Early Treatment for Early Onset Scoliosis

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Early surgical treatment for early onset scoliosis (EOS) may be justified in select cases based upon etiology of deformity, spine growth, pulmonary development, complications, "diminishing return", implant related deformity and quality of life.

Etiology of Deformity

The thorax is the 4th dimension of the spine. Depending upon whether the primary problem is the spine or the chest wall, the choice of treatment may differ.

If the etiology of the scoliosis is related to a chest wall disorder, then early surgical intervention (i.e. rib-based distraction) should be considered in order to facilitate chest wall growth and potentially improve upon pulmonary development. The primary problem in congenital scoliosis may be fusion of the ribs and that efforts to correct/maintain scoliosis may not be successful if these rib tethers are not addressed surgically. The dilemma is, that by waiting to intervene, the chest wall deformity may become too severe to reverse; however, by intervening too early, growth friendly treatment may cause spontaneous fusion (Emans, 2012).

Severe scoliosis, without rib fusions, may also create chest wall deformity. The spinal penetration index (endothoracic hump) has been defined as a way of quantifying this induced deformity (Dubosset, 2002). This has also been demonstrated in a growing rabbit model in which a unilateral deformity of the ribs or of the spine creates chest wall deformity and scoliosis with asymmetric lung volumes (Metha, 2006).

Spine Growth

One of the goals of treatment of EOS is the maintenance of spine growth. A short, straight spine is not necessarily better than a long, curved spine. Treatment of EOS should focus upon preventing progression of scoliosis while maintaining normal spine growth.

Spine growth is bimodal and has it’s first peak from birth to 5 years of age. During this time, sitting height increases by 27 cm. Thoracic height (T1-T12) increases from 12 cm at birth to 18 cm at age 5 years, to 22 cm at age 10 years, and to 28 cm at adulthood (Dimeglio, 1993). Thoracic height of at least 22 cm has been defined as the threshold for which FVC improves significantly in patients treated for EOS (T1-T12 less than 22 cm had average FVC of 63% predicted vs. T1-T12 greater than 22 cm had an average
FVC of 85% predicted). "Clearly, the shorter the thoracic spine, the smaller the forced vital capacity and the greater the likelihood of pulmonary restrictive disease" (Karol, 2008).

T1-S1 Growth is highest between birth to 5 years of age (2.2 cm/yr) and diminishes from 5 to 10 years of age (0.9 cm/yr). Spine height (T1-S1) is normally 20 cm at age 5 years, 29 cm at age 10 years, and 45 cm at adulthood (Dimeglio, 1993).

Early growth friendly surgery can capitalize on the first peak of spine height velocity.

**Pulmonary Growth**

The rib-vertebral-lung complex is an elastic and structural unit in which scoliosis can increase the rigidity of this unit and prevent normal lung development.

The "golden" period for thoracic spine and chest growth occurs between birth and 8 years of age. Treatment should maintain the normal alveolar development (number of alveoli grows by a factor of 10 from birth to 4 years of age). Patients with early onset scoliosis have less alveoli than normal and these alveoli have been found to be emphysematous. A growth modulation concept has been described and can be used to justify treatment of thoracic deformities before the age of 5 years in order to preserve pulmonary growth.

For patients with rib fusions, opening wedge thoracostomy can increase the thoracic volume ("parasol effect") and should be performed prior to the end of bronchial tree development (8 years of age) (Campbell, 2003).

**Why the recent growing trend away from early surgery?**

There is some evidence that although surgical expansion of the thorax can increase volume, it may not necessarily increase pulmonary capacity (Redding, 2011). There is also potential that surgery may restrict chest wall movement, reduce hemi-thorax size, which may have a detrimental effect on alveolar and capillary development (Mehta, 2006).

This conflicting pulmonary data, coupled with growing rod complication and diminishing returns data, has impacted the enthusiasm for early intervention with growth friendly surgery. It has also created an interest in serial casting as an alternative to growth friendly surgery (Fletcher, 2012, Johnston, 2013).

**Diminishing Returns**

Sankar et al. published that the T1-S1 gain after initial spinal growing rod lengthening surgery decreased significantly with repeated lengthening. Beyond the 7th lengthening, very little additional length was observed (Sankar, Spine’11). It was presumed that this law of diminishing returns for growing rods was related to auto-fusion of the spine. Additional explanations for this phenomena are currently being studied.

The Chest Wall and Spine Deformity Study Group (CWSDSG) performed a study examining spine growth related to rib-based distraction surgery. It was found that 150% of predicted T1-S1 growth was observed for the first five lengthenings and then 50% of predicted T1-S1 growth was observed until the 15th lengthening. When examined by age of the patient, 173% of predicted T1-S1 growth was observed
for lengthenings performed under the age of 5 years versus 32% of expected growth for lengthenings performed between ages 6 and 10 years (El-Hawary, 2013).

