

# Chest Wall Surgery for Asphyxiating Thoracic Surgery

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## BACKGROUND

Asphyxiating thoracic dystrophy (ATD), also known as Jeune Syndrome is an autosomal recessive disease involving deformities of the thorax, pelvis, and phalanges with varying degrees of renal dysfunction. Estimates suggest that approximately 40 patients per year are born in the United States and over 600 worldwide.

## ANATOMY AND PATHOPHYSIOLOGY

While the small, bell shaped thorax is the most striking finding in these patients, there are other factors contributing to respiratory insufficiency. The ribs are short and tend to be horizontally oriented. This causes a basically rigid thoracic wall because the external intercostal muscles do not have the ability to move the ribs anteriorly and laterally during normal inspiration as they do in the normal thorax. Thus, breathing is almost entirely diaphragmatic. There are also typical changes that occur in the pulmonary parenchyma. The changes are characterized on computerized tomography (CT) by bronchial crowding associated with atelectasis causing streaky opacities in various lobes. It is not certain whether the pulmonary changes are secondary to constriction of otherwise normal lungs or an intrinsic part of the pathology of ATD. Previous pathologic studies have suggested that changes are secondary to the chest wall pathology (1). Anecdotally, we have noted progression on CT scan that would support the hypothesis that the parenchymal changes are secondary to constriction. The question is an important one, and has implications for the timing of surgery.

## SURGERY FOR JEUNE SYNDROME

Several approaches to thoracic expansion for ATD have been suggested, but most reports are anecdotal and long term follow-up is minimal.

For the last ten years we have been working to develop an operative approach that expands the lateral aspect of the chest wall in a way which does not compromise diaphragmatic function and heals without ultimate reliance on any prosthetic material. We first reported this procedure, called "lateral thoracic expansion" (LTE), in

1995 (2). A subsequent report in 2001 (3), updated technical details of the procedure, confirmed that the LTE could be accomplished safely in young children and reported our mid-term follow-up experience. Our procedure divides the chest wall using staggered osteotomies and expands the thorax by opposing the long ends of adjacent ribs. Opposed rib ends are stabilized using titanium plates. Expanded ribs heal at the level of the titanium plates. Liberated periosteum is pulled across the open areas between the short rib segments. This results in new bone formation and "new ribs" over the subsequent months (4). We have generally enlarged both sides in a two stage procedure separated by 6 to 24 months.

## RESULTS OF SURGERY

At the time of this summary, we had performed 13 procedures on 8 infants and young children with ATD (mean age 16.7 months, range 5–47 months). Two mortalities occurred early in the series. Unilateral LTE procedures were done in these 2 patients at 8 and 9 months of age. Both patients had severe underlying parenchymal lung disease and required chronic ventilator support preoperatively. Both went on to develop progressive respiratory insufficiency following surgery. These two experiences led us to caution against the use of the procedure in patients less than two years of age in our initial follow-up report (3). We have, however, subsequently performed 5 procedures in 3 ventilator dependent patients under two years of age with success in terms of enlarging the thorax and reducing ventilator dependency.

Using innovative CT and infant pulmonary function methods (5–6) we have obtained pre and post operative volumetric measurements on our patients with ATD. Preoperatively, mean total lung capacity for the 8 patients was 47.4 % (range 35–69) predicted and mean functional

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residual capacity 62.1% predicted (range 48–93). In ATD patients we have obtained comparable CT and lung function studies before and after (6 months to one year) LTE of one or both sides in 5 patients. Lung function and CT volumetric measurements both demonstrate increases in lung size over time following surgery. Lung volumes do not, however, increase in proportion with increases in height resulting in declines percent of predicted volumes. CT volumetric measurements parallel these changes. All of the 6 surviving patients have, however, done well clinically. All have had dramatic decreases in ventilatory support required and improvements in resting blood gases. One patient, who was ventilator dependant from birth, has now at age 3 been decanulated and requires no ventilatory support. Measurements of lung function and structure have not provided an explanation for the clinical improvement observed in patients. Some mechanism other than simply increasing the size of the thorax may be playing a role. An expanded circumference of the chest may enhance the diaphragm's ability to be effective in some way, and thus improve ventilation, even though the percent of predicted lung volumes for body size decline. Whatever the explanation, our experiences suggest the need for continued long term follow up and caution regarding long term outlooks for these patients.

## SUMMARY

Our experience to date suggests that LTE is a safe and effective treatment of symptomatic patients with ATD. Patients uniformly are clinically improved. Measured increases in thoracic volume are offset by overall increases in body size keeping the percent of predicted volumes low, suggesting mechanisms other than pure thoracic volume increase to explain the observed clinical improvement. Long term follow-up is mandatory.

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