Operative Treatment of Spinal Deformities in Patients with Cerebral Palsy or Mental Retardation

An Analysis of One Hundred and Seven Cases*†

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ABSTRACT: **Many controversial areas exist in the treatment of spinal deformities in patients with cerebral palsy or mental retardation, including the benefits of surgery, the use of traction for preoperative curve correction, the need for a combined anterior and posterior approach, the need to fuse to the sacrum, and the moral question of operating on these severely handicapped patients. To help to clarify these questions, the surgical treatment of spinal deformities in patients with cerebral palsy or mental retardation was analyzed in 109 patients who were treated from May 1948 through December 1979 at the Twin Cities Scoliosis Center, Minneapolis and St. Paul, Minnesota. Seventy-seven (71 per cent) of the patients had cerebral palsy and thirty-two (29 per cent) had only mental retardation. One patient had lordosis only and one had kyphosis only. Of the 107 patients with scoliosis, forty-four had Group-I (double balanced) curves and sixty-three had Group-II (large unbalanced lumbar or thoracolumbar) curves. The incidence of the two curve types was found to differ in those patients with only mental retardation, in ambulatory patients, in patients who lived at home, and in patients with pelvic obliquity.**

The treatment programs in use during the thirty-one years that are covered by this study were: cast correction and posterior fusion followed by a long postoperative supine period; posterior fusion and Harrington instrumentation; and a two-stage combined anterior and posterior fusion and instrumentation with a very short postoperative supine period. The indications for surgery were curve progression (63 per cent), loss of function (35 per cent), and the magnitude of the curve (77 per cent). Traction was found to be of no use for correcting the curve, but was very useful in controlling the uncooperative patient.

The length of follow-up averaged 4.5 years (range, two to twenty-nine years). All but ten of the patients achieved a solid spine fusion. Eight of the ten had painless pseudarthroses without loss of correction and two had pseudarthroses with loss of correction. The Group-II curves were better treated by the two-stage combined approach, which gave better correction of the scoliosis and a lower rate of pseudarthrosis compared with posterior fusion and instrumentation alone. The improvement in the results using the combined approach caused us to use this approach also in selected Group-I curves in the presence of a significant lumbar component. Fusion to the sacrum was necessary only when pelvic obliquity was present or sitting balance was absent. One patient was functionally worse postoperatively, eighty-two showed no change, and twenty-four showed improvement.

The complication rate was high (81 per cent). The most frequent complications were pressure sores, wound problems, instrumentation problems, and an increase in the length of the curve. Pseudarthroses occurred in 17 per cent and infection, in 5 per cent of the patients. Three patients died and one became paraplegic. In our opinion, surgery can be of benefit in this group of severely handicapped patients.

Spinal deformities in patients with cerebral palsy or mental retardation are often progressive and disabling[5]. Although this problem is best managed by early recognition and control of the curve by bracing before the deformity becomes severe, many patients do not come to the attention of the orthopaedic surgeon until a severe and progressive spinal deformity has developed, function has been lost, and pain is present. Prior to the introduction of new techniques and instruments, the surgical treatment of these patients was difficult and prone to failure[6]. Even with recent advances, controversy still exists concerning the indications and methods of treatment for spinal deformity in these patients. Typical of the statements relating to this controversial subject are these: (1) patients with severe cerebral palsy or severe mental retardation, or both, do not benefit from the surgical correction of scoliosis, and thus surgery is not indicated in this group of patients; (2) traction is useful in the treatment of these curves; (3) it is very difficult, if not impossible, to obtain a solid spine fusion in these patients unless combined anterior and posterior fusion is done; and (4) as patients with cerebral palsy have neuromuscular scoliosis, a spine fusion must include the sacrum.

The purpose of this paper is to review our experience

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with the operative treatment of spinal deformity in patients with cerebral palsy or mental retardation alone, and to examine these controversial statements.

Review of the Literature

There has been little awareness of the true incidence of scoliosis in cerebral palsy in the earlier literature. James, in 1967, reported only two patients with cerebral palsy in a series of 3000 scoliotic patients. Keats stated that scoliosis was seen infrequently in cerebral palsy and was non-progressive when present. Other reviews, however, have reported the incidence of scoliosis in cerebral palsy and mental retardation to range from 6.5 to 67 per cent, which is higher than in the general population, and to be highest in non-ambulatory patients[2,14,18-20].

One of the earliest and most interesting discussions of the treatment of spinal deformity in cerebral palsy was by Haas, who successfully treated a patient with lumbar scoliosis and pelvic obliquity with a variety of muscle-transfer procedures. The results of spine fusion in patients with cerebral palsy have been reported by Balmer and MacEwen and by Baumann. MacEwen reported the results following posterior spine fusion and Harrington instrumentation in fifteen patients, in whom the average curve was 72.5 degrees, the average correction was 50 per cent, and the pseudarthrosis rate was 20 per cent. Bleck noted the difficulty in managing these patients with a Milwaukee brace and, based on the experience of Bonnett et al.[5], recommended combined posterior and anterior fusion.

The results of different operative techniques for the treatment of scoliosis in thirty-three patients with cerebral palsy were reported in 1976 by Bonnett et al.[5]. These authors concluded: "Only the combined procedure appeared to give adequate correction and a low incidence of pseudarthrosis."

The early experience with patients with cerebral palsy from the Twin Cities Scoliosis Center was presented in 1976[1] and again in 1978[12]. Both studies suggested that the best method of operative treatment for lumbar and thoracolumbar curves in these patients is by a combined anterior and posterior fusion.

Materials and Methods

At the outset, an understanding as to what is included in the terms cerebral palsy or mental retardation is necessary. Brain damage before, during, or after birth has an effect on motor, sensory, cognitive, and emotional development, the important ones for classification being motor and cognitive development[15]. If there is delay in motor development *with* neural (motor) signs (increased muscle tone, spasticity, or hyperreflexia) these children are classified as having cerebral palsy, which may or may not be accompanied by mental retardation (cognitive involvement). In pure mental retardation, cognition is delayed and motor delay is possible, but there are *no* neural (motor) signs, although there may be a tendency toward hypotonicity. In reviewing the treatment of spinal deformities in these two groups of patients in our institution, we have found that the presentation, curve pattern, and treatment were generally the same.

From May 1948 through December 1979, 114 patients with the diagnosis of either cerebral palsy or mental retardation alone were operated on for a spinal deformity at the Twin Cities Scoliosis Center (Fairview and the University of Minnesota Hospitals, Minneapolis, Minnesota, and Gillette Children's Hospital, St. Paul, Minnesota). Five patients were excluded from the study because of inadequate follow-up or incomplete records, leaving 109 patients available for review. One hundred and seven patients had scoliosis as the primary deformity, while one patient had lordosis only and another had kyphosis only. The latter two patients are not included in the review. The cases of all patients were analyzed by chart and radiograph review. The functional status was evaluated by discussion with the parents or with the physical therapist and personnel who were caring for the patients in home-care facilities or institutions. The average length of follow-up was 4.5 years (range, two to twenty-nine years).

