Original Article

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Surgical treatment of scoliosis in Ullrich Congenital Muscular Dystrophy: a case series of 3 patients

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SUMMARY Scoliosis in Ullrich Congenital Muscular Dystrophy (UCMD) is very common, with a reported incidence of more than 50%, and it is rapidly progressive. There are no previous studies which specifically focus on scoliosis surgery in UCMD patients. This article reports three cases of scoliosis surgery in UCMD, focusing on operative course, clinical and radiological results achieved, fusion area and complications, with a 2-year follow-up. The surgical technique adopted for vertebral arthrodesis included: high-density pedicle screw systems, asymmetric rods contouring and direct vertebral rotation. The summary results shown a significative correction of the coronal deformity, with a reduction of the mean Cobb angle from 49° to 25° post-operatively. Mean pelvic tilt remained stable, while L5-tilt showed a decrease from 10° to 6°. Mean screw density was 1.92. None of the patients required extended fixation to S2. No major complications were reported, and patients maintained their pre-operative walking ability. All the patients reported a subjective improvement in quality of life, with a better sitting comfort. In conclusion, posterior spinal fusion with high-density pedicle screw systems and direct vertebral rotation may be safe and effective in surgical correction of scoliosis in UCMD. If pelvic obliquity and L5-tilt are less than 15°, could be possible to achieve an optimal spinal and pelvic balance even without sacral or pelvic fixation.

Keywords neuromuscular scoliosis, ullrich congenital muscular dystrophy, spine surgery

1. Introduction

Ullrich Congenital Muscular Dystrophy (UCMD) is a rare muscular dystrophy firstly described by Otto Ullrich in 1930 (*1*). It is caused by a mutation in one of the COL6A1-A2-A3 genes, which leads to a deficiency of collagen VI in the extracellular muscular matrix (*2-4*). The prevalence of UCMD is reported to be 1.3 per million (*5*) and scoliosis is present in more than 50% of them (*6*).

Surgical treatment of scoliosis in patients with UCMD is more challenging than idiopathic scoliosis, because of both the patients' respiratory comorbidities and the characteristics of the curve. In fact, scoliosis in UCMD has an early onset (reported average age of 6.5 years at diagnosis (*7*)) and is rapidly progressive (reported maximum progression rate of 16.2 ± 10° per year (*6*)). This eventually results in highly rigid, severe, and extended scoliotic curves, with high peri-operative risks and surgical complexity. Moreover, scoliosis in UCMD may be associated with pelvic obliquity, which makes sitting position difficult and worsens the disability of the patients. Therefore, long fusions to the sacrum and/or pelvis are often required to correct pelvic obliquity and to restore an optimal sitting balance. This poses an additional problem, since the poor bone quality and the large loads found in the lumbosacral region may lead to instrumentation failure and pseudoarthrosis.

Because of the rarity of UCMD, there are currently no studies that focused on the results of the surgical treatment of scoliosis in this specific disease. The aim of this article was to review the clinical and radiological results of scoliosis surgical treatment in 3 UCMD patients treated at our Institute over the last 5 years.

2. Patients and Methods

Three consecutive patients with UCMD diagnosis who underwent scoliosis surgery at our institution between 2015 and 2018 were included.

To make the initial diagnosis, genomic DNA from UCMD patients and unaffected parents was extracted from peripheral lymphocytes after informed consent following standard diagnostic methods (*8*): PCR primers

were designed to amplify all the 107 exons of COL6 genes and their flanking intronic regions. Skeletal muscle biopsies were collected during surgery and fresh frozen samples were used to confirm genetical disorder.

Patients were clinically and radiologically reviewed pre-operatively, post-operatively and after 2 years of follow-up.

Radiologic measurements were performed by two independent operators on the radiographs to evaluate the scoliotic deformity, obtaining the following parameters: Cobb angle, flexibility index, pelvic obliquity (using the Maloney method), L5-tilt (calculated as the angle between a line across the superior endplate of L5 and the intercristal line), thoracic kyphosis, lumbar lordosis, and vertebral rotation (according to Nash and Moe) (*9*). All measurements were done with the help of software (Carestream Health Italy, Inc., Genova, Italy).

