The First Generation of Early Onset Scoliosis Care

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Abstract:
Reports of the prevalence and natural history of spinal deformity in younger pediatric patients became part of the orthopaedic literature in the middle of twentieth century. Formal use of the term “Early Onset Scoliosis” to describe a wide range of spinal pathology based on age of onset did not gain popularity until much later. Early reviews of the natural history of these deformities detailed which patients were at risk of progression, and which patients may benefit from intervention rather than simple observation. However, long-term follow-up of the application of adult spine deformity management principles in skeletally immature patients demonstrated a significant risk of both spinal and associated pulmonary complications over time. Reports of efforts to alter the natural history of these conditions through surgical treatment that attempted to control the deformity, while still allowing spinal growth, emerged in the late 1970s and 1980s.

Key Concepts:
• Early onset scoliosis is defined as a progressive spinal curvature in children less than 10 years of age and can be a result of multiple etiologies.
• The last 90 years has seen an evolution of our knowledge base regarding these challenging patients.
• Progressive deformities in these young patients is associated with a natural history marked with increases in morbidity and mortality.
• Practitioners and treatments have evolved over this period in time and growth friendly strategies, while not perfect; currently offer the best treatment.
• Much work remains in developing strategies and principles for this heterogenous patient population.

Introduction
Early Onset Scoliosis (EOS) is a descriptive term of relatively recent acceptance that includes multiple etiologies of spinal deformity in young children. While the primary deformity is recognized most often in the coronal plane, rotational or abnormal sagittal plane deformities often exist concomitantly. EOS is considered frequently to be idiopathic without a known or obvious underlying cause. However, EOS may be associated with neuromuscular conditions (cerebral palsy, myelodysplasia), congenital or structural abnormalities of the vertebral and/or chest wall, or syndromic disease associated with poor bone health or bone malformation (skeletal dysplasia, neurofibromatosis). At this time, EOS is classified as a progressive spinal deformity, regardless of etiology, diagnosed before the age of 10 years.¹

Early attempts at management of EOS patients were focused primarily on those with idiopathic or congenital
deformities, as the medical fragility of patients with neuromuscular or syndromic etiologies frequently precluded, or greatly influenced, treatment of any associated spinal deformity. As such, this review will be limited to the historical and early published literature regarding the diagnosis and treatment of patients with EOS secondary to congenital spinal abnormalities and infantile idiopathic scoliosis. The authors hope to provide a historical context regarding the development of understanding of the natural history of these conditions as well as the rationale and results of early attempts at treatment of EOS patients prior to the advent of “modern” growth-friendly interventions.

Infantile Idiopathic Scoliosis: Etiology, Diagnosis, and Natural History

In 1936, Harrenstein reported a series of pediatric patients with scoliosis in the French literature. All patients were under the age of 2 years with no apparent rib or vertebral deformities, and he described the entity as “infantile” scoliosis. He noted a female preponderance in this cohort, that the deformities were primarily right thoracic curves, and that there was a good response to nonoperative treatment. In addition, there were a number of patients in whom the deformity appeared to correct spontaneously without intervention. Twenty-five years later, James formalized the term “infantile idiopathic scoliosis” (IIS) as patients diagnosed with a progressive spinal deformity under the age of 3 years without any associated vertebral or chest wall deformities. In contrast to Harrenstein’s earlier report, the majority of James’s patients were male, most had a left thoracic curve with significant rotation, and spontaneous resolution occurred in only 12% of cases (Figure 1).

Scott and Morgan subsequently reviewed 1200 patients with a diagnosis of scoliosis in 1955. In that series, the authors found 28 patients between the ages of 0 and 2 years of age that met the criteria for a diagnosis of infantile idiopathic scoliosis. There was no difference in the percentages of males and females, and the curves were almost exclusively left thoracic. They reported a rate of spontaneous resolution of 25%, but deformities greater than 30 degrees at diagnosis tended to progress at a rate of approximately 5 degrees/year. Of note, the authors commented that, “It seems that in America this type of scoliosis is either rare, incompletely recognized, or confused with congenital scoliosis.” As late as 1965, Lloyd-Roberts and Pilcher reported on 100 patients with onset of IIS prior to 1 year of age. They were unable to differentiate between patients at risk of progression and those who may spontaneously stabilize or resolve, and as such, recommended simple observation and repeat examination. The authors noted that the deformity resolved without treatment in greater than 90% of the patients in the study.

