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Skeletal deformities in the form of pectus excavatum or scoliosis are unavoidable sequalae of large diaphragmmatic defects irrespective of the technique of repair used at the time of primary procedure. These sequelae lead to cosmetic and functional issues in the rapidly growing skeleton of a young child. The crux of management is adequate correction of these deformities with minimal residual functional impairment. A customized case-based approach using the modified Ravitch procedure is an excellent technique to repair these pectus defects. We describe such a procedure in a 3-year-old

girl. *Ann Pediatr Surg* 13:95–98 © 2017 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2017, 13:95-98

Keywords: congenital diaphragmmatic hernia, pectus excavatum, Ravitch procedure

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Received 11 June 2016 accepted 19 July 2016

#### Introduction

Congenital pectus excavatum is the most common chest anomaly of childhood. It affects one in every 400–1000 live births [1]. However, acquired or iatrogenic pectus excavatum has also been reported infrequently and is generally a recurrent form after a failed initial pectus excavatum repair or because of improper development of the chest wall following a repair for congenital diaphragmmatic hernia (CDH) in early childhood [2]. Most of the acquired varieties need one or more corrective procedures [3]. Here we present a case of iatrogenic unilateral pectus excavatum in a 3-year-old child, which was secondary to a CDH repair done in childhood. The deformity was corrected by the use of a modified Ravitch procedure.

## **Case history**

A 3-year-old girl presented to us with a known case of right CDH. The CDH repair was performed at one month of age in her home country. She was now brought by parents because of a deformed right-sided chest wall, which was progressively increasing in severity, and also for progressively increasing scoliosis. There were no other associated respiratory or cardiovascular symptoms. On clinical examination, there was an obvious pectus deformity noted in the right chest wall extending in the parasternal region from the fourth rib down to the eighth rib. The pectus was extending from the mid clavicular line up to the sternal margin on the right side (Fig. 1).

A computed tomography scan of the chest confirmed the clinical diagnosis and showed the abnormal development of the ribs and also the partial fusion of the costal cartilages of the fourth to eighth rib (Fig. 2). The diaphragm appeared to have grown well and did not show any residual defect.

A two-dimensional echo of the heart did not reveal any significant functional abnormality.

The patient was taken up for unilateral pectus excavatum repair. Given the iatrogenic nature of the problem and the unilaterality of the deformity, the decision to proceed with a modified Ravitch procedure was taken. The thorax was approached and opened from the previous thoracoabdominal incision. Numerous nonabsorbable sutures were found around the affected ribs. The sutures were excised. The dissection then proceeded both above and below the diaphragm in the extrapleural and extraperitoneal space, respectively. Once the diaphragmmatic integrity was confirmed, the costochondral junction of the fourth to eighth rib was disconnected using a bone cutter (Fig. 3).

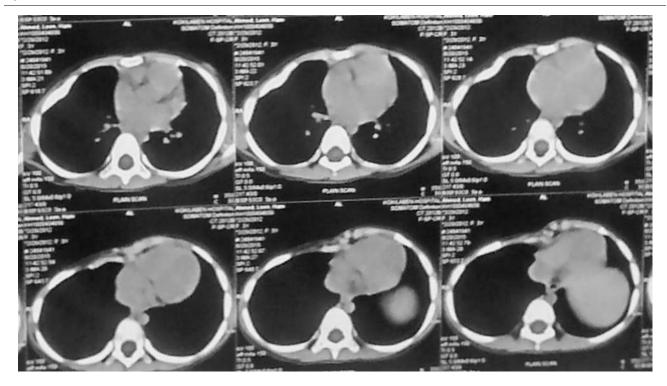
Once freed, the ribs were further opened up like a book using a chest spreader in the axis at right angles to the long axis of the ribs. This ensured that the contour of the affected ribs now matched the contour of the ribs above the defect. Once the contouring was done, the pectoralis muscle and the rectus abdominis muscle flaps, which were raised to gain access to the affected ribs, were now used to cover the gap between the ribs and the costal cartilages, thus ensuring an airtight closure.

Fig. 1



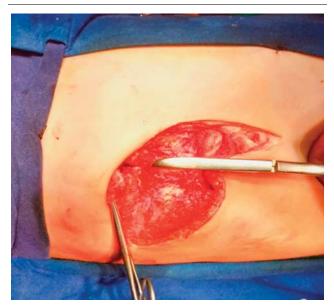
Lateral view of the pectus deformity.

