REVISTA BRASILEIRA DE MEDICINA DE FAMÍLIA E COMUNIDADE
RBMFC

www.rbmfc.org.br
ISSN 2197-7994

Clinical Case

# *Pectus carinatum* approach in Primary Health care and its impact on the emotional health of the patient: case report

## Abordagem de *pectus carinatum* na Atenção Primária e seu impacto na saúde emocional do paciente: relato de caso

## *Abordaje del Pectus Carinatum en Atención Primaria y su impacto en la salud emocional del paciente: caso clínico*

Luiza Carla Migliavacca Pian[1], Laura Vitória Xavier[1], Thais Caroline Fin[1], Juliana Frighetto[1], Hellany Karolliny Pinho Ribeiro[2]

[1]Universidade de Passo Fundo – Passo Fundo (RS), Brazil.
[2]Universidade Federal do Piauí – Teresina (PI), Brazil.

## Abstract

**Introduction:** Pectus carinatum (PC) is an unusual deformity of the chest wall that usually arises in childhood and becomes more pronounced in adolescence. This malformation can lead to aesthetic and self-esteem disorders, in addition to respiratory problems, due to the restriction of the chest wall. There are few Brazilian studies dedicated exclusively to PC, and most are directed at pectus excavatum or the investigation of both conditions jointly. **Presentation of the case:** In this case, a 13-year-old male sought medical attention in primary health care (PHC) because of the appearance of tumefaction in the parasternal region. He denied physical symptoms but reported embarrassment and aesthetic discomfort with the presence of the prominence. **Conclusions:** The multidisciplinary approach of thoracic deformity is fundamental in acceptance and treatment, with PHC playing an essential role, since all the team must assist and provide, through treatment, a better aesthetic, physical and emotional quality of life to the patient, and these factors led us to document this case.

**Keywords:** Primary health care; Thoracic wall; Pectus carinatum; Self-concept.

**Corresponding author:**
Luiza Carla Migliavacca Pian
E-mail: luizacarlapian@gmail.com
**Funding:**
No external funding.
**Ethical approval:**
CAAE 25126719.8.0000.5342
**Provenance:**
Not commissioned.
**Peer review:**
External.
Received: 07/08/2022.
Approved: 10/11/2022.
**Guest Editor:**
Leonardo Ferreira Fontenelle

**How to cite:** Pian LCM, Xavier LV, Fin TC, Frighetto J, Ribeiro HKP. Pectus carinatum approach in Primary Health care and its impact on the emotional health of the patient: case report. Rev Bras Med Fam Comunidade. 2022;17(44):3495. https://doi.org/10.5712/rbmfc17(44)3495

## Resumo

**Introdução:** O *pectus carinatum* (PC) é uma deformidade incomum da parede torácica, que geralmente surge na infância e se acentua na adolescência. Essa malformação pode levar a distúrbios estéticos e de autoestima, além de problemas respiratórios, que se devem à restrição da parede torácica. Existem poucos estudos brasileiros dedicados exclusivamente ao PC, e a maioria desses trabalhos direciona-se ao estudo do *pectus excavatum* (PE) ou de ambas as condições de forma conjunta. **Apresentação do caso:** Apresenta-se o caso de um paciente de 13 anos, masculino, que procura atendimento na Atenção Primária à Saúde (APS) por surgimento de tumefação em região paraesternal. Nega sintomas físicos, porém relata constrangimento e desconforto estético com a presença da proeminência. **Conclusões:** A abordagem multidisciplinar da deformidade torácica é fundamental na aceitação e no tratamento, tendo a APS um papel essencial Toda a equipe deve auxiliar e proporcionar, por meio do tratamento, melhor qualidade de vida estética, física e emocional ao paciente, fatores estes que nos impulsionaram a documentar este caso.


**Palavras-chave:** Atenção primária à saúde; Parede torácica; Pectus carinatum; Autoimagem.


## Resumen

**Introducción:** El pectus carinatum (PC) es una deformidad poco frecuente de la pared torácica, que suele surgir en la infancia y se acentúa en la adolescencia. Esta malformación puede provocar trastornos estéticos y de autoestima, así como problemas respiratorios, que se deben a la restricción de la pared torácica. Hay pocos estudios brasileños dedicados exclusivamente al CP, y la mayoría de los trabajos están dirigidos al estudio del Pectus Excavatum (PE) o al estudio de ambas condiciones conjuntamente. **Presentación del caso:** Presentamos el caso de un paciente masculino de 13 años que acude a Atención Primaria de Salud (APS) por la aparición de una tumefacción en la región paraesternal. Niega síntomas físicos, pero refiere malestar y vergüenza estética con la presencia de prominencia. **Conclusiones:** El abordaje multidisciplinar de la deformidad torácica es fundamental para su aceptación y tratamiento, teniendo la APS un papel fundamental, ya que todo equipo debe asistir y proporcionar, a través del tratamiento, una mejor calidad de vida estética, física y emocional al paciente, factores que nos han llevado a documentar este caso.