All the patients received pre-operative echocardiography and pulmonary function tests: none of them had cardiac impairment, but all the patients had restrictive lung disease. All the patients received spinal CT-scan and MRI before surgery, to help the surgical planning and to rule-out any underlying neural axis abnormality.

All the patients underwent posterior instrumented spinal arthrodesis, performed by the same surgeon (C.F). The surgical technique used for all patients consisted of high-density pedicle screws constructs, various combinations of translation maneuver over differently shaped cobalt chrome rods (according to deformity) and direct vertebral rotation. Titanium rods were used in 2 cases, cobalt-chrome rods in 1 case (*10-12*).

During all surgeries, Somatosensory and Motor Evoked Potentials (SEPs and MEPs) were monitored.

After surgery, all the patients were monitored in intensive care unit. A full-time TLSO brace was prescribed to all the patients for two months, followed by progressive weaning.

3. Results

All the patients had a single large thoracolumbar curve: 2 patients presented the curve convexity on the right side, 1 patient on the left side. Average age at surgery was 13.3 years (range 10.8-15.6); pre-operative radiographic values are reported in Table 1. None of the patients showed intellectual impairment. Two patients had a limited deambulatory autonomy of a few meters; one had been never able to walk.

3.1. Case 1 (female): *De novo COL6A3 heterozygous c.6465G>A* (*novel mutation*)

The patient referred to our division presenting a thoracolumbar curve of 55°, convex to the right (Figure 1, A and B), with left coronal imbalance requiring two sub-

Table 1. Pre-operative radiographic measurements

PATIENT	FUSION AREA	POST-OPERATIVE					
		Cobb Angle	Thoracic Kyphosis	Lumbar Lordosis	Nash Moe	Pelvic Obliquity	L5-tilt
Case 1	T4-S1	21°	48°	61°	II	6°	9°
Case 2	T4-L4	11°	26°	68°	II	6°	2°
Case 3	T4-L4	43°	36°	54°	II	10°	8°
Mean Value	-	25°	36°	61°	II	7°	6°

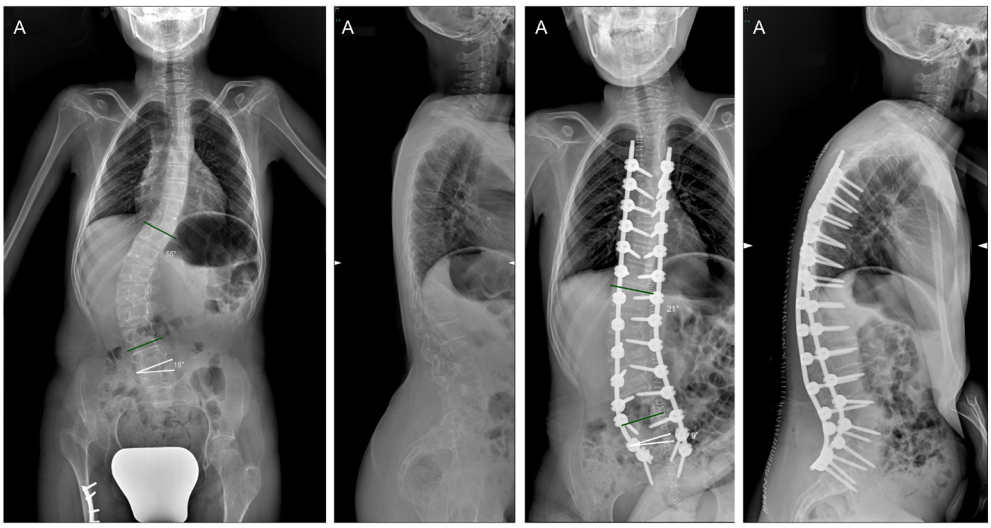

Figure 1. Case 1: Pre-operative radiographs, anteroposterior (A) and lateral (B) view; post-operative radiographs, anteroposterior (C) and lateral (D) view. Both pre- and post-operative Cobb angle (grey lines) and L5-tilt (white lines) measurement are reported.

axillary side pads when seated.

