



Case Report

## Pectus Deformities: Dilemma in Diagnosis

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### ABSTRACT

Pectus deformities are among those congenital skeletal deformities which can be easily identified just by physical examination, but the actual problem arises when there is a need to classify them depending on their skeletal presentation. Therefore there is always a need for radiological evaluation along with physical examination of the patient to justify the type of pectus deformity. Present article mainly focusses on correlating the external appearance of the anterior chest wall with the radiological imaging of ribs and sternum.

**Key words:** Pectus carinatum; Pectus excavatum; Chondro-gliolar deformity; Chondro-manubrial deformity

### INTRODUCTION

Pectus deformities are among the commonest anterior chest wall deformities. They have an incidence of approximately 1:1000 and the genetic studies have proved a congenital origin. Chest-deformities must not always be present at birth; they can develop gradually in the first 5 years of life.

[1] According to Goretzky et al; congenital chest wall anomalies fall into two groups: those with overgrowth of the rib cartilages causing either a depression or protuberance, and those with varying degrees of either aplasia or dysplasia. Pectus excavatum, a chest wall depression, accounts for more than 87% of the deformities and is by far the most common chest wall deformity, as its name suggests, it presents with an

excavated, sunken or funnel chest. Pectus carinatum, a chest wall protuberance, constitutes approximately 5% of chest wall deformities and combined excavatum/carinatum deformities constitute 6.1% of chest wall anomalies. [2,3]

During the course of our research from the year 2008-2014, we were exposed to a wider spectrum of pectus deformity presentations. In this period of six years we observed that certain cases of chest deformities initially seemed like a misfit into the basic three categories of chest wall deformities which was later proven otherwise. We realized that to understand and to identify a particular type of pectus deformity, there are two important criteria's:- a) External appearance of the

anterior chest wall, b) radiological imaging of the skeletal components of the chest. Therefore this present report series would be first of its kind, in an effort to correlate the external appearance of the chest with radiological imaging of the chest wall. We present six case reports of chest deformities which required special attention for they were posing difficulties in concluding the diagnosis for the specific category of chest deformity.

## **MATERIALS AND METHODS**

During the past of six years, our group of anatomist and radiologist with special interest in the field of chest deformities collaborated along with paediatric and the orthopaedic departments for patient referral. After their initial assessment patients were sent for further evaluation; spirometric and radiological analysis. Radiological data concerning the patients were collected mainly from two medical centres; KVG Medical College and hospital and K S Hegde medical college and hospital. Physical examination reports and radiological imaging concerning only to thorax has been explained, others area are not included as they were out of the scope of this particular article.

## **CASE REPORTS WITH DISCUSSION**

Pectus deformities are broadly classified into three major categories depending on the skeletal presentation: A) pectus excavatum B) pectus carinatum Type 1 [chondro-gladiolar deformity] C) pectus carinatum Type 2 [chondro-manubrial deformity]. Description like funnel chest, chicken breast and pigeon breast are also used to describe each of these deformities. Some of the cases mentioned in this article initially seemed like a mismatch when tried to pinpoint the deformity either only by the method of radiological imaging or by simple physical examination of chest wall.

Therefore effort has been made to merge the radiological findings and external skeletal presentation of each case to reach final diagnosis.

### ***Pectus carinatum (Type 1):***

*Case 1A:* 7 year old male child visited the department of paediatrics with a bulge in the upper anterior chest wall [Fig 1]. Parents were questioned about the medical and family history of the child. There was no history of fall or trauma. Physical examination of the child showed a convex shaped bulge in the upper chest wall, with a prominent sternal angle. Respiratory pattern was predominantly abdominal during chest wall motion, with no difficulty for expansion for breathing. Lung sounds were clear, no murmurs were heard. No tenderness was elicited on palpation and the curvature of the thoracic spine was normal. Radiological evaluation was recommended for ruling out and mediastinal masses or lung abnormalities. CT images showed pectus carinatum type 1 deformity [Fig 2]

*Case 1B:* 10 year girl child was brought to the department of paediatrics with complaint of protruding upper chest wall [Fig1]. Physical examination showed that the child had a prominent upper chest wall protrusion with widely spaced nipples, retracted scapula and externally rotated shoulders. Examination of spine showed a reduced thoracic curvature and exaggerated lordotic lumbar curve. Thoracic expansion was fairly good with a reduced diaphragmatic expansion. CT images showed a pectus carinatum type 1 deformity [Fig 2].

