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Editorial

Ultra-long-term follow-up of pediatric spinal deformity problems: 23 patients with a mean follow-up of 51 years

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Abstract
Background. The aim of this study was to analyze the true outcomes of a unique cohort of patients with spinal deformities who were treated as children and followed for 40 or more years.
Methods. Altogether, 23 patients were reviewed who had been originally treated in our community, whose original charts and radiographs were still available, and who had undergone recent evaluation.
Results. The diagnoses were congenital deformity in eight, adolescent idiopathic scoliosis in four, poliomyelitis in three, infantile idiopathic scoliosis in two, spondylolisthesis in two, and one each of tuberculosis and dwarfism. Sixteen had undergone fusion surgery.
Conclusions. Early spine fusion for deformity produced far better results than delayed fusion. A solid fusion at the end of growth remained unchanged. Degenerative changes outside the fusion area were rare and seldom required further surgery. In summary, 23 patients with a mean follow-up of 51 years after treatment are presented. Early fusion was far superior to delayed or nonsurgical treatment.

Introduction

As professionals, we as physicians and surgeons have the desire to determine the outcome of our interventions, whether nonsurgical or surgical. Our problem is how and when to measure such outcome. The standard for adults is generally a minimum of two years. For children the standard is usually the end of growth. This does not, however, measure the outcome after the child enters adult life, employment, marriage, and childbearing. This review constitutes the longest mean follow-up (51 years) ever reported for pediatric spinal problems.

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Material and methods

Included in this review were 23 patients, all of whom were originally seen and treated in our community, most of them at our local crippled children's hospital. This hospital has never discarded any chart or radiograph, making them still available when we subsequently see one of these patients.

The 23 patients ranged in age from 1 to 17 years when first seen. The mean follow-up was 51 years (range 40–76 years). The earliest initial record was from 1923 and the most recent from 1967.

The diagnoses were idiopathic scoliosis in eight (two infantile, two juvenile, four adolescent), congenital deformity in eight, poliomyelitis scoliosis in three, spondylolisthesis in two, tuberculosis in one, and spondyloepiphyseal dysplasia in one. Sixteen of the patients had undergone fusion surgery, all posterior, and only one with instrumentation (1967). The youngest patient to have fusion was 1 year old, and the oldest was 17 years.

The reasons we were able to see them for long-term follow-up were variable. Thirteen were seen because of our specific efforts to find patients for long-term follow-up (not because they had any complaint or problem). Five were seen because of cor pulmonale, two because of neck pain, one because of low back pain, one because of coronal decompensation, and one for unrelated problems.

The original charts and radiographs were available for all of the patients in this series as were those prepared at the time of their long-term follow-up. All radiographs were in excellent condition and easily measured.

Results

Because these patients essentially are defined by a series of case reports with different diagnoses and different

treatments, statistical analysis is not possible. Furthermore, because our inclusion criterion was a follow-up of 40 years or more, patients who died of their curvatures in their twenties or thirties are not included. Several of the patients included in this article have been reported in the literature as single case reports, so they are not reported in detail here. The patients are presented in order, from shortest to longest follow-up.

Case 1 (40-year follow-up)

A 14-year-old girl presented with 47° right thoracic adolescent idiopathic scoliosis. She underwent Harrington instrumentation and fusion T4-L1 in 1967. She was seen in 2007 with complaints of low back pain and was found to have degenerative spondylolisthesis of L4/5. The intervening discs were normal.

Case 2 (40-year follow-up)

A 7-year-old girl was first seen in 1933 with 30° thoracic juvenile idiopathic scoliosis (Fig. 1). Her curve progressed rapidly despite brace treatment, reaching 112° by age 11. A posterior fusion was done elsewhere but only of the apical 7 segments of an 11 vertebrae curve. The scoliosis continued to progress, reaching 124° by age 15. She was seen in 1973 at age 47, which was 40 years after the first visit and 36 years after the fusion. The curve was 142°. She presented with major respiratory insufficiency and early cor pulmonale. Her case illustrates failure to perform adequate fusion surgery at a more appropriate time.

