Revision of surgical correction of scoliosis in pediatric patients affected by neuromuscular pathology using Unit Rod instrumentation

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Abstract

Purpose The purpose of this study is to present a large consecutive series of pediatric patients affected by neuromuscular scoliosis treated with Unit Rod instrumentation.

Methods Retrospective review of 160 children treated with Unit Rod instrumentation between 2003 and 2010. This review analyzed complications, radiographic outcome, time surgery, average blood loss, hospital staying and functional outcome of patients and parents after surgery. The follow-up of 80 patients is at more than 5 years.

Results Scoliosis was corrected from a mean of 56° range 21° to 76° Cobb (68 %). Pelvic obliquity was corrected from a mean of 24° to 5° (74 %). Major complications are not related to the technique but to the pathology.

Conclusion Unit Rod instrumentation is a common standard technique, is considerably less expensive than the other system, and remains a good indication for the treatment of the spinal deformity in children affected by neuromuscular disease.

Keywords Neuromuscular disease · Spine deformity · Surgical management · Unit Rod instrumentation · Scoliosis

Purpose

In children affected by neuromuscular disease in particular: Duchenne muscular dystrophy (DMD), spinal muscular atrophy (SMA), cerebral palsy (CP), Rett syndrome,

scoliosis represents the most common pattern of spinal deformity. Scoliosis in these patients is not responsive to orthotic management and causes significant trunk and pelvic imbalance [1–6]. The pelvic obliquity and the curve stiffness represent factors that worsen the quality and expectative of life in these children. The scoliosis associated with this pathology progresses rapidly during pubertal growth 2°–4° per month, curve stiffness increases as well and progression occurs after skeletal maturity [7].

Instrumentation for the treatment of scoliosis in neuromuscular disease evolved over time. Luque developed the concept of segmental spinal fixation with sublaminar wire [8] and Galveston improved the technique for the pelvic fixation [9, 10]. The Unit Rod instrumentation was developed to use the Galveston pelvic fixation and sublaminar wire with a single rod to improve construct stiffness [11, 12]. Previous report has indicated the good results with the use of Unit Rod in treatment of neuromuscular scoliosis [13]. The goal of this study is to report the incidence of surgical complication, degree of deformity correction, the caretaker's and parents perceived outcome and to prove that Unit Rod is still a valid instrumentation in 2013 for the treatment of children affected by neuromuscular scoliosis.

Materials and methods

The two senior authors reviewed Medical records, spine radiographies and subjective questionnaires of 160 consecutive patients with neuromuscular spine deformity. They were treated with Unit Rod instrumentation with fusion to the pelvis, between 2003 and 2010 at the “V. Buzzi” Children Hospital. The study included 85 patients affected by Cerebral Palsy: 25 with SMA 2, 18 with Myelomeningocele, 17 with Duchenne muscular

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dystrophy, 9 with Rett Syndrome, 1 with Poliomyelitis, 1 with Friedrieichs Atassia, 1 with Arthrogryposis, 4 with Neuroblastoma, 8 patients were able to walk. We treated 71 male, 85 female, aged between 10 and 19. In 21 patients, we associated the anterior surgery.

The surgical complications were divided into intraoperative and post-operative.

Spine radiographies with the patients supine in anteroposterior (AP) and latero-lateral (LL) projection were taken before surgery and at 1–3–6–12 months of follow-up and then once a year for 5 years. All the patients were evaluated by the first author using Cobb method.

A non-validated questionnaire and Scoliosis Resource Society (SRS)-22 assessing patient's functional improvement after spinal arthrodesis were formulated and administered to the parents by an independent investigator.

The questionnaire focused on assessing the effectiveness of the surgery in correcting spinal deformity, improving patients' physical appearance, ability to lie on bed, sitting tolerance, ability to stand or walk, head control, ability to eat or feed, respiratory function, back pain, mental capacity, ability to use hands and ease of care.

Procedure

The operative indications on the basic disease in patients affected by CP were for scoliosis greater than 60° in DMD 30° in SMA 50°. We associated anterior release when the curve was greater than 100° or when the lordosis with retroversion of the pelvis was too stiff.

The instrumentation used in this study is Unit Rod precontoured quarter inch (6.25 mm) diameter Stanley Steel rod, from 31 to 41 cm in length. We fused from T2 or T3 into to the pelvis, preparing the tunnel from the posterior spine into iliac crest. Decortication of the transverse processes and lateral laminae with extensive facetectomies was routinely performed. We use sublaminar wires at each level and contour the 1.2 mm diameter wires during surgery and double them at L5 and T2.

