Distraction Osteogenesis of Multiple Ribs for the Treatment of Acquired Thoracic Dystrophy

Merisa L. Piper, MD, Lawrence Delrosario, MD, William Y. Hoffman, MD

Acquired thoracic dystrophy is a complication associated with early open repair of pectus excavatum resulting from extensive cartilage resection. The condition can cause serious functional and physiologic impairments, including cardiac compression and restrictive pulmonary function. We describe a 17-year-old boy with acquired thoracic dystrophy after Ravitch repair of pectus excavatum during infancy, whom we treated with distraction osteogenesis. The patient had a marked deformity of the chest wall and general hypoplasia of the central portion of the rib cage, with resultant symptomatic dyspnea on exertion and reduced pulmonary function. After osteotomies and distraction osteogenesis of bilateral ribs 4–8 using customized distraction devices, he had improved thoracic contour, resolution of dyspnea, and decreased restrictive pulmonary symptoms. This case suggests that distraction osteogenesis, already used extensively in craniomaxillofacial and orthopedic surgery, may be a novel method for management of this condition.

abstract

Acquired thoracic dystrophy is a complication associated with early open repair of pectus excavatum resulting from extensive cartilage resection. The condition can cause serious functional and physiologic impairments, including cardiac compression and restrictive pulmonary function. We describe a 17-year-old boy with acquired thoracic dystrophy after Ravitch repair of pectus excavatum during infancy, whom we treated with distraction osteogenesis. The patient had a marked deformity of the chest wall and general hypoplasia of the central portion of the rib cage, with resultant symptomatic dyspnea on exertion and reduced pulmonary function. After osteotomies and distraction osteogenesis of bilateral ribs 4–8 using customized distraction devices, he had improved thoracic contour, resolution of dyspnea, and decreased restrictive pulmonary symptoms. This case suggests that distraction osteogenesis, already used extensively in craniomaxillofacial and orthopedic surgery, may be a novel method for management of this condition.

Surgical correction of pectus excavatum has evolved to be less invasive and cause less morbidity.1,2 Nevertheless, it is still associated with a variety of complications. A hypoplastic, abnormally developed thorax has been described in patients after repair by the open Ravitch technique or other modified open repair techniques on young children.3–5 The proposed etiology of this thoracic dystrophy is decreased longitudinal rib growth resulting from extensive resection of rib cartilage and injury to the costochondral junction in young patients during repair.3,5 The disruption of the cartilage growth plate limits the growth and proper development of the chest wall. This results in decreased thoracic volume and may cause restricted pulmonary function and cardiac compression.3,5,6 The optimal treatment approach to increase thoracic volume by surgical intervention has been controversial and must accommodate chest wall expansion as children continue to grow.7–13

Distraction osteogenesis, first presented by Codivilla in 1905 and revisited by Ilizarov starting in the 1950s,14 describes the phenomenon in which separation and gentle traction of skeletal elements leads to metabolic activation and eventual bone formation between the separated skeletal elements.15,16 Distraction osteogenesis has been used in a wide variety of orthopedic and craniomaxillofacial applications, particularly after traumatic injury or from congenital deformities. Here, we present the case of a 17-year-old boy with symptomatic acquired thoracic dystrophy after repair of pectus excavatum who was successfully treated with distraction osteogenesis.
PATIENT DESCRIPTION

The patient initially presented to the plastic surgery service at University of California, San Francisco, at age 11 years for consultation for arrested sternal growth after a Ravitch repair of pectus excavatum at age 2. This deficient growth led to inadequate bony protection of the heart and upper abdominal viscera. Because a short sternum is not typical with this condition, we speculate that it is related to the early age of his initial surgery and presumed devascularization of the sternum. He subsequently underwent methyl methacrylate chest wall reconstruction with rectus abdominis muscle coverage to address the hypoplastic sternum and complex chest wall deformity. However, he returned after a hiatus of several years with a marked deformity of his chest wall and general hypoplasia of the central portion of the ribcage (Fig 1). He had symptomatic dyspnea on exertion, was unable to participate in sports, which negatively impacted his self-esteem, and was self-conscious of his appearance. Pulmonary function tests revealed a 50% reduction in vital capacity and forced expiratory volume. After extensive discussions with our pediatric pulmonology and pediatric surgery colleagues, we decided to pursue bilateral distraction osteogenesis of ribs 4–8 when the patient was 17 years old to increase his overall thoracic volume.

OPERATIVE TECHNIQUE

After the patient was positioned supine with arms extended, lazy-S incisions were made bilaterally over the anterior axillary line. The serratus anterior was divided in 3 locations to permit access to the ribs of interest. A small incision was made at the lower border of each rib to dissect a small periosteal pocket, permitting us to divide the rib while maintaining the periosteum as a sleeve to promote osteogenesis. Osteotomies of bilateral ribs 4–8 were performed by using a reciprocating saw.

The 10 rib distractors used were custom manufactured by KLS Martin (Jacksonville, FL), based on a design originally used for placement on metacarpals. They were chosen over midface distractors because of their increased rigidity; the custom manufacturing extended their length to 3 cm. The distraction device was placed across the osteotomy gap in a supraperiosteal position, using 4 screws (2-mm diameter, 11-mm total length) on each side. The activating rods were brought out anteriorly through small separate skin incisions. All distractors were placed in a similar fashion with the exception of the fourth rib on the patient’s right side. The fourth rib was located slightly inferior to the third rib and pectoralis muscle, and consequently the activating rod would not reach the skin. An osteotomy was performed on the right fourth rib but no distractor was placed, with the expectation that it would follow the distraction of the other four ribs on that side. After excellent fixation of all devices was obtained, muscle and soft tissue were closed in a layered fashion (Fig 2).

After a latency period of 5 days, distraction began at a frequency of twice a day, for 1 mm of total daily lengthening for each of the 9 distractors. The patient was discharged on postoperative day 9, and the family continued distraction at home until the total distraction distance of 3 cm was achieved. Two
months after discharge, the patient returned to the operating room for removal of the activator rods to minimize the risk of ascending infection. A computed tomography scan was obtained before removal to evaluate the ossification and distraction length (Fig 3). Seven months later, the distraction devices were removed during a third operation once computed tomography documented sufficient bone growth (Fig 4).

**OUTCOME**

One month after the activator rods were removed, the patient underwent pulmonary function analysis (Table 1). Compared with levels from 9 months preoperatively, his forced vital capacity increased from 53% to 67% of predicted, forced expiratory volume increased from 55% to 63% of predicted, vital capacity increased from 50% to 67% of predicted, and total lung capacity increased from 70% to 80% of predicted. At his most recent follow-up visit 4 years later, although he remains asymptomatic without dyspnea, his forced vital capacity was 57%, forced expiratory volume was 59%, vital capacity was 55% and total lung capacity was 61%. All measurements were still higher than preoperative values, except for the total lung capacity, but to a lesser degree.

We calculated lung volumes pre- and postoperatively based on computed tomography. Before expansion, the left lung volume was 2.44 L and the right was 2.92 L (5.36 L total). Six months after removal of the distractor devices, the left lung volume was 2.72 L and the right was 3.48 L (6.20 L total). This reflects a 16% increase in overall lung volume.

The patient had no early or late postoperative complications. He was initially seen monthly, and now is seen on an annual basis. At his most recent postoperative visit, the patient still had a mild hourglass deformity of the chest wall. This is exaggerated by the abrupt end of the pectoralis muscles (Fig 5). However, the patient reported no restrictive pulmonary symptoms, such as exertional dyspnea or cough. He rides a bike without shortness of breath and has no pain. Overall, he is pleased with his functional and aesthetic outcomes.

**DISCUSSION**

Acquired thoracic deformity can be a significant cause of morbidity after early open repair of pectus excavatum, resulting in cardiac compression and restrictive pulmonary function. In this case...
PIPER et al report, we present a novel method for managing this complication using distraction osteogenesis, a technique often used in craniomaxillofacial and orthopedic surgery. The particularly challenging aspect of this case was to attempt distraction with the constant respiratory movement of the ribs. The distractors proved capable of maintaining fixation under these conditions and allowing for new bone formation. The success of distraction osteogenesis in restructuring the thoracic cavity acts as a proof of principle that this technique may be used in a broader scope to treat other acquired and congenital thoracic conditions.

However, as with all surgical procedures, this procedure has drawbacks. First, it requires 3 separate surgeries: distractor placement, activator rod removal, and distractor removal. This means 3 episodes of general anesthesia, potential hospitalizations, and periods of postoperative recovery. Second, this technique is painful during the distraction process, as well as after the 3 surgeries. Third, the distractor devices are expensive, ∼$5000 per piece, although we did receive a bulk discount. Finally, there is bilateral scarring, which may be unsightly. However, the alternative surgical options also have drawbacks. Sternal split with rib graft placement improves pulmonary function, but leaves the intrathoracic organs vulnerable to injury and does not adequately address cosmetic concerns.13 A vertical expandable prosthetic titanium rib requires a large incision, has minimal benefit in children older than 8 years, and can cause significant tension on the soft tissues, leading to erosion.7 Risks of anterior chest wall reconstruction with release and elevation of the sternum include supporting retrosternal bar migration, migration and exposure of the rib grafts and sternal wires, and difficulty in

<table>
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<th>Test</th>
<th>Preoperative, %</th>
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<tr>
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<td>67</td>
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<td>Forced expiratory volume</td>
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<tr>
<td>Vital capacity</td>
<td>50</td>
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<tr>
<td>Total lung capacity</td>
<td>70</td>
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Values are expressed as age-adjusted percentages of predicted values.

FIGURE 4
Three-dimensional reconstruction of patient’s thoracic cavity 6 months after removal of distractor devices, demonstrating adequate ossification of ribs 4–8 bilaterally. Upper left, anterior view of chest wall. Upper right, coronal view of chest wall. Bottom left, left lateral view of chest wall. Bottom right, right lateral view of chest wall.

FIGURE 5
A, Anterior view of the patient’s chest wall before distraction osteogenesis. B, Anterior view of the patient’s chest wall 4 years after distraction osteogenesis completed; note the abrupt end of the pectoralis muscles. C, Appearance of scarring 4 years after distraction osteogenesis completed; lazy-S scar from rib access and punctate scars from distractor rods.
controlling hemorrhage as the sternum is separated from scar tissue.17

Despite the drawbacks, our patient had substantial functional and aesthetic improvement, as evidenced by higher levels in his pulmonary function tests, increased lung volumes, and greater self-esteem. His pulmonary function never normalized, however, and in fact declined over time after initial postoperative improvement. Although this was suboptimal, he remains asymptomatic. The decline may indicate additional growth of the patient without commensurate growth of the chest wall. Therefore, there may be a theoretical benefit to performing this operation when the patient is younger, so that the ribs and lungs may continue to grow together. We believe this outcome could also be improved by increasing the length of distraction, and propose to do this in future cases or even to perform distraction at 2 points in the rib (for example, anterior and posterior axillary lines). In addition, a device that follows the curvature of the ribs could improve both the aesthetic, as well as the functional, outcome. Overall, the symmetry of this patient's chest wall has remained stable over 4 years despite continued growth and skeletal maturity.

Distraction osteogenesis may provide a safe and effective alternative for surgical management of acquired thoracic dystrophy.

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REFERENCES


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