt²

Severity

- Haller Index (HI)
 - It is defined as the ratio of the transverse diameter (the horizontal distance of the inside of the ribcage) and the anteroposterior diameter (the shortest distance between the vertebrae and sternum)
 - Normal HI is 2.5, severe >3.25
- Cardiorespiratory symptoms
- Psychological and QoL effect



Presentation

- Most common presentation is due to cosmetic concern either from patient or parents

Symptoms³

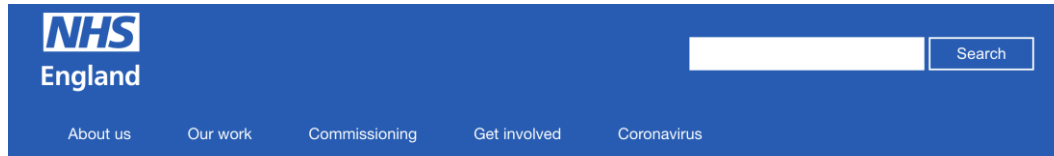
- Exercise intolerance (82%)
- Chest pain/discomfort (68%)
- Shortness of breath (42%)
- Palpitations

Signs

- Resting tachypnea
- Tachycardia
- Functional systolic murmurs

- Limited epidemiological data re prevalence of symptoms, data is from patients evaluated for surgery however *clear subset of patients with PEx who suffer from cardiorespiratory symptoms*
- Case series evidence suggests worsening of symptoms with age⁴

- **Controversy:** Quantifying these subjective complaints w/ qualitative evidence of impairment
- NHS previous decommissioning of surgical repair in UK citing insufficient evidence for improvement following surgical repair



Surgery for pectus deformity (all ages)

Document first published: 22 February 2019
Page updated: 22 February 2019

Topic:
Publication type: [Policy or strategy](#), [Report](#)

NHS England will not routinely commission surgery for pectus deformity in accordance with the criteria outlined in this document.

In creating this policy NHS England has reviewed this clinical condition and the options for its treatment. It has considered the place of this treatment in current clinical practice, whether scientific research has shown the treatment to be of benefit to patients, (including how any benefit is balanced against possible risks) and whether its use represents the best use of NHS resources.