Multiple early authors noted a generally poor prognosis for patients with a diagnosis of IIS. In their patients, most deformities progressed relentlessly, and treatment options were limited. In many cases, the growing spinal deformity was associated with the development of severe chest wall and rib deformities, and the authors commented on worsening pulmonary status and diminishing vital capacity/lung function. In 1992, Pehrsson et al. reviewed the natural history and mortality of patients born between 1902 and 1937 with untreated scoliosis, looking specifically at age of onset of spinal deformity. They found an increase in mortality associated with an underlying diagnosis of scoliosis, but not in those patients with adolescent-onset disease. Those patients with onset...
of a progressive deformity before the age of the 3 years of age were at greatest risk of early mortality, most commonly associated with pulmonary and/or cardiovascular disease. Branthwaite\textsuperscript{9} similarly reported disabling dyspnea or cardio-respiratory failure attributable to unfused idiopathic scoliosis diagnosed early in life. In that study, 10/11 patients with mortality due to cardiopulmonary failure and associated unfused idiopathic deformity were diagnosed with scoliosis at less than 5 years of age.

The highly variable rates of spontaneous deformity correction reported in early studies of IIS in conjunction with the poor natural history of those deformities that did not stabilize or regress, made it imperative to understand which patients are at risk of developing severe deformity without treatment and which may have limited risk of progression or even the possibility of some element of spontaneous resolution. In 1972, Mehta\textsuperscript{10} proposed the concepts of “rib phase” at the apex of the deformity and the measurement of the “Rib Vertebral Angle Difference” (RVAD), which was also measured at the apex of the deformity on an early AP or PA radiograph (Figures 2a and 2b). Mehta demonstrated that IIS patients with deformities that measured at least 25 degrees and had what she termed “Phase 2 ribs” or an RVAD > 20 degrees, were unlikely to show resolution of their scoliosis, and that these curves should be considered progressive (Figures 3a and 3b). She stressed the importance of early diagnosis and treatment for patients with apparently progressive IIS deformities. Few, if any, other reports of casting for IIS exist in the early literature.\textsuperscript{11} Many of the principles, measurements, and techniques espoused by Mehta remain foundations of the diagnosis and treatment of IIS to this day and will be reviewed in greater detail by Welborn et al., \textit{JPOSNA Vol. 3, No 2}.

\textbf{Congenital Scoliosis: Classification and Natural History}

Congenital scoliosis is the result of an error in embryogenesis in the first 6 weeks of gestation that may result in malformations of single or multiple vertebra,

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\includegraphics[width=\textwidth]{image}
\caption{PA radiographs of a 19-month patient demonstrating the determination of Rib/Vertebral Angles. In this case, the Rib/Vertebral Angle Difference (RVAD) is 76-51 or 25 degrees. An RVAD greater than 20 degrees correlates with a higher risk of progressive deformity in IIS patients.}
\end{figure}

intervertebral disc spaces, or both. Congenital scoliosis can affect any of the three columns of the spine and is a heterogeneous disorder that can present as an isolated deformity at a single vertebral level or may involve multiple spinal segments. Wynne-Davies\textsuperscript{12,13} found that there was often a hereditary component in patients with multiple congenital spinal anomalies, but that the incidence of single abnormalities was generally sporadic in nature. There have been multiple reports in animal models of relationships between intrauterine exposures,
including hypoxia, carbon monoxide, and other substances, and the development of congenital spinal anomalies.\textsuperscript{14,15} In addition, the association of congenital scoliosis and other congenital abnormalities, particularly of the renal system and neural axis, have been well documented in the literature.\textsuperscript{16}

Similar to early reports of IIS, the initial studies addressing the natural history of congenital scoliosis emerged in the 1950s. Kuhns and Hormell\textsuperscript{17} reviewed 165 patients with congenital scoliosis in 1952, and Billings\textsuperscript{18} described 14 patients in 1955. These authors concluded that, in most cases, any resulting spinal curvature had a relatively benign prognosis. They felt that these patients progressed slowly, if at all, and that any progression would result in little significant deformity or functional effect. The work of Winter et al.\textsuperscript{19} in 1968 provided evidence to the contrary and demonstrated a more problematic prognosis for a large percentage of congenital scoliosis patients. They found that congenital scoliosis was found most commonly in the thoracic spine, and deformities in that area had a greater risk of progression than those originating in the cervical or lumbar region. In addition, they felt that there was an elevated risk of deformity progression regardless of the specific underlying congenital anomaly.

McMaster and Ohtsuka\textsuperscript{20} confirmed many of the findings of Winter et al. in their landmark paper in 1982. That study provided a comprehensive review of the natural history of congenital scoliosis describing 216 patients with a minimum 5-year follow-up. The authors reported specific rates of spinal deformity progression depending on the patient’s age at presentation as well as the location and type of congenital bony abnormality. They emphasized that those patients with unbalanced congenital malformations, such as unilateral hemivertebrae or unilateral areas of failure of segmentation, are at the highest risk of progressive deformity. Overall, the works of Winter et al. and McMaster and Ohtsuka highlighted the importance of defining the anatomy of the congenital lesions in order to determine the level of concern regarding deformity progression with future growth. Classification systems like that illustrated in Figure 4A-C, have proven essential for understanding the true natural history of congenital scoliosis in the skeletally immature patient population.

**Early Onset Scoliosis: Historical Nonoperative and Surgical Treatment**

Until later in the 20th century, the principles of management of patients with EOS, regardless of the underlying origin, were relatively similar from institution to institution. Milder deformities were observed or underwent initial attempts at bracing. Patients who presented with larger curves, or progressed despite bracing or casting, were managed surgically with early, definitive fusion procedures. McMaster and Macnicol described the “Edinburgh Policy” for IIS in 1979.\textsuperscript{21} At their center, the goal was to identify patients with a progressive thoracic curve early, and manage the deformity with bracing or Risser turnbuckle casting until the age of 10 years, if possible. The patient would then undergo posterior spinal fusion, in some cases utilizing single Harrington (or equivocal) Rod instrumentation (Figure 5). Overall, the
authors felt that this course of treatment that utilized both nonoperative and operative interventions was an effective approach in this patient population.

Hefti and McMaster reviewed further experiences with posterior fusions in younger patients with significant idiopathic deformities in 1983. They included both infantile and juvenile patients in their cohort. They noted that the posterior fusions inhibited longitudinal growth of the solidly fused segments, but that the continued anterior growth associated with the eventual adolescent growth spurt caused the vertebral bodies to “bulge laterally toward the convexity and to pivot on the posterior fusion, giving rise to a loss of correction, increasing vertebral rotation and recurrence of the rib hump.” Dubousset et al. reviewed the Texas Scottish Rite Hospital (TSRH) and Miami Children’s Hospital experiences with early spinal fusions in 1989. They also highlighted the persistence of anterior spinal growth in young patients despite an apparently solid posterior fusion that was associated with worsening rotational deformity and termed this the “Crankshaft Phenomenon.” The authors recommended that early fusions should always include anterior and posterior procedures to minimize the
risk of this progressive secondary deformity. The Crankshaft Phenomenon can also occur in patients who have had a longstanding posteriorly based distraction device (Figure 6).

Although there are reports of acceptable functional long-term results with minimal complications after definitive early fusion in patients followed into adulthood, multiple authors have demonstrated significant issues with the concept of generating a “short and straight” rather than a “long and crooked” spine. Goldberg et al. reviewed 43 patients treated with early fusion for congenital scoliosis. Over a quarter of their patients, including patients who underwent circumferential fusion procedures, required revision or reoperation for further deformity progression. Karol, et al. reported on the TSRH experience with early fusion, also including patients who had anterior/posterior procedures. These authors demonstrated a reoperation rate of close to 40%. Vitale et al. assessed 21 patients who had undergone what was expected to be a definitive early fusion for congenital scoliosis. They reported that almost 24% of the cohort had required more than one procedure to achieve fusion, and their follow-up was relatively short at the time of publication. Overall, the success of early fusion from the standpoint of the index procedure correcting and controlling the deformity, even when a circumferential fusion was performed, often proved to be problematic.

Multiple authors have explored the effects of attempted early definitive fusion on pulmonary status. Much of this concern has been focused by the works of Campbell and Dimeglio detailing chest and lung development with growth. Goldberg, et al. looked at pulmonary function testing of 23 patients after early fusion for infantile deformities. They found a significant difference in vital capacity in those patients fused prior to age 10 versus patients who underwent fusion at an older age. Bowen et al. compared pulmonary function tests between a group of
patients who underwent early fusion for congenital scoliosis and a group treated without surgery. They found that both groups had a diminished vital capacity but no difference between the two groups. However, they did find a correlation between a higher thoracic apex of deformity and lower vital capacity. This was similar to one of the conclusions in the study by Karol et al.27 from TSRH. Finally, Vitale et al.28 found vital capacity that averaged 64.2% of predicted values after early definitive fusion for congenital scoliosis with a thoracic apex. In addition, these authors correlated lower pulmonary volumes and diminished quality of life scores. While there is certainly evidence that underlying chest and spinal pathology present in patients with EOS prior to surgery may be associated with abnormal pulmonary function data, multiple studies have demonstrated the further negative effects of attempts at early, definitive fusion in this patient population.