Case 3 (41-year follow-up)

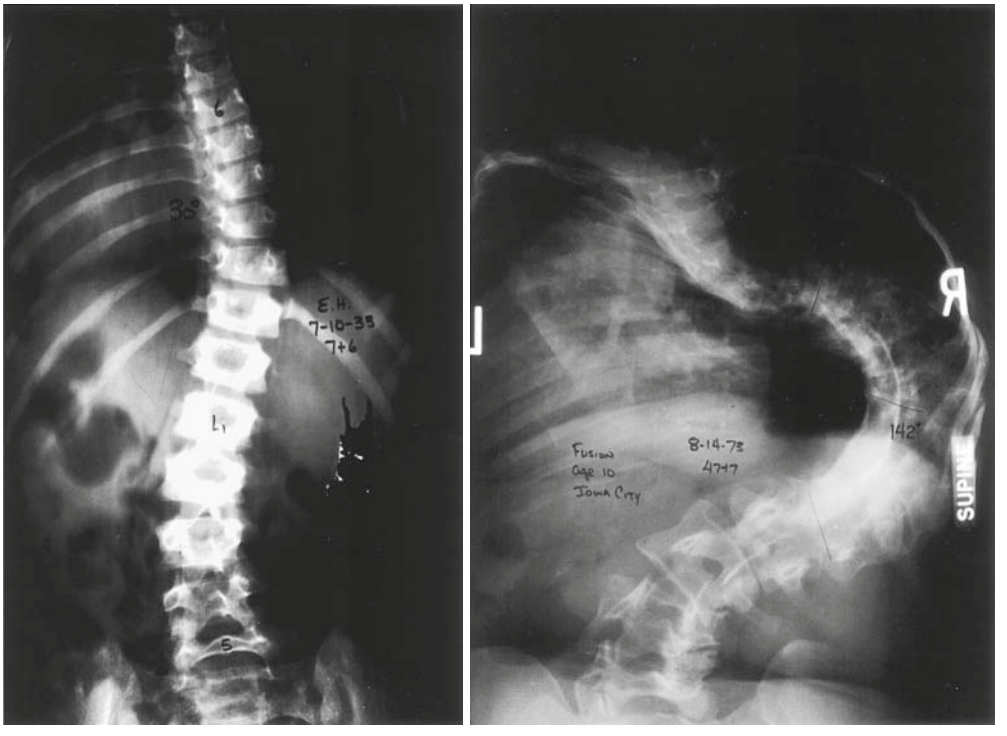
A 15-year-old boy was seen in 1943 with grade IV isthmic spondylolisthesis at L5-S1. He underwent L3-S1 posterior spine fusion with cast immobilization (no reduction). He was seen in 1984 as part of a long-term follow-up study. His fusion was solid, and he had done hard labor as a farmer for the entire 41 years since surgery. He was married, had three children, and had no back or leg complaints.

Case 4 (42-year follow-up)

A 15-year-old girl was first seen in 1925 with a diagnosis of "dwarfism with scoliosis." She had 35° thoracic scoliosis and 60° thoracic kyphosis. No treatment was given. She was seen again in 1967 as part of a long-term follow-up of patients with dwarfing disorders. She was diagnosed as spondyloepiphyseal dysplasia. Her curve was now 52° scoliosis and 78° thoracic kyphosis. Other than cosmetic dissatisfaction, she had no spinal complaints.

Case 5 (44-year follow-up)

A 13-year old boy was first seen in 1942 with isthmic spondylolisthesis of L5-S1. He had a 31% slip with a 10° slip angle (measured from the superior endplate of L5).



A B

Fig. 1. A Case 2, a girl with 30° juvenile idiopathic scoliosis. **B** at age 47, the patient had 142° scoliosis. She had undergone a short posterior fusion at age 11 when her curve had reached 112°. She was in cor pulmonale at this time and died soon afterward

No treatment was given. When seen in 1947, the slip had increased to 60%, with a 35° slip angle. He underwent L3-S1 posterior spine fusion with cast immobilization but no reduction. When seen in 1984 as part of a long-term follow-up study, he had a solid fusion but a slip of 76% and a slip angle of 41°. He was asymptomatic.

Case 6 (44-year follow-up)

An 11-year-old girl was first seen in 1928 with a 142° curve due to infantile idiopathic scoliosis. No treatment was given. By age 14, her curve was 160°. In 1972, she presented with cor pulmonale that was refractory to medical treatment, and she died. She illustrates the tragedy of failure to apply fusion to progressive infantile idiopathic scoliosis.