There were sixty-eight female and thirty-nine male patients. The average age at surgery was sixteen years (range, six to thirty-four years). Seventy-six patients (71 per cent) had cerebral palsy and thirty-one (29 per cent) had mental retardation alone. Of the patients with cerebral palsy, none had pure athetosis, fourteen had mixed athetoid and spastic cerebral palsy, fifty-two had spastic quadriplegia, and thirteen had monoplegic, hemiplegic, or diplegic involvement. Fifty-three patients in the cerebral palsy group were also mentally retarded, so that a total of eighty-four (79 per cent) of the patients in this series had some degree of mental retardation. Sixty-one patients were ambulatory. Sixty-two lived at home, twenty-seven were institutionalized, and eighteen were cared for in residential facilities.

The average of all of the presenting scoliotic curves was 86 degrees (range, 30 to 185 degrees). Forty-six patients also had a kyphotic deformity of more than 50 degrees (average, 83 degrees; range, 52 to 161 degrees). In the majority of patients the kyphotic deformity was in the area of the scoliotic curve, but was not a true kyphosis. The kyphotic deformity was due to spinal rotation, and represented the kyphosing scoliosis described by Stagnara et al. Lumbar lordosis of more than 60 degrees was seen in thirty-five patients (average, 79 degrees; range, 60 to 115 degrees) (Table I).

The preoperative curve patterns could be divided into two major groups that showed distinct differences in the patient population, in the treatment given, and in the results obtained.

Group-I curves were double curves with both a thoracic and a lumbar component (Fig. 1). Either these curves were well balanced and compensated, or the thoracic curve was the more significant curve with a fractional and poorly compensated lumbar curve below it.

TABLE I

CLINICAL AND RADIOGRAPHIC PRESENTATION

	Group I	Group II	Total
No. of patients	44 (40%)	63 (58%)	107
Average age at surgery (*yrs.*)	14	18	16 (6 to 34 yrs.)
Sex distribution	33 F, 11 M	35 F, 28 M	68 F, 39 M
No. with cerebral palsy	24 (55%)	52 (83%)	76 (71%)
No. with pure mental retardation	20 (45%)	11 (17%)	31 (29%)
No. with spastic quadriplegia	16 (36%)	36 (57%)	52 (49%)
Total no. with mental retardation	36 (82%)	48 (76%)	84 (79%)
Home care (*no.*)	31 (70%)	31 (49%)	62 (58%)
Institutional care (*no.*)	5 (11%)	22 (35%)	27 (25%)
Ambulatory (*no.*)	34 (77%)	27 (42%)	61 (56%)
Non-ambulatory (*no.*)	10 (23%)	36 (58%)	45 (44%)
Hip contractures (*no.*)	12 (27%)	28 (44%)	40 (38%)
Hip dislocation or subluxation (*no.*)	7 (16%)	15 (23%)	22 (20%)
Average scoliosis (*degrees*)	72	95	86 (30 to 185)
Average kyphosis (*degrees*)	64 (14 pts.)	90 (32 pts.)	83 (46 pts.) (52 to 161)
Average lordosis (*degrees*)	74 (15 pts.)	82 (20 pts.)	79 (35 pts.) (60 to 115)
Pelvic obliquity (*no.*)	26 (59%)	59 (94%)	85 (79%)
Average pelvic obliquity (*degrees*)	7	20	16 (1 to 63)
Average decompensation (*cm*)	3.8	5.4	4.8 (0.5 to 15)

Forty-four patients had a Group-I curve.

These curve patterns were seen most often in patients with only mental retardation and in patients who were ambulatory and lived at home, and were associated with fewer hip contractures and fewer subluxated or dislocated hips. The average curve was smaller and fewer patients had pelvic obliquity, which was usually minor (Table I).

The curves in Group II were more severe lumbar or thoracolumbar curves with marked pelvic obliquity. Sixty per cent of these patients had a short fractional curve between the end of the curve and the sacrum, and in 40 per cent the curve continued into the sacrum (Fig. 1). Sixty-three patients had a Group-II curve. These curves occurred more frequently in the cerebral palsy patients with spastic quadriplegia, in the non-ambulatory patients, and in the patients who were not cared for at home. The average curve was larger, and nearly all of these patients had significant pelvic obliquity. When the two subtypes of curve patterns are compared, the patients with a fractional lumbosacral curve showed a higher incidence of mental retardation alone and were more likely to be ambulatory. On the other hand, the patients in whom the sacrum was part of the curve had a higher incidence of spastic quadriplegia and of dislocated hips, and their pelvic obliquity was greater.

As in the Group-I patients, a kyphotic deformity,

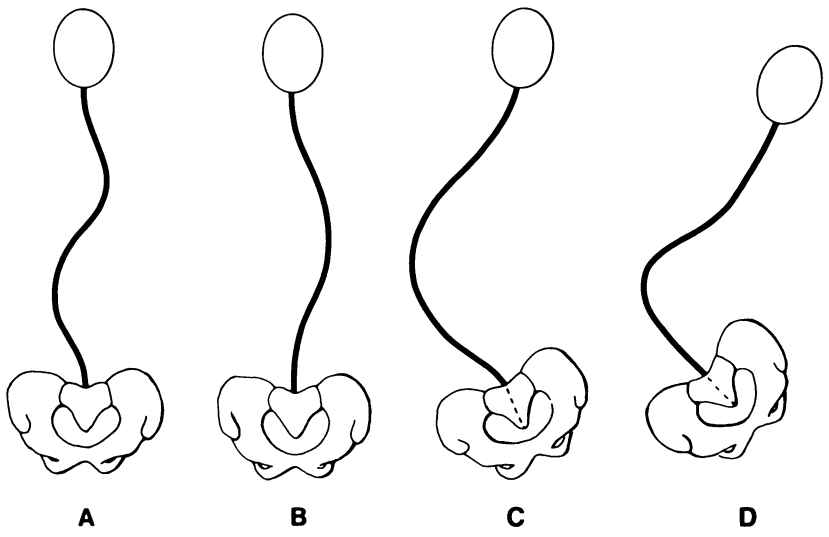

FIG. 1

Examples of curve patterns. Group-I curves are double curves with a thoracic and lumbar component. There is little pelvic obliquity and either the patient is well balanced (*A*) or the thoracic curve is more significant, with a fractional, poorly compensated curve below it (*B*). Group-II curves are large lumbar or thoracolumbar curves with marked pelvic obliquity. There may be a short fractional curve between the end of the curve and the sacrum (*C*) or the curve may continue into the sacrum (*D*).

when present, was due to rotation in the area of the scoliotic curve, and represented the kyphosing scoliosis described by Stagnara et al.

When the patients with mental retardation alone were compared with the patients with cerebral palsy, it was found that the majority of those with mental retardation had a Group-I curve, 85 per cent being true thoracic curves, while the curves of the patients with cerebral palsy were mainly in Group II. The patients with mental retardation were more often ambulatory, only four of the thirty-one (all in Group II) being unable to walk. The clinical and radiographic findings on preoperative evaluation were similar to the average for all of the patients, as noted in Table I, except that the average curve in the patients in Group II with mental retardation alone averaged 117 degrees.