CT sagittal images of 11 year old female (case1A) child showed the manubrium along with the entire body of sternum protruding forwards with the xiphoid process joining it without producing any angulation with an obvious increased sterno-vertebral distance. On the other hand CT plain sagittal and axial images of a 7 year male child (case 1B) showed the mal-

aligned manubrium and gladiolus and the increased sterno-vertebral distance. here the manubrium and the upper sternebrae are protruding forwards and the lower two sternebrae are curving backwards and joining the xiphoid process without producing any angulation, making the entire sternum appear convex forwards and with the ribs been more horizontally placed [Fig 3,4]. Radiological evidences were sufficient to pronounce both cases as pectus carinatum.



Figure 1: Photographic images of case 1A - 7 year old female child (A) and case 1B - 11 year old female child (B) with pectus carinatum type 1 deformity.

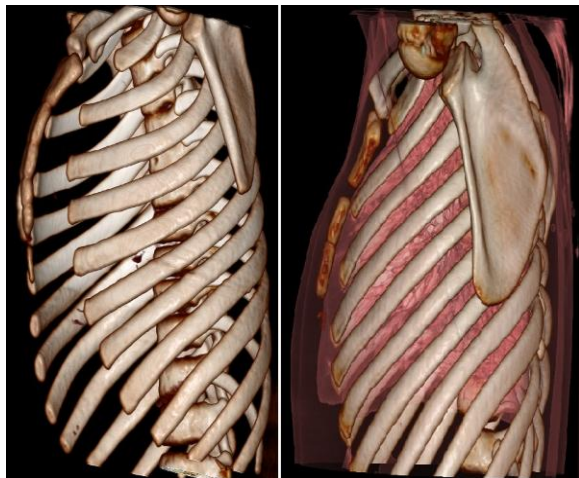


Figure 2: CT reconstructed images showing the difference in the alignment of manubrium and the body of sternum, also the inclination of ribs between two cases (cases 1A and B).



Figure 3: CT plain sagittal images showing difference in the shape of sternum. A – Case 1A shows incurving (convex anteriorly) sternum. B – Case 1B shows straight, forward protruding sternum.

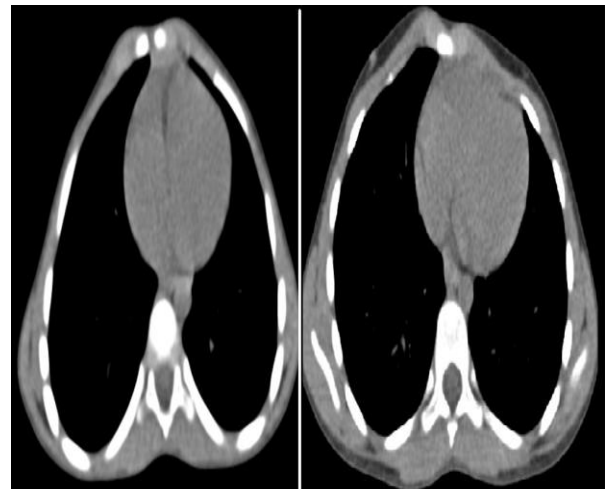


Figure 4: CT axial images at the level of xiphi-sternum, showing increased sterno-vertebral distances in both cases 1A and 1B.

Both the children explained here had an evident pectus carinatum deformity but there was a gross difference in their external presentation as well as the alignment of bony components of sternum. Robisek et al explains Pectus Carinatum type 1 (Chondrosternal Prominence, Chondrogladiolar Prominence) as the protrusion of the sternum and symmetrical prominence of the involved costal cartilages. The sternum itself is elongated and joins the xiphoid process in an abnormal sharp angle that makes the sternal protrusion even more evident. [4] When comparison was done

between the classical presentation as explained by robisek et al and the two present reports, case 1A shows close resemblance with the classical chicken breast appearance, but case 1B showed difference in external appearance as well as alignment of the bony components, especially sternum. The position of *manubrium and upper two sternebrae* seem to be vital. These two bony structures are protruding forwards in both cases, the angulation *at the manubriosternal junction* becomes less prominent and but the key difference is the position of lower sternebrae which is curving in instead of protruding forwards. anatomical variations between two cases of pectus carinatum type 1 are detailed in table 1.