She had the first diagnosis of scoliosis at the age of 9, unsuccessfully treated with full-time bracing. In anamnesis she had developmental right hip dysplasia, torticollis and talus foot, both surgically corrected. She suffered from severe restrictive lung disease (Forced Vital Capacity FVC: 58% of the predicted value) and allergic asthma.

We performed a T4-S1 arthrodesis at the age of 13.6 years (Figure 1, C and D). S1 was chosen as the lowest instrumented vertebra because of the high L5-tilt (18°), despite the relatively low pelvic obliquity (8°). Screw density was 2.0; operative time was 210 minutes. No major complications occurred intra- and post-operatively. After 2 years, she reported a subjective improvement in quality of life, with a better sitting comfort and the same indoor walking ability.

3.2. Case 2 (female): *Inherited COL6A1 heterozygous c.896G > A*

She referred to our division presenting a curve of 42°, convex to the left side, complaining recurrent low back pain and discomfort in the sitting position in her wheelchair. Notably, the patient's stepsister was affected by UCMD carrying the same genotype, but never required spine surgery. Conversely, the father was asymptomatic, strongly suggesting the occurrence of paternal germline mosaicism (*13*).

Physical examination of the patient revealed generalised muscle weakness, predominantly in trunk and proximal limb musculature, joint contracture of elbows, knees and ankles, and a single large thoracolumbar scoliotic curve. In anamnesis, bilateral clubfoot (left foot corrected with a triple arthrodesis). She was able to walk for a few meters. She suffered from severe restrictive lung disease (FVC 44%) treated with mechanical insufflation-exsufflation machine ("cough machine") twice a day and non-invasive ventilation during night-time.

We performed T4-L4 arthrodesis at the age of 15.6 years. Screw density was 1.92; operative time was 300 minutes. Neither intra-operative nor post-operative complications occurred. After 2 years, the patient reported a more balanced sitting posture and maintained her indoor walking ability.

3.3. Case 3 (male): *De novo COL6A2 homozygous c.2572C>T.*

The patient referred to our division presenting a 52° thoracolumbar curve with convexity to the right, diagnosed at the age of 8 and unsuccessfully treated with bracing.

At the physical examination, the patient showed a trunk and proximal limb muscles weakness, lower and upper limbs areflexia, right knee joint flexion contracture and hyperlaxity of the wrists. In anamnesis, he had bilateral developmental right hip dysplasia treated with Pavlik harness and then corrective surgery. He had never walked, although at 36 months he was able to stand erect with aid: unfortunately, this ability was lost after a few months. He suffered from an early onset restrictive lung disease, with an FVC of 29% of the predicted value. Therefore, since the age of 4, he used non-invasive ventilation during night-time and mechanical insufflation–exsufflation machine twice a day.

We performed T4-L4 fusion at the age of 10.8 years. Screw density was 1.84 and operative time was 170 minutes. No intra-operative complication occurred. After extubation, the patient required prolonged respiratory support with non-invasive ventilation. On post-operative day 2, fever and productive cough occurred, therefore an empirical antibiotic therapy with Ceftriaxone was administered, suspecting a post-operative pneumonia. The patient was discharged after 17 days, after general conditions returned optimal. After 2 years, the patient reported better quality of life, with an improvement in sitting posture comfort.

Average operative time was 226 minutes (range 170-300), and average blood loss was 1190 mL (range 642-1580 mL). Average screw density was 1.92. Post-operative results are reported in Table 2. A satisfactory reduction of Cobb angle was obtained, from the average pre-operative value of 49° to the post-operative value of 25°, with an average reduction of 48% (24°) of the pre-operative value. Average lumbar lordosis improved from 56° to 61° and Nash-Moe decreased from III to II in all the patients. Pelvic obliquity remained stable, with a slight decrease to an average value of 7°. L5-tilt was reduced to a mean value of 6°. None of the patients required fixation to S2 nor to the pelvis.