Case 7 (44-year follow-up)

A 15-year-old boy had severe residual problems of poliomyelitis, being essentially quadriplegic with no functional use of his arms or legs. His scoliosis was severely progressive, reaching 112° by age 15 (Fig. 2). He was treated by halo-cast reduction to 55° and posterior spine fusion C4-S1 in 1960. At a 2-year follow-up the fusion was solid and the curve 74°. When seen electively in 2004 at age 59, the curve measured 77°. Despite his severe handicap, he was a successful attorney, married, and supporting his parents as well as his wife and himself.

Case 8 (44-year follow-up)

A 3-year-old girl with rapidly progressive congenital scoliosis underwent posterior spine fusion from T2-L3 at that age. She was seen subsequently for elective follow-up at age 47. Despite agenesis of the left lung and the early fusion, her vital capacity was 71% of normal. She has been previously reported.[1]

Case 9 (44-year follow-up)

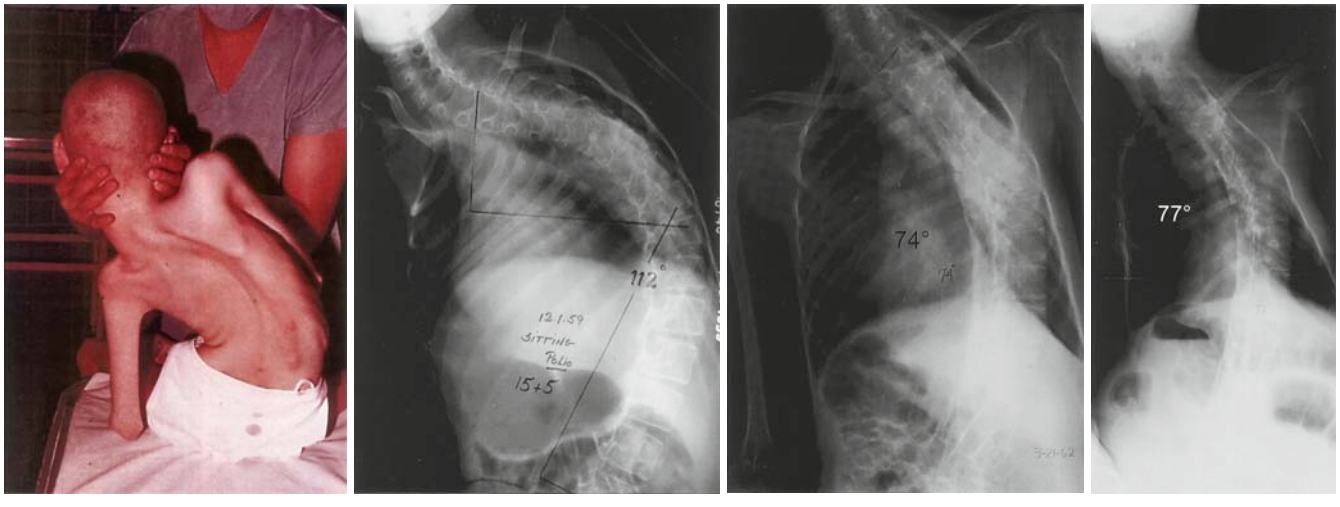
A 17-year-old girl was seen in 1953 for progressive adolescent idiopathic scoliosis. She had an 83° thoracic curve and a 62° lumbar curve. She underwent selective thoracic fusion with cast correction to 56°. The unfused lumbar curve balanced at 50°. When seen in 1997 for elective follow-up, the curves were unchanged at 57° and 50°. She had no low back pain. She has been previously reported.[2]

Case 10 (44-year follow-up)

A male infant had 37° congenital scoliosis of the thoracic spine with a unilateral unsegmented bar. He underwent posterior spine fusion with cast correction at age 12 months. When seen as an adult with major neck complaints, the fusion was solid, the curve was 26°, and his vital capacity was 70%. He has been previously reported.[3]

Case 11 (47-year follow-up)

A female patient had progressive scoliosis due to poliomyelitis. She underwent posterior spine fusion in 1949 at age 17. Unfortunately, the fusion did not fully solidify, and her original correction was lost. When seen in 1995 at age 64, she had primarily neck pain complaints, and her curve measured 91°.