Proposed Mechanism

? Respiratory

- Reduced thoracic volume
- Restrictive pulmonary defect

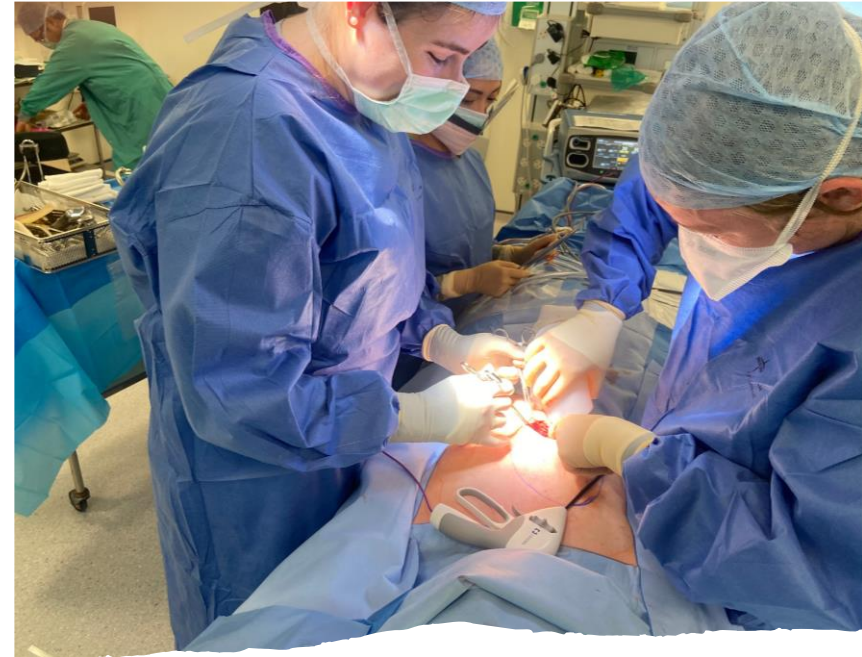
? Cardiac

- Mechanical compression of RV
- Reduced SV

? Both

? At rest vs dynamic in response to exercise

? Change with BMI or age



Evaluation

- **Controversy:** Quantifying these subjective complaints w/ qualitative evidence of impairment
- Recent re-look at policy and re commissioning of service – why?

Classification: Official

Publication reference: PR00394



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

Summary

The proposition is: 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.

Evidence Base

Evidence based off

- (1) Case reports of severe cases
- (2) Retrospective reviews of operative series
- (3) Small prospective cohorts
- (4) Systematic reviews of above

N.B. Data from patients considered for surgery, prone to inherent selection bias

Cases of PEx with severe cardiorespiratory compromise well documented

- Ventricular arrhythmias⁶
- Mitral valve prolapse⁷
- IVC compression⁸

Pectus Excavatum: A Case Study

Uchechi Eunice Okani DNP,¹ Peggy Mancuso PhD,²

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<https://doi.org/10.1016/j.nurpra.2012.05.021>

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Abstract

This case study presents the diagnosis and treatment of a child with pectus excavatum (PE), a common congenital abnormality of the anterior chest wall characterized by depression of the lower sternum. This depression ranges in severity from a minor dent to a deep, concave hollow capable of displacing the heart and intrathoracic structures. PE could present as a mild, asymptomatic condition, primarily with cosmetic implications. PE

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REVIEW ARTICLE

Pectus Excavatum With Compression of the Inferior Vena Cava

PDF/EPUB

A Rare Cause of Recurrent Syncope

James A. White, Nowell M. Fine and Yaron Shargall

Originally published 27 Oct 2009 |
<https://doi.org/10.1161/CIRCULATIONAHA.109.866400> |
Circulation. 2009;120:1722-1724

Jump to

Footnotes

JOURNAL ARTICLE

Ventricular arrhythmia solved by surgical correction of pectus excavatum

Joana Pimenta, António Vieira, Tiago Henriques-Coelho

Interactive Cardiovascular and Thoracic Surgery, Volume 26, Issue 4, April 2018, Pages 706–708, <https://doi.org/10.1093/icvts/ivx397>

Published: 12 December 2017 Article history

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Abstract

Pectus excavatum is generally considered a benign problem with a wide spectrum of impact in body image. Some articles had highlighted that this deformity can be associated with important clinical problems. We report a case of a 14-year-old boy with a severe pectus excavatum associated with palpitations who was

? Pulmonary function tests (at rest)⁹

- 21 studies pre and post repair
- Often low normal range, not correlated to severity of defect
- Improvements (if any) post surgery small and can not explain symptomatic improvement post-op



Thoracic surgery

BMJ Open
Respiratory
Research

Systematic review of physiological and psychological outcomes of surgery for pectus excavatum supporting commissioning of service in the UK

Jamie Walsh ¹, Ross Walsh,¹ Karen Redmond²

To cite: Walsh J, Walsh R, Redmond K. Systematic review of physiological and psychological outcomes of surgery for pectus excavatum supporting commissioning of service in the UK. *BMJ Open Respir Res* 2023;10:e001665. doi:10.1136/bmjresp-2023-001665

► Additional supplemental material is published online only. To view, please visit the journal online (<http://dx.doi.org/10.1136/bmjresp-2023-001665>).

Received 9 February 2023
Accepted 28 July 2023

ABSTRACT

Background Pectus excavatum (PEX) is the most common congenital chest wall abnormality affecting 1 in 400 births in the UK. PEX is associated with significant physiological and psychological impairment. While readily surgically correctable, the benefits that surgery can bring have been debated and proven difficult to objectively measure. In the UK, this has led to the decommissioning of PEX surgery. The aim of this review is to conduct a systematic search of the literature on PEX surgery to assess physiological and psychological outcomes.

