Ashford and St. Peter's Hospitals
NHS Foundation Trust

Pectus Anomalies:

A clinical guideline for management in CYP

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Guideline History		
Date	**Comments**	**Approved By**
01.07.2002	Reviewed by Mr Okoye	Mr Okoye
14.11.2022		Paediatric Guidelines Committee

Patients first • Personal responsibility • Passion for excellence • Pride in our team

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Pectus Guideline

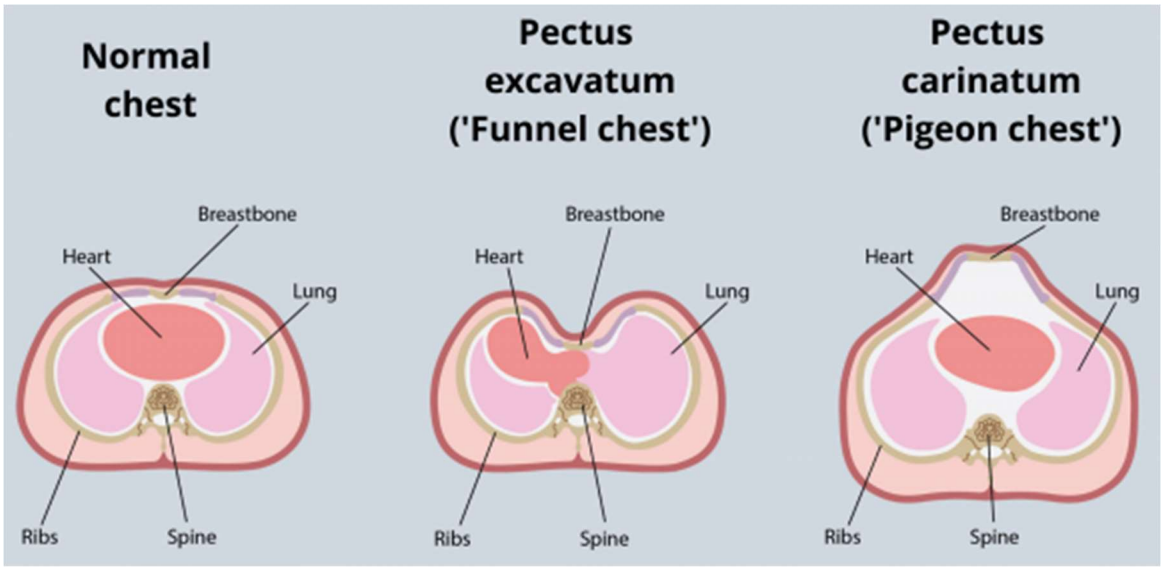
Introduction

Pectus anomaly is deformity of the sternum. It is the most common congenital wall deformity, occurring in 4 in 1,000. It is four times more common in boys. It can be present from birth but more often becomes more evident with rapid growth in early adolescence. 25% of diagnosed individuals have an affected family member.

Pectus anomalies are thought to be caused by poorly coordinated growth of the costal cartilages, in that they buckle and push the sternum inwards (pectus excavatum) or outwards (pectus carinatum).

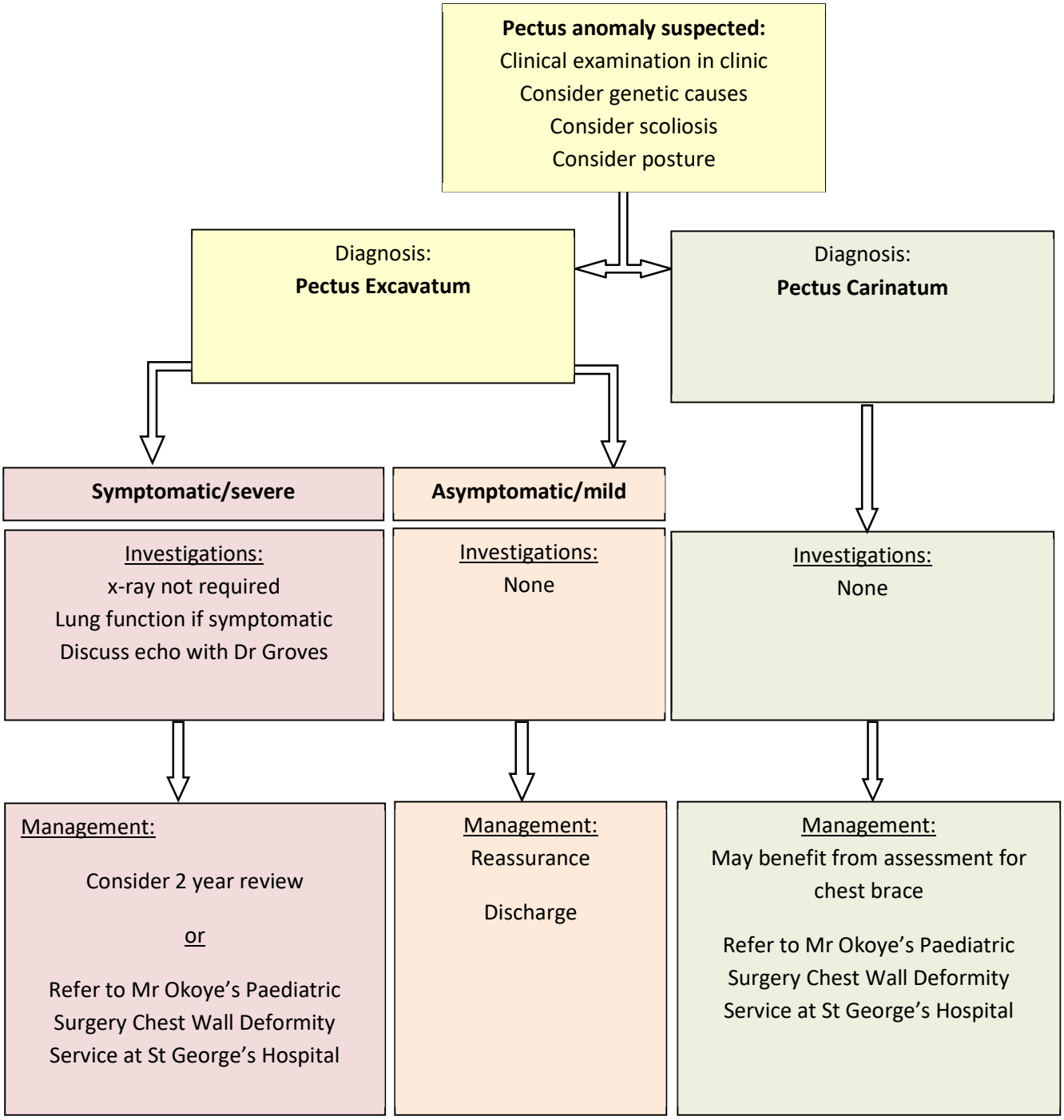
There are two main types of anomaly:

- **Pectus excavatum** (also known as "funnel chest"). The sternum is sunken inwards and the chest looks hollow. Affects 1 in 400 children and accounts for 87% of pectus deformities.
- **Pectus carinatum** (also known as "pigeon chest"). The sternum is raised outwards and the chest looks bulging and bird-like. Sometimes there is a depression (dip) on one side and a protrusion (bulge) on the other. Affects 1 in 1500 children and accounts for 5% of deformities. Most children do not have any symptoms beyond the cosmetic appearance of the chest.



Pectus arcuatum (also known as "horseshoe chest" or Currarino-Silverman syndrome) is a rarer deformity. There is a curved ridge across the upper portion of the sternum and the remaining chest lies flatter. It is often misdiagnosed as a combination carinatum-excavatum.

Flowchart


Pectus anomaly suspected:
Clinical examination in clinic
Consider genetic causes
Consider scoliosis
Consider posture
Diagnosis:
Pectus Excavatum
Diagnosis:
Pectus Carinatum
Symptomatic/severe
Asymptomatic/mild
Investigations:
x-ray not required
Lung function if symptomatic
Discuss echo with Dr Groves
Investigations:
None
Investigations:
None
Management:
Consider 2 year review
or
Refer to Mr Okoye's Paediatric Surgery Chest Wall Deformity Service at St George's Hospital
Management:
Reassurance
Discharge
Management:
May benefit from assessment for chest brace
Refer to Mr Okoye's Paediatric Surgery Chest Wall Deformity Service at St George's Hospital

Associated Symptoms

Common symptoms	Less frequent symptoms
• Chest pain • Back pain	• Shortness of breath on exertion • Syncope • Palpitations

Associated conditions

Musculoskeletal disorders	Connective tissue disorders	Genetic disorders

• Scoliosis
• Kyphosis
• Poland syndrome
	• Marfan syndrome
	• Ehlers-Danlos syndrome
		• Noonan syndrome
		• Turner syndrome
		• Loeys-Dietz syndrome
		• Homocystinuria
		  ○ Flushed cheeks
		  ○ Myopia
		  ○ Tall, thin build

Diagnosis and assessment

Pectus anomalies are diagnosed by clinical examination and inspection.

Full clinical examination should be undertaken, with focus on symmetry/asymmetry of the deformity, rib-flaring and features of associated conditions (above).

Assessment of posture is important to identify spinal deformity and "pectus posture" (rolled shoulders and lumbar lordosis attributed to shortening of the pectoral muscles.)

Investigations

Most children do **not** require any investigations if they are asymptomatic.

Imaging

X-ray: AP and lateral chest x-ray are **not** required at ASPH to make a referral to Paediatric Chest Wall Deformity Service (see flow chart). May be performed locally to help assess degree of excavatum and scoliosis.

Non-contrast chest CT: No routinely performed at ASPH. Chest deformity service may arrange to assess the Haller Index (the intrathoracic width divided by depth between sternal notch and vertebrae as seen on non-contrast CT).

Functional assessment

Lung function testing: Not required but may be helpful in **symptomatic** children to identify other causes of breathing difficulty (e.g. asthma). Basic spirometry, exhaled nitric oxide and peak flow can be performed at ASPH. Lung function testing is usually only possible in a cooperative child over about 8 years old.

