

COTREL-DUBOUSSET INSTRUMENTATION IN NEUROMUSCULAR SPINAL DEFORMITY

A FIVE TO ELEVEN-YEAR FOLLOW-UP STUDY

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Thirty patients with neuromuscular spinal deformity treated with Cotrel-Dubousset instrumentation were followed for 5 to 11 years. Fusion was extended to the pelvis in 21 patients. Mean frontal correction was 56% postoperatively and 51% at final follow-up. Thoracic kyphosis was reduced from an average 44° to 33° in nine patients; kyphosis of the entire spine in 21 patients was converted to a balanced sagittal plane. Average pelvic obliquity and torso decompensation improved from 24° to 13° and from 273 to 204 mm respectively. Complications included infection (10 cases) and loss of hardware fixation (4 cases); except for one infection, all complications presented in the pelvic fixation group. Surgery was considered to have improved quality of life and would be recommended in all cases. Surgical treatment of neuromuscular spinal deformity with Cotrel-Dubousset instrumentation allows a long-lasting correction and improves quality of life, but is associated with a significant complication rate.

Keywords : neuromuscular scoliosis ; Cotrel-Dubousset instrumentation.

Mots-clés : scoliose neuro-musculaire ; instrumentation de Cotrel-Dubousset.

INTRODUCTION

Patients with neuromuscular disorders are at high risk for the development of severe spinal deformity. Treatment of neuromuscular spinal deformity (NSD) presents a great challenge owing not only to its progressive nature but also to frequently associated extraspinal abnormalities that include pelvic obliquity, hip dislocation, limited balance or ability to sit, back pain and, in some circumstances, a serious decrease in pulmo-

nary function (2, 3). As NSD is difficult to control with a brace, surgical correction and fusion are generally recommended for these patients, even though it is a complex procedure not devoid of complications (1-3, 9, 14, 15). In many cases, fusion to the pelvis is required and poses one of the most challenging instrumentation problems (10).

Because of the severe osteopenic bone commonly found in these patients, segmental fixation techniques are considered preferable to single distraction rod constructs. Luque-type sublaminar wire systems have been used in the majority of recently reported series on surgical treatment of NSD with good clinical results (1-3, 9, 14, 15). The successful use of variable hook-rod systems (such as Cotrel-Dubousset (CD) instrumentation) in idiopathic deformities (6, 8) has prompted their use in neuromuscular scoliosis, and these instrumentations are presently accepted alternative systems to augment fusion in NSD (13). However, there are few reports on the long-term results with these more recent systems, and in particular CD instrumentation, for neuromuscular patients (10, 11).

The purpose of this study is to evaluate the results, complications and long-term outcome of arthrodesis augmented with CD instrumentation to treat neuromuscular spinal deformity.

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MATERIAL AND METHODS

From 1986 to 1993, spinal arthrodesis with CD instrumentation was performed at our institution on 30 patients with NSD. Their medical and radiographic records were complete, and they were all personally interviewed and examined by the authors at the latest follow-up evaluation, performed at an average of eight years after surgery (range, five to 11 years).

There were seven male and 23 female patients. The average age at operation was 17 years (range, nine to 33 years). Nine patients (30%) had cerebral palsy, eight (26,7%) had poliomyelitis, six (20%) had myelomeningocele and the remaining seven had other neuromuscular conditions. The pattern of scoliosis was thoracic in five, thoracolumbar in three, lumbar in seven, and a long c-curve was present in 15 patients. Twelve of the long c-curve patients had associated rotational thoracolumbar kyphosis. The reasons for operation were progression of the curve and deterioration of functional status in all cases. Two patients with poliomyelitis had significant spinal pain prior to surgery.

Anterior arthrodesis was performed as a first stage in 18 patients one week before the posterior procedure. The average number of segments that were released and included in the arthrodesis anteriorly was 5,9 (range, three to seven). Anterior Zielke instrumentation was used in one patient with poliomyelitis. The remaining 12 patients were considered to require only posterior fusion. Corrective forces included intraoperative traction, rod rotational maneuvers and segmental distraction and compression. Arthrodesis was extended to the pelvis in 21 patients. The indications for pelvis fixation were curve extension into the sacrum, fixed pelvic obliquity greater than 15° and limited balance when sitting. Several strategies were used for sacral fixation during the years when surgery was performed : bilateral sacral screws in seven cases, sacral screws and staples in four, iliosacral screws in six, bilateral sacral staples in two and Chopin blocks in two cases. Traction was not used preoperatively in any case ; it was used between operations in six of the patients who required both anterior and posterior surgery. Bone autograft was supplemented with femoral head allograft only in the eight posterior fusions most recently performed. External immobilization was used postoperatively on four patients for six months and in one patient for one year mainly because of such factors as osteopenic bone, tenuous instrumentation fixation or poor compliance of the patient.