Methods A systematic review of the MEDLINE (PubMed), Embase and Cochrane databases was performed. Articles were sought which included patients undergoing surgery for PEX and reported on changes in cardiopulmonary measures, symptoms, quality of life and psychological assessments before and after surgical repair. Last search was performed in July 2022 and relevant findings were synthesised by narrative review.

Results Fifty-one articles were included in qualitative

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ The benefits of surgical repair of pectus excavatum (PEX) are debated and changes in cardiopulmonary function has been difficult to objectively measure—this has led National Health Service UK to recently decommission surgery for PEX, affecting access to services.

WHAT THIS STUDY ADDS

⇒ This paper is an updated review of physiological and psychological outcomes following PEX surgery.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Cardiopulmonary exercise testing (CPET) has demonstrated improvements in exercise limitation following surgery both in adults and children however clinical correlation with patient's symptoms is necessary before minimally clinically important dif-


? Echo⁹

- 12 studies of TTE/TOE in PEx
- RV compression with improvement in RV end diastolic diameter and SV following repair
- Small subset of patient, has not been correlated with symptoms

Thoracic surgery

BMJ Open Respiratory Research

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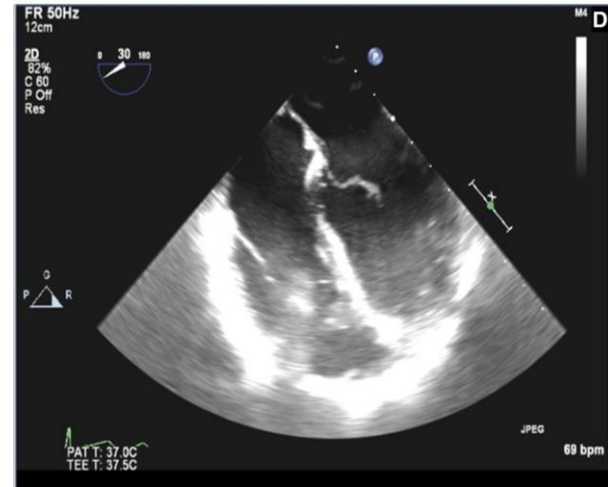
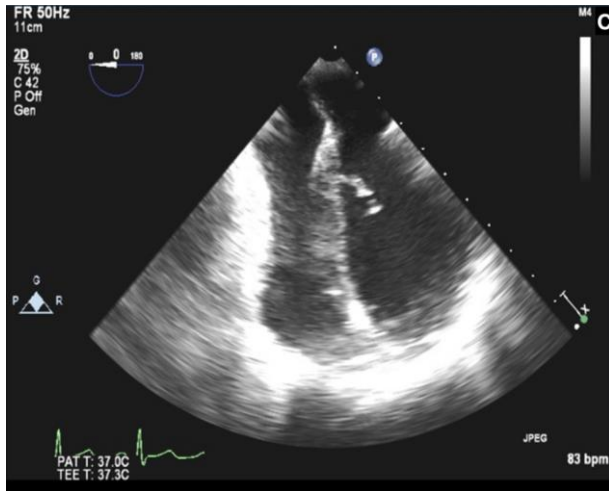
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Transoesophageal Echo

? Intraoperative transoesophageal echo during repair

- Complete release of RV compression and improvement of tricuspid annulus and right ventricle diameters after NUSS repair