Two patients were extubated after surgery in the

Table 2. Post-operative radiographic measurements

PATIENT	AGE AT SURGERY	RISSER STAGE	POST-OPERATIVE						
			Cobb Angle	Flexibility index	Thoracic Kyphosis	Lumbar Lordosis	Nash Moe	Pelvic Obliquity	L5-tilt
Case 1	13.6	3	55°	43%	38°	67°	III	8°	18°
Case 2	15.6	5	42°	38%	14°	59°	III	5°	3°
Case 3	10.8	0	52°	45%	33°	42°	III	11°	10°
Mean value	13.3	2.7	49°	42%	28°	56°	III	8°	10°

operating room, with a smooth transition to spontaneous breathing; Case 3 required prolonged non-invasive respiratory support. Also, none of the patients had mechanical or septic complications. No neurological injury occurred.

Patients were discharged after an average hospital stay of 15 days. At 2-year follow-up, no long-term complications, and no deformity progression or loss of correction occurred. All patients who had a pre-operative walking capacity, although limited, maintained it.

4. Discussion

Scoliosis in UCMD is characterized by an early onset, a rapid progression and resistance to brace treatment (*14,15*). This is confirmed by the low Risser stage (average value 2.66) that our patients had at the time of surgery. Regarding curve type, all the patients had a single large thoracolumbar curve. These characteristics are different from those seen in in Adolescent Idiopathic Scoliosis (AIS) and are more similar to those described by Karol *et al.* (*16*) in Duchenne Dystrophy. A large, gentle curve develops, with the apex at the thoracolumbar junction; with its rapid progression, the curve involves the whole thoracic and lumbar spine, leading to pelvic obliquity (*16*). This highly progressive scoliosis accelerates the progression of physical disability, making even simple activities such as standing, sitting, and walking difficult. On the other hand, by reducing chest wall compliance, scoliosis contributes to the restrictive respiratory dysfunction that usually afflicts these patients. For these reasons, scoliosis treatment in UCMD is crucial.

In our case series, we adopted a surgical technique derived from AIS surgery, achieving satisfactory clinical and radiological results. Infact, while sublaminar instrumentation and hybrid constructs are often used in neuromuscular scoliosis surgery, we preferred high density pedicle screws. Pedicle screws provide strong and stable 3-column fixation, allowing to perform strong corrective maneuvers such as direct vertebral rotation, achieving better curve correction, reduced blood loss and reduced surgical time compared to hybrid constructs (*17,18*). Moreover, as Hitchcon *et al.* (*19*) demonstrated, pedicle screws offer greater pull-out strength compared to sublaminar instrumentation. High-density screw constructs allow to distribute the corrective forces to every instrumented vertebra, avoiding pedicle breakage and screw pull-out during the corrective maneuver. This is crucial, considering that patients with neuromuscular scoliosis tend to have osteoporosis and osteopenia due to D-hypovitaminosis (*20*).

There are no studies in current literature that specifically focus on the surgical treatment of scoliosis in UCMD. Only one study conducted by Takaso *et al.* (*21*) described scoliosis surgery in a series of 10 patients affected by various congenital muscular dystrophies: 3 of their patients had UCMD. We believe that the rarity of UCMD is the reason why there are no previous studies focusing on scoliosis surgery in these patients: in fact, the vast majority of research on neuromuscular scoliosis involves patients with more common neuromuscular diseases such as Duchenne Dystrophy, Cerebral Palsy and Spinal Muscular Atrophy. On the other hand, research on neuromuscular scoliosis is less developed than that on AIS because surgery in neuromuscular scoliosis was developed later, due to cardiological and respiratory comorbidities. Moreover, many patients still have a certain, albeit limited, degree of motor autonomy and they refuse surgery for fear of losing it. This is particularly true in UCMD, in which scoliosis onset typically precedes the loss of ambulation, as Yonekawa *et al.* (*6*) and Nadeau *et al.* (*7*) studies revealed; on the contrary, scoliosis in Duchenne Dystrophy usually develops after patients become wheelchair dependent (*22*).

In our case series, 2 patients were able to walk for a few meters in an indoor setting and the surgery did not compromise motor autonomy in any of the cases. Indeed, post-operatively, all patients reported an increased comfort in the sitting position and higher confidence in walking, due to the restoration of a better global spinal balance, both in sagittal and coronal planes.