Preoperative, postoperative and final functional ambulation was classified according to Hoffer *et al.* (7)

In addition, patients were requested to fill in an outcome questionnaire consisting of ten questions (table I). The patient's caregiver was requested to help filling in the questionnaire when the patient was considered unable to answer the questions by himself. Medical charts were reviewed for complications.

Coronal and sagittal plane corrections, pelvic obliquity and torso decompensation were measured on upright radiographs made preoperatively, immediately postoperatively and at final follow-up evaluation. In patients who had poor balance when sitting, the upright radiograph was made with the minimal possible support. Decompensation of the torso was measured as described by Osebold *et al.* (12) and pelvic obliquity by measuring the angle formed by the intersection of the intercrestal line and a horizontal line parallel to the inferior margin of the radiograph.

RESULTS

Correction of deformity and functional ambulation

Coronal plane correction improved from an average preoperative curve of 78° degrees (range, 50° to 130°) to an average immediate postoperative curve of 34° (range, 10° to 65°, 56% correction) and average final curve of 38° (range, 11° to 75°, 51% correction). In nine patients, thoracic kyphosis improved from an average 43° preoperatively to 30° immediately after operation and 33° at final follow-up, and lumbar lordosis remained basically unchanged. Kyphosis of the entire spine in 21 patients was converted to a balanced thoracic kyphosis and lumbar lordosis (fig. 1).

In the group of 21 patients with fusion to the pelvis, the pelvic obliquity averaged 24° (range, 15° to 38°) prior to surgery, improved to a mean of 11° (range, 0° to 28°) postoperatively and averaged 13° (range, 0° to 30°) at final follow-up. Decompensation of the torso in this group improved from an average of 270 mm (range, 50 to 250 mm) preoperatively to a mean of 200 mm (range, 25 to 210 mm) postoperatively and 204 mm (range, 25 to 215 mm) at final follow-up.

Preoperatively, nine patients could walk with aids and 21 used a wheelchair to some extent ; 16 of these required support for balance when sitting. Postoperatively, nine additional patients could sit in a wheelchair without aids ; functional

Table I. — Outcome questionnaire answers

Question	Choices	Number
1. Occupied / Employed	a. No for problems unrelated to the spine b. No for spine-related problems c. Yes (Specify : _____).	9 3 18
2. Spinal pain	a. Yes b. No.	3 27
3. Dependence / Aids for ambulation	a. Able to walk. b. Unassisted sitting. c. Assisted sitting. d. Unable to sit.	5 11 14 0
4. Memories of hospital stay	a. Good b. Bad. c. Indifferent	— 24 6
5. How were complications faced by the patient and family ?	a. There were no complications b. They seemed bearable c. They seemed unbearable	13 13 4
6. Did spinal surgery improve quality of life and patient management initially ?	a. Yes b. No.	30 —
7. Has the benefit from surgery persisted until present time ?	a. Yes b. No.	27 3
8. Is the spine the limiting factor for activity and quality of life at present time ?	a. Yes b. No.	9 21
9. Was spinal surgery worth being performed ?	a. Yes b. No.	30 —
10. Would you recommend this type of surgery to another patient in the same circumstances ?	a. Yes b. No.	30 —

ambulation remained unchanged in the rest of the cases. At final follow-up, five patients with poliomyelitis were able to walk with aids, 11 patients reported using a wheelchair without assistance and 14 required assistance for wheelchair use.