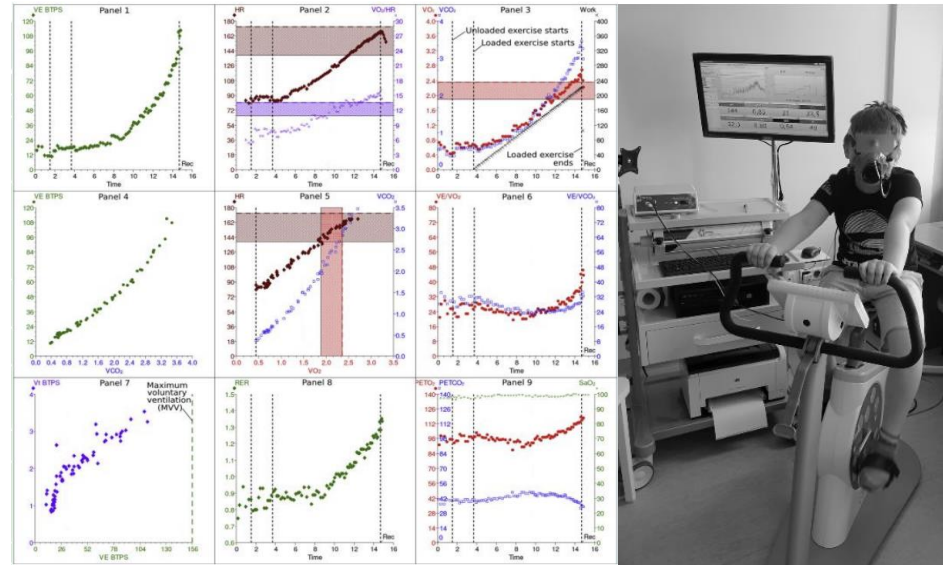
Cardiopulmonary Exercise Testing (CPET)

? CPET

- Assess functional capacity
- Better assessment for PEx where there is a dynamic cardiorespiratory limitation

Measures

- $\dot{V}O_{2\max}$
- O_2 Pulse
- Anaerobic threshold



Jaroszewski et al¹⁰

- Large cohort of 392 adult patients evaluated for NUSS Repair
- 68% abnormal CPET pre-op
- VO_{2max} and O_2 pulse sign improved post-op, correlated w/ HI

JAHA
Journal of the American Heart Association

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- DISCUSSION
- CONCLUSIONS
- Sources of Funding

Cardiopulmonary Outcomes After the Nuss Procedure in Pectus Excavatum

Dawn E. Jaroszewski, Juan M. Farina, Michael B. Gotway, Joshua D. Stearns, Michelle A. Peterson, Venkata S. K. K. Pulivarthi, Peter Bostoros, Ahmad S. Abdelrazek, ... See all authors

Originally published 4 Apr 2022 | <https://doi.org/10.1161/JAHA.121.022149> | Journal of the American Heart Association. 2022;11:e022149

This article is commented on by the following: [Other version\(s\) of this article](#)

Abstract

Background

Pectus excavatum is the most common chest wall deformity. There is still controversy about cardiopulmonary limitations of this disease and benefits of surgical repair. This study evaluates the impact of pectus excavatum on the cardiopulmonary function of adult patients before and after a modified minimally invasive

Table 3. Changes in Percentage of Predicted Relative VO_2 Max According to Prespecified Subgroup Analysis

Subgroup	Preoperative % of predicted VO_2 max	Postoperative % of predicted VO_2 max	P value (95% CI for the difference)
Sex			
Women	75.4±15.3	90.4±17.0	<0.001 (10.7–19.2)
Men	71.6±15.5	81.4±21.6	<0.001 (6.2–13.2)
Age, y			
≤32	68.5±13.7	79.3±19.8	<0.001 (6.4–15.0)
>32	77.0±16.0	89.0±20.4	<0.001 (8.5–15.6)
Inspiratory Haller index			
>3.25	72.1±15.1	83.3±20.2	<0.001 (8.3–14.2)
≤3.25	76.8±17.1	88.9±22.5	0.005 (4.0–20.3)

- ¿ Cardiopulmonary exercise testing (CPET)⁹
- 16 studies
 - Consistent impairment in response to exercise testing, correlated with anatomical severity
 - Significant improvements in $\text{VO}_{2\text{max}}$ and O_2 pulse seen following repair both in adults and children