Despite the fact that early formal fusion, initially by posterior-only procedures and then eventually with combined circumferential approaches, was the primary means of addressing those EOS patients who failed nonoperative procedures or presented with large deformities, there were some limited early efforts at surgical deformity control without actively generating a fusion. While these attempts apparently began with Harrington33 in 1962, many of the reports exist only in meeting abstracts or personal communications.34-36 In 1982, Luque37 reported on segmental wiring of single Harrington rods without fusion in 47 patients with paralytic scoliosis 8 years of age or less. He exposed only the instrumented side of the deformity and did not utilize any external support. A number of patients underwent multiple operative procedures, but the indications for these interventions were unclear. Overall, the complication rate, including rod failure, was high. The author reported some growth of the instrumented segments but felt it was primarily due to curve correction at the time of initial insertion and repeated Harrington Rod distraction. Of note, variations and modifications of this technique became known colloquially as a “Luque Trolley.”

Moe et al.38 reported on 20 patients managed with a subcutaneous, modified Harrington Rod for early onset deformity (Figure 7A-D). The average age at time of surgery was 8 years 11 months, with the youngest patient 4.5 years of age. Over half of the patients had failed bracing prior to surgery. All were placed in a full-time Milwaukee brace postoperatively. The rods were lengthened intraoperatively every 4-6 months. Complications
were noted in 10 patients, including rod fracture and hook displacement. Nine patients had undergone formal fusion at the time of the report, and 9 patients continued to undergo lengthening procedures. The authors reported an average growth of the instrumented area of 2.9 cm for all patients over the course of treatment. They concluded that the method allowed for growth while avoiding early fusion in some patients as well as providing and maintaining curve correction.

Long-term results of early attempts at what is now known as “growth-friendly” or “fusion-less” surgery are extremely limited. Mardjetko et al. reviewed the Chicago experience with revision of previous “Luque Trolley” procedures. Patients in the cohort had undergone segmental instrumentation without fusion from 1982-84, at an average age of 9 years. The authors reported that the average gain in spinal height appeared to have occurred outside of the instrumented segments, and that this method of instrumentation without fusion was not effective in controlling progression of the underlying deformity (Figure 8A-C). At the time of revision, all patients showed areas of autofusion, and the procedures were complicated by these areas of fusion as well as by scarring and poor bone quality. Pratt et al. compared a series of “Luque Trolley” procedures with and without a concomitant convex hemiepiphyseodesis in EOS patients. They found that the Trolley procedure alone did not prevent curve progression while addition of the convex hemiepiphyseodesis appeared to stabilize or improve the deformity in some patients, which suggested some element of growth effect over time.

Conclusion

The initial era of diagnosis and management of EOS was complicated due to the heterogeneity of the patients, the relatively limited prevalence of the diagnosis, an early underappreciation of the natural history of the deformity, and a limited available treatment armamentarium. Comprehensive natural history studies performed in the 1970s and 1980s, coupled with rigorous stratification of deformity based on etiology, anatomic location, and age at presentation, allowed clinicians a better understanding of the risk of progression and determination of those patients requiring intervention.

Long-term follow-up of early efforts at surgical intervention, even novel techniques that were thought to provide deformity correction and control while allowing some element of spinal growth, often led to significant complications and potentially disastrous results. Early posterior spinal fusions led to retardation of the growth of the thorax, often leading to further pulmonary compromise. Additionally, posterior fusion alone in the face of continuing anterior growth often led to worsening spinal and thoracic deformity.

These concerns led some investigators to devise other “growth-friendly” strategies to control early onset spinal deformity and permit some element of continued spine
and thoracic growth. The first generation of these efforts succeeded to a limited degree but at the risk of high short and long-term complication rates and significant iatrogenic difficulties at the time of revision or final fusion. Future generations of surgical innovators would go on to use the lessons learned through these early efforts to better address both the issues of deformity progression and minimize the potential complications of treatment.

References