Treatment

There have been numerous improvements in the treatment of scoliosis over the thirty-one years covered by this study (Fig. 2). Surgically, there has been greater use of two-stage combined anterior and posterior fusions, more frequent use of internal stabilization with multiple Harrington rods, and more use of heterogenous bank bone when autologous bone was inadequate. Postoperatively, a two-piece body jacket has replaced the cast, and mobilization of the patient begins as soon as the brace is fitted, usually seven to ten days postoperatively.

Nevertheless, the indications for surgery in patients with cerebral palsy or mental retardation remain controversial. Our present indications for operative treatment are the following, either singly or in combination: (1) curve progression documented on serial radiographs; (2) loss of functional status, either noted on follow-up examinations or reported by parents or attendants caring for the child; and (3) the magnitude of the curve. The size of the curve alone was not used as an indication for surgery except in children with near-normal intelligence, in whom any curve of more than 60 degrees was operated on. Usually the curve magnitude was a co-indication, combined with curve progression or loss of function. Sixty-eight patients (63 per cent) had surgery because of curve progression; thirty-eight (35 per cent), because of deterioration of function; and eighty-four (77 per cent), because of the magnitude of the curve. No patients in this group had complaints of pain or respiratory problems as an indication for surgery.

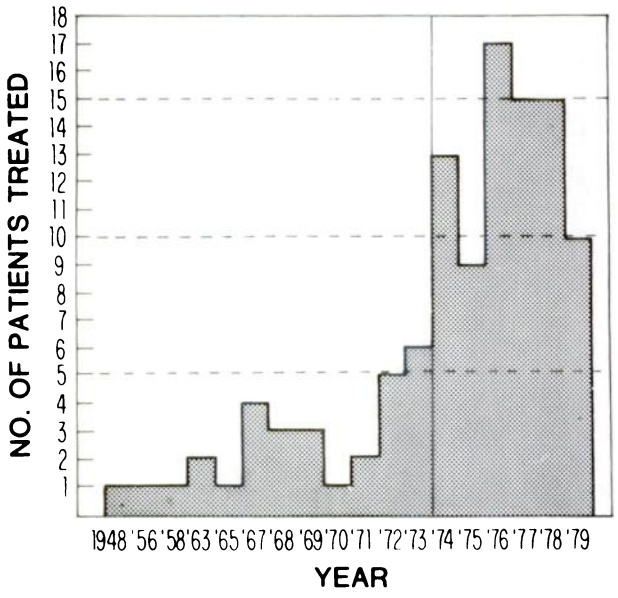

FIG. 2

Bar graph showing the number of patients treated each year. The graph is divided into the two treatment periods: 1948 to 1973 (thirty patients) and 1974 to 1979 (seventy-seven patients).

Twenty-four patients had had non-operative treatment for scoliosis prior to surgery, twenty-two with a brace and two with a sitting-support orthosis. The brace, usually a two-piece body jacket, had controlled the curve initially, but with growth or the onset of puberty the curve had progressed. Thirteen per cent of the patients also required surgery for the treatment of hip contractures or subluxation and dislocation. Seventy-two patients (67 per cent) had traction as part of the surgical procedure.

Group I (Figs. 3-A and 3-B)

Forty-one of the forty-four patients in this group had a posterior fusion with concomitant Harrington instrumentation, and three of them had a two-stage combined anterior and posterior fusion and instrumentation. One patient with a severe kyphotic deformity had an anterior interbody fusion without instrumentation. The other two patients, both of whom had a severe lumbar component in the double curve pattern, had an anterior fusion and instrumentation.

The average number of segments fused anteriorly was seven (range, six to eight segments) and the average blood loss (1919 milliliters) was 58 per cent of the estimated blood volume. The average number of segments fused posteriorly was thirteen (range, seven to seventeen). In six patients the fusion extended to the fifth lumbar segment and in eleven patients, to the sacrum. A single distraction rod was used in thirty-two patients and multiple distraction rods, in nine patients. Nine patients had compression rods, with transverse-loading cross-linking devices used in four. One patient had a Luque segmental fixation rod used in addition to other fixation, and multiple segmental sublaminar wiring was used with the Harrington distraction rod in two patients. The average blood loss (2215 milliliters) was 79 per cent of the estimated blood volume. Two patients who had a posterior fusion and instrumentation had an inadequate autologous bone graft, and they underwent exploration at six months to reinforce the fusion. At that time one was found to have a pseudarthrosis, which was repaired.

Postoperatively, the patients were kept at bed rest for an average of 4.6 weeks. With improved techniques, this time was reduced from seventeen to 1.5 weeks. The average period of immobilization (bed rest and cast or brace-

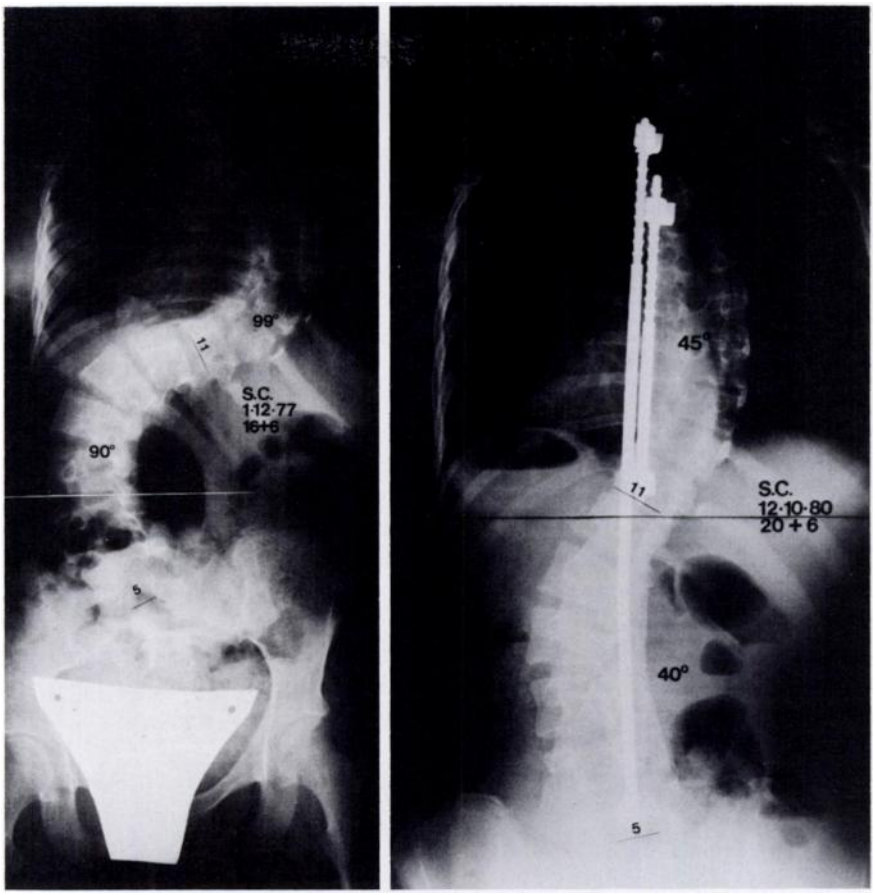

FIG. 3-A FIG. 3-B

Figs. 3-A and 3-B: Example of treatment of a Group-I curve. A girl with mental retardation alone was seen in 1977, at the age of sixteen years and six months, with increasing scoliosis. Functionally she was an unassisted community ambulator.

