Learning objectives

- Identify common chest wall malformations and its pathology
- Understand common surgical correction procedures
- Identify post-surgical appearances

Background

Congenital chest wall deformities are due to anomalies of chest wall growth or prominence, leading to sternal depression, or deformities related to failure of normal spinal or rib development. Cross-sectional imaging allows appreciation of the involved structures, assessment of displacement or deformity of adjacent but otherwise normal structures, and differentiation between anatomical deformity and neoplasia. In some cases, CT is also useful for surgical planning. In this pictorial essay, we discuss the radiological features of congenital chest wall deformities and also illustrate pre and post-surgical appearances for more conditions requiring surgical correction.

Imaging findings OR Procedure details

Pectus excavatum

Pectus excavatum (funnel chest) is the most common congenital deformity of the sternum. Its incidence is approximately 1 in 400 births, afflicting men more than women. It is uncommon among African Americans and Latinos (1). Although the majority of pectus excavatum cases are congenital, around 15% of cases appear later during development and these are frequently associated with abnormalities of connective tissues such as Marfan and Ehlers-Danlos syndrome (2). Pectus excavatum is characterised by the presence of deep sternal depression causing the ribs on each side to protrude more anteriorly than the sternum (3). The sternal and cartilaginous depression causes a reduction in the prevertebral space, which gives rise to a leftward displacement and axial rotation of the heart. Other features include decreased density of the heart, an indistinct right heart border, horizontal posterior ribs, and vertical anterior ribs (4). These features can be seen on chest radiographs (Fig. 1 on page 5). While pectus excavatum is usually detected clinically, CT maybe used to quantify the severity of the deformity especially when surgical intervention is being considered (Fig. 2 on page 6, Fig. 3
on page 6, Fig. 4 on page 7, Fig. 5 on page 8). Severity may be guided using the Haller index, which is based on the ratio of lateral and antero-posterior thoracic diameters (Fig. 6 on page 9). Haller et. al. have suggested that an index of greater than 3.25 would require surgical correction (5). In normal children the Haller index ranges from 1.9 to 2.7. The Haller index for children under 2 years of age is markedly lower than that in older children, and girls between the ages of 0-6 and 12-18 years may have a higher index than their male counterparts (6). Different phases of respiration may affect the Haller index, and may yield a value significantly lower during inspiration. This reflects advances in CT scanner technology, in comparison to Haller’s day, when scans were performed over multiple breath holds (7).

Pectus excavatum is easily diagnosed in childhood, but is commonly ignored. Recent literature suggest that many patients experience detrimental cardiovascular and respiratory physiological changes as they mature. The exact reason remains elusive, but this maybe due to decreasing chest wall compliance with increasing age (8). Pectus excavatum correction may improve the quality of life in patients, both physically and psychosocially (9).

The two most common surgical procedures used to correct pectus excavatum deformities are the Nuss procedure and the modified Ravitch procedure. The Nuss procedure is also known as minimally invasive repair of pectus excavatum (MIRPE).

The Nuss procedure is a minimally invasive procedure which consists of implanting two curved retrosternal metallic bars, inserted through small lateral incisions (Fig. 7 on page 10, Fig. 8 on page 11) (2). This procedure allows immediate correction of the deformity since in childhood, chest wall structures are less rigid. The Nuss bar is usually removed after 3 years. The advantage of the Nuss procedure is its cosmetic advantage, with smaller skin incisions which maybe more acceptable. However, it is associated with a higher complication rate and cost when compared to the modified Ravitch (10).

The original Ravitch procedure is a radical repair, freeing the sternum from all restrictions. He resected all sternal attachments and the whole length of the deformed costal cartilages. The sternum is then bent sharply anteriorly, fracturing the posterior cortical lamella in the process (11). However, without external traction, which was popular then, his technique had an increased recurrence rate. The modified Ravitch procedure is more extensive when compared to the Nuss procedure, and involves the resection of the deformed anterior costal cartilages and then fixation of the sternum into a more normal position using metal bars (Fig. 9 on page 12, Fig. 10 on page 13) (2). The metal bars are wired to the adjacent sternum and are typically removed after 2 years.
Chest radiographs are useful to visualise the position of the Nuss and Ravitch bars, and to detect any complications. The complications include wound infections, haematomas, bar migration and pneumothorax (Fig. 1 on page 5, Fig. 10 on page 13, Fig. 11 on page 13, Fig. 12 on page 14).

