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Repair of pectus excavatum in a toddler with Prune Belly syndrome and left bronchus compression

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ABSTRACT

A 2-year-old boy with prune-belly syndrome and severe pectus excavatum experienced recurrent pulmonary infections. A CT scan of the chest demonstrated compression of the left mainstem bronchus and leftward shift of the heart. The bronchial compression resulted in left upper lobe collapse and left lower lobe air-trapping requiring two hospitalizations for respiratory distress and pneumonia. The child underwent minimally invasive repair of his pectus excavatum and has not experienced any further pulmonary events. The pectus bar was removed 3 years post-operatively and at seven years following surgery he has a sustained repair.

1. Introduction

Prune-belly syndrome is a rare congenital disorder characterized by the triad of abdominal muscle deficiency, urinary tract abnormalities, and bilateral cryptorchidism. The first case was described in 1839 in a male infant with a “defect of lateral abdominal muscles,” undescended testicles, and concurrent pectus carinatum [1]. Associated comorbidities have been reported including pulmonary hypoplasia, pneumonia, urinary tract infections, musculoskeletal abnormalities, chest wall defects, connective tissue disease and scoliosis [2–5]. Lung hypoplasia in combination with pectus excavatum in patients with Prune Belly syndrome, has also been described [6]. One study reported successful surgical repair of pectus excavatum in 2 children with Prune-belly syndrome using the Ravitch repair [7]. In this report, we describe the successful repair of pectus excavatum in a child with Prune-belly syndrome using the Nuss procedure. Although repair was performed at an unusually young age, we felt this was necessary due to the patient's severe respiratory symptoms.

2. Case report

A 2-year-old boy with a history of prune-belly syndrome status post abdominoplasty with recurrent respiratory distress and pneumonia was referred for surgical evaluation of pectus excavatum deformity. In addition to his abdominoplasty, our patient had previously undergone bilateral orchiopexy for undescended testicles and bilateral ureteral reimplantations for vesicoureteral reflux. He had not experienced respiratory distress at birth but at two months of age he presented to an outside hospital with cough and hypoxia. During chest physiotherapy he became apneic and underwent rapid sequence intubation before being transferred to our institution. On admission there was collapse of the left lung thought to be caused by a mucous plug and Pulmozyme™ therapy was started. The infant was extubated on hospital day 1 and weaned to room air. He was discharged in stable condition with persistent left sided atelectasis and mediastinal shift to the left on chest X-ray. Ten months later the infant was again admitted for respiratory distress with RSV positive viral cultures (Fig. 1). Chest CT scan demonstrated pectus excavatum (Fig. 2) with left main stem bronchial compression (Fig. 3A–D) and left upper lobe collapse. He was treated successfully with Pulmozyme and chest physiotherapy after which he was discharged home. He required two additional hospitalizations for respiratory distress. Follow up CT examination at 24 months of age again demonstrated severe pectus excavatum

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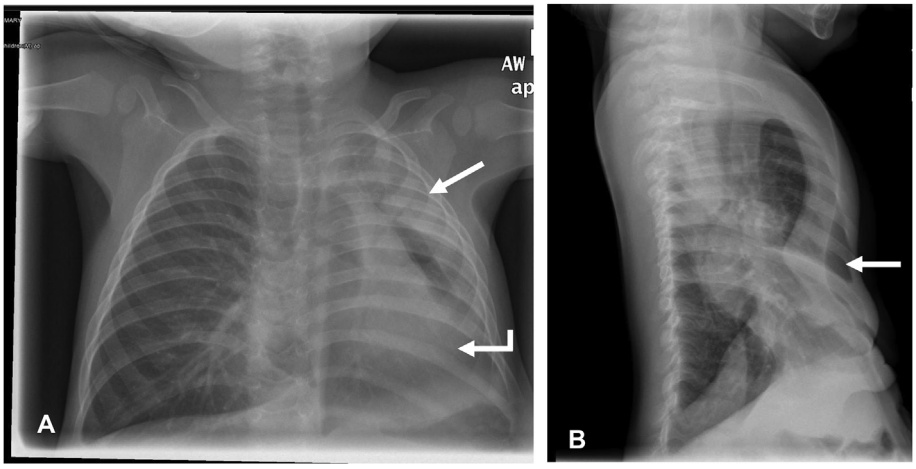

