Fusionless spinal surgery in children with spinal muscular atrophy type 1 with bipolar system: a preliminary communication

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Spinal muscular atrophy (SMA) is a severe childhood neuromuscular disorder caused by degeneration of lower motor neurons, leading to muscle atrophy. SMA type 1 (SMA1) is the most severe form and the leading genetic cause of infant mortality. While recent therapies such as nusinersen and onasemnogene abeparvovec have improved survival and ventilation-free time, affected children develop pelvic asymmetry and progressive spinal deformity, impairing the sitting position. Minimally invasive fusionless surgery (MIFS) using the Bipolar system has shown promising outcomes in SMA types 2 and 3, but evidence in SMA1 remains limited. This retrospective study reviewed medical records of SMA1 patients treated with MIFS using the Bipolar system between July 2023 and January 2025. Pre- and post-operative parameters were compared using paired Student's *t*-tests. Sixteen SMA1 patients (mean age: 8.1 ± 2.2 years; mean weight: 18.0 ± 3.2 kg) underwent MIFS with no surgical or anesthesiologic complications. Cobb angle improved from 71.8 ± 8.7 to 43.2 ± 9.2 ° ($P < 0.001$), pelvic obliquity from 12.7 ± 9.2 to 7.8 ± 6.1 ° ($P = 0.0035$), kyphosis from 62.9 ± 16.5 to 44.9 ± 14.1 ° ($P < 0.001$), and lordosis from 58.1 ± 15.3 to 43.5 ± 11.5 ° ($P < 0.001$). T1–S1 spinal length increased from 27.1 ± 1.8 to 30.9 ± 2.0 cm ($P < 0.001$), and thoracic width from 167.4 ± 12.0 to 181.5 ± 15.8 mm ($P = 0.0017$). The bipolar system appears to be a safe and effective surgical option for managing scoliosis in SMA type 1 patients, achieving significant correction of spinal and pelvic parameters without complications. *J Pediatr Orthop B* **XXX: XXXX–XXXX**
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Introduction

Spinal muscular atrophy (SMA) is a severe childhood monogenic disease resulting from loss or dysfunction of the gene encoding survival motor neuron 1 [1]. The incidence of this disease is approximately 1 in 10 000 live births, with a carrier frequency of 1 in 54.1 SMA is characterized by the degeneration and loss of lower motor neurons, which leads to muscle atrophy. The disease is divided into four subtypes (1–4) on the basis of the age at onset and milestone achievement. SMA type 1 (SMA1) is the most severe form and most common genetic cause of death among infants [1]. All patients with SMA1 have a precipitous decline in respiratory and swallowing functions after birth and ultimately require mechanical nutritional support (through a nasogastric or gastrostomy tube) to maintain adequate nutrition and reduce the respiratory risks associated with aspiration.

Recent therapeutic options for SMA1 patients include nusinersen [2], which is administered by means of repeated intrathecal injections, onasemnogene abeparvovec [3], given as a one-time intravenous administration that delivers a copy of SMN in a self-complementary adeno-associated viral serotype, and Risdiplam, an orally administered, small molecule that modifies *SMN2* pre-mRNA splicing and increases levels of functional SMN protein [4]. Patients receiving these therapies have shown longer survival and ventilation-free time than the historical cohorts with this disease [5].

Those children who survive and do not achieve or lose walking ability will develop typically a pelvic asymmetry and a progressive spinal deformity, which interferes with sitting position [6]. Scoliosis is an important complication in these children because it impairs respiratory function, which is already reduced because of intercostal muscle weakness and fatigue. Although improvement in motor function has been proven by different trials and in real-life studies, most patients do not achieve walking ability,

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and scoliosis remains a major complication in them from very early ages [6].

Minimally invasive fusionless surgery (MIFS) is a promising approach for neuromuscular scoliosis, offering an alternative to spinal fusion by promoting gradual spinal ankylosis. This technique has shown positive results in SMA 2 and 3 patients [7]. However, there's a lack of literature on its application in children with SMA 1, who often present with rapid and severe kyphoscoliosis due to advancements in treatment that have extended their lifespan. This presents a unique challenge, as these deformities can significantly impact respiratory function, necessitating early surgical intervention.