**Pectus carinatum**

Pectus carinatum (pigeon chest) is the second most common congenital deformity of the sternum, with its incidence estimated to be 5 times lower than pectus excavatum (12). It is characterised by an abnormal anterior protrusion of the sternum and costochondral joints (Fig. 13 on page 15, Fig. 14 on page 16, Fig. 15 on page 18, Fig. 16 on page 18, Fig. 17 on page 19, Fig. 18 on page 20). Pectus carinatum can be classified according to the location of the protrusion. Type 1 (inferior or chondo-gladiolar), which is the most frequent type, involves protrusion of the inferior or mid-sternum, while Type 2 (superior or chondro-manubrial) involves the manubrium or superior aspect of the sternum (2). Surgical correction is reserved for moderate or severe deformities. Initial lateral chest radiographs will demonstrate sternal protrusion, but subsequent imaging with CT allows quantification of the severity using the Haller index, similar to its use in pectus excavatum (13). A study quoted the average Haller index of patients requiring surgical correction as 1.8 (14). In pectus carinatum, a lower index indicates more severe deformity. Surgical correction of pectus carinatum is similar to pectus excavatum in which the Ravitch and Nuss procedures are modified appropriately to correct the protruding defect (15). Recent literature has suggested the increasing popularity of non-invasive bracing as an alternative treatment (16).

**Pectus arcuatum**

Pectus arcuatum means 'wave-like' chest deformity and it is a rare condition with an unknown aetiology. The term is used to describe mixed deformities which contain both excavatum and carinatum either along a longitudinal or axial axis, and is also known as a pouter pigeon chest (16) (Fig. 19 on page 21). It contains a protrusion at the upper part of the sternum involving the manubriosternal junction and the second to fifth rib cartilages, with premature sternal ossification (1). There may be an associated excavatum deformity at the lower sternum in up to one third of cases. The pouter pigeon chest can be appreciated on sagittal CT images although traditionally lateral chest radiography have been used. Imaging with CT allows calculation of the angle of Louis in order to determine the severity of the deformity. The normal angle of Louis is between 145-175°. Surgical correction is recommended in patients with an angle of #130° (18).

Surgical correction involves a wide wedge transverse sternotomy at the angle of Louis and subperichondrial resection of the adjacent costal cartilages (1).
### Poland syndrome

Poland syndrome is a non-genetic congenital abnormality and the aetiology is unknown. It is characterised by partial or total absence of the pectoral muscles and is most commonly unilateral (2). There is a range of breast involvement, varying from mild hypoplasia to complete absence (amastia). Poland’s syndrome is associated with rib cage anomalies in up to 60% of cases including aplasia or hypoplasia of the ribs (13). Other associations include hand involvement (varying from mild shortening of the phalanges to syndactyly), lung herniation and dextrocardia (17).

On chest radiography, the affected side appears of increased transradiancy due to reduction in overlying soft tissues. CT is useful to demonstrate the absence of the greater pectoral muscle and any associated musculoskeletal anomalies of the chest wall (Fig. 20 on page 22, Fig. 21 on page 23) (3). Surgical correction is only required if there is a rib defect large enough to cause a lung herniation or if there are concerns of injury to the heart or lungs (19). Adolescent females with amastia may also require cosmetic reconstruction.

### Rib Exostosis

Rib osteochondromas can be solitary or associated with multiple hereditary exostoses. Up to 50% of patients with multiple hereditary exostoses may have rib osteochondromas (20). Exostoses projecting outwards maybe palpable, whereas internal exostoses may be an incidental finding on imaging only (21) (Fig. 22 on page 24, Fig. 23 on page 25, Fig. 24 on page 26).