Fig. 2



Preoperative computed tomography image showing the abnormal development of the ribs and also the partial fusion of the costal cartilages of the fourth to eighth rib.

Fig. 3



Costochondral disarticulation.

An intercostal drain was kept on the right side. The incision was closed in layers. The final appearance confirmed the correction of the pectus deformity (Fig. 4).

An epidural catheter was placed to ensure postoperative pain relief. The patient was extubated and shifted to the ICU. Postoperative recovery was uneventful. The International Classification of Diseases was removed on day 2

Fig. 4



Final appearance showing correction of pectus deformity.

and the patient was mobilized from day 3 and discharged

At 1-year follow-up, she has no complaints (Figs 5 and 6).

#### **Discussion**

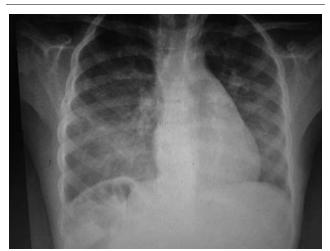
Iatrogenic pectus excavatum has been described as a consequence of overenthusiastic correction of the deformity during primary surgery and also as a sequel of diaphragmmatic hernia repair [3]. The severity of the defect has been correlated to the severity of the primary condition – that is, the size of the defect in the diaphragm at the time of primary surgery for diaphragmmatic hernia [4]. However, no convincing correlation has been found in the type of operative technique used for the primary closure. Regardless of whether the defect was closed primarily or with the

Fig. 5



Clinical photograph of the patient at 1-year follow-up.

Fig. 6



Chest radiography of the patient at 1-year follow-up.

use of a local muscle flap or with the use of a patch, the final outcome with respect to the presence and severity of the pectus deformity was unaffected [4-6].

On prolonged patient follow-up, the presence of abnormal anthropometric parameters such as weight and BMI along with the presence of skeletal abnormalities such as pectus and scoliosis points toward the fact that CDH not only affects normal lung growth but has wide-ranging implications on the overall growth and development of the child in later years. As mentioned previously, the skeletal deformities are more common in patients with large defects in the diaphragm, and no correlation has been found with the type of repair carried out [6]. Moreover, it is also hypothesized that tension at the time of repair may interfere with normal development of the thoracic cage and promote asymmetric chest wall development. The creation of more negative intrathoracic pressure because of the increased work of breathing in these patients may also promote retraction in the most compliant portion of the chest wall, thus contributing to the pectus deformity [5,6].

The persistence of these chest wall deformities leads to changes that are not just cosmetic. Differential growth of the chest wall consequent to abnormal growth and fusion of costal cartilages leads to progressive scoliosis and changes in the cardiac and lung function secondary to the scoliotic deformity [4–6].

Despite the apparent recognition of pectus deformity as an obvious sequel of CDH repair in early childhood, very little evidence is available in literature regarding correction of these deformities.

Contrary to the congenital pectus, the iatrogenic pectus is more of a focal and unilateral problem that needs novel customized approach to its treatment. The crux of the resolution relies on the correct anatomical interpretation of the anomaly.

Hence, it is axiomatic that a customized approach be chosen for each patient after adequate radiological imaging to map the deformity.

Nuss procedure as the treatment of choice for congenital pectus is unlikely to work in an iatrogenic pectus primarily because the defect is more often than not unilateral, and it affects the ribs and not the sternum.

It is our belief that a modified Ravitch procedure works best in such cases. The approach can be from the scar of the previous surgery and allows excellent access to the affected ribs. A costochondral disarticulation ensures that the ribs regain normal contour and allow normal growth in the future. As the disarticulation is limited to the affected ribs, conventional problems associated with Ravitch procedure such as acquired thoracic dystrophy are unlikely to occur.

# Conclusion

Acquired or iatrogenic pectus deformity is generally secondary to the repair of a large diaphragmmatic repair in early childhood. The basic pathology is the restricted growth of the affected ribs with costochondral fusion. A modified Ravitch procedure provides a viable option for repair of these deformities, ensuring good cosmetic and functional outcome.

# **Conflicts of interest**

There are no conflicts of interest.

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