While magnetic-controlled growing rods (MCGR) are commonly used in MIFS, they may not be suitable for SMA 1 patients. These children typically have smaller spinal dimensions, making it difficult to accommodate the rigid central portion of MCGRs, particularly in the presence of significant kyphosis. Moreover, their low body mass necessitates a low-profile system for pelvic stabilization. Lastly, early surgical intervention requires a system capable of lengthening beyond the 4.8 cm limit of MCGRs. The NEMOST system, with its potential for spontaneous lengthening up to 8 cm, addresses these challenges.

This study aims to present the outcomes of MIFS using the NEMOST system in a cohort of SMA 1 patients treated between July 2023 and January 2025.

Methods

The medical charts of the patients who underwent MIFS in the given period were retrospectively reviewed, and data on demographic characteristics as well as nutritional status, motor and respiratory function were extracted after parental consent.

The used implants are provided by Euros.

Surgical technique

Two midline skin incisions are made: one proximal at the upper thoracic spine (T2) and one distal at the lumbosacral junction. The fixation system extends from T2 to the sacrum, proximally anchored with eight hook claws and distally by iliosacral screws engaging the sacral domino. Two short, precontoured rods are inserted into the hooks and stabilized with two crosslinks to create a strong proximal foundation.

The L5–S1 space is exposed to prepare for the insertion of the sacral domino. An osteotomy is performed to create a hole that accommodated the domino, positioned cranially and laterally to the S1 foramen. The domino is connected to a compass with a cannula to guide the direction of the iliosacral screws.

The iliosacral screws are inserted percutaneously from the posterior iliac bone to the S1 body, engaging the sacral domino in an oblique posterior-to-anterior direction. The compass is bent 20 ° medially to avoid the spinal canal. Insertion of the 7.0 screws is challenging without navigation, and 3D CT planning is used to ensure adequate space in S1. The patient's pelvis exhibits significant retroversion and lateral tilt, increasing the difficulty of screw insertion.

Precontoured medial long rods are inserted from the proximal to the distal incision, tunneling subfascially with a chest tube. They are fixed proximally in the hook claws and distally to lateral short rods using side-to-side closed connectors.

The short rods, prebent for lordosis, are Nemost rods, designed to lengthen 5 or 8 cm spontaneously with patient growth. They are inserted and fixed inside the iliosacral connectors. The side-to-side connectors are first locked at the ends of the medial long rods. Correction was achieved by distraction maneuvers on the concave side between the iliosacral connector and the side-to-side connector, which migrated proximally with the long rod. At the end of distraction, the side-to-side connector was locked to the short lateral rod. Similar rods are then inserted and fixed on the convex side.

Eight-centimeter NEMOST rods are chosen due to the patient's age and expected growth. The Nemost rods inserted from the lumbar incision are cranially tunneled with a chest tube to prevent malpositioning under the ribs in the thorax. Finally, the ability of the Nemost rods to lengthen and produce distraction was checked. If they do not function correctly, the rod rotation is adjusted until they move freely.

Anesthesiologic management

Patients are fasted according to standard guidelines and infused with intravenous balanced crystalloids with glucose since the evening before surgery. After standard monitoring [heart rate, noninvasive blood pressure, SpO2, bi-spectral index (BIS)], anesthesia is induced intravenously with propofol and remifentanil in raising concentrations until a BIS of 40–50 is reached with the patient in spontaneous breathing; patients are then intubated with videolaringoscopy and invasively monitored with arterial and central venous lines. Anesthesia is maintained with a totally intravenous technique in order not to alter Sensory and Motor Evoked Potentials. Before incision, an ultrasound-guided erector spinae plane block is performed in 4 points [8]. Tranexamic acid is administered in a bolus (15 mg/kg) at skin incision and maintained in continuous infusion (3 mg/kg/h) up to skin closure. Patients are then transferred intubated to the ICU and extubated on the same day, transitioning to noninvasive ventilation (NIV) for the first hours and then as clinically needed.