Fig. 1. Frontal chest radiograph at 1 year of age demonstrates collapse of the left upper lobe and lingula (straight arrow, A). Marked leftward mediastinal shift (angled arrow, A) is caused by a severe pectus excavatum deformity, as shown on the lateral image (arrow, B).

deformity with a Haller index of 10.1 (Fig. 4) and compression of the heart and left mainstem bronchus. The patient subsequently underwent minimally invasive Nuss repair of his pectus excavatum at 27 months of age (Fig. 5) and the bar was removed 3 years later (Fig. 6). Rigid bronchoscopy prior to the pectus repair demonstrated compression of the left main bronchus with inability to pass a 3.5 mm bronchoscope into the distal bronchus. Following repair, bronchomalacia was identified in the left main bronchus but the bronchoscope could be passed distally. At 4 years following removal of the bar, he has not experienced any respiratory events requiring hospitalization and has a sustained repair.

3. Discussion

Prune-belly syndrome is frequently associated with musculoskeletal abnormalities however, pectus excavatum deformities are less common [4]. In this case, the severe pectus excavatum deformity caused compression of the left mainstem bronchus resulting in numerous admissions for respiratory distress and left lung collapse. The current literature describes two cases of pectus excavatum with concomitant prune-belly syndrome. These defects were successfully repaired using the Ravitch procedure. We chose the Nuss procedure due to the child's young age and concern for future growth of the chest wall. Although repair of pectus excavatum is typically recommended around the time of the adolescent growth spurt (10–15 years of age), our patient's respiratory compromise required earlier intervention. The use of the Nuss technique has previously been described in a 4-year-old with tracheomalacia and multiple respiratory infections due to tracheal compression from the innominate artery and pectus excavatum with good results [8]. To our knowledge, this is the first report describing the successful use of the Nuss procedure in a toddler with prune-belly syndrome. Follow up is limited to 4 years after bar removal, however, our patient's physiologic and cosmetic results have been durable. While the patient is currently a preadolescent and remains at risk for recurrence during the adolescent growth spurt, this case illustrates the utility of the Nuss technique

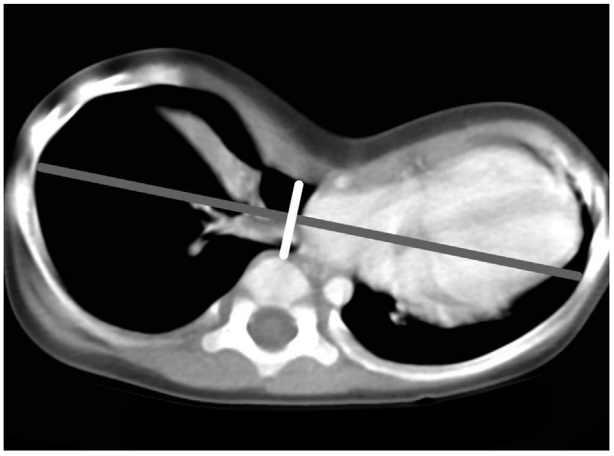

Fig. 2. Axial contrast-enhanced chest CT obtained at 12 months of age demonstrates severe pectus excavatum. The Haller index (8.5) is obtained by dividing the transverse diameter (grey line) of the chest by the AP diameter (white line) at the level of the deepest concavity on the patient's chest wall. Note how the sternum compresses the heart and deviates it toward the left.