Fig. 3-A: An anteroposterior radiograph made with the patient standing shows a right thoracic curve of 99 degrees and a left lumbar curve of 90 degrees. A posterior fusion with Harrington instrumentation from the second thoracic to the fifth lumbar vertebra was done. She was immobilized in a Risser underarm cast and was allowed to walk fourteen days postoperatively. The total duration of cast immobilization was twelve months.

Fig. 3-B: Three and one-half years after operation, the fusion was solid. The fusion mass was excellent, and correction was maintained at 45 degrees and 40 degrees. The patient's functional status was unchanged postoperatively.

wear) was thirty-nine weeks (range, twenty-six to fifty-two weeks). A Risser-Cotrel cast was used in thirty-five patients; a halo cast, in five; and a two-piece body jacket or thoracolumbosacral orthosis, in four patients.

The hospital stay of all of these patients averaged fifty days. If the nine patients who were operated on early in the series are excluded, the average hospital stay decreased to twenty-eight days.

Group II (Figs. 4-A through 5-E)

Twenty-seven patients in this group of sixty-three had a posterior fusion alone, and thirty-six had a combined anterior and posterior fusion and instrumentation. In all but two patients, the anterior fusion was performed first and was followed two weeks later by the posterior fusion. The other two patients had had an anterior fusion and Dwyer instrumentation alone as the initial operative procedure, but because of curve progression above or below the area of the anterior fusion, posterior fusion and instrumentation was necessary two years later.

Nineteen patients had Dwyer and six had Zielke instrumentation. Anterior instrumentation was theoretically contraindicated in this group of curves because of the apparent kyphosis. However, this was not a true kyphosis, but a kyphosing scoliosis due to severe rotation of the scoliotic curve. Correct placement of the screws during instrumentation will cause derotation as the scoliosis is corrected, with reduction in the rotatory kyphosis[11]. The average number of segments fused anteriorly was seven (range, four to nine), and the average blood loss (1803 milliliters) was 54 per cent of the estimated blood volume.

A variety of preoperative, intraoperative, and postoperative traction methods were used in fifty-four of the sixty-four patients. Because of the rigidity of these curves, however, traction was found to have no effect on curve correction and was used mainly to control the patient and to facilitate nursing care.

There were a few patients in Group II in whom the posterior fusion was staged. This was either planned from the outset or was done because the initial posterior procedure had to be terminated due to excessive blood loss. There was thus a total of seventy posterior procedures in this group (excluding posterior surgery for the treatment of complications). The average number of segments fused

was fifteen (range, nine to eighteen), with the fusion in four patients extending to the fifth lumbar vertebra and in forty-nine, to the sacrum. As the curves in this group were larger and tended to be more rigid, multiple Harrington rods were used in 46 per cent of the patients. Eleven patients had compression rods with transverse linking (usually wire loops) between the compression and distraction rods. A Luque rod and segmental wiring was used in one patient and multiple segmental sublaminar wires, in three other patients. The average blood loss (2629 milliliters) was 84 per cent of the estimated blood volume. Two patients who had a posterior fusion without instrumentation had an inadequate autologous bone graft and underwent exploration at six months to reinforce the fusion. At that time both had pseudarthroses, which were repaired.

The length of bed rest postoperatively averaged ten weeks. When the patients who were operated on early in the series are excluded, the average time in bed decreased from twenty-three to two weeks. The average period of postoperative immobilization was forty weeks. A Risser-Cotrel cast was used for thirty patients; a halo cast, for nineteen; and a two-piece body jacket or thoracolumbosacral orthosis, in fourteen patients.

The hospital stay for all of the patients in this group averaged ninety-six days. If the nineteen patients who were operated on early in the series are excluded, the average total hospitalization time was forty-six days.

Results

All of the patients were followed for a minimum of two years, with an average follow-up of 4.5 years. A solid fusion was obtained in all but ten patients. All of these ten patients had asymptomatic radiographic defects in the fusion mass, but only two showed a loss of correction.

The scoliosis was corrected from an average curve of 86 degrees preoperatively to 32 degrees after surgery (63 per cent correction). There was an average loss of correction at follow-up of 7 degrees. The kyphosis was corrected from an average of 83 degrees to 38 degrees and the lordosis, from 79 to 58 degrees. In the eighty-five patients with pelvic obliquity, the average preoperative obliquity of 16 degrees was corrected to 5 degrees at follow-up. Decompensation was corrected from an average of 4.8 to 2.0 centimeters. The results in Group I and Group II were very similar, except for a slightly higher percentage correction of the scoliosis and greater correction of pelvic obliquity in

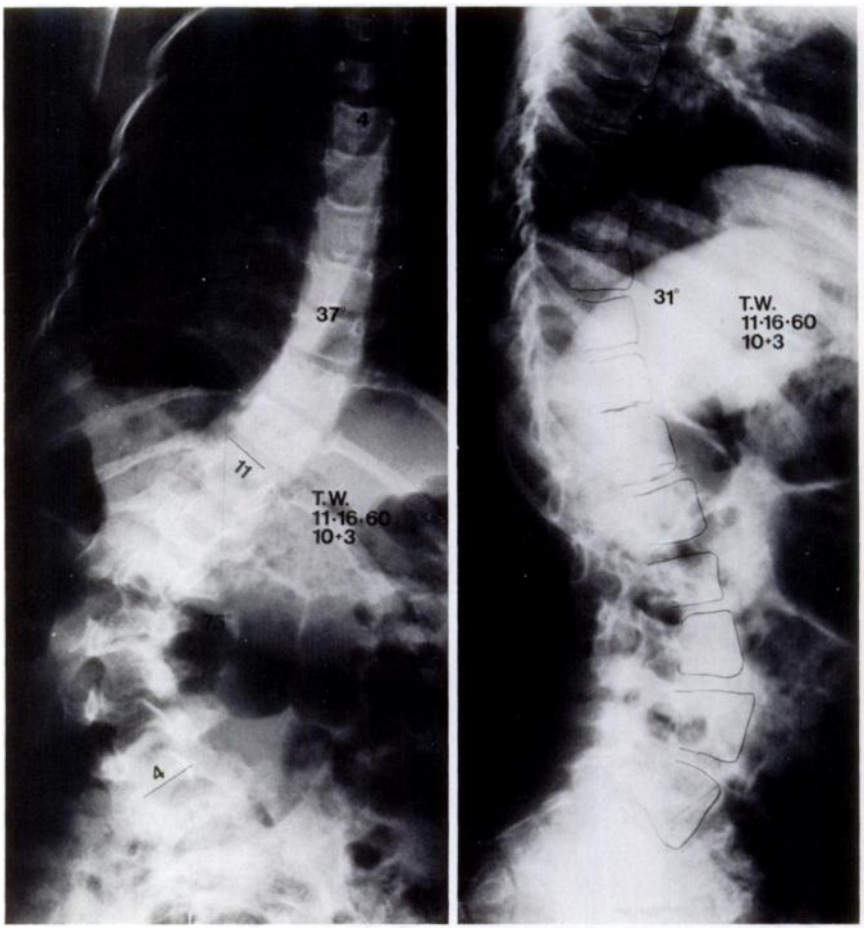

FIG. 4-A

FIG. 4-B

Figs. 4-A through 4-F: Example of treatment of a Group-II curve by posterior surgery. A girl with spastic quadriplegia and mental retardation was first seen in 1960, at the age of ten years and three months.