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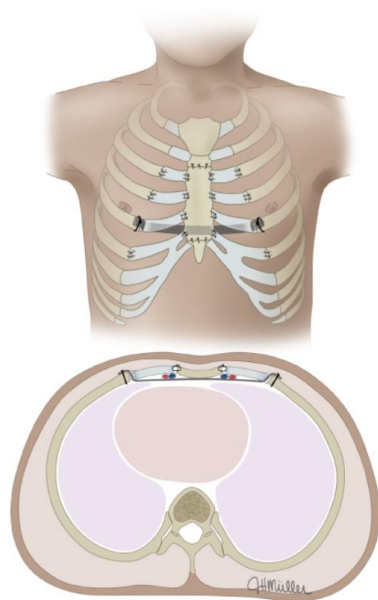
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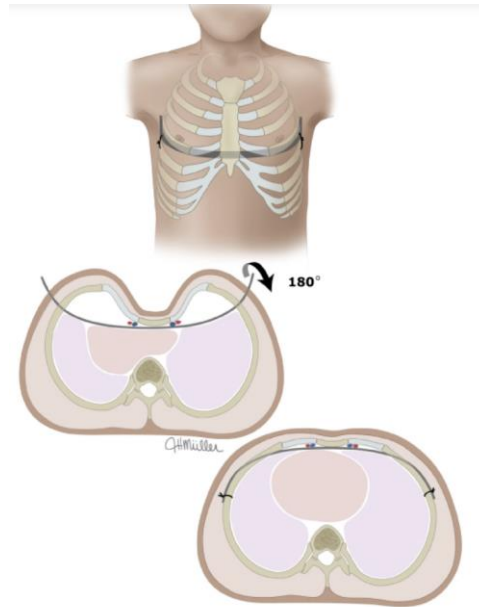
Surgical Considerations Corrective Surgery

Repair

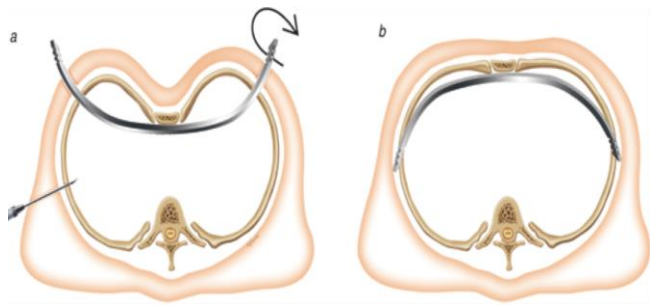
Open (modified Ravitch)