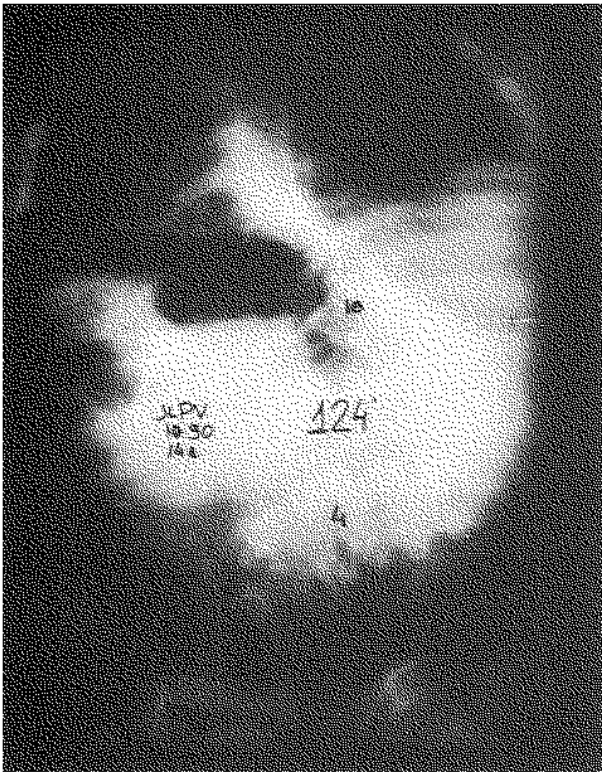
Complications

There were 18 complications in 17 patients (56.7%) (table II). Early complications included femorocutaneous nerve dysfunction in two cases, one hemothorax and one acute respiratory failure. All resolved without sequelae. Ten patients presented deep infection ; in nine of them, the pelvis had

been included in the arthrodesis and the primary diagnosis was either myelomeningocele (six cases) or poliomyelitis (three cases). The remaining infection presented eight years after surgery in a

Table II. — Postoperative complications

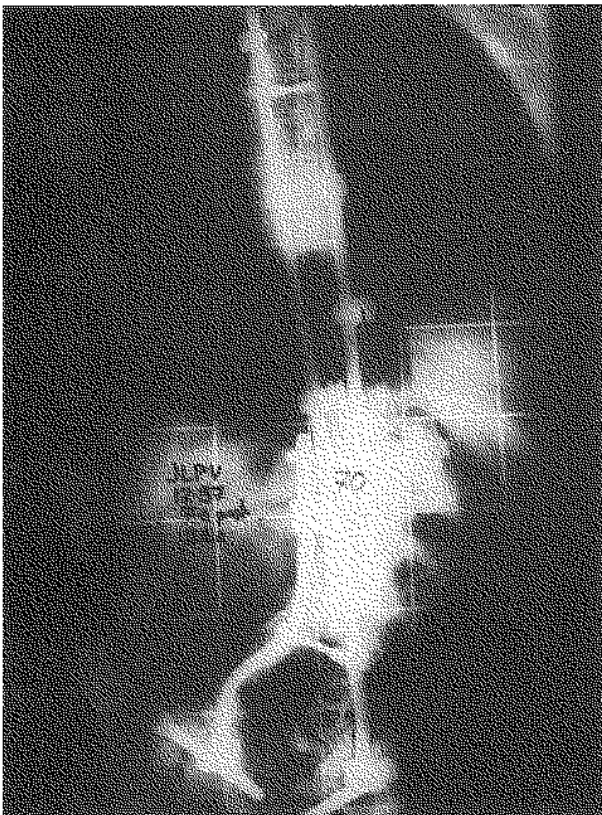
Early	Hemothorax	1
	Acute respiratory failure	1
	Transient femorocutaneous dysfunction	2
Late	Infection	10
	Hardware failure	4