Figs. 4-A and 4-B: There was a left lumbar scoliosis of 75 degrees (Fig. 4-A) and the kyphosis measured 31 degrees (Fig. 4-B). The patient was able to walk with assistance and was not treated.

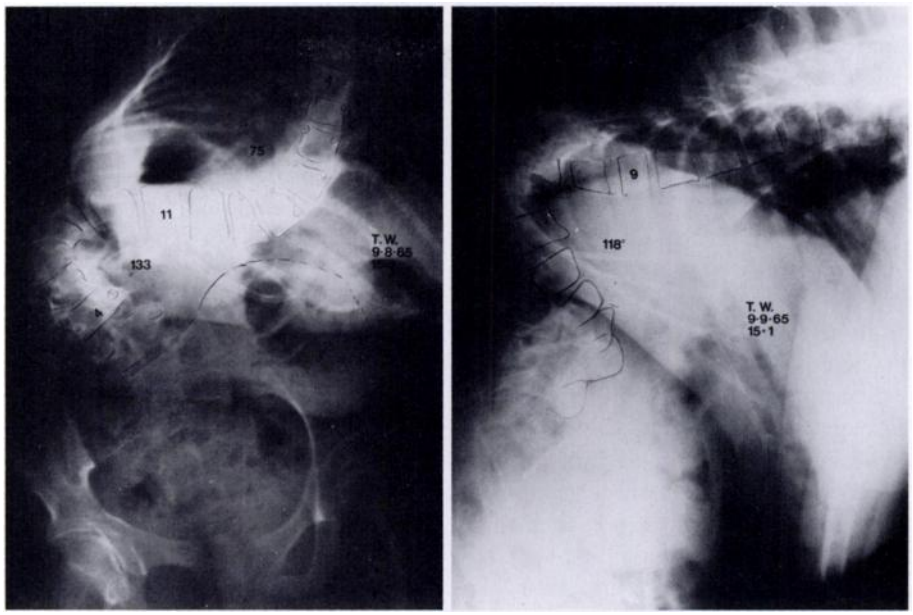

FIG. 4-C FIG. 4-D

Five years later, the scoliosis had progressed to 133 degrees (Fig. 4-C) and the kyphosis measured 118 degrees (Fig. 4-D). The patient was treated with halo-femoral traction for nine weeks, followed by posterior fusion and instrumentation. She had multiple pseudarthroses, stiff hips and knees, pressure sores, and curve extension. Five procedures (four posterior and one anterior) were done on the spine, and the patient was immobilized for a total of three years. She spent a total of 3.9 years in the hospital.

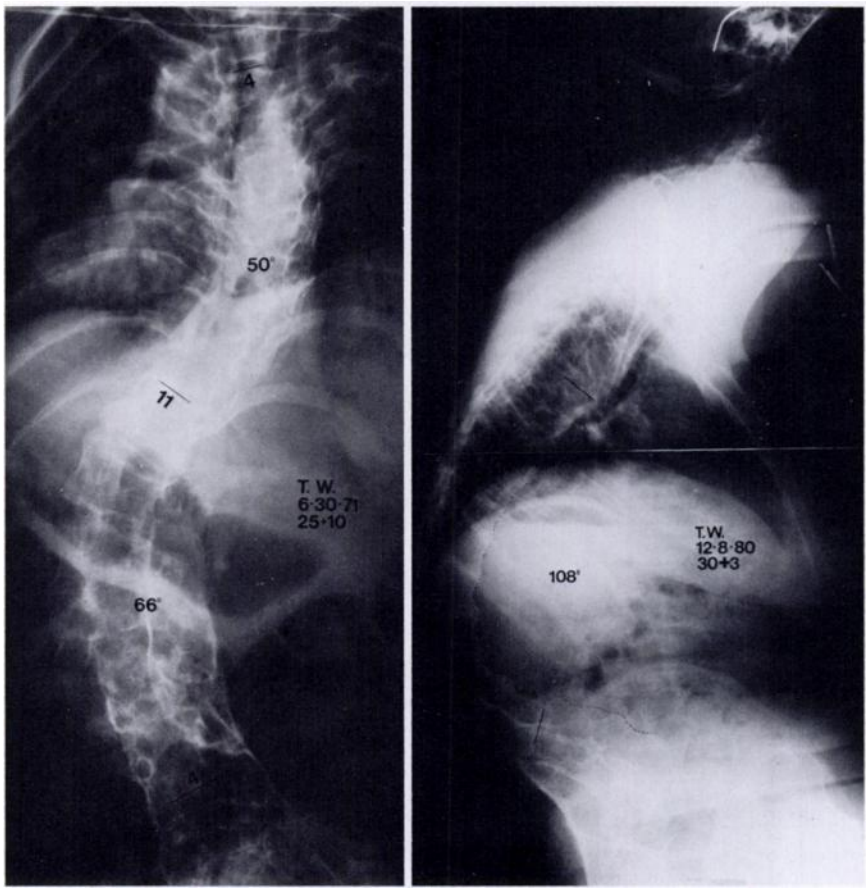

FIG. 4-E FIG. 4-F

Fig. 4-E: In 1971, the scoliosis measured 66 degrees and the kyphosis, 80 degrees. A residual pseudarthrosis caused further kyphotic deformity.

Fig. 4-F: In 1980 the kyphotic deformity measured 108 degrees. The patient had back pain and was able to walk with assistance. The early treatment plan for this Group-II curve did not achieve a solid fusion, and in addition it resulted in numerous complications, with prolonged hospitalization and high treatment costs.

Group II. The results in the patients with mental retardation were very similar to those in the patients with cerebral palsy except for a slightly lower percentage of correction of the scoliosis in the former.

To assess the patients' functional ability, a modified Rancho Los Amigos classification[8] was used, in which Group-1 patients were independent walkers or community ambulators; Group 2, household or physical therapy ambulators; Group 3, independent sitters; Group 4, dependent sitters; and Group 5, non-sitters or bedridden. Using this classification, eighty-two patients remained unchanged, twenty-four patients improved one or more levels, and only one patient deteriorated, going from Group 2 preoperatively to Group 3 postoperatively. As seen in Figure 6, the biggest change was found in the patients who were in Group 4 preoperatively (the dependent sitters). Such patients are wheelchair-bound, often requiring additional support such as side-pads or a sitting-support orthosis to maintain an upright sitting posture. With increasing curvature and loss of sitting ability, more attention is required from the people taking care of the child, as constant repositioning is necessary to keep the child upright.

Postoperatively there was an improvement in functional ability in more than half of these patients, while the remainder required far less care to keep them in the upright position. This improvement in functional ability is difficult to measure, but the greater ease of functioning and the less difficulty and time required by personnel in the care of these children was commonly remarked on in all functional groups. Other factors, such as endurance and the ability to use the hands, were difficult to measure, but were also improved in the majority of patients.

Complications

Two hundred and six complications (Table II) occurred in eighty-seven patients (81 per cent). Thirty patients were in Group I and fifty-seven patients, in Group II.

FIG. 5-A FIG. 5-B FIG. 5-C

Figs. 5-A through 5-E: Example of treatment of a Group-II curve by the combined approach. This girl had spastic quadriplegia and mental retardation.

Fig. 5-A: When the patient was first seen in 1965, at the age of nine years and nine months, the right thoracolumbar curve measured 15 degrees. No treatment was recommended, and she was lost to follow-up.

Figs. 5-B and 5-C: When the patient was next seen, in 1978, she had lost the ability to sit independently and the curve had progressed to 92 degrees (Fig. 5-B). A lateral radiograph made with the patient sitting showed a 74-degree kyphotic deformity (Fig. 5-C). Note that the apical vertebrae are seen in an anteroposterior projection due to vertebral rotation, this being an example of so-called kyphosing scoliosis, and thus anterior instrumentation was possible. An anterior fusion with Zielke instrumentation was done, followed two weeks later by a posterior fusion with multiple Harrington rods. The patient was immobilized postoperatively in a Risser cast. The total hospitalization lasted thirty-two days and the total immobilization time was ten and one-half months.

Pressure Sores

The most common complication was a pressure sore. A pressure sore developed in a total of thirty patients (28 per cent): in two patients while they were in traction and in twenty-eight patients during the immobilization period. In all but four patients, the pressure sore was successfully treated by cast or brace modifications and local treatment; the other four patients required a skin graft or secondary closure.

Minor Wound Problems

In twenty-eight patients (26 per cent) minor wound problems developed, including wound hematoma, wound drainage without infection, wound separation, superficial infection, and retention of a Hemovac tube. The retained Hemovac tube was removed surgically, and all of the remaining minor wound problems were successfully treated with local wound care.

Instrumentation Problems

Twenty-five patients had problems related to the instrumentation. The most common problem was a dislocated hook, which occurred in seventeen patients. In seven patients the hook dislocated at surgery, requiring a change of hook site, and in eleven patients the dislocation occurred six days to one year after surgery. One patient had both an intraoperative and a postoperative dislocation. Surgery for relocation of the hooks was performed in seven patients, none of whom had a pseudarthrosis, while in one of the four patients whose hook was not reinserted an asymptomatic pseudarthrosis developed. Twelve of these seventeen patients with a dislocated hook had a straight rod inserted without contouring for pre-existing kyphosis or lordosis. Two patients had breakage of a Dwyer cable postoperatively, and both had a pseudarthrosis. The average curve in this group of patients was not significantly different from that of the total group.

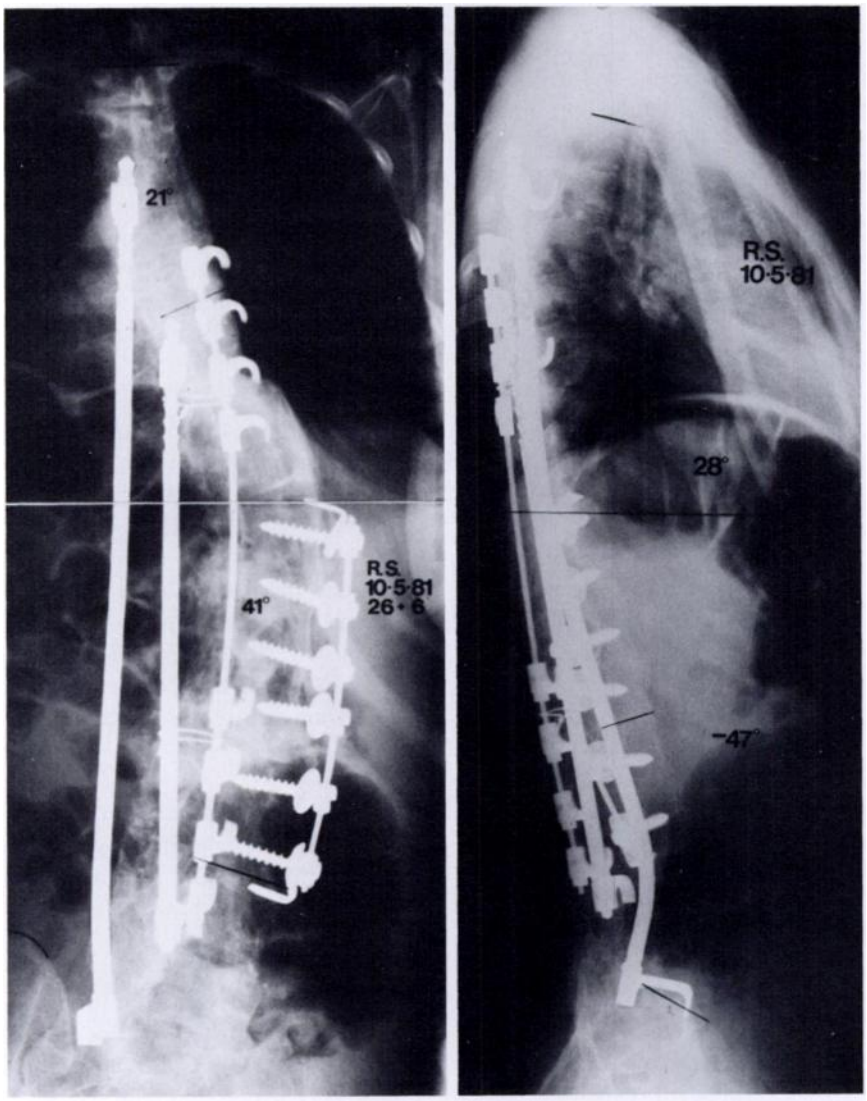

FIG. 5-D FIG. 5-E

Three years postoperatively the fusion was solid. The scoliosis was corrected to 41 degrees (Fig. 5-D) and the kyphotic deformity measured 28 degrees (Fig. 5-E). Note the rod contouring for lumbar lordosis on this lateral radiograph, with a solid lumbar fusion. The patient's functional level had improved and she was an independent sitter.

TABLE II

COMPLICATIONS

	Group I	Group II	Total
No. of patients	30 (68%)	57 (91%)	87 (81%)
Pressure sores	8 (18%)	22 (35%)	30 (28%)
Minor wound problems	7 (16%)	21 (33%)	28 (26%)
Instrumentation problems	7 (16%)	18 (28%)	25 (23%)
Pin problems	4 (9%)	13 (20%)	17 (16%)
Curve extension	7 (16%)	11 (17%)	18 (17%)
Pseudarthrosis	9 (20%)	9 (14%)	18 (17%)
Pulmonary	0	13 (20%)	13 (12%)
Genito-urinary	5 (11%)	6 (10%)	11 (10%)
Gastrointestinal	5 (11%)	1 (1.5%)	6 (5%)
Deep wound infection	1 (2%)	5 (8%)	6 (5%)
Death	0	3	3 (2.8%)
Paraplegia	1	0	1 (0.9%)
Others*	8 (18%)	16 (25%)	24 (22%)

* Cutaneous rash, six; blood-transfusion reaction, six; excessive bleeding, three; stiff hips and knees, two; transient nerve palsy, two; flatback, two; dural laceration, two; sinus tachycardia, one; seizure, one; and anorexia, one.