Cardiopulmonary exercise testing (CPET) may be the most sensitive test for detecting functional consequences of physiologic impairments due to pectus excavatum and may be requested by cardiothoracic surgeons at a tertiary centre. This is not available at ASPH.

Echocardiogram: may be considered in severe cases if considering a tertiary referral or if associated cardiac problems are suspected. Please discuss individual cases with Dr Groves.

Other Investigations

Genetic testing: Consider genetic workup if suggestive features on history or examination.

Management

Most CYP with mild pectus excavatum **do not require any treatment or a referral**.

1. Asymptomatic CYP (i.e. no signs of cardiorespiratory compromise or pain):

   Reassure, watch and wait, especially if CYP is < 10 years old. CYP may be discharged with patient initiated review or offer 2 year review if concerns. Surgery is complex and often deferred until adulthood.

2. Symptomatic CYP or severe deformity

   Refer to Mr Okoye. Paediatric Surgery Chest Wall Deformity Service at St George's Hospital.

Exercises and physiotherapy may be help for CYP with mild pectus, especially "pectus posture". A postural deformity can be corrected by repositioning. A true pectus is a fixed deformity and will not be benefitted by physiotherapy.

Investigations for pectus deformity might reveal a **scoliosis**. Scoliosis must be referred to Orthopaedics. The orthopaedic team sometimes refer these CYP for physiotherapy.

Some CYP with pectus carinatum can be corrected with custom made orthotic braces. Braces can be an effective treatment but must be used long term. Braces are provided by the orthotic service at Queen Mary's Hospital in Roehampton. Referrals to this service are via Paediatric Chest Wall Deformity Service. Private management may be provided outside of the NHS. ASPH does not make any private referrals or recommendations.

Supporting References

https://www.baps.org.uk/subspecialties/thoracic/chest-wall-deformity/

https://www.rbht.nhs.uk/our-services/pectus-anomaly

https://www.nice.org.uk/guidance/ipg310

1675-Policy_Surgery-for-pectus-deformity.pdf (england.nhs.uk)

Krasopoulos G, Goldstraw P. 2011 Minimally invasive repair of pectus excavatum deformity. European Journal of Cardio-thoracic Surgery. February;39(2):149-58.

Goretsky MJ, Kelly RE, Croitoru D, Nuss D.2004. Chest wall anomalies. Pectus excavatum and pectus carinatum AdolescMed. 2004;15(3):455–71.

Kelly RE, Jr., Cash TF, Shamberger RC, Mitchell KK, Mellins RB, Lawson ML, et al. 2008. Surgical repair of pectus excavatum markedly improves body image and perceived ability for physical activity: Multicenter study. Pediatrics. Dec 2008

3. Supporting relevant trust guidelines

2. Guideline Governance

a. Scope

This guideline is relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department.

b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

All healthcare professionals responsible for the care of all children 0-18years should be aware of practice according to this guideline.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.
ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.
iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 3 yearly basis by the appropriate persons.
b. If new information comes to light prior to the review date, an earlier review will be prompted.
c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background • Who was involved in the Equality Impact Assessment
Author and the supervising consultants.
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion • Provide a summary of the overall conclusions
There is no evidence of discrimination.
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document: Pectus Anomalies: A clinical guideline for management in CYP

Policy (document) Author: Dr T Maycock

Executive Director: N/A

		Yes/No/ Unsure/NA	Comments
1.	**Title**		
	Is the title clear and unambiguous?	**Yes**	
	Is it clear whether the document is a guideline, policy, protocol or standard?	**Yes**	
2.	**Scope/Purpose**		
	Is the target population clear and unambiguous?	**Yes**	
	Is the purpose of the document clear?	**Yes**	
	Are the intended outcomes described?	**Yes**	
	Are the statements clear and unambiguous?	**Yes**	
3.	**Development Process**		
	Is there evidence of engagement with stakeholders and users?	**Yes**	
	Who was engaged in a review of the document (list committees/ individuals)?	**Yes**	**Mr B Okoye (St George's Hospital)** **Departmental Guidelines Meeting**
	Has the policy template been followed (i.e. is the format correct)?	**Yes**	
4.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	**Yes**	

		Yes/No/ Unsure/NA	Comments
	Are local/organisational supporting documents referenced?	**Yes**	
5.	**Approval**		
	Does the document identify which committee/group will approve/ratify it?	**Yes**	
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?	**NA**	
6.	**Dissemination and Implementation**		
	Is there an outline/plan to identify how this will be done?	**Yes**	
	Does the plan include the necessary training/support to ensure compliance?	**NA**	
7.	**Process for Monitoring Compliance**		
	Are there measurable standards or KPIs to support monitoring compliance of the document?	**NA**	
8.	**Review Date**		
	Is the review date identified and is this acceptable?	**Yes**	
9.	**Overall Responsibility for the Document**		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?	**Yes**	
10.	**Equality Impact Assessment (EIA)**		
	Has a suitable EIA been completed?	Yes	

Committee Approval (Paediatric Guidelines Group)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	**Dr Claire Mitchell**	**Date**	**14/11/2022**
Ratification by Management Executive (if appropriate)			
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner			
Date: n/a			