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Radiological data

For each patient, the following radiologic parameters were reviewed before surgery and at first follow-up control after surgery: major coronal curve, pelvic obliquity, thoracic kyphosis, lordosis, T1-S1 distance, and thoracic width.

Statistical analysis

Descriptive data are shown as absolute numbers and percentages for categorical variables, and mean (SD) or median (IQR) according to distribution for continuous ones. Equality of pre- and post-operative measures was tested with paired Student's *t* tests. A linear regression model was conducted on pulmonary function tests with thoracic width as a covariate.

Results

Sixteen patients affected by SMA 1 were treated with MIFS in the specified period. Supplementary Fig. 1, Supplemental digital content 1, *https://links.lww.com/JPOB/A133* presents the flowchart of patient enrollment. Nine patients were treated with nusinersen and seven with risdiplam. All of them are followed by the Hospital Pediatric Palliative Care Service and are ventilated noninvasively at home; in 15/16 (94%) cases, NIV was performed routinely during nighttime, and in one case, the patient used the ventilator only during respiratory tract infections.

Demographic data are reported in Table 1. All patients presented with abnormal preoperative pulmonary function tests; the degree of reduction in forced expiratory volume in the first second was close to correlation with thoracic width (Coeff. 0.54; 95% confidence interval: −0.05 to 1.14; $P = 0.06$).

Eight patients received blood transfusions. All patients were extubated on the same day as surgery and put on home NIV. Hospital length of stay was 9.0 days.

All patients receiving Nusinersen therapy were able to continue it after surgery.
Orthopedic data (pre- and post-operative) are reported in Table 2. Mean control after surgery was 9.8 (±4.95) months. Left rod lengthening was 5 (3–10) cm, and right rod lengthening was 4 (3–10) cm.

Figure 1 presents the X-ray (pre- and post-operative) of one patient and the relative appearance of the back after surgery. Figure 2 visually displays the improvement in orthopedic measures after surgery. Table 3 reports the available papers that used MIFS up to now and the populations of patients in whom they were tested.

Table 1 **Demographic and surgical data**

	Total
	N = 16
Age	8.1 (2.2)
Weight, kg	18.0 (3.2)
Height, m	1.2 (0.1)
BMI, kg × m^{-2}	12.9 (1.4)
Male sex	8 (50%)
Fev1 (Z score)	−5.4 (1.2)
Fev1 (%)	36.4 (11.7)
Fvc (Z score)	−5.6 (1.3)
Fvc (%)	35.2 (12.5)
Nusinersen treatment	9 (60%)
Risdiplam treatment	7 (40%)
Surgery duration, minutes	214.5 (25.9)
Need for blood transfusion	8 (50%)

Data are presented as mean (SD) for continuous measures, and *n* (%) for categorical measures.
FVC, forced vital capacity; FEV1, forced expiratory volume in the first second.

Table 2 **Results of paired Student's *t*-test conducted on pre- and post-operative orthopedic measures**

	Pre	Post	*P* value
Cobb angle, degrees	71.8 (8.7)	43.2 (9.2)	<0.001
Pelvic obliquity, degrees	12.7 (9.2)	7.8 (6.1)	0.0035
Kyphosis, degrees	62.9 (16.5)	44.9 (14.1)	0.0001
T1-S1 height, cm	27.1 (1.8)	30.9 (2.0)	<0.001
Lordosis, degrees	58.1 (15.3)	43.5 (11.5)	<0.001
Thoracic width, mm	167.4 (12.0)	181.5 (15.8)	0.0017

Discussion

In recent years, the management of spinal deformities in patients with SMA has entered a new era, driven by therapeutic advances and the need for early surgical solutions. Our paper explores the evolving role of fusionless techniques in a population affected by SMA type 1, which is an emerging indication for scoliosis correction. Until a few years ago, in fact, this kind of patients had no therapy and would be candidates for scoliosis correction at later ages, with a different system.