A,B C,D

Fig. 2. **A** Case 7, a 15 year-old boy with severe residual problems of poliomyelitis. His scoliosis was rapidly progressive. **B** This sitting radiograph was obtained just prior to surgery when the patient was 15 years of age. **C** Upright radiograph at age 17 (2 years after surgery). **D** Upright radiograph at age 58 (43 years after surgery)

Case 12 (47-year follow-up)

A 17-year-old boy was first seen in 1938 with double thoracic congenital scoliosis of 60° (T1/5) and 80° (T5/12). No treatment was given. When seen again in 1985 at age 64 for a routine follow-up, his curves were 60° (T1/5) and 105° (T5/12). He was functioning well with no problems.

Case 13 (48-year follow-up)

An 11-year-old girl was first seen in 1928 with 142° scoliosis due to infantile idiopathic scoliosis. By age 14, the curve had reached 160°. No treatment was given. When seen in 1976 at age 59, she was in terminal cor pulmonale and died shortly thereafter.

Case 14 (50-year follow-up)

A 3-year-old girl was first seen in 1955 with severe congenital lumbar kyphosis and spinal stenosis. She underwent posterior spine fusion from T12 to S1 at age 3 and pseudoarthrosis repair at age 4. When seen for routine follow-up in 2005 at age 53, she had no spine complaints. She has been previously reported.[4]

Case 15 (50-year follow-up)

A 15-year-old girl with scoliosis due to poliomyelitis was first seen in 1957 with a 115° curve. She underwent posterior spine fusion, but it did not solidify. Repeat surgery in 1968 also failed, and she had progressive deformity to 140°. In 1994, at age 46, she went into acute cor pulmonale but was salvaged with multiple osteotomies, traction, anterior fusion, and posterior fusion with instrumentation. In 2008 she is nearing the end owing to recurrent cor pulmonale.

Case 16 (51-year follow-up)

A 13-year-old girl was seen in 1925 with 40° congenital scoliosis. No treatment was given. When seen in 1976 at age 65, her scoliosis was 60°, but she was severely decompensated to one side owing to a progressive lumbosacral anomaly.

Case 17 (54-year follow-up)

A 6-year-old boy was first seen in 1943 with 57° congenital thoracic scoliosis. No treatment was given, and the curve progressed to 115° by age 15. At that time he underwent posterior spine fusion with cast correction to 90°. He was seen in 1997 with early signs of cor pulmonale. His curve was still 90°.

Case 18 (55-year follow-up)

A 3-year-old girl was seen in 1923 with spinal tuberculosis. She was treated with bed rest, diet, casting, and bracing. She obtained complete cure of the tuberculosis and solid spontaneous fusion of the diseased area. When seen in 1978 at age 58 for another, unrelated problem, she had mild residual gibbus but no pain or neurological problems.

Case 19 (59-year follow-up)

An 11-year-old girl was seen in 1948 with 60° right thoracic idiopathic scoliosis. She underwent posterior spine fusion from T4 to L3 with cast correction to 30°. When seen in 1977 for a long-term follow-up project, her curve was 35°, and she had no symptoms. When seen in 2007, the curve was the same, and she reported only occasional, mild back pain.

Case 20 (59-year follow-up)

A 4-year-old boy was first seen in 1948 with congenital scoliosis of 36° at T5/11. No treatment was given, and the curve progressed. He was first seen at our center in 1957 at age 13 with a 54° T5-L2 curve. He underwent T1-L2 posterior spine fusion with cast correction to 30°. At a 4-year follow-up, the fusion was solid at 37°. When contacted in 2007 (50 years after surgery), the curve remained the same, and he was having minimal low back pain and some shoulder pain. He was married, had children, and was successfully employed.