Pin Problems

Seventeen patients had a superficial infection or loosening of either the halo pin or the femoral pin, in traction or in the halo cast. All were successfully treated by local treatment of the infection and replacement of loose pins.

Curve Extension

Eighteen patients had curve extension or progression above or below the original curve. The most common reason was too short an initial fusion. Four patients with a pseudarthrosis had progression of the curve in the fusion area.

Of interest are two patients, both in Group II, who were treated initially by anterior fusion and Dwyer instrumentation alone, one being operated on at our center and one, elsewhere. One had a pseudarthrosis, but in both severe decompensation and progression of the curve developed in the unfused spine, and both required second-stage posterior fusion and Harrington instrumentation two years after the anterior fusion.

Pseudarthrosis

In eighteen (17 per cent) of the 107 patients a pseudarthrosis developed. Three patients were found to have a pseudarthrosis at the time of the six-month exploration and reinforcement procedure. In the remaining fifteen patients the diagnosis was made radiographically, only one patient having a moderate amount of pain associated with the pseudarthrosis. There was no correlation between the magnitude of the scoliosis or the lower level of the fusion and the development of a pseudarthrosis.

The type of surgical procedure, however, did correlate with the development of a pseudarthrosis. A pseudarthrosis developed in fifteen of the sixty-eight patients who had a posterior fusion alone, for a rate of 22 per cent, while only two (5.4 per cent) of the thirty-seven patients who had the combined approach as the initial procedure had a pseudarthrosis; in one patient a pseudarthrosis developed after a Dwyer procedure alone. Of the Group-I curves, all nine patients in whom a pseudarthrosis developed had a posterior spine fusion with or without Harrington instrumentation. Of the Group-II curves, six (22 per cent) of the twenty-seven patients who had a posterior spine fusion alone had a pseudarthrosis, while only two (5.9 per cent) of the thirty-four patients who had the combined approach as the initial procedure had a pseudarthrosis. There was also a difference in the pseudarthrosis rate depending on the date of surgery — the rate was 33 per cent before 1974 and 10 per cent after 1974 — and the curve pattern, the rate decreasing from 33 to 17 per cent in Group-I curves and from 37 to 4.5 per cent in Group-II curves.

A total of twenty-two procedures were done in fourteen patients for treatment of a pseudarthrosis. The other four patients, all with a Group-I curve, had an asymptomatic pseudarthrosis without loss of correction and thus did not undergo pseudarthrosis repair. The repair was posterior in all but three patients; these were the only three with multiple attempts at repair. One patient had five procedures and still had a residual pseudarthrosis at the time of writing, and the other two patients had three procedures each. At follow-up, eight patients had a solid fusion and ten patients had an asymptomatic pseudarthrosis radiographically, with curve progression in two patients of 29 degrees of scoliosis in one and 19 degrees of scoliosis and 24 degrees of kyphosis in the other.

Deep Wound Infection

Six patients (5 per cent) had a deep wound infection. Infection developed in the posterior incision in five patients and in the anterior thoracotomy incision in one patient. All of these patients were successfully treated: four had débridement and insertion of irrigation and suction tubes and one had only débridement of the thoracotomy incision. In the other patient, débridement and the use of suction-irrigation tubes did not cure the infection. The wound had to be treated open, with subsequent secondary closure and multiple skin grafts.

Death

The over-all mortality rate was 2.8 per cent. Three patients with spastic quadriplegia died of bronchopneumonia at five to nine months following surgery, in the postoperative immobilization period.

Paraplegia

Paraplegia developed in one patient postoperatively. This girl had a posterior spine fusion at the age of eleven years and five months, but the scoliosis and kyphosis continued to progress, and required posterior osteotomies and Harrington-rod instrumentation five years later. The operative procedure and immediate postoperative course were uneventful, but following an episode of hypotension

in the recovery room a sixth thoracic-level paraplegia developed. The rod was immediately removed, but no motor recovery occurred.

As seen in Table II, complications were more common in Group-II patients with significant spastic quadriplegia, either alone or with an athetoid component. Patients with mental retardation alone had fewer complications than those with cerebral palsy. The complication rate has also fallen since 1974, even though the majority of patients were treated in this six-year time-period. The only complications that were more frequent after 1974 were pulmonary, perhaps reflecting the fact that more severely handicapped patients are now being treated.

Discussion

The patient population in cerebral palsy clinics is changing. With the passage of Public Law 42-142, which legislates the opportunities for education that must be available to all children, children with more severe involvement are being seen for care. In addition, many of these children are being cared for in residential homes rather than in state institutions. The mental and physical involvement in these children varies greatly, with many being seen who have no spasticity, but instead have severe mental retardation and a mental age of less than six months. The series presented here, from a large cerebral-palsy spine clinic, reflects this patient population, with 29 per cent having mental retardation alone and 49 per cent having spastic quadriplegia.

The surgical management of this group of patients is very controversial, especially in light of the expense of surgery in patients with severe mental retardation. The questions that we have attempted to answer in this review are: Is surgery of benefit in these patients? Is traction useful for the treatment of these curves? Do all of these patients require a combined anterior and posterior fusion? Is fusion to the sacrum always necessary? Should surgery be performed in patients with severe mental retardation?

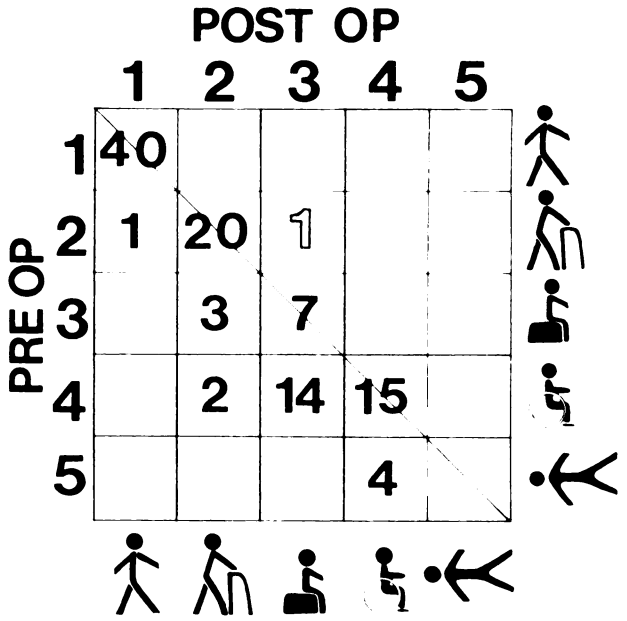

FIG. 6

Preoperative and postoperative functional ability using a modified Rancho Los Amigos classification[8]. The figures on the diagonal axis indicate the number of patients with unchanged function; the figures on the left, those with improved postoperative function; and the figure on the right, a patient who had loss of function postoperatively, going from a dependent walker to an independent sitter.