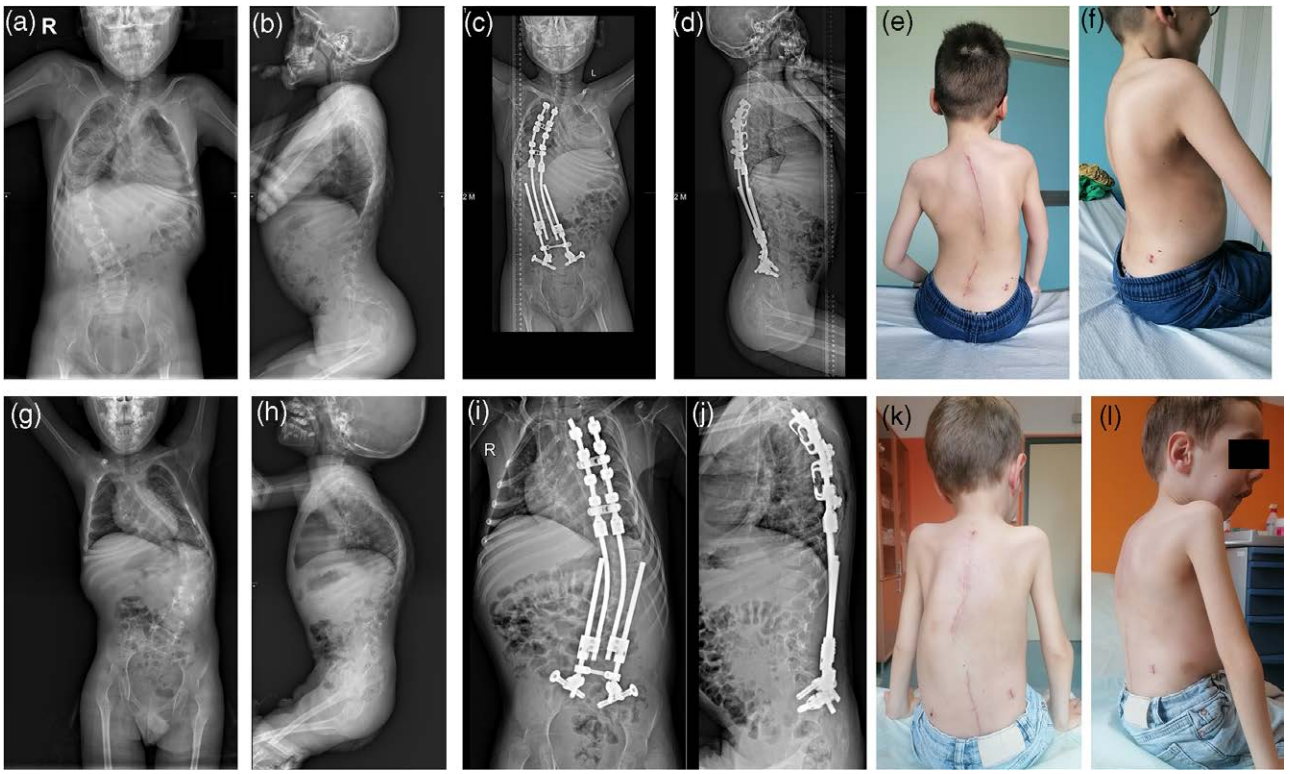
Historically, the traditional growing rod (TGR) system required repeated surgical procedures every 6 months to lengthen the rods—a process that, while effective in curve control, carries a high burden of infection, anesthesia-related complications, and respiratory compromise, particularly in fragile SMA patients. Bilateral constructs were favored due to their lower complication rates.

MCGR systems offered a major advancement. These allowed noninvasive lengthening through an external magnetic controller, performed in outpatient settings while the patient remains awake. Based on Alain Di Meglio's data, the expected spinal growth between ages 6 and 12 is approximately 15 mm per year. Our team developed a guideline for lengthening MCGR every 3 months by 3.5 mm [14].

