Lateral Thoracic Expansion for Jeune's Asphyxiating Dystrophy: A New Approach

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A new procedure is described for thoracic expansion in Jeune's asphyxiating dystrophy. The chest wall is enlarged by division of ribs and underlying tissue in a staggered fashion so that either rib or periosteum covers the lung. New bone formation has been demonstrated so that a viable enlargement has been obtained. The clinical result is excellent to date.


Asphyxiating thoracic dystrophy, also known as Jeune's syndrome, is a rare autosomal recessive malformation characterized by a disturbance of endochondral bone formation in utero. Jeune's syndrome exists clinically as a triad composed of small thoracic

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cage, pelvic, and phalangeal deformities. Histologically this syndrome demonstrates the features of a chondrodyplasia [1, 2]. Alveolar hypoventilation is the most common and important clinical feature of this syndrome related to impairment of chest expansion by the short, horizontally positioned ribs. Pulmonary hypoplasia, recurrent pneumonia, and pulmonary hypertension result in significant morbidity and ultimate mortality in the severe form of this syndrome. Previously, pathologic studies have suggested that lung alveolar growth potential was normal in children with asphyxiating thoracic dystrophy, with pulmonary hypertension and subsequent arterial vessel wall thickening related (or secondary) to the severity of the chest wall hypoplasia [2].

If lung growth potential is normal, surgical expansion of the rib cage should allow further growth of the lung and partial resolution or stabilization of pulmonary hypoplasia and associated clinical problems. This hypothesis has led to the development of several surgical techniques for chest wall expansion, including resection of costochondral cartilage and procedures involving splitting the sternum, with interposition rib grafts or prosthetic material used to maintain expansion and protect the anterior mediastinum [3, 4].

In the present report, we describe a new technique for lateral rib cage expansion using staggered subperiosteal rib osteotomies and rigid titanium miniplate augmentation and stabilization with primary skin closure. This method allows rib cage expansion of approximately 3 cm, which is comparable with midline sternotomy splitting procedures, yet retains normal anatomic protection of the anterior mediastinum. Follow-up at 10 months demonstrates early rib formation in areas of subperiosteal rib advancement, and substantial clinical improvement of respiratory status.

A 4-year-old boy was diagnosed with achondroplasia at birth. His medical history included hydrocephalus treated with a ventriculoperitoneal shunt and multiple hospital admissions for apnea, aspiration, and recurrent pulmonary infections. The additional diagnosis of Jeune's syndrome became apparent with time. Respiratory insufficiency was chronic and progressive. Preoperatively he received continuous positive airway pressure via a tracheostomy at night and gentamicin aerosols for tracheitis prophylaxis.

Physical examination at the time of operation demonstrated a chest circumference of 41 cm at the inferior edge of the areola bilaterally, with significant rib contour depressions in the anterolateral areas of the rib cage. Pulmonary examination demonstrated diffuse rhonchi and decreased lung volume. His limbs were markedly shortened, but phalangeal abnormalities were not evident. Preoperative ventilation/perfusion scan demonstrated normal distributions despite the small thoracic size bilaterally. Unfortunately, preoperative pulmonary function studies could not be performed because of technical difficulties. His tracheostomy had a significant air leak, which could not be quantified, and this would invalidate the results of any volumetric or flow studies.

A transverse incision was made over the lateral right rib cage with extensive skin flaps raised. Ribs four through nine were exposed in the area of greatest depression. Using a periosteal elevator, the ribs were separated from the surrounding tissues including peristium, intercostal muscles, and parietal pleura. Ribs five through eight were divided at alternating levels, with one rib divided anteriorly and the next divided 4 to 5 cm posteriorly. The underlying peristium, muscle, and pleura were then divided in an opposite fashion, with the result being a staggered arrangement of rib and soft tissue when expansion was complete, allowing continuous coverage of underlying lung tissue. Ribs four and nine were simply divided to allow for chest wall expansion. Titanium miniplate fixation was used to maintain expansion (Fig 1). Approximately 3 cm of expansion was easily obtained. A single chest tube was placed and the skin was closed primarily.

The patient was weaned from the ventilator shortly after operation, and his chest tube was removed on the fourth postoperative day. His respiratory status improved with decreasing respiratory rate and reduced oxygen requirements, and he was successfully weaned from continuous positive airway pressure. Throughout the year after operation these improvements persisted and his activity level improved. Postoperative radiographs obtained at 10 months demonstrate rib formation in areas of subperiosteal rib advancement (Fig 2). He recently electively underwent a second procedure on the opposite side, with a similar hospital course and preliminary results.
Comment

This technique allows safe lateral rib cage expansion in children with severe forms of Jeune’s asphyxiating dystrophy. In contrast to techniques involving a sternotomy and mediastinal fixation with struts, lateral rib cage expansion maintains anatomic protection of the anterior mediastinal structures, and subperiosteal rib osteotomies allow regrowth of bone to stabilize the rib expansion. This procedure also has theoretical advantages over simple removal of costochondral cartilage because it actually enlarges the volume of the thoracic cage by making a new area with osseous support. The future growth potential of this area remains speculative.

Although still relatively short, early follow-up demonstrates dramatic clinical improvement in this patient’s quality of life. Longer follow-up and experience with additional patients will be required before we establish the ultimate place and optimal timing for this procedure in the treatment of Jeune’s syndrome.

References


INVITED COMMENTARY

Jeune’s syndrome is a complex malformation. The principal feature is a small, narrow chest with shortened, broad ribs and abnormal costal cartilages. The fourth through the ninth ribs bend inward, usually at the costal chondral junction. The pleural cavity is greatly reduced in size. Transverse computed tomographic scan cuts of the chest reveals thin, crescent-shaped pleural cavities with very little room for lung expansion. A serious associated abnormality involves the kidneys. Many children have bilateral microcystic disease and gradual development of renal failure.

In my experience there are actually two forms of Jeune’s syndrome. In one form, fatal during infancy, there is respiratory distress immediately after birth. Some infants require immediate mechanical ventilation and cannot be weaned. Others have rapid development of progressive respiratory failure and after bouts of recurrent pneumonia require intubation and ventilatory support. Without some kind of operative intervention these babies die of respiratory failure as their lungs become increasingly damaged by long-term ventilation.

A second group of patients have almost the identical chest configuration but have only moderate respiratory symptoms at birth and can be managed for years without mechanical ventilation. I believe that operation is indicated in the first group of patients because failure to intervene will result in progressive pulmonary damage and eventually death. I am not convinced that operation benefits the group that do not require ventilation.

In the operative approach introduced by Davis and associates the patient is 4 years old and has not required mechanical ventilation, just continuous positive airway pressure at night. It is difficult to assess the severity of the pulmonary compromise because no pulmonary function tests were done. I find it very helpful to evaluate patients with Jeune’s syndrome with preoperative and postoperative computed tomographic scans. These show the actual size of the pleural cavities and the changes with operation. Serial pulmonary function tests are equally important.

Nevertheless, the new technique may have some merit. First, it avoids splitting of the sternum. Although sternal split does increase the size of the thoracic cavity, fixation has been difficult and over the years I have seen the sternal fixation break down. The growth of bone in the rib technique appears to be a positive factor, although long-term follow-up will be necessary to see if growth continues and whether the plates limit growth. In my experience the lung is restricted not only by the chest wall but also by the pleura. This procedure does appear to divide the pleura and allow for expansion. This technique may also be useful in children who have severe scoliosis with unilateral chest wall compression.

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