To date, spine height has been measured primarily on coronal plane radiographs. By not taking into account other dimensions, these time-related changes in height have been ignoring other factors. Growth friendly surgeries are kyphogenic. As a result, over time, distraction surgery will contribute more to sagittal plane growth (i.e. kyphosis) at the expense of the standard height measurements obtained on PA radiographic views. Efforts are being made to quantify sagittal plane and "true" spine growth related to growth friendly surgery. If growth is maintained when all dimensions are considered, then efforts at modifying implant design may be able to better maintain coronal plane spine height and minimize unwanted increases in kyphosis.

**Complications**

In 2010, the publication of complication data related to spine-based distraction has contributed to the diminished enthusiasm for early surgical intervention. Studying 140 patients treated with growing rods, it was found that 58% of patients had at least one surgical complication. It was also observed that the risk of complications decreased by 13% for each year of increased patient age at the initiation of treatment. The complication risk increased by 24% for each additional surgical procedure performed (Bess, 2010).

A new classification to report complications in growing spine surgery is now being utilized. Severity Grade (SV) I is a complication that does not require unplanned surgery, SVII requires an unplanned surgery(ies), and SVIII is a complication that alters the planned course of treatment. In a study of 65 patients from 5 institutions treated with growth friendly surgery, only 9% of complications were severe enough to alter the planned course of treatment (Smith, 2012).

While it has been found that complications are common with growth friendly surgery, less than 10% of complications were significant enough to change the treatment.

**Quality of Life**

The recently validated Early-Onset Scoliosis 24 item Questionnaire (EOSQ-24) demonstrated improvements in multiple domains of health-related quality of life after growth friendly surgery in 68 patients with an average age of 6.2 years (range 0 -11 years) (Matsumoto, 2012).

Nutritional status has also been observed to improve after insertion of growth friendly implants. This was especially true for those children who had failed to thrive (<5th percentile body weight).

**Casting**

Serial casting is a promising method to prevent scoliosis progression and to delay surgical interventions such as growth friendly surgery and fusions.

The effects of serial casting on the pulmonary system is not yet completely understood. A recent publication from the Dupont Institute revealed that intra-operatively, peak inspiratory pressure was 16 cm H2O before casting, 32 cm H2O after cast application, and 20 cm H2O after making windows. This was a 106% increase after casting and 32% increase after window cutout from the baseline (Dhawale,
2013). Similar adverse effects were found in another study for lung compliance (decreased with cast, increased with window), airway resistance (increased with cast, decreased with window), and tidal volume (decreased with cast, increased with window) (Shafer, 2012). Intra-operatively, there are certainly untoward effects on the respiratory system that seem to return close to baseline by the end of the casting procedure.

In a study of pulmonary compliance on 24 patients, 6 were studied during a 2nd cast application. Those 6 patients had similar intra-operative findings as described above; however, all of those parameters completely normalized by the time of the 2nd cast application (Decoursey Jenson, 2012).

The long term effects of serial casting (as a "delay tactic") on pulmonary development requires further study.

**Proximal Junctional Kyphosis**

PJK rates associated with growth friendly have varied depending upon the definition of PJK. CWSDSG studied a group of 40 children treated with either spine-based or rib-based surgery and found a 27.5% rate of PJK. Two significant findings for the PJK group were pre-operative hyperkyphosis and a higher age at implantation (7.1 years vs. 5.0 years) (El-Hawary, 2011). In a separate study, children with EOS had a trend towards higher thoracic kyphosis for the 6-10 year age group than for the 0-5 year age group (40 degrees vs. 37 degrees) (El-Hawary, 2013). There may be a (weak?) link that by delaying implantation of growth friendly implants, that these older patients may be at a higher risk of developing implant related problems. This association requires further study.

**Summary**

Early treatment of EOS with growth friendly implants should be considered in select patients in order to prevent progression of scoliosis and to maximize spine height, minimize chest wall deformity, and to improve upon thoracic volume. It is not clear if these positive effects on growth will translate to improved pulmonary capacity.

The law of diminishing returns may not hold true for rib-based distraction systems and may also be related to implant design and sagittal-plane effects of repeated lengthening surgeries, rather than on presumed autofusion alone.

The high complication rate associated with growth friendly surgery may only cause significant changes in management in less than 10% of patients.

Multiple domains of health-related quality of life have improved with growth friendly surgery.

The use of serial casting as a "delay tactic" to surgery has become quite popular; although, the long-term effects of serial casting on pulmonary development are not yet fully understood.

The delay of growth friendly surgery may be at the expense of implant related complications such as proximal junctional kyphosis.
References


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