Regarding the deformity correction, we obtained a 48% coronal correction of the curve, while Takaso *et al.* (*21*) reported a higher 76% coronal correction. This can be explained by comparing the flexibility of the curves: in Takaso's case series, the curves had a flexibility index of 75%, whereas our patients had stiffer curves, with a flexibility index of 42%. It is also important to remember that deformity correction in neuromuscular scoliosis is less important than in patients with AIS, therefore higher radiological correction of the deformity may not be clinically relevant.

Instead, pelvic obliquity is a keypoint in neuromuscular scoliosis surgery: in order to correct or prevent it, routinely long fusion to the sacrum is a mainstay for many authors (*23-26*). Conversely, considering the challenges that sacropelvic fixation poses in these patients (including instrumentation failure due to osteoporosis and biomechanical stress, increased blood loss and longer operative time) others (*27-31*) support shorter fusions. In particular, Modi *et al.* (*31*) and Takaso *et al.* (*27*) reported excellent results with fusion up to L5 in patients with pelvic obliquity less than 15° and L5-tilt less than 15°. As we support this view, we performed fusion up to S1 only in one patient, whose L5-tilt was 18°. Infact, pelvic obliquity is often caused by supra-pelvic effects of scoliosis, such as asymmetric retraction of the muscles connecting trunk and pelvis (*32*): therefore, correction of the scoliotic curve may also reduce the pelvic obliquity, as Frischhut *et al.* (*33*) noted. In our experience, pelvic obliquity remained stable, but patients reported improved quality of life, with a

balanced and comfortable sitting position.

Respiratory complications have been reported as the most frequent complication following neuromuscular scoliosis surgery (*34-36*) and their frequency was found to be related to pre-operative FVC (*37,38*). In particular, Kang *et al.* (*37*) reported that patients with a pre-operative FVC of < 39.5% of the predicted value are more likely to develop a post-operative pulmonary complication. In our series, one patient (whose FVC was 29%, the lowest in our series) developed a suspected post-operative respiratory infection, which was successfully treated with Ceftriaxone. This is consistent with the Literature and shows how a multidisciplinary approach can be successful in controlling respiratory complications. None of our patients had infectious, cardiological, neurological, or implant-related complications.

The present study is affected by several limitations. First, the sample size is small, but UCMD is an extremely rare disease. Second, the follow-up period is too short to detect long-term complications. Further research is needed in order to assess the impact of scoliosis surgery on long-term pulmonary function of these patients as well.

5. Conclusion

Posterior spinal fusion with high density pedicle screws and direct vertebral rotation is safe and effective in the surgical correction of scoliosis in UCMD. If pelvic obliquity and L5-tilt are less than 15°, optimal balance could be achieved even without sacral or pelvic fixation, avoiding the risks of instrumentation failure, increased blood loss and operative time involved in sacropelvic fixation.