***Pectus carinatum type 2 (Chondro-Manubrial deformity):***

*Case 2A:* A 24 year old male presented himself to the department of orthopaedics with cosmetic discontent of his chest wall appearance [Fig 5]. Physical examination showed patient was well built, with prominent pectoral muscles masking the deformity, anterior chest wall showed protruding manubrium and sunken body of sternum above the level of nipples, giving an appearance false appearance of pectus excavatum. Diaphragmatic excursion was less with no paradoxical movements of chest wall. 3D CT reconstructed images revealed chondro-manubrial deformity rare type 2 pectus deformity [Fig 6].

*Case 2B:* 17 year male child visited the orthopaedic outpatient department with chief complaints of deformed chest and spine and with breathlessness with physical exertion. Physical examination showed that that child was malnourished with deformed ribcage also revealed a midline depression in the anterior chest wall at the level of nipples followed by a protruding abdomen [Fig 5]. Further evaluation also revealed severe kypho-scoliotic deformity. Thoracic

expansion was limited. Radiological images showed an abnormal acute angulation at the level of third costal cartilage. Several rib abnormalities were noted with kypho-scoliotic deformity of the spine [Fig 6].



Figure 5: Photographic images of case 2A - 24 year old male (A) and case 2B - 17 year old male child (B) with pectus carinatum type 2 (chondro-manubrial) deformities.

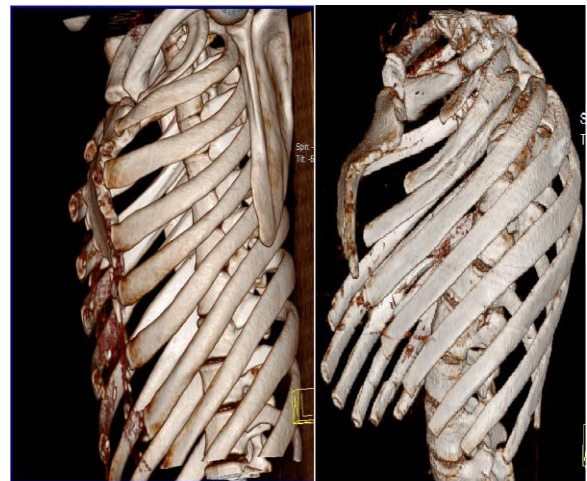


Figure 6: CT reconstructed images showing the difference in the alignment of manubrium and the body of sternum with the angulation, also the inclination of ribs between two cases (cases 2A and 2B).

According to Balasubramaniyam; chondro-manubrial prominence with chondro-gladiolar depression also known as pectus carinatum type 2 or pouter pigeon (Currarino-Silverman (C-S) syndrome). This infrequently seen anomaly consists of protrusion of the manubrium sterni and the adjacent two pairs of costal cartilages and a

typical excavatum deformity involving the lower part of the sternum and is probably caused by premature fusion of some of the sternal ossification centres and by obliteration of the manubrio-sternal joint. This results in an abnormally short sternum with forward angulation at the manubrio-sternal junction. [5,6]

In the present report, CT bone reconstructed and plain sagittal images of the case 2A; 24 year old male shows a short sternum, fused manubriosternal joint, having an acute angulation at the level of 3<sup>rd</sup> costal cartilage. [7] Manubrium and the upper sternbrae are protruding forwards with the backward bend of the remaining part of the gladiolus. A similar case of chondro-manubrial deformity, 17 year old boy (case 2B) with a severe kypho-scoliotic deformity presented with forward protruding manubrium similar to case 2A, but the difference was in the position of gladiolus which did not show gross backward bend therefore the angulation was less prominent in comparison to the case 2A, giving an false impression of pectus carinatum type 1 deformity (chondro-gladiolar deformity). Child also show gross rib deformations like fused ribs (2<sup>nd</sup>, 3<sup>rd</sup> and 4<sup>th</sup>) with a count of only ten ribs. Both cases showed a classical pectus excavatum in the lower part of sternum [Fig 7, 8].