**Palabras clave:** Atención primaria de salud; Pared torácica; Pectus carinatum; Autoimagen.

# INTRODUCTION

Pectus carinatum (PC), whose classic presentation is known as “pigeon chest”, is a rare deformity of the chest wall, with an incidence of one in 1,500 individuals, and it predominantly affects males.[1] PC is a protrusion of the anterior chest wall, which appears or is accentuated during puberty, when the entire body is growing, and usually reaches its maximum at age 16 in girls and 18 in boys.[2]

It manifests itself in an oligosymptomatic way and with progressive growth in most cases, with demand for medical attention due to aesthetic and emotional implications. Symptoms are often the result of associated diseases or psychological disturbances, which lead these patients to be introverted and withdrawn.[3] Cardiorespiratory symptoms such as palpitations, dyspnea and wheezing may be present, related to reduced chest wall circumference due to decreased lateral dimension.[4]

Because it is approximately five times less prevalent than pectus excavatum (PE), it has not received the same degree of interest, and most repair reports combine patients with PC and PE, with little specific discussion of PC. Its approach is unknown by many professionals, which results in a lower number of patients referred for treatment.[2]

Thus, the scarcity of reports from Brazilian patients with this deformity, its implication in the physical and emotional well-being of affected individuals and its diagnosis and multidisciplinary follow-up still in primary health care were the motivation for this work. We present an approach to a case of PC in a male adolescent patient, who sought medical attention at the basic health unit (UBS) because of the appearance of the deformity and aesthetic embarrassment.

## CASE PRESENTATION

A 13-year-old male, Caucasian, attended the nursing screening of our UBS in August 2020 with a complaint of the appearance of a prominence in the parasternal region on the right for about ten days. He reported never having observed a previous deformity in the thoracic region and denied local pain or acute symptoms. On physical examination, the thoracic region was observed with accentuated curvature of the ribs in the right hemithorax, without skin changes or local fluctuation. The patient was referred for medical evaluation.

About ten days later, in the consultation with the family and community doctor (MFC) of the unit, he reported an intermittent rise in the local temperature, without other phlogistic signs on the skin and local fasciae. He denied being physically limited in walking, sleeping or other activities. He also denied previous comorbidities, continuous use of medications and a family history of similar pathologies. The main complaint reported by the patient regarding the prominence was the aesthetic discomfort/embarrassment with the presence of the chest protrusion, and the patient's mother was concerned about the situation. On physical examination, a swelling was observed in the parasternal region, with a hard consistency, fixed to the planes, without phlogistic signs, measuring approximately 7–10 cm (Figures 1 and 2). Laboratory tests and chest radiography were requested for investigation and diagnosis.