We retrospectively analyzed 34 SMA type two patients treated with MCGR between November 2012 and April 2023 at our institution (unpublished data). All patients, aged 6–12 years, underwent surgery with the same team using a construct extending from T2 to the sacrum. Proximal fixation involved T2 claw hooks, with the rod tunneled subfascially and anchored distally with pedicle screws at L4–L5 and bilateral iliac screws. Outpatient

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Fig. 1



X-ray and clinical appearance of two operated patients. Patient 1: (a) anteroposterior before correction; (b) lateral before correction; (c) anteroposterior after correction; (d) lateral view after correction. (e) Posterior view of the sitting patient; (f) lateral view of the sitting patient. Patient 2: (g) Anteroposterior before correction; (h) lateral before correction; (i) anteroposterior after correction; (j) lateral view after correction. (k) Posterior view of the sitting patient; (l) lateral view of the sitting patient.

lengthenings were performed every 3 months. Follow-up involved ultrasound (pre- and post-lengthening) and annual radiographs. The average maximum rod elongation plateaued after 18 months, reaching 40.5 mm for standard rods and 38.9 mm for compensatory rods. Revision surgeries revealed mechanical issues and soft tissue metallosis. These outcomes, combined with the unique kyphoscoliotic deformity pattern in SMA type 1, prompted a shift toward a new surgical approach.

OWSER was selected as a low-profile, minimally invasive alternative to TGR and MCGR with the goal of obviating the need for further surgery while correcting the spine deformity and preserving the spine and thoracic growth. Also, in contrast to MCGR's central magnet (10 cm unbendable core), OWSER can be contoured along rigid spines in small patients.

This self-lengthening system showed spontaneous bilateral growth of approximately 1 mm per month. Though early, these results are promising.

Pelvic fixation in neuromuscular patients is essential to ensure sitting balance. The bipolar configuration of the OWSER includes a direct sacroiliac screw fixation to S1, eliminating the need for connecting rods, which is advantageous in low BMI SMA patients prone to implant prominence and skin breakdown. The tricortical screw in S1 ensures strong, deep fixation even in osteoporotic bone.

Crosslinks between the rods enhance stability and serve as a powerful lever arm for correcting pelvic obliquity. As Miladi has reported, the bipolar construct allows definitive correction without the need for final fusion [12].

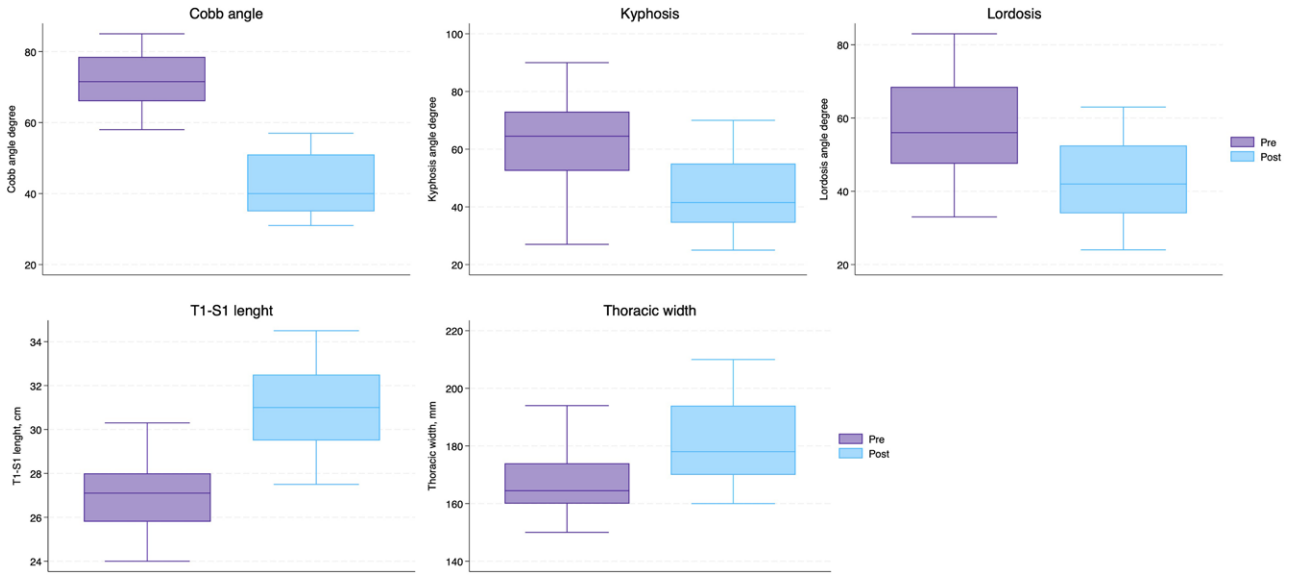
Until now, all previous publications on OWSER system come from the same Institution, 'Hopital Necker' in Paris [7,9,10,12,15]. This is understandable, as they are the ones who developed the system. They gained considerable experience in treating different diseases, and we learned from them, applying the technique to this very specific and fragile population. Our work is the first, to our knowledge, performed on a homogeneous group of patients.