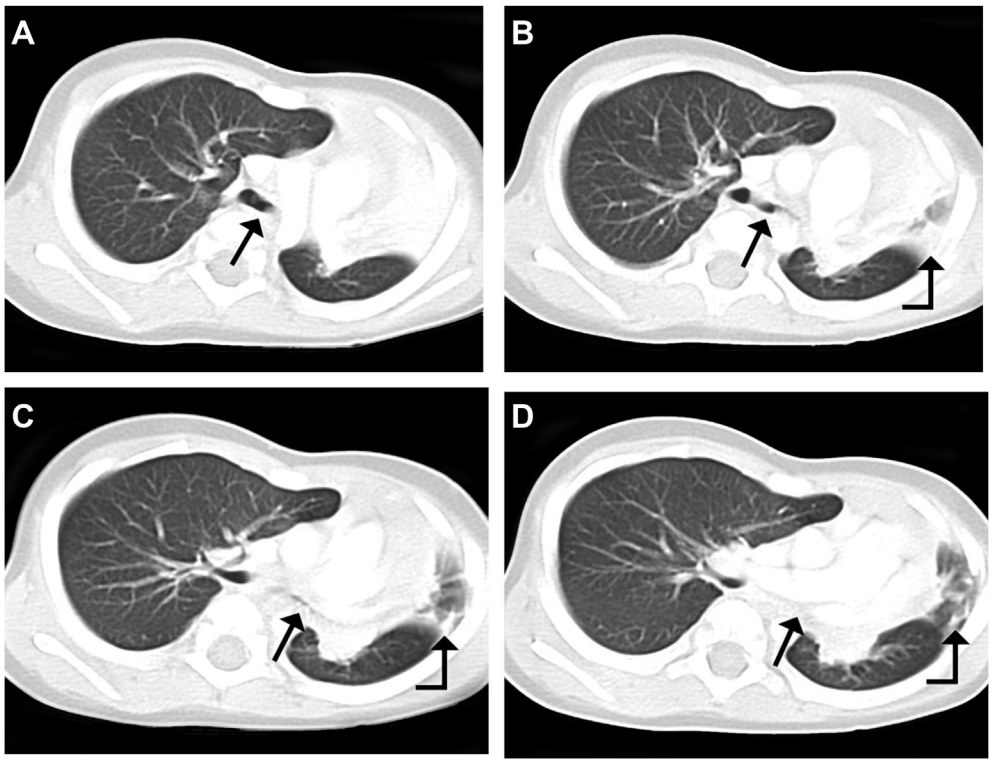

Fig. 3. A–D: Sequential lung-window images from the same study as Fig. 2 reveal severe narrowing of the left main bronchus (straight arrows, A–D). Volume-averaging artifact makes it appear that the bronchus is completed occluded, when in fact it is still partly open, allowing some aeration of the lower lobe, but near-complete atelectasis of the left upper lobe (angled arrow, B–D).

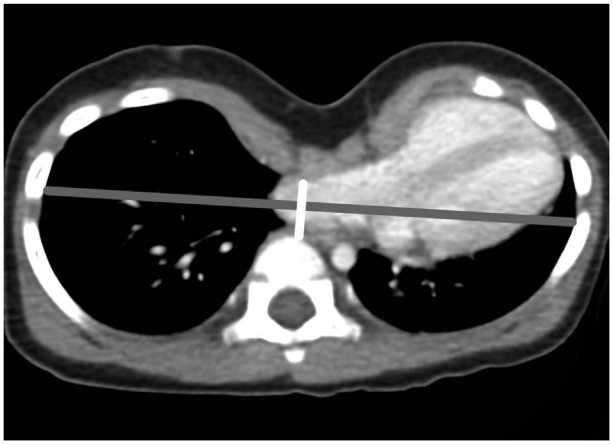

Fig. 4. A follow-up CT obtained at 24 months of age reveals interval worsening of the child's pectus deformity, with a Haller index (grey line/white line) of 10.1.

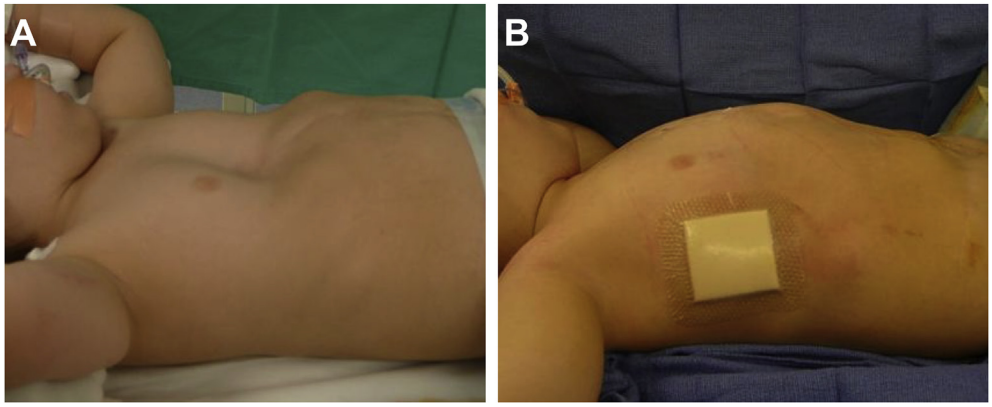

Fig. 5. Before (A) and after (B) placement of a single bar to correct the pectus deformity at 27 months of age.

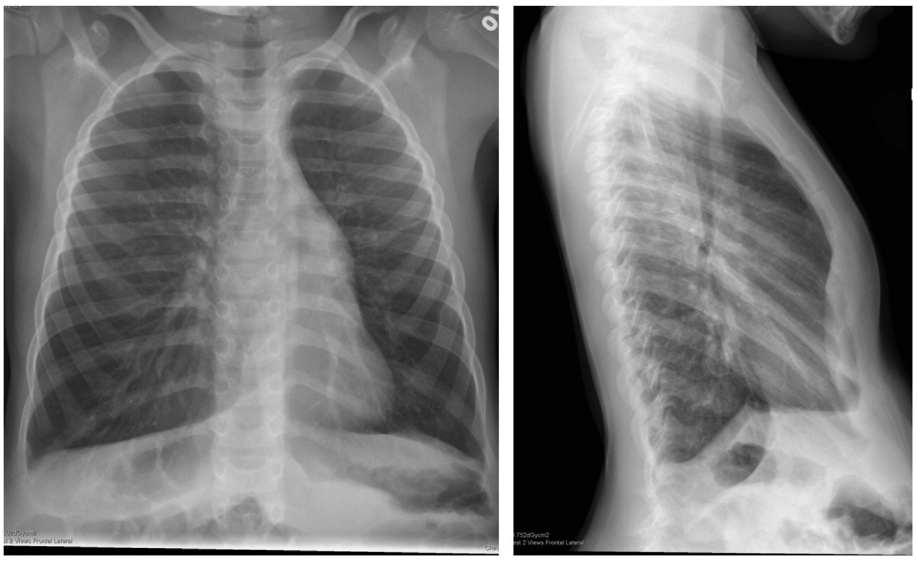

Fig. 6. Frontal and lateral chest radiograph one year following bar removal (6 years old) demonstrating a well expanded left lung with minimal residual mediastinal shift and correction of the pectus deformity.

for repair of pectus excavatum in a very young child with anatomical abnormalities causing severe respiratory compromise.

Author contribution

Shawn T. Liechty: literature review, chart review, manuscript creation.

Henry J. Baskin: imaging review, imaging interpretation, manuscript revision.

Rebecka Meyers: figures creation, manuscript revision.

Michael D. Rollins: literature review, chart review, figures creation, manuscript revision.

Evidence: Level V.

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