Case 21 (63-year follow-up)

A 15-year-old girl was seen in 1939 with 68° adolescent idiopathic scoliosis. She underwent posterior spine fusion down to L3. At age 50 this was extended to L4 because of junctional degeneration. When seen in 2002 at age 78, she had degenerative changes in the L4-S1 area with mild stenosis symptoms not requiring surgery. She has been previously reported.[5]

Case 22 (72-year follow-up)

A 6-year-old girl was first seen in 1927 with progressive infantile idiopathic scoliosis of 60°. She was treated in casts and braces until age 13, when the curve was 132°. Posterior spine fusion was performed in 1935 without correction. A solid fusion was obtained. She was seen in 1985 at age 64 because of a painful bursa between her rib prominence and the scapula. Thoracoplasty solved this problem. When seen again in 1997 at age 76, she was in cor pulmonale and on full-time oxygen. She died in 1999 at age 78. Her curve measured 140°. Although

Table 1. Patients' history

Patient	Age when first seen (years)	Age at follow-up (years)	Length of follow-up (years)	Diagnosis	Treatment	Reason for follow-up
1	14	54	40	Adolescent idiopathic scoliosis	Fusion	Low back pain
2	7	47	40	Juvenile idiopathic scoliosis	Fusion	Cor pulmonale
3	15	56	41	Spondylolisthesis	Fusion	Routine follow-up
4	15	67	42	Dwarfism	None	Routine follow-up
5	13	57	44	Spondylolisthesis	Fusion	Routine follow-up
6	11	55	44	Infantile idiopathic scoliosis	None	Cor pulmonale
7	15	59	44	Polio	Fusion	Routine follow-up
8	3	47	44	Congenital scoliosis	Fusion	Routine follow-up
9	17	61	44	Adolescent idiopathic scoliosis	Fusion	Routine follow-up
10	1	45	44	Congenital scoliosis	Fusion	Neck pain
11	17	64	47	Polio	Fusion	Neck pain
12	17	64	47	Congenital scoliosis	None	Routine follow-up
13	11	59	48	Infantile idiopathic scoliosis	None	Cor pulmonale
14	3	53	50	Congenital scoliosis	Fusion	Routine follow-up
15	15	65	50	Polio	Fusion	Cor pulmonale
16	13	64	51	Congenital scoliosis	None	Decompensation
17	6	60	54	Congenital scoliosis	Fusion	Routine follow-up
18	3	58	55	Tuberculosis	Bed rest, braces	Unrelated problem
19	11	70	59	Adolescent idiopathic scoliosis	Fusion	Routine follow-up
20	4	63	59	Congenital scoliosis	Fusion	Routine follow-up
21	15	78	63	Adolescent idiopathic scoliosis	Fusion	Routine follow-up
22	6	78	72	Infantile idiopathic scoliosis	Fusion	Cor pulmonale
23	11	87	76	Juvenile idiopathic scoliosis	Fusion	Routine follow-up

the fusion was done far too late, it did prevent an early death.

Case 23 (76-year follow-up)

An 11-year-old boy was first seen in 1929 with rapidly progressive juvenile idiopathic scoliosis of 48°. He underwent posterior spine fusion with tibial autografts and casting. At 6 months after surgery, his fusion looked solid, and the curve was 46°. The curve progressed over the next 5 years to 100° owing to "crank-shafting" and a too short fusion proximally. He was then fused down to L3. When seen in 1978 at age 61, he had low back pain complaints and was requesting early retirement. The curve measured 120°. Our last contact was by telephone in 2002 at age 84. He stated that he had no back pain, no neck pain, no respiratory problems, and was "much too busy to come in for an appointment." He died in 2007 at age 89 of natural causes. He has been previously reported.[6]

Discussion

Such a diverse group of patients does not lend itself to any type of statistical analysis, but much can be learned by these long-term outcomes (Table 1). When dealing with scoliosis, whether idiopathic, neuromuscular or congenital, it is crystal clear that early surgery gives far better results than "end-of-growth" surgery.

Solid fusions with some correction can be achieved without instrumentation, something orthopedic surgeons knew very well during the 1940s and 1950s but seem to have been forgotten in this era of implants. The two patients with severe isthmic spondylolisthesis were fused "in situ" and have excellent long-term results.

Neglected early-onset scoliosis, regardless of diagnosis, led to cor pulmonale and early death. This is a lesson from the past—and one we need to remember.

Although difficult to accomplish, such long-term follow-ups give us valuable insight into the effect of our treatments over the lifetime of our patients, something that short-term follow-ups of 2 or 5 years do not provide.

Conclusion

Failure to treat early-onset scoliosis leads to early death due to cor pulmonale. It is well to remember that surgical procedures that appear "primitive" to us today still provided life-saving outcomes.

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