Lateral Thoracic Expansion for Jeune Syndrome: Evidence of Rib Healing and New Bone Formation

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Background. Lateral thoracic expansion is a procedure that has been described to enlarge the thoracic cage in patients with Jeune's asphyxiating thoracic dystrophy. The procedure involves separating ribs from their periosteum and platting them in an expanded fashion with titanium struts. We have speculated that the ribs heal in this situation, despite the absence of surrounding periosteum, and that new rib formation occurs in the liberated periosteum.

Methods. Radiographic studies of patients who have undergone lateral thoracic expansion were reviewed for evidence of rib healing and periosteal new bone formation.

Results. This study presents radiologic evidence that rib healing actually occurs, as does periosteal ossification.

Conclusions. Lateral thoracic expansion creates additional chest wall that is formed of autologous tissue, fully healed, and not ultimately dependent on titanium struts.

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Radiographic signs of bone healing in children have been described in detail in the literature both clinically [1] and experimentally [2]. These include periosteal reaction, callus formation, bridging, and remodeling. Specifics of the location of these signs in ribs have been explored in the setting of trauma [3]. The importance of the proximity of the periosteum has been suggested [4]. In 1965 we reported using an operation called lateral thoracic expansion (LTE) for the treatment of Jeune syndrome or asphyxiating thoracic dystrophy [5]. With time we have modified the procedure and reported midterm results and technical modifications [6]. The essence of the LTE procedure involves the separation of a segment of six ribs from their underlying periosteum followed by staggered osteotomies. The periosteum and adjacent intercostal muscle bed is divided in the opposite direction. The segment is spread apart and the expansion maintained by bringing together the long segments of stainless steel plates. Healing of the newly created, longer ribs has been anticipated. Periosteal bridges are pulled together to fill in the gap to create the new bone formation. Healing has been confirmed on two previously undocumented events: the healing of rib in a child in the absence of surrounding periosteum, and periosteal new bone formation in Jeune syndrome in the absence of rib tissue.

In most instances, we have performed the LTE bilaterally as two independent procedures separated by 6 months to a year. Before the second stage, we perform a complete evaluation of the patient, which includes plain chest radiographs as well as computed tomographic scans of the chest. Therefore, we have the opportunity to evaluate the status of the first-stage procedure done many months prior. The purpose of this study is to describe the radiographic evidence of rib healing and periosteal new bone formation in this situation.

Patients and Methods

We reviewed our series of patients to identify those who had radiologic evaluation at any period of time after an LTE procedure on one side. Sixteen sites in 6 patients were available for review regarding rib healing and periosteal new bone formation from 3 weeks to 2 years after the procedure. Plain chest radiographs as well as computed tomographic scans were evaluated by two board-certified pediatric radiologists. Particular attention was paid to the previously operated area for evidence of rib healing and periosteal new bone formation by comparing immediate post-stage one radiographs with pre-stage two radiographs of the same area. Bone was considered healed if the plated sites (in the absence of periosteum) if there was evidence of cortical and cancellous bone bridging the two bone ends held together by the plate. Periosteal new bone formation was confirmed by the underlying new cortical and cancellous bone bridging the surgically created space where the periosteum was placed, and immediate postoperative films had shown no bone.

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Fig 1. Axial 1-mm-thick image in a 16-month-old girl after bilateral chest expansion. Arrow indicates complete fusion of bone ends at the site of the titanium struts.

Results

All evaluated areas confirmed healing of bone at the plated site or showed evidence of new bone formation in the periosteal beds. Figure 1 is a coronal view of the site of plating of the long segments of two adjacent ribs, thus creating a single elongated rib 5 months previous to this study. The arrow points out the bone healing bridging the previous gap. This area had no periosteum adjacent to the bone to support the healing. Figure 2 is a detail from a three-dimensional computed tomographic scan 5 months after an LTE procedure. ribs 6 and 8 are the ribs where the staggered osteotomy occurred posteriorly. The gap created laterally had been filled with periosteum, and the arrows point to new bone formation in that area. This gives the appearance of synostosis.

Figure 3 gives information about the time frame of the new bone formation. This particular patient was the only one who had loss of expansion. He returned 15 weeks after a left side LTE with fracture of the struts and collapse of the repair. The smaller struts were replaced with larger ones that we now use on all of our patients. Because of this extra step we have multiple radiographs for a 3-week period from the original procedure. Figure 3A is an early postoperative picture showing the short end of divided ribs after the first left-sided expansion. Unseen radiographically, as expected, is the periosteum that continues laterally and is tethered to the expanded rib below. Also of note is the fracture in the upper strut. Figure 3B shows the same patient after reexpansion with larger struts, now 3 weeks after the first expansion. This image demonstrates new bone already present in the area of the periosteum.

Comment

These findings indicate that nont healing and new bone formation occur in the setting of the LTE procedure.

Fig 2. Coronal reformatted maximal intensity projection image obtained from helical computed tomographic study in the same patient as in Figure 1. Arrows indicate the completed new bone formation in the area of periosteum attached laterally in the short posterior ends of ribs 6 and 8.
Periosteal new bone formation is evident as early as 3 weeks after the procedure. Several implications and considerations follow from this observation.

The first is that the creation of a new and expanded area on the lateral aspect of the thorax would be expected to be stable in the long term. Furthermore, as the repair is based on autogenous tissue that has healed, there is no concern about the long-term fate of the titanium plates, which stabilize the repair in the short term during healing. This explains why we have had late strut fractures that seemed to have no clinical consequence to the operation. The only problem with late strut fractures we have observed is local irritation of the back of the skin, which resolved with strut removal. Throughout the follow-up period we have not observed any anunion or pathologic fractures.

There are also implications regarding ventilatory mechanics. Both the nature of the procedure itself, which detaches and reattaches entire areas of intercostal musculature, and the radiographic appearance of synostosis raise the question of the effect of LTE on regional chest wall motion. It is our assumption that the involved region of chest wall, while expanded in volume, is frozen mechanically and contributes little motion to the respiratory effort. Jeune syndrome has characteristically small, short, horizontally oriented flat ribs, suggesting that most ventilation is by means of diaphragmatic contraction and not thoracic wall motion. This is compatible with our clinical observations in these patients. For this reason, the beneficial effects of increased thoracic volume outweigh any loss of chest wall motion, because that was minimal to begin with. This would, in part, explain the positive clinical findings we have experienced with the LTE. As an aside, it is worth noting that despite the appearance of synostosis created by the periosteal new bone formation (Fig 5), no evidence of suckosis has been observed in follow-up.

The final consideration is growth potential. The LTE procedure is carried out in the midportion of the ribs, and therefore the area of the growth plates is not affected. This allows overall growth of the hemithorax to continue. We have noted that growth occurs in some of the patients with milder forms of the Jeune syndrome whom we have followed without operation through the years. These individuals continue to grow, albeit at a slow rate and to a diminutive end point, but their respiratory capacity seems to keep up with their somatic demands. To date, no patient has required reoperation for recurrent respiratory insufficiency. Once the patients have made it through the first few years, with good nutrition they can continue to grow and have a good quality of life. Our first patient, operated on at 4 years of age for repeated and severe respiratory episodes, is now 13 years old and doing well without further intervention. Others who came to us dependent on ventilators are now not dependent on ventilators and continuing without the need for further intervention.

In conclusion, the LTE procedure results in expansion of the chest wall by actual healing of the ribs in the expanded area and ossification of the periosteal bed liberated from the rib. This newly created area of chest
wall expands the thoracic volume without being ultimately dependent on the titanium struts that maintain the expansion during healing.

References