Early Onset Scoliosis

Editorial Comment

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Over the last decade, probably more than in any other condition in orthopaedics, we have witnessed almost exponential changes in treatment philosophy for early-onset scoliosis (EOS), a spine deformity of early childhood that is potentially fatal as a result of respiratory impairment associated with chest wall deformity. Defined as spinal deformity of any etiology presenting under the age of 5 years, it is now accepted that traditional orthopaedic management of this condition—spinal fusion—may produce morbidity of a severity similar to that associated with completely untreated deformity for reasons well described by the leading article, which reviews what is known about patients' outcome from this traditional method. To put it simply, “definitive fusion” under the age of 5 years of more than five to six thoracic segments is an oxymoron and can no longer be recommended, because the spinal deformity is unlikely to be controlled, thoracic growth will be limited to less than 50% of normal, and restrictive lung disease (forced vital capacity [FVC] less than 50% predicted) will likely ensue, especially if the most proximal thoracic vertebrae (T1 and T2) are included in the fusion.

Growth-sparing procedures using “growing” distraction-based instrumentation constructs anchored to spine or ribs, which are periodically lengthened, thus requiring serial reoperation, have been widely accepted as the alternative to a growth-ending spinal fusion for EOS. However, almost no consensus exists for the application of these treatment modalities for specific diagnoses and deformity patterns, an untenable situation for both patients and spinal surgeons and a topic thoroughly discussed in the second article. The

Fig. 1 Charles E. Johnston, MD, is shown.

poor correlation between deformity correction (Cobb angle) and any improvement in pulmonary function (measured as FVC % predicted) after expansion thoracoplasty is analyzed by the third article, a sobering objective outcome measure considering the accepted importance of expanding thoracic volume to combat thoracic insufficiency syndrome (TIS), a term coined by Campbell et al. [1], the originators of the expansion procedure, describing the inability of the thorax to support normal lung growth and respiration. However, in a rabbit model of TIS published in this symposium [3], histologic and histochemical

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evaluation of postexpansion thoracoplasty lung tissue has demonstrated improvement in what are otherwise alveolar and capillary abnormalities, which heretofore have been described in postmortem studies of patients dying of respiratory failure from untreated EOS, thus providing experimental evidence that expansion of a constricted or hypoplastic thorax has a salutary effect.

Thereafter, the symposium contains research articles on the creation of an appropriate large animal (porcine) scoliosis model using flexible cables to tether the chest wall and spine unilaterally during growth and a second report of creating and correcting experimental porcine deformity by inserting pedicle screws across the neurocentral synchondrosis on one side of the spine and then later on the other. Underlying both these studies is the attempt to develop a growth guidance method of deformity correction, a welcome advancement to the serial reoperation distraction-based procedures currently in vogue.

Lastly, the symposium contains five current clinical studies. The first documents the problems of proximal anchor failure, proximal junctional kyphosis (PJK), and global sagittal plane imbalance in applying the Vertical Expandable Prosthetic Titanium Rib (VEPTR™) device (Synthes, Inc, West Chester, PA) specifically to patients with kyphoscoliosis, problems that are all too common and for which a solution remains elusive. Two studies describe outcomes of modification of VEPTR™ implantation to combat anchor failure and PJK. A new method of achieving convex hemiepiphyseodesis without anterior spine surgery using pedicle screw fixation is presented. Last but not least, a modern revival of the growth guidance method originally described by Eduardo Luque [2] using a minimally invasive implantation technique shows great early promise as an alternative to serial lengthening procedures.

In reality, the contributions in this symposium represent but a small fraction of the active clinical and basic research directed toward ameliorating the life-threatening condition that is EOS. I have recruited these contributions with the goal of presenting where we have come from, what the current liabilities of accepted treatment methods are, how the latest in treatment method modifications address some of these liabilities, and where we might be going with animal models of TIS and the reversal of its potentially fatal outcome. However, ultimately, until we have successfully harnessed and/or stimulated spinal growth by an effective internal guide to autocorrect the spine and chest wall deformity and expanded hypoplastic thoraces without creation of respiration impairment by chest wall ankylosis, the long-term outcome for the patient with EOS may remain less than satisfactory as a result of diminished quality of life in a multiply operated scenario. These are exciting times with new clinical and experimental approaches to TIS being developed constantly, but there remains much to be done for this most serious of nonmalignant orthopaedic conditions.

References

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