The indications for surgery in this group of patients varied, the most common being the magnitude of the curve, curve progression, or loss of function. Usually a combination of these indications was present. With increasing curvature, the sitting ability of the patient changes and support is needed for sitting. An independent sitter loses function, while a dependent sitter needs more support and constant repositioning to an upright posture. Although the majority of patients showed no postoperative change in their functional ability, a significant change was found in those who had been dependent sitters. More than half of these patients showed improvement in their functional ability, while the remainder needed much less support and repositioning to be maintained in an upright sitting position (Fig. 6). In addition, the parents or attendants caring for these patients usually noted that less time and difficulty was experienced in caring for a patient with a stable, balanced spine. Also, with the improvement in sitting, the use of communication devices or electric wheelchairs was possible.

Over the many years that this series covers, there have been changes in the surgical and postoperative protocol, with the most dramatic changes occurring in 1974. At that time, a more systematic approach to the care of the spine deformities in these patients was developed, with the establishment of a cerebral palsy spine service in 1977. Also, the combined two-stage anterior and posterior surgical approach was more commonly used, with multiple Harrington rods posteriorly, rod-contouring, and the use of heterogenous bank bone from the femoral head as supplementary bone graft. In addition, even with a fusion extending to the sacrum, patients were mobilized and returned to their preoperative functional status as soon as the postoperative cast or brace was applied. The most dramatic effect of this early mobilization was a reduction of the time spent in the horizontal position postoperatively from twenty-one to 1.9 weeks. In addition, the average hospital stay was reduced from 184 to thirty-eight days.

Although halo-femoral traction was used initially for curve correction, it was found to be ineffectual. No more correction can be obtained in the spine than can be demonstrated on a radiograph made with the patient supine while manual traction is being applied. We still use halo-femoral traction for selected uncooperative patients, to help to control the patient and to facilitate nursing care, but *not* for curve correction. For these uses, the traction is applied a day or two before the anterior fusion or during the same anesthetic session as the anterior fusion. This selective, limited use of halo-femoral traction has also helped to reduce the length of hospitalization.

The early return of the patient to the upright position postoperatively did not have an adverse effect on the rate of pseudarthrosis, even when the fusion extended to the sacrum, as the rate of pseudarthrosis fell from 33 per cent before 1974 to 10 per cent after 1974. In addition, with a more systematic approach to the care of these patients the incidences of all but one complication fell. The only complication whose incidence increased after 1974 was pulmonary problems, which was related to the greater use of the anterior approach after this date as well as to the increase in the number of more severely handicapped patients who were operated on.

The patterns of scoliotic curves could be divided into two distinct groups, which differed in their clinical presentation and treatment programs. Group-I patients had a higher incidence of mental retardation alone, and more of the children were able to walk and were cared for at home. Pelvic obliquity was less common and less severe in this group. A posterior fusion alone was usually sufficient to achieve a solid, balanced spine, with fusion to the sacrum rarely being necessary. The patients in Group II had large decompensated lumbar and thoracolumbar curves, with a higher incidence of spastic quadriplegia, and fewer patients were able to walk. The scoliosis was, on average, more severe than in Group I, and nearly all of the patients had a pronounced pelvic obliquity. The treatment of Group-II patients was either by a posterior fusion alone (twenty-seven patients) or by a combined approach (thirty-six patients). In addition, 80 per cent of the patients had a fusion that included the sacrum. When these two treatments for Group-II patients were compared, it was found that the combined approach gave better correction of the scoliosis (70 per cent compared with 55 per cent), slightly greater correction of the pelvic obliquity (75 per cent compared with 68 per cent), and a lower rate of pseudarthrosis (5.9 per cent compared with 22 per cent). The combined approach is thus the treatment of choice in Type-II curves, confirming the findings of Bonnett et al. The anterior approach with fusion and instrumentation of the lumbar curve, and placement of the lower screw in the fourth or fifth lumbar vertebra, has previously been shown to be very effective in correcting paralytic pelvic obliquity[16,17].

The lower pseudarthrosis rate that was achieved with the use of the combined approach in Group II suggests a role for this approach in selected Group-I curves. Group-I curves with a significant lumbar component, especially when there is decompensation, may benefit from an anterior fusion to more certainly achieve a solid fusion. Currently our approach is to use the combined approach in Group-I curves when there is a significant lumbar component. Anterior instrumentation is used in these curves only when there is no danger of overcorrecting the lumbar curve, with a resultant loss of balance and decompensation.

In reviewing the cases of these patients, it was found that it was possible to obtain a solid fusion and that the combined approach is not necessary in all patients. The combined approach did give markedly improved results in Group-II curves and is thus indicated for this curve type. Fusion to the sacrum was not necessary in all patients, being rarely indicated in Group-I curves and very common in Group-II curves. Fusion to the sacrum was performed in patients with pelvic obliquity or when sitting balance was absent and it was necessary to anchor the spine using the pelvis as a base.

The complication rate in this group of patients was very high. With a more systematic plan of care and with greater experience on the part of the physicians and the nursing staff, the number of complications has decreased. Contouring of the Harrington rod in the sagittal plane has been found to be important, especially in the lumbosacral area, as the insertion of a straight rod to the sacrum reduces lumbar lordosis, and the sacral fixation is tenuous. If the rod is contoured and a square-ended rod and square-holed sacral hook are used, the lumbar lordosis is maintained and the sacral hook is less likely to dislodge with the pelvic rotation that occurs with sitting.

With advances in surgical techniques and in postoperative care and immobilization, it is possible to successfully fuse scoliotic curves in patients with cerebral palsy and pure mental retardation. In these patients, especially when there is severe mental retardation and a mental age of less than six months, the moral question arises as to whether one *should* embark on a plan of surgical treatment.

It is our opinion that with the functional improvement and stabilization seen in these patients, even in light of the high complication rate, stabilization of the spine should be performed when indicated. The improvement that is obtained in sitting balance leads to a significant reduction in the care needed to constantly reposition the patient in the sitting position, which lessens the demands on those who care for these patients. In addition, an improved ability to communicate and useful hand function are possible when the patient is not constantly attempting to maintain an upright sitting position.

Conclusions

On reviewing our experience with the surgical treatment of scoliosis in patients with cerebral palsy and in those with mental retardation alone, we concluded that scoliosis is a significant problem for these patients. When surgery was indicated, it frequently was possible to obtain a solid posterior fusion alone, and the combined anterior and posterior approach was not necessary in the majority of patients. The combined approach, however, was the treatment of choice in Group-II curves, and resulted in a marked reduction of the rate of pseudarthrosis. Extension of the fusion to the sacrum was indicated only when pelvic obliquity was present or when the patient was not able to sit independently.

Traction was found to be of no benefit in correcting a curve preoperatively, but was found to be invaluable in

controlling difficult patients. The complication rate was very high, but was reduced by a more systematic plan of care and new surgical techniques. A gratifying improvement in the patients' functional abilities occurred after the spine fusion. We concluded that when indicated, surgery should be performed even in patients with severe mental retardation.

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