The reason these two cases were included was because of some key variation in their external presentation and structural malformation differing from classical chondro-manubrial deformity. Keys structural variations are detailed in table 2.

**Asymmetrical pectus carinatum:**

*Case 3A:* 14 year old male child visited the orthopaedic department with chief complaints of deformed spine, skin lesions and painful joints. There was no family history of spine deformities or joint disorders. Physical examination showed short stature child with deformed thoracic

wall, skin lesions with discolouration were noted [Fig 9]. Ribs showed bulge on either side of the sternum and the thoracic cage appeared to be stretched anteroposteriorly, more prominently on the left side. Joint stiffness was noted; kypho-scoliotic deformity was also seen. CT bone reconstructed images showed a forward protruding gladiolus with fused manubriosternal joint indicating the deformity as Type 1 Pectus carinatum [Fig 10].



Figure 7: CT plain sagittal images showing difference in the level of angulation. A – Case 2A shows acute angulation at 3<sup>rd</sup> costal cartilage. B – Case 2B shows lesser angulation at the same level.

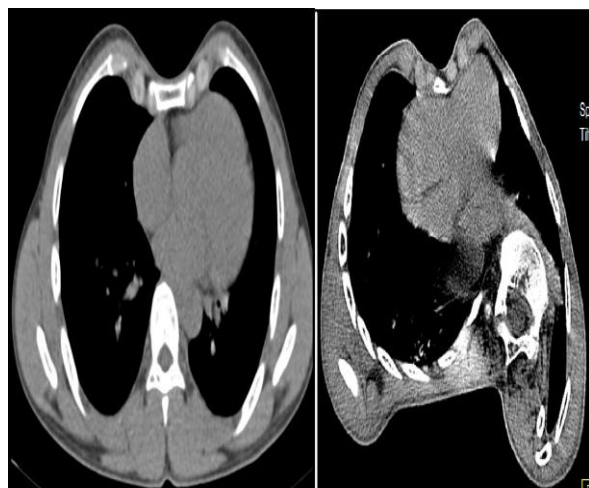


Figure 8: CT axial images at the lower end of sternum, showing reduced sterno-vertebral distances in both cases 2A and 2B mimicking pectus excavatum.



Figure 9: Photographic images of case 3A - 14 year old female child (A) and case 3B - 11 year old female child (B) with asymmetrical pectus carinatum type 1 deformity.

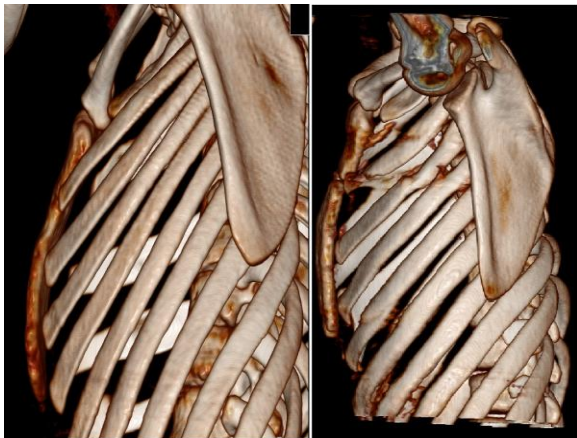


Figure 10: CT reconstructed images showing the difference in the alignment of manubrium and the body of sternum, also the inclination of ribs between two cases (cases 3A and B).

*Case 3B:* 11 year old child was brought to outpatient department of KVG Medical College and hospital with complaints of swelling on the right anterior chest wall and discomfort on prone lying. Child had no history of trauma or fall. Physical examination showed a protrusion on the right chest wall near the 3<sup>rd</sup> and 4<sup>th</sup> intercostal space [Fig 9]. Mild tenderness was experienced on palpation near the costochondral junctions especially near the 3<sup>rd</sup> and the 5<sup>th</sup> rib. His lung sounds were clear and no complaints of respiratory discomfort on deep inspired. Spine curvatures were normal. No skin lesions were found. Radiological examination was conduct to

rule out any chest wall tumours. CT 3D reconstructed images showed an asymmetrical pectus carinatum with tilted sternum and rib deformations [Fig 10].