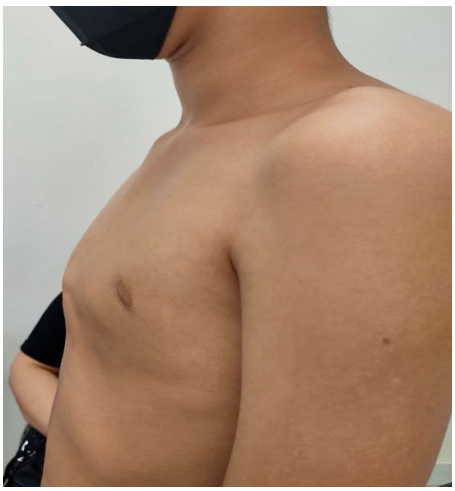

**Figure 1.** Lateral view of *pectus carinatum.*

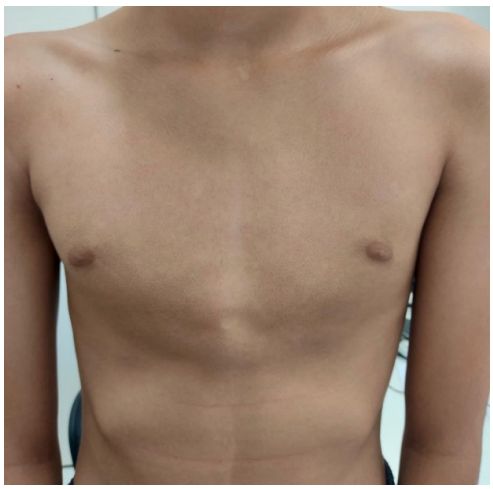

**Figure 2.** Anterior view of *pectus carinatum.*

During the follow-up of the patient at the UBS, it was found that, with the emergence of prominence and the resulting aesthetic embarrassment, there was a decline in his social life, especially in the school environment, in addition to the emergence of internal conflicts. This picture had an impact on his learning. Psychological follow-up based on humanistic psychotherapy with a Person-Centered Approach (PCA) was then started at the health unit, in 12 sessions with a clinical psychologist. There were not enough anxious or depressive symptoms for psychiatric referral. Nutritional monitoring was also started at the unit.

The patient's mother also used the psychological care service to expand her ability to provide emotional support to her child, given her condition as a single mother and complicated divorce with the father. This was because it was noted that the mother was self-critical about not having detected the

deformity in her son before and also about blaming the father for the genetic inheritance of PC. Conceptions of negative self-concept, such as "my bad luck, it had to happen to my son" were brought up spontaneously by her and managed according to the humanist basis.

About two months after the first medical visit, the patient reported increased prominence. Radiography did not show significant changes in the thoracic anatomy, but the patient was referred to the orthopedist for surgical correction of the deformity, due to aesthetic and emotional problems, although he denied related physical discomfort.

During this entire period, the patient was and continued to be monitored at the UBS by the multidisciplinary team, composed of a family doctor, nurse, psychologist and nutritionist, to maintain health care on the whole and maintain close monitoring of the patient.

The study was approved by the Ethics Committee, under approval No. 3.761.832 and Certificate of Presentation for Ethical Assessment (CAAE) No. 25126719.8.0000.5342, as recommended in Resolution 466/2012, and an informed consent form was signed by the person responsible for the patient in question.

## DISCUSSION

PC consists of a spectrum of anomalies that vary in anatomy and severity, with chondrogladiolar/pigeon chest prominence (projection of the middle and lower portions of the sternum) being the most common type, while chondromanubrial prominence (projection of the upper portion of the sternum, with protrusion of the manubrium) accounts for only 5% of cases.[2,4] Despite being considered rare (5–10% of all chest wall anomalies), it is the second most frequent malformation of the chest wall, and emerging data from chest wall anomalies have indicated that PC is at least as common as PE.[4]

Anatomical deformity of the PC and PE can be considered as a disproportionate growth of the costal cartilages compared to the rest of the bony thoracic skeleton, exerting pressure on the sternum to cause protrusion, depression, or a combination of both. It most often affects whites and Latinos, being uncommon in blacks and Asians, and most patients with PC and PE have an asthenic body habitus.[4,5]

At least 25% of patients with PC have another family member with a chest wall deformity, thus suspecting this deformity of having a genetic basis.[5] The monogenic syndromes most associated with PC and PE are Marfan syndrome, Noonan syndrome and Turner Syndrome, with the last being the main chromosomal one. No important genetic cause for non-syndromic PC or PE has yet been described, and it is possible that all or most of the familial cases represent a multifactorial inheritance.[6] The associated diseases most commonly found and responsible for respiratory symptoms in these patients are asthma and chronic bronchitis, which occur in 16.4% of cases.[2,7]

Although the onset of PC usually coincides with the beginning of the growth spurt in adolescence, the anomaly can be congenital and diagnosed in newborns and children under ten years of age (<10% of cases). However, as PC is rarely noticed at birth, its occurrence is believed to be more acquired than congenital.[2,4] In most children, the protrusion is smaller and remains stable during the first decade of life, and treatment is rarely indicated during this period, where only observation is considered.[4]

Treatment is mainly associated with a reduction in social disability, by improving body image, self-esteem and mental health.[3,4] Currently, treatment options for PC, in increasing order of invasiveness, include observation, orthoses, minimally invasive techniques and Ravitch's invasive surgical procedure.[4,7] The orthosis is a reliable non-surgical therapy, but using it for a prolonged period is not easy for an adolescent, with concerns about comfort, clothing and embarrassment.