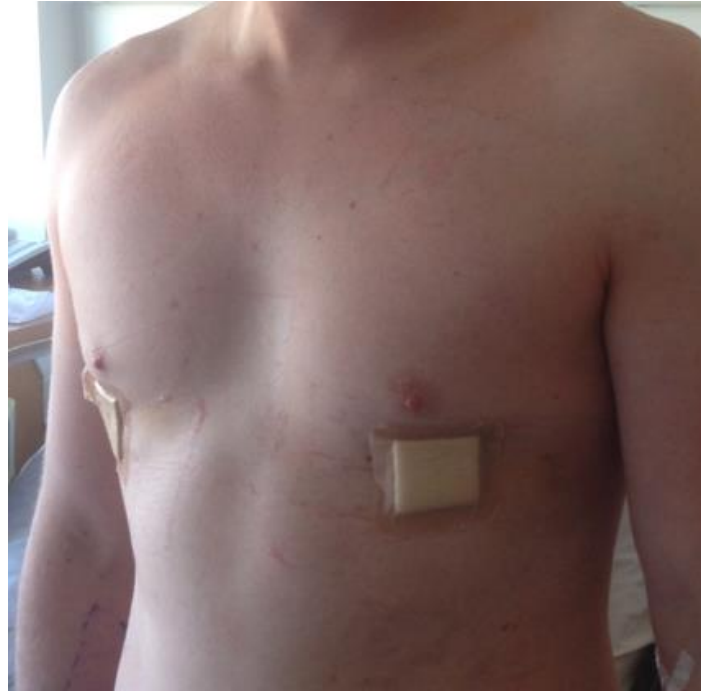


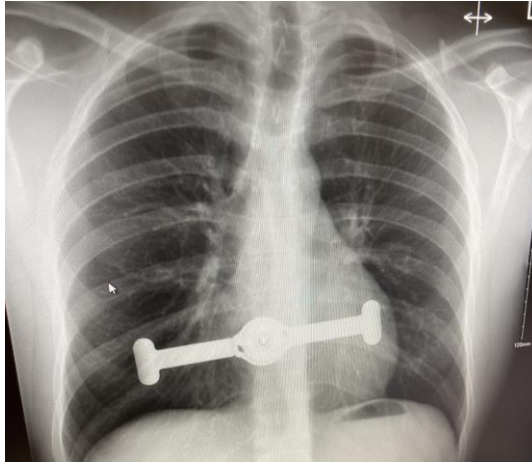
Minimally invasive (NUSS)



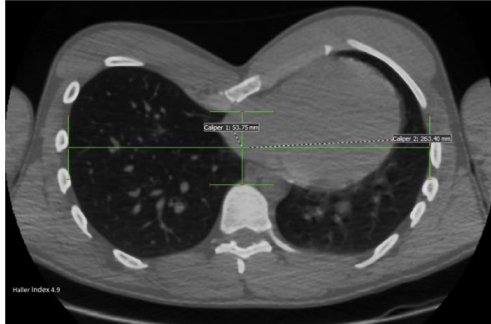
NUSS



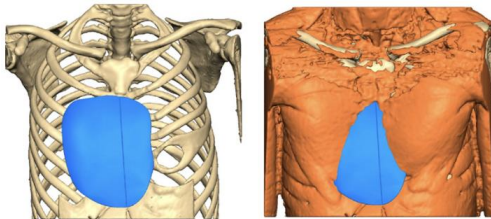




Surgical Considerations Non-Corrective Surgery Implant



Chest CT showing a significant pectus excavatum



Front bony view and with muscle showing implant in place

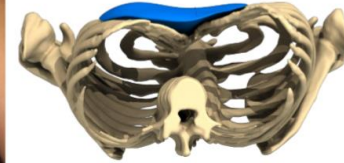


Before (left image) and 6 weeks following non-corrective surgery with a pectus implant (right image)

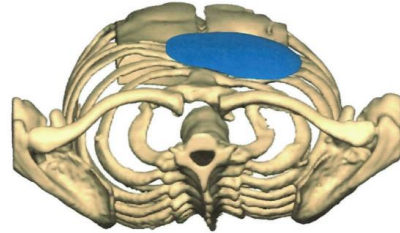
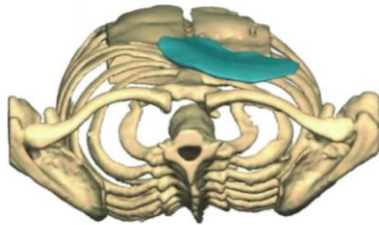
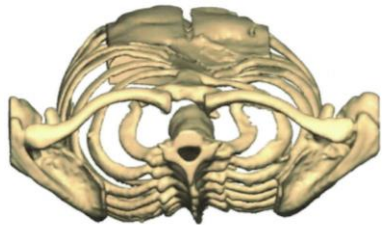