### Normal variation

There are normal asymptomatic variations of the anterior chest wall that maybe incidental on imaging. They may include a tilted sternum, convex anterior ribs or prominent costal cartilages (22).

### Images for this section:
Fig. 1: A 24 year old female patient presented with pectus excavatum. Three consecutive radiographs demonstrates subsequent corrective surgeries. (a) Pre-operative chest radiograph. (b) The patient initially underwent a Nuss procedure. Note the post-operative right sided pneumothorax and subcutaneous surgical emphysema. (c) She subsequently needed the Nuss bar removed, which was then exchanged for a Ravitch bar.

Fig. 2: (a) Axial CT of the same 24 year old patient demonstrating pectus excavatum. Note the sternal depression and tilting, with leftward displacement of the heart. (b) Sagittal reconstructions.
Fig. 3: 3D surface reconstructions of the same 24 year old patient, showing sternal depression and tilting.
Fig. 4: 3D reconstruction of the same 24 year old patient with pectus excavatum, demonstrating anterior chest wall depression.
Fig. 5: 3D cutaway of the same 24 year old patient with pectus excavatum.
Fig. 6: Haller index of the same 24 year old patient (ratio of A to B).
Fig. 7: CT of a 37 year old male patient with a Nuss bar deep to the sternum.
**Fig. 8:** 3D reconstructions of the same 37 year old patient with a Nuss bar.
**Fig. 9:** 3D surface reconstructions of a 21 year old patient presenting with pectus excavatum.

**Fig. 10:** (a) Pre-operative CT of the same 21 year old female patient. (b) Post modified Ravitch procedure CT. The right anterior end of the Ravitch bar had migrated inferiorly and invaginated the lung parenchyma. The bar was subsequently removed.
Fig. 11: 51 year old man with a previous modified Ravitch procedure presented with chest tightness. An initial chest radiograph demonstrated migration of the Ravitch bar inferiorly and eccentric to the right.
Fig. 12: 3D surface reconstruction of the same 51 year old patient with migration of the Ravtich bar.
**Fig. 13:** Radiograph of a 34 year old patient with pectus carinatum. The PA appearances are almost normal, but the lateral CT reconstructions will demonstrate the "pigeon chest" much clearer.
**Fig. 14:** Sagittal reconstructions of the same 34 year old patient with pectus carinatum, belonging to the most common type 1.

![Sagittal reconstructions of the same 34 year old patient with pectus carinatum.](image)

**Fig. 15:** CT of the same 34 year old patient with pectus carinatum.

![CT of the same 34 year old patient with pectus carinatum.](image)
Fig. 16: 3D surface reconstruction of the same 34 year old patient with pectus carinatum.
Fig. 17: 24 year old male patient with asymmetrical pectus carinatum affecting the right costal cartilages
Fig. 18: 3D reconstructions of the same 24 year old patient with asymmetrical pectus carinatum.
Fig. 19: A female patient with mixed deformity consisting of both pectus excavatum and carinatum, creating a "rolling" appearance.
**Fig. 20:** Poland syndrome with absence of right pectoral muscles causing chest wall asymmetry.
Fig. 21: MIP images of the same patient with Poland syndrome. In addition to the absence of right sided pectoral muscles, there is also abnormal sternal protrusion.
**Fig. 22:** Right 5th rib exostosis directed into the thoracic cavity.
**Fig. 23:** Coronal reformats of the same patient with exostosis of the right 5th rib.
**Fig. 24:** 3D reconstruction of the same patient with a right 5th rib bony exostosis.
Conclusion

In summary, there is a variety of congenital conditions affecting the chest wall which commonly presents at birth or childhood. We have reviewed the characteristic radiographic and CT appearances of these conditions. Familiarity with the different congenital chest wall deformities allows accurate diagnosis and provides helpful information for appropriate patient treatment.

References


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Personal Information