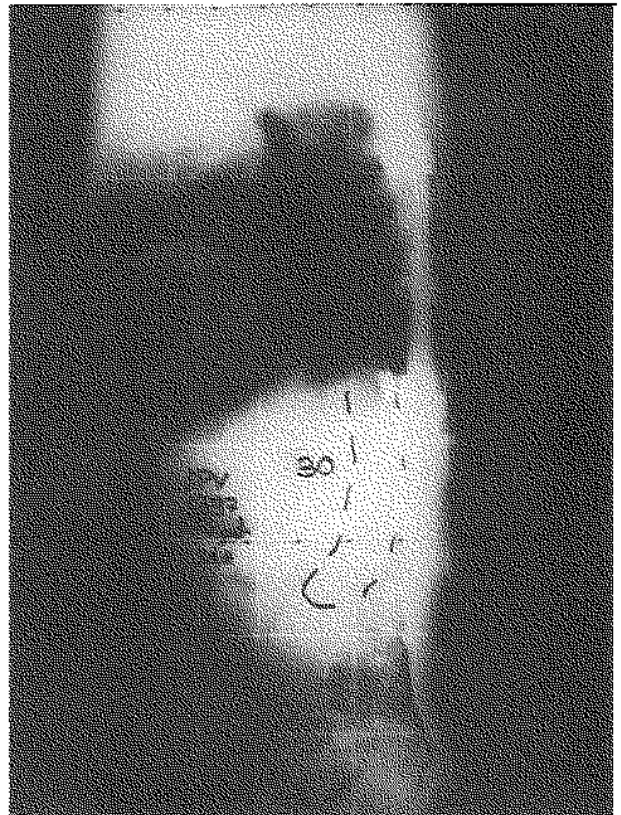
A



B



C



D

Fig. 1. — Preoperative anteroposterior (A) and lateral (B) radiographs of a patient with severe neuromuscular spinal deformity. Seven years after surgery, satisfactory frontal (C) and sagittal (D) spinal alignment is maintained. Note loss of sacral fixation on one side.

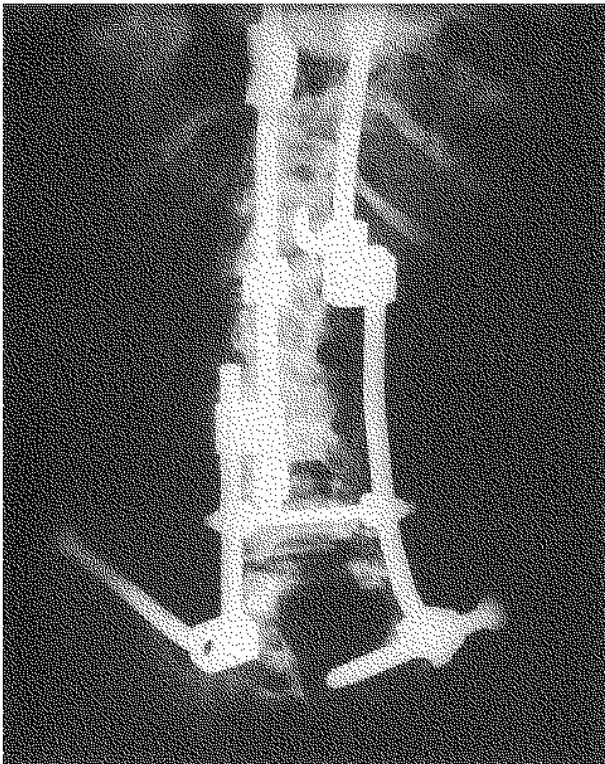


Fig. 2. — Anteroposterior radiograph of the lumbosacral spine shows left iliosacral screw loosening.

patient who had been treated without pelvic fusion. There were four hardware fixation failures, all related to pelvic fusion and none to infection. The two Chopin blocks used in this series failed; one became loose and the other was dislodged from the instrumentation with simultaneous distal DTT breakage. There were also two cases of iliosacral screw loosening (fig. 1, 2). All cases developed solid fusions. At final follow-up, hardware removal had been performed in eight patients with and one without sacropelvic fusion.

Outcome questionnaire

All poliomyelitis patients filled in the outcome questionnaire by themselves. Myelomeningocele patients answered the questionnaire by themselves with the help of their caregivers for some of the questions. The remaining questionnaires were filled out almost completely by patients' caregivers. All caregivers were relatives of the patients and had

taken care of them throughout follow-up. Table I summarizes the answers to the questionnaire in all 30 cases.

Eighteen patients reported an occupation or employment at final follow-up evaluation. Eleven were students and the remaining seven had various sedentary jobs. All receive at least some disability compensation. The spine was considered the factor mainly responsible for not having an occupation or employment in only three of the remaining 12 cases. Spinal pain was referred by only three patients, all with poliomyelitis as the primary diagnosis. Five patients with poliomyelitis were able to walk with aids, 11 referred using a wheelchair without assistance, 14 required assistance for wheelchair use and no patient were bedridden.

The hospital stay was remembered as a bad experience in most cases. Thirteen of the 17 patients who had complications found it difficult to cope with them; the remaining four thought complications had been bearable. Spinal surgery was considered to have improved quality of life and patient management initially in all cases. Only three poliomyelitis patients considered that the benefit from surgery had not continued until the time of most recent evaluation. The spine was considered the limiting factor for activity and quality of life at present time in only 9 cases; associated extraspinal abnormalities were thought to be more limiting than the spine itself in the remaining 21 patients. In all cases spinal surgery was considered worth while and would be recommended to another patient in the same circumstances.

DISCUSSION

Cotrel-Dubousset instrumentation was designed to provide bilateral segmental fixation of the spine while achieving a two-dimensional and possibly three-dimensional correction of the spinal deformity (6). After the satisfactory use of CD instrumentation for the surgical treatment of idiopathic scoliosis (8), its indications were expanded to include the more complex neuromuscular spinal deformities (13), but there is little information about the long-term outcome of spinal arthrodesis

augmented with CD instrumentation in neuromuscular scoliosis (10, 11). According to our results, substantial correction of the deformity, an excellent fusion rate and adequate patient and caregiver satisfaction can all be achieved with CD instrumentation in NSD, but at the price of significant complication and reoperation rates.

There are several features that make NSD more difficult to correct than idiopathic scoliosis. Neuromuscular patients frequently have osteopenic bone that provides poor instrumentation fixation. In addition, demands placed on the instrumentation system are significantly increased if fusion needs to be extended to the pelvis because of dysraphic posterior lumbosacral segments, severe torso decompensation or fixed pelvic obliquity that impairs sitting ability. Patient noncompliance and extraspinal problems also contribute to endanger correction (1-3, 9, 14, 15).

The preferred instrumentation system for these complex deformities has traditionally been the Luque segmental system, using the Galveston technique when pelvic fixation is required (1-3, 9, 14, 15). Several authors have reported acceptable results with Luque instrumentation in NSD. Broom *et al.* (3) reported on 74 patients with neuromuscular scoliosis treated using Luque-rod segmental instrumentation, with Galveston fusion to the pelvis in 38 (51.4%) cases. The mean curve was corrected from 73° preoperatively to 38° postoperatively (52.1% correction) and there was a mean loss of correction of 4° at an average follow-up of 42 months (range, 2 to 7.3 years). Complications included one death, three deep infections, six sets of broken rods and one case of distal rod migration. Boachie-Adjei *et al.* (2) reported on 46 patients with neuromuscular scoliosis treated with Luque segmental instrumentation, with fusion extended to the sacrum in 39 (84.8%) cases. The mean curve was corrected from 74° preoperatively to 39° at an average follow-up of three years (52.7% correction). Their complication rate was 48%, with 15% rod failure and 6.5% nonunion rate. Modifications of surgical technique and greater experience seem to have improved the results of Luque system in NSD (1, 14, 15).

Reported short and midterm experience with CD instrumentation in NSD seems to compare favorably with above-mentioned results using Luque segmental instrumentation. Neustadt *et al.* (11) published their results on 18 patients with neuromuscular spinal disorder treated using CD instrumentation into the pelvis. Several pelvic fixation devices were used; double sacral screws were the most common. The mean frontal plane curve was corrected from 70° preoperatively to 38° postoperatively (54.3%) and there was a mean loss of correction of 3° at an average follow-up of 28 months. Pelvic obliquity was improved in nine patients from a preoperative mean of 22° to 11° at follow-up. There was a single case of perioperative hardware failure and one deep infection, but no nonunion or rod breakage. Miladi *et al.* (10) reviewed 154 patients with neuromuscular scoliosis and pelvic obliquity treated with CD instrumentation. Pelvic fixation was achieved with iliosacral screws in all cases. Frontal correction ranged from 53% to 70% with a loss of 3% to 14% after an average follow-up of five years. Correction of pelvic obliquity ranged from 60% to 84% and loss of correction was mild. Complications included five deep infections, two nonunions, four iliosacral screw dislodgements and two other instrumental failures.