On the other hand, open repair, despite being a more standardized surgical correction, progresses with the resulting chest scar.[1,4]

The presence of the deformity may reflect on the choice of clothing or poor body posture, in addition to restricting the practice of social activities and sports. Steinmann et al.[3] concluded, based on a case-control study carried out with a total of 90 patients, all male, 71 with PE and 19 with PC, that physical quality of life is more affected in PE patients because of the presence of symptoms. On the other hand, mental quality of life and social activities have a greater negative impact on patients with PC, who are less satisfied with their appearance than those with PE. Body image, however, is disturbed in all patients. Such data corroborate the situation of the case presented here, in which the patient did not complain of physical symptoms, but reported embarrassment regarding his body image, and the cognitive and emotional findings collected during the 12 psychotherapy sessions reinforced the risk that a person CPC has of developing a disturbed body image.

Quality of life, in turn, is correlates with family support.[8] Generally, parents are anxious, stating that the defect appeared a few weeks ago, attributing it to trauma or weight loss in the child or adolescent,[2] such as the concern of the patient's mother in the case presented above. The way in which this mother accepted her feelings regarding the unknown of her child's deformity during the psychotherapy sessions also allowed her to perceive herself to be more responsive and less demanding with him in the face of general adolescent situations: at the beginning of the sessions, she had complaints regarding the way the child eats, dresses and interacts, and at the end, her speech about this was acceptance of the child's way of being. The management of these issues also helps in the willingness to wait for the time it takes to carry out the surgical procedure in the Unified Health System. It is worth mentioning that, thanks to the humanistic basis centered on the person, there was the possibility for this mother to achieve greater confrontation with reality.

The therapeutic approach of our patient resulted from the communication between the different specialties present at the UBS, which allowed a joint approach from investigation to the referral for surgical correction with the orthopedic, as well as for psychological and nutritional follow-up. However, despite the joint and multidisciplinary work of the entire team, the process until the corrective surgery is carried out can be lengthy.

## CONCLUSION

On the basis of the case presented, we conclude that body image concerns may be even more relevant to the treatment decision-making process than physical restrictions, and systematic body image assessment is essential in the evaluation and correction of the deformity. Minimally invasive options are increasingly available for the therapeutic approach, with the appropriate selection being reviewed with the patient and their family. Primary care plays an essential role in the comfort and referral of these patients, from diagnosis to treatment options. Moreover, the scarcity of data on the psychological impact on patients with the malformation underscores the importance of further research on the approach and treatment of the deformity.

## CONFLICT OF INTERESTS

Nothing to declare.

## AUTHORS' CONTRIBUTIONS

LCMP: Conceptualization, Writing – original draft, Writing – review & editing, Investigation, Methodology, Software, Visualization. LVX: Conceptualization, Writing – first draft, Investigation, Methodology. TCF: Project administration, Formal analysis, Supervision, Validation. JF: Conceptualization, Data Curation, Resources. HKPR: Data Curation, Resources.

## REFERENCES

1. Yuksel M, Lacin T, Ermerak NO, Sirzai EY, Sayan B. Minimally invasive repair of pectus carinatum. Ann Thorac Surg 2018;105(3):915-23. https://doi.org/10.1016/j.athoracsur.2017.10.003
2. Coelho MS, Guimarães PSF. Pectus carinatum. J Bras Pneumol 2007;33(4):463-74. https://doi.org/10.1590/S1806-37132007000400017
3. Steinmann C, Krille S, Mueller A, Weber P, Reingruber B, Martin A. Pectus excavatum and pectus carinatum patients suffer from lower quality of life and impaired body image: a control group comparison of psychological characteristics prior to surgical correction. Eur J Cardiothorac Surg 2011;40(5):1138-45. https://doi.org/10.1016/j.ejcts.2011.02.019
4. Emil S. Current options for the treatment of pectus carinatum: when to brace and when to operate? Eur J Pediatr Surg 2018;28(4):347-54. http://doi.org/10.1055/s-0038-1667297
5. Fonkalsrud EW. Surgical correction of pectus carinatum: lessons learned from 260 patients. J Pediatr Surg 2008;43(7):1235-43. http://doi.org/10.1016/j.jpedsurg.2008.02.007
6. Cobben JM, Oostra RJ, van Dijk FS. Pectus excavatum and carinatum. European Journal of Medical Genetics 2014;57(8):414-7. http://doi.org/10.1016/j.ejmg.2014.04.017
7. Nuchtern JG, Mayer OH. Pectus carinatum. UpToDate [Internet] 2020 [accessed on: Aug 20. 2021]. Available at: https://www.uptodate.com/contents/pectus-carinatum
8. Bustamante MI. Busca de evidências de validade: escala para avaliação de tendência à agressividade [tese de doutorado]. Itatiba: Universidade São Francisco; 2014. Available at: https://www.usf.edu.br/galeria/getImage/427/12848697690782283.pdf