3D custom-made implants

3D CT Reconstructed Tailored Pectus Implants in Women



Redo Surgery



Pectus Carinatum

- Also called **pigeon chest**, is a malformation of the chest characterized by a protrusion of the sternum and ribs
- In moderate to severe cases of pectus carinatum, the chest wall is rigidly held in an outward position
 - Thus, respirations are inefficient and the individual needs to use the accessory muscles for respiration, rather than normal chest muscles, during strenuous exercise
 - This negatively affects gas exchange and causes a decrease in stamina
 - Children with pectus malformations often tire sooner than their peers due to shortness of breath and fatigue
 - Commonly concurrent is mild to moderate asthma
- Some children with pectus carinatum also have scoliosis (i.e., curvature of the spine).
- Some have mitral valve prolapse, a condition in which the heart mitral valve functions abnormally, may be due to associated connective tissue disorders

Marfan Risk Calculator

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Systemic Score

Click one or more physical characteristics to Include in the Calculation

Wrist AND Thumb Sign (+3)	Wrist OR Thumb Sign (+1)
Pectus Carinatum Deformity (+2)	Pectus Excavatum or Chest Asymmetry (+1)
Hindfoot Deformity (+2)	Plain Flat Foot (+1)
Spontaneous Pneumothorax (+2)	Dural Ectasia(+2)
Protocio Acetabulae (+2)	Scoliosis or Thoracolumbar Kyphosis (+1)
Reduced Elbow Extension (+1)	3 of 5 Facial Features (+1)
Skin Striae (+1)	Severe Myopia (+1)
Mitral Valve Prolapse (+1)	

Reduced Upper Segment / Lower Segment & Increased Arm span / Height (+1)

To learn more about each physical characteristic, click on the accordion links below:

1. Wrist and/or thumb sign

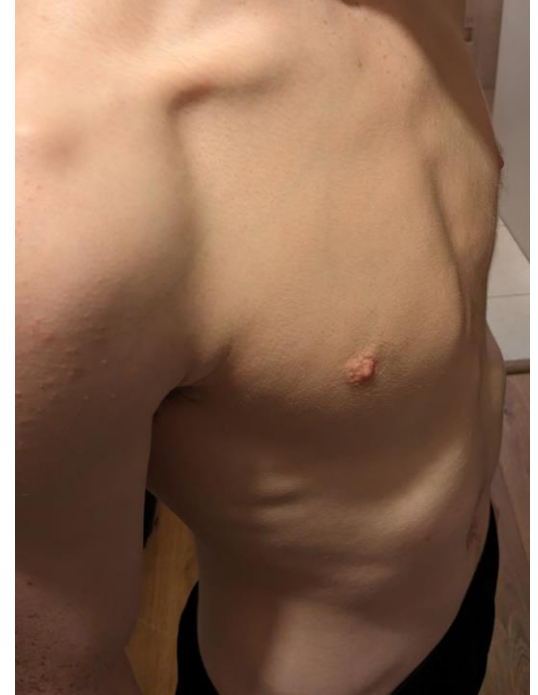
2. Anterior chest wall deformity

3. Hindfoot deformity

4. pneumothorax

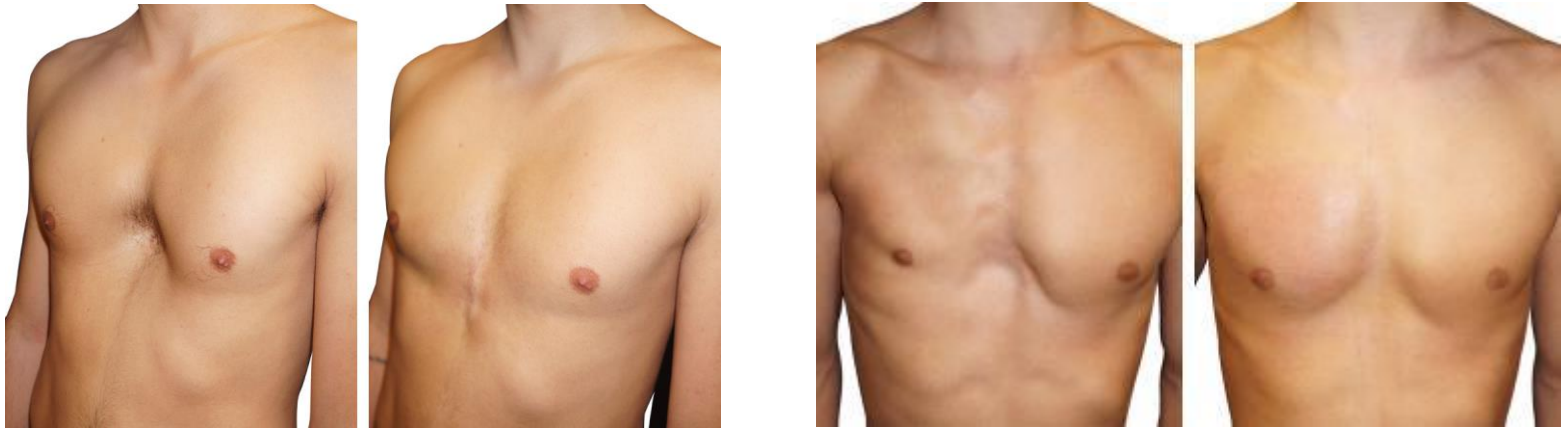
5. Dural ectasia

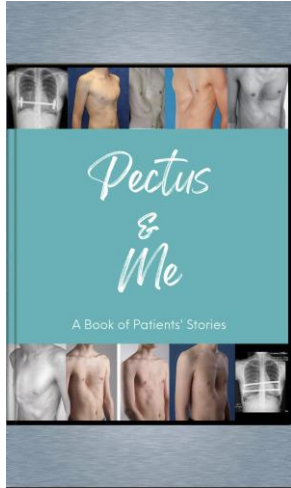
Pectus Carinatum Bracing



Pectus Arcuatum / Poland Syndrome

- A less common variant of pectus carinatum is PAr, produces a manubrial and upper sternal protrusion at the sternal angle
- Poland syndrome is a birth defect characterized by an underdeveloped chest muscle and short webbed fingers on one side of the body





- What is Pectus Deformity? ▾
- Effects of Pectus Conditions ▾
- What are the treatments available? ▾
- What are the benefits and risks of having surgery? ▾
- What is recovery like? ▾
- What is the referral pathway for treatment in the Republic of Ireland? ▾
- What is the referral pathway for treatment in Wales? ▾
- What is the referral pathway for treatment in England? ▾
- What is the referral pathway for treatment in Scotland? ▾
- What is the referral pathway in Northern Ireland? ▾
- Support Groups ▾

SCTS Website 'Pectus and Me'
Pectus support group / charity 'Pectus Matters'
Commissioning of 3 surgical units with St. Bart's National MDT
Regional units offering non-surgical care including bracing and VB therapy
The Joint Societies Best Practice Guidelines For The Treatment Of Patients With Pectus Abnormalities
NIHR-funded RESTORE Trial commissions 12 units



Referrals

Email

info@sshi.ie

admin@thoracictransplant.com

Fax-to-email

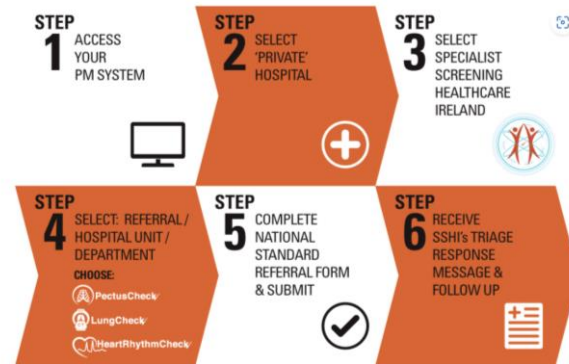
01-5563415

Healthlink e-Referral

Telephone

01-2645744

085-2562200



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