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References

1. Ullrich O. Kongenitale, atonisch-sklerotische Muskeldystrophie, ein weiterer Typus der heredodegenerativen Erkrankungen des neuromuskulären Systems. Zeitschrift für die gesamte. Neurol and Psychiatr. 1930; 126:171-201. (in German)
2. Ishikawa H, Sugie K, Murayama K, Ito M, Minami N, Nishino I, Nonaka I. Ullrich disease: Collagen VI deficiency: EM suggests a new basis for muscular weakness. Neurology. 2002; 59:920-923.
3. Ishikawa H, Sugie K, Murayama K, Awaya A, Suzuki Y, Noguchi S, Hayashi YK, Nonaka I, Nishino I. Ullrich disease due to deficiency of collagen VI in the sarcolemma. Neurology. 2004; 62: 620-623
4. Okada M, Kawahara G, Noguchi S, Sugie K, Murayama K, Nonaka I, Hayashi YK, Nishino I. Primary collagen VI deficiency is the second most common congenital muscular dystrophy in Japan. Neurology. 2007; 69:1035-1042.
5. Norwood FLM, Harling C, Chinnery PF, Eagle M, Bushby K, Straub V. Prevalence of genetic muscle disease in Northern England: In-depth analysis of a muscle clinic population. Brain. 2009; 132 (Pt11):3175-3186.
6. Yonekawa T, Komaki H, Okada M, Hayashi YK, Nonaka I, Sugai K, Sasaki M, Nishino I. Rapidly progressive scoliosis and respiratory deterioration in Ullrich congenital muscular dystrophy. J Neurol Neurosurg Psych. 2013; 84:982-988.
7. Nadeau A, Kinali M, Main M, Jimenez-Mallebrera C, Aloysius A, Clement E, North B, Manzur AY, Robb SA, Mercuri E, Muntoni F. Natural history of Ullrich congenital muscular dystrophy. Neurology. 2009; 73:25-31.
8. Tagliavini F, Pellegrini C, Sardone F, Squarzoni S, Paulsson M, Wagener R, Gualandi F, Trabanelli C, Ferlini A, Merlini L, Santi S, Maraldi NM, Faldini C, Sabatelli P. Defective collagen VI α6 chain expression in the skeletal muscle of patients with collagen VI-related myopathies. Biochim Biophys Acta. 2014; 1842:1604-1612 .
9. Nash CL Jr, Moe JH. A study of vertebral rotation. J Bone Joint Surg Am. 1969; 51:223-229.
10. Faldini C, Perna F, Geraci G, Pardo F, Mazzotti A, Pilla F, Ruffilli A. Triplanar correction of adolescent idiopathic scoliosis by asymmetrically shaped and simultaneously applied rods associated with direct vertebral rotation: clinical and radiological analysis of 36 patients. Eur Spine J. 2018; 27;165-174.
11. Faldini C, Perna F, Ruffilli A, Mazzotti A, Panciera A, Traina F. Surgical correction in AIS. Eur Spine J. 2019; 28:6-8.
12. Faldini C, Perna F, Chehrassan M, Borghi R, Martikos K, Mazzotti A, Cristofolini L, Traina F. Simultaneous double rod and en-bloc direct vertebral rotation technique for correction of main thoracic adolescent idiopathic scoliosis: retrospective analysis of 14 cases. J Biolog Reg Homeost Agents. 2016; 30:181-186.
13. Armaroli A, Trabanelli C, Scotton C, Venturoli A, Selvatici R, Brisca G, Merlini L, Bruno C, Ferlini A, Gualandi F. Paternal germline mosaicism in collagen VI related myopathies. Eur J Paediatr Neurol. 2015; 19: 533-536.
14. Olafsson Y, Saraste H, Al-Dabbagh Z. Brace treatment in neuromuscular spine deformity. J Ped Orthop. 1999; 19:376-379.
15. Kotwicki T, Jozwiak M. Conservative management of neuromuscular scoliosis: Personal experience and review of literature. Disabil Rehabil. 2008; 30:792-798.
16. Karol LA. Scoliosis in patients with Duchenne muscular dystrophy. J Bone Jt Surg. 2007; 89 Suppl 1:155-162.
17. Arun R, Srinivas S, Mehdian SMH. Scoliosis in Duchenne's muscular dystrophy: A changing trend in surgical management : A historical surgical outcome study comparing sublaminar, hybrid and pedicle screw instrumentation systems. Eur Spine J. 2010; 19:376-383.
18. Mattila M, Jalanko T, Puisto V, Pajulo O, Helenius IJ. Hybrid versus total pedicle screw instrumentation in patients undergoing surgery for neuromuscular scoliosis: A comparative study with matched cohorts. J Bone Joint Surg Br. 2012; 94:1393-1398.
19. Hitchon PW, Brenton MD, Black AG, From A, Harrod JS, Barry C, Serhan H, Torner JC. In vitro biomechanical comparison of pedicle screws, sublaminar hooks, and