In asymmetrical type of chest deformity, the external appearance is seen as an abnormal bulge on one side of the chest with or without a tilted sternum. The internal thoracic dimensions, especially the antero-posterior dimension of thorax are altered. Distance from sternal end of the rib to the vertebral end of the same rib may vary when compared to the opposite side. The causes may vary from overgrowth of costal cartilages, deformations of ribs, intra-thoracic masses and spine deformities like scoliosis. Present report we have included two such cases of asymmetrical pectus carinatum with a varying etiology to produce asymmetry of chest.

CT bone reconstructed, axial and sagittal images confirm the deformity as well as the asymmetry in both the cases [Fig 10, 11, 12]. It is common finding of pectus carinatum been associated with deformities of spine. kypho-scoliosis is a congenital abnormality affecting the spine. Altered curvature of the spine can affect the internal thoracic dimensions and the shape of the thoracic cage. [2] But the bifid rib producing an asymmetry is a rare possibility. Bifid rib or sternal bifidum is a congenital deformity with the sternal end of the rib cleft into two halves. Ribs deformations are usually associated with Gorlin goltz syndrome. It occurs in 1.2 % of the population and is usually unilateral. [8,9]

Anterior chest wall deformities are rarely reported in India probably because of fewer symptomatic cases encountered and most of them only been cosmetic disfigurement. Present report series could aid in providing better understanding of variations presented by the anterior chest wall deformities especially in Indian population and also enforces the need for

through external physical examination to be correlated to radiological changes in the skeletal components of the rib cage.

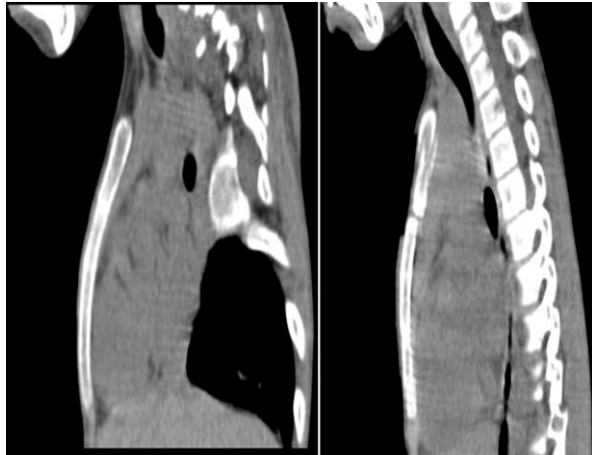


Figure 11: CT plain sagittal images showing difference in the shape of sternum.  
A – Case 3A shows incurving (convex anteriorly) sternum. B – Case 3B shows tilted sternum with mild protrusion.

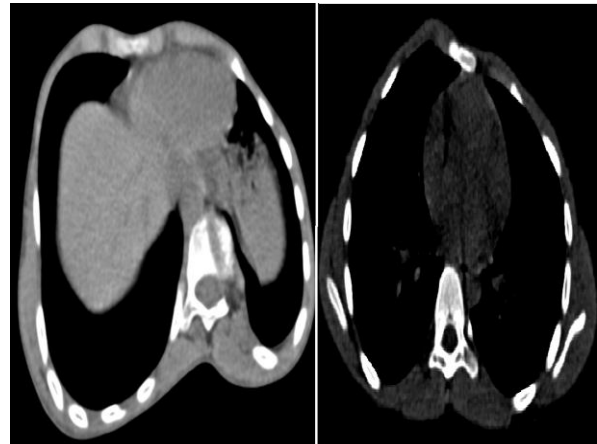
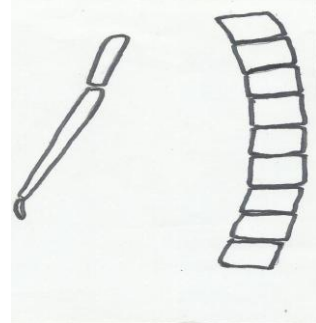
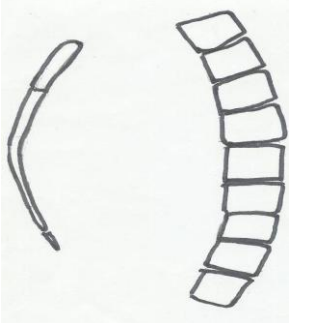


Figure 12: CT axial images at the lower end of sternum, showing increased sterno-vertebral distance on one side.  
A- Asymmetry due to deviation of thoracic spine to the left, B - due to bifid rib seen on one right side.