Our results with a longer follow-up parallel those of Neustadt *et al.* (11) and Miladi *et al.* (10) in terms of deformity correction and fusion rate. Corrections averaged 51% in the frontal plane and 54% in pelvic obliquity 5 to 11 years after surgery. Kyphosis of the entire spine was converted to a balanced thoracic kyphosis and lumbar lordosis in 21 patients, and average thoracic kyphosis was corrected from 43° to 33° in nine patients. In addition, none of the patients developed nonunion or permanent neurological deficits. However, our rate of instrumentation failure was 13.3%, in the range of reported figures using the Luque technique. This is in accordance with a biomechanical and clinical study that reported the Galveston method of sacral fixation to be superior to CD pelvic devices (4). The two Chopin blocks and two of the iliosacral screws used in our patients failed. Infection was also very frequent in our series,

which might in part be related to the high number of myelomeningocele patients included. It should be emphasized that this study includes the first neuromuscular patients treated with CD instrumentation at our institution and that the various pelvic fusion devices employed were in evolution at the time of application; the learning curve surely played a role in the high complication rate, as has been the case using alternative systems (1).

Patient-perceived results are not always taken into consideration when assessing the benefits of a given surgical treatment in a neuromuscular patient population. The answers to the questionnaire used in this study show that spinal surgery is considered beneficial for neuromuscular spinal deformity, even though some patients still consider the spine their limiting factor and complications were difficult to face when they happened. Comstock *et al.* reported similar patient and caregiver satisfaction rates in a cerebral palsy population treated with Luque instrumentation despite the surgical complexity and expected complications associated with the procedure (5).

In conclusion, CD instrumentation allows a safe and long-lasting correction of NSD with high fusion rates and patient and caregiver satisfaction. However, CD pelvic fixation devices do not always provide solid pelvic fusion in neuromuscular patients, and a higher complication rate should be anticipated when arthrodesis extension to the pelvis is required. Selective use of the most effective pelvic fusion devices and greater experience with the system is expected to improve the results of CD instrumentation in NSD.

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SAMENVATTING

J. SANCHEZ-SOTELO, F. J. SANCHEZ PEREZ-GRUESO. Cotrel Dubousset instrumentatie voor neuromusculaire wervelkolomafwijkingen : een 5 tot 11 jaar follow-up.

Dertig patiënten met een neuromusculaire wervelkolomafwijking werden behandeld met een Cotrel Du-

bousset instrumentatie en 5 tot 11 jaar later teruggezien. Bij 21 was er fusie tot op het bekken. De gemiddelde frontale correctie bedroeg 56% postoperatief en 51% op de laatste follow-up. De thoracale kyfose werd gereduceerd van 44° tot 33° bij 9 patiënten; kyfose van de globale wervelkolom werd omgebouwd tot een gebalanceerd sagitaal plan. De gemiddelde bekken inclinatie en torso verbeterde van 24° tot 13° en van 273 tot 204 mm. Bij 10 was er een infectie en bij 4 was een falen van de hardware. Behalve één infectie waren alle complicaties in de bekken groep.

De ingreep verbeterde de levenskwaliteit en werd door allen aangeraden naar andere patiënten toe. Chirurgische behandeling van spinale deformaties met Cotrel Dubousset instrumentatie geeft een blijvende correctie, verbetert de levenskwaliteit maar gaat gepaard met een belangrijke complicatie ratio.

RÉSUMÉ

J. SANCHEZ-SOTELO, F. J. SANCHEZ PEREZ-GRUESO. L'instrumentation de Cotrel-Dubousset dans les déformations neuromusculaires du rachis. Résultats avec un suivi de 5 à 11 ans.

Les auteurs rapportent avec un recul de 5 à 11 ans, les résultats obtenus dans le traitement de déformations

neuromusculaires du rachis avec l'instrumentation de Cotrel-Dubousset. L'arthrodèse a été étendue au bassin chez 21 patients. La correction dans le plan frontal a été en moyenne de 55% en post-opératoire et de 51% au dernier examen. La cyphose thoracique a été réduite d'une valeur moyenne de 44° à 33° chez neuf patients; 21 patients avaient une cyphose globale et ont recouvré un rachis équilibré dans le plan sagittal. L'obliquité pelvienne moyenne est passée de 24° à 13°, le déséquilibre du tronc a été réduit de 273 à 204 mm. Comme complications, on a relevé 10 cas d'infection et 4 cas de défaillance de la fixation; à l'exception d'une infection, toutes ces complications sont survenues dans le groupe avec arthrodèse étendue au pelvis. Les patients considéraient que l'opération avait amélioré leur qualité de vie et ils la recommanderaient à d'autres. Le traitement chirurgical des déformations neuromusculaires du rachis avec l'instrumentation de Cotrel-Dubousset permet une correction de longue durée et améliore la qualité de vie, mais est associé à un taux de complications élevé.