We always reinforced fusion bone site with freeze-dried granulated corticocancellous bone allograft mixed with autogenous bone harvested from the spinous processes. All patients received prophylactic antibiotic treatment repeated after 4 h. During distraction, they received cortisone 30 mg/kg and we monitor nerves' root answer with Somatosensory evoked potentials SEP [17]. Patients were transferred to the Intensive Care Unit (ICU) in the immediate post-operative period. Nutritional support was provided by central venous hyper alimentation.

No post-operative immobilization was used and physical therapy program started on the 5th day. They return in wheelchair after 10 days without brace (Figs. 1, 2).

Results

Surgery time ranged between 3.45 and 8.15 h when associated with anterior release.

Before surgery, the mean value of scoliosis was 78°, between 40° and 120°, correction from a mean of 56° range 21° to 76° Cobb (68 %). Pelvic obliquity before surgery

Fig. 1 Pre- and post-surgery in a patient affected by CP

Fig. 2 X-ray pre- and post-surgery in patients affected by CP with ITB

was between 10° and 56° and it was corrected range 24° – 5° (74 %). Loss of correction of 5° during the first year was 12 %. In posterior procedure, the average blood loss was 2.1 l. ICU stay was 3 days, and the hospital stay was 14 days. Mean time surgery was 4 h. In the case with anterior and posterior associated procedure, blood loss was 2.8 l, mean time surgery was 6 h, ICU stay was 4 days and hospital stay was 16 days.

Major complications included 1 intraoperative death (affected by DMD), 1 death after 43 days for lungs infection, 1 instrumentation removed for deep infection, 11 cases of superficial infection treated with prolonged antibiotic management. There was one case of pancreatitis in patients affected by cerebral palsy, one kidney insufficiency from the blood saver, one glomerular damage by therapy, one thoracic duct rupture during thoracoscopy, 4 ileo paralysis that resolved in 4 weeks, 4 fractures of the pelvis, 6 breakages of the rods in patients able to walk, one neurologic bowel in a case with associated Ponte osteotomy, 4 cases of prominent proximal end of the rod that was removed.

Eleven cases of broken sublaminar wires noted at the X-ray, 3 consecutive rupture of the rod in the same patients affected by myelomeningocele in which we did not perform anterior release.

We analyzed the variables to determine significant relationship. Higher pre-operative scoliosis was correlated with increased surgical time and number of days in hospital. In lumbar lordosis over 60° we faced increased time of surgery and complications with pelvic fixation, stiff kyphosis also increased time of surgery and in many cases

we bent the rod in situ. We did not have in time crank shafting [14, 15].

Caretakers, parents and patients satisfaction evaluated with SRS questionnaire and subjective modified questionnaire reported an improvement for 74 %, of cases, no worsening, (0 %), SRS mean value 4.2 (Fig. 3).

We stopped using SRS score because it is not specific for neuromuscular scoliosis, even if validate. Since 2 years we have been using a modified Bridwell questionnaire more specific for children affected by neuromuscular scoliosis even if not validated.

The more sensitive parameters improved are: pain, sitting tolerance, patients physical appearance, ease of care (dressing, bathing), digestion and respiratory function [16]. We did not have any complications related to the instrumentation after 5 years follow-up, except for the rod breakage in those patients able to walk 4 years after surgery and one we re-intervened because she was symptomatic.

Conclusion

The use of the Unit Rod instrumentation for correction of neuromuscular scoliosis has been previously reported in smaller patients' population: 34 pediatric patients [12] with good results.

The present study demonstrates the possibility to gain excellent correction of the spinal deformity using the Unit Rod instrumentation with reduced operative times. The results are comparable to the ones in the literature and with

Fig. 3 Results of the SRS 22 6 month after surgery

the new third generation instrumentation using multiple pedicle screw and iliac bolt. When new instrumentation is being introduced, there is also a significant increase in implant cost, which should be balanced by benefits obtained by the use of these new techniques.

Since there is a well-documented height degree satisfaction of caretakers after deformity correction, the selection of the appropriate operative technique and type of instrumentation should depend on achieving optimum correction of the curvature while minimizing the complication rate. In contrast, the specific instrumentation used is not likely to reduce the risk for the common medical

complication related to such a major surgical undertaking. The Unit Rod instrumentation may be considered the gold standard technique and the primary instrumentation system for the treatment of pediatric patients with neuromuscular scoliosis, because it is simple to use and it provides good stability with the sublaminar wires also in presence of osteoporosis, although it is technically more difficult in patients with hyperlordosis. Unit Rod instrumentation is a common standard technique, is considerably less expensive than the other system, and remains a good indication for the treatment of the spinal deformity in children affected by neuromuscular disease (Figs. 4, 5).

Fig. 4 Patient affected by myeloblastoma with severe hyperlordosis we associated anterior release and posterior fusion pre-operative and post-surgery

Fig. 5 X-ray of the same case

Conflict of interest None.

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