sublaminar cables. J Neurosurg. 2003; 99:104-109.
20. Joyce NC, Hache LP, Clemens PR (2012) Bone health and associated metabolic complications in neuromuscular diseases. Phys Med Rehabi Clin N Am. 2012; 23:773-799.
21. Takaso M, Nakazawa T, Imura T, Okada T, Ueno M, Saito W, Takahashiz K, Yamazaki M, Ohtori S. Surgical correction of spinal deformity in patients with congenital muscular dystrophy. J Orthop Sci. 2010; 15: 493-501.
22. Archer JE, Gardner AC, Roper HP, Chikermane AA, Tatman AJ. Duchenne muscular dystrophy: the management of scoliosis. J Spine Surg. 2016; 2:185-194.
23. Allen BL, Ferguson RL. The galveston technique of pelvic fixation with L-rod instrumentation of the spine.Spine (Phila Pa 1976). 1984; 9: 388-394.
24. Peelle MW, Lenke LG, Bridwell KH, Sides B. Comparison of pelvic fixation techniques in neuromuscular spinal deformity correction: Galveston rod versus iliac and lumbosacral screws. Spine (Phila. Pa. 1976). 2006; 31:2392-2398; discussion 2399.
25. Phillips JH, Gutheil JP, Knapp DR. Iliac screw fixation in neuromuscular scoliosis. Spine (Phila Pa 1976). 2007; 32:1566-1570.
26. Miladi LT, Ghanem IB, Draoui MM, Zeller RD, Dubousset JF. Iliosacral screw fixation for pelvic obliquity in neuromuscular scoliosis: A long-term follow-up study. Spine (Phila Pa 1976). 1997; 22:1722-1729.
27. Takaso M, Nakazawa T, Imura T, Fukuda M, Takahashi K, Ohtori S. Segmental Pedicle Screw Instrumentation and Fusion only to L5 in the Surgical Treatment of Flaccid Neuromuscular Scoliosis. Spine (Phila Pa 1976). 2018; 43:331-338.
28. Mubarak SJ, Morin WD, Leach J. Spinal fusion in duchenne muscular dystrophy — fixation and fusion to the sacropelvis? J Pediatr Orthop. 1993; 13:752-757.
29. Wild A, Haak H, Kumar M, Krauspe R. Is sacral instrumentation mandatory to address pelvic obliquity in neuromuscular thoracolumbar scoliosis due to myelomeningocele? Spine (Phila Pa 1976). 2001; 26:E325-E329.
30. Mccall RE, Hayes B. Long-term outcome in neuromuscular scoliosis fused only to lumbar 5. Spine (Phila Pa 1976). 2005; 30:2056-2060.
31. Modi HN, Suh SW, Song HR, Yang JH, Jajodia N. Evaluation of pelvic fixation in neuromuscular scoliosis: A retrospective study in 55 patients. Int Orthop. 2010; 34:89-96.
32. Vialle R, Thévenin-Lemoine C, Mary P. Neuromuscular scoliosis. Orthop Traumatol Surg Res. 2013; 99:S124-S139.
33. Frischhut B, Krismer M, Stoeckl B, Landauer F, Auckenthaler T. Pelvic tilt in neuromuscular disorders. J Pediatr Orthop Part B. 2000; 9:221-228.
34. Mohamad F, Parent S, Pawelek J, Marks M, Bastrom T, Faro F, Newton P. Perioperative complications after surgical correction in neuromuscular scoliosis. J Pediatr Orthop. 2007; 27:392-397.
35. Sharma S, Wu C, Andersen T, Wang Y, Hansen ES, Bünger CE. Prevalence of complications in neuromuscular scoliosis surgery: A literature meta-analysis from the past 15 years. Eur Spine J. 2013; 22:1230-1249.
36. Cognetti D, Keeny HM, Samdani AF, Pahys JM, Hanson DS, Blanke K, Hwang SW. Neuromuscular scoliosis complication rates from 2004 to 2015: A report from the Scoliosis Research Society Morbidity and Mortality database. Neurosurg Focus. 2017; 43:E10.
37. Kang GR, Suh SW, Lee IO. Preoperative predictors of postoperative pulmonary complications in neuromuscular scoliosis. J Orthop Sci. 2011; 16:139-147.
38. Almenrader N, Patel D. Spinal fusion surgery in children with non-idiopathic scoliosis: Is there a need for routine postoperative ventilation? Br J Anaesth. 2006; 97:851-857.

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