Table 1: Summary of keys structural changes in both cases of pectus carinatum type 1

Structures:	Case 1A (classical type 1 pectus carinatum)	Case 1B (variant)
Manubrium	Protruding and prominent	Protruding and prominent
Upper sternebrae	protrudes	More prominent protrusion
Lower sternebrae	Protruding forwards	Curves backwards
Xiphoid process	Produces a angulation	Joins without producing an angulation
Diagrams		

## CONCLUSION

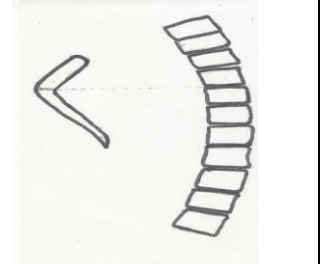
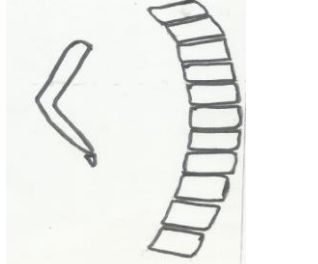

To conclude, present report series could provide not only an insight into the variants in presentation of each category of pectus deformity and also would aid in better understanding of external presentation of each category. Present series also emphasizes on the need for radiological examination, not only to identify the type but also to understand the morphological

changes in each type of pectus deformity so as to provide concrete evidence to evaluate post operative surgical prognosis.

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Table 2: Summary of key differences between classical and present cases of chondro-manubrial deformity.

	Classical	Case 3A	Case 3b
Level of angulation	At the level manubriosternal joint.	Below the manubriosternal joint. at the level of third costal cartilage	Below the manubriosternal joint. at the level of third costal cartilage
Degree of angulation	Acute angulation	Acute angulation	Less acute angulation
Sterno-vertebral distance at the lower end of sternum	Reduced	Reduced	Unable to asses.
Diagrams			

## REFERENCES

1. Willital GH, Saxena AK, Schu'tze U, Richter W. 2011. Chest deformities: a proposal for a classification. *World J Pediatr.* 7(2):118–123
2. Michael J. Goretsky, Robert E. Kelly, Jr, Daniel Croitoru, Donald Nuss. 2004. Chest wall anomalies: pectus excavatum and pectus carinatum. *Adolesc Med.* 15 (2004) 455–471.
3. Stephen j, Mathes, Alan E, Seyfer, Edward P, Miranda. *Congenital Anomalies of the Chest Wall*, chapter 134, pages 1-81
4. Robicsek F, Daugherty HK, Mullen DC, Harbold NB Jr, Hall DG, Jackson RD et al. 1974. Technical considerations in the surgical management of pectus excavatum and carinatum. *Ann Thorac Surg.* 18(6):549–564
5. Balasubrahmanyam C, Ashok V.1992. Currarino–Silverman syndrome (pectus carinatum type 2 deformity) and mitral valve disease. *Chest.* 3:102
6. Currarino G, Silverman FN. 1958. Premature obliteration of the sternal sutures and pigeon-breast deformity. *Radiology.* 70:532–540
7. Allwyn Joshua S, Lathika Shetty, Pare V, Roopa Sebastian. 2013. Chondro-manubrial deformity and bifid rib, rare variations seen in pectus carinatum: a radiological finding. *Surg Radiol Anat.* Jul; 35(5): 443-7
8. Glass R, Norton K, Mitre S, Kang E. 2002. Pediatric ribs: A spectrum of abnormalities. *RadioGraphics.* 22:87–104
9. Rai S, Gauba K. 2007. Jaw cyst-basal cell nevus-bifid rib syndrome: a case report. *J Indian Soc Pedod Prev Dent.* 25:137–139.

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