Why fusionless is the future for spinal muscular atrophy type 1

Patients with SMA type 1 now survive longer due to disease-modifying therapies [2,11,13]. These children

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Fig. 2



Modifications of Cobb angles degrees (°), Kyphosis degree (°), Lordosis degree (°), T1-S1 distance (cm), and thoracic width (cm) from presurgery to postsurgery period.

Table 3 Available publications on bipolar system with their relative populations of use

Author	Year	Population type	Total number of patients	Complications
Miladi L [9]	2018	Cerebral palsy (61), Muscular dystrophy (10), SMA (22, type not specified), other NM disorders (7)	100	Nerve roots perioperative alerts (5); Blood transfusion (24); wound infection (16); implant dislodgement (12); superior mesenteric artery syndrome (2), pneumonia (1); anaphylactic shock (1)
Miladi L [10]	2020	Idiopathic (6), Cerebral palsy (3), Rett (3), Neurofibromatosis (2), Syndromic (3), Muscular dystrophy (1), SMA (2, type not specified)	20	Deep surgical site infection (2); Proximal junctional kyphosis (2); bulkiness (2); device-related complications (5); unrelated to device (6)
Gaume M [11]	2021	Cerebral palsy (11), Muscular dystrophy (5), SMA (3, type not specified), other NM disorders (2)	21	Mechanical (3); infections (5)
Gaume M [12]	2021	Cerebral palsy (45), Muscular dystrophy (10), SMA (21, type not specified), other NM disorders (7)	83	Mechanical (12); infections (15)
Gaume M [13]	2021	SMA 1c (9), SMA 2 (47), SMA 3 (3)	59	Blood transfusion (6); wound infection (%); mechanical complication (4); unplanned surgeries (6)
Gaume M [14]	2023	Cerebral palsy (95), Muscular dystrophy (8), SMA (15, type not specified), Spina Bifida (6), other NM disorders (43)	167	Mechanical (16)
Gaume M [7]	2023	Cerebral palsy (6), SMA 2 (3), Syndromic (4)	13	–
Gaume M [15]	2024	Cerebral palsy (12), SMA 2 (1), Syndromic (6)	19	–
Colombo	2025	SMA 1(16)	16	Blood transfusions (8)_

Numbers in brackets refer to the number of patients/events.
SMA, spinal muscular atrophy.

develop early kyphoscoliosis, often requiring surgery at very young ages. Fusion at such an early age would halt thoracic and spinal growth, compounding respiratory issues. Fusionless techniques like OWSER offer correction with ongoing spinal elongation.

In our series, we successfully implanted OWSER in patients with very low BMIs, as low as 11.5 kg/m^2. The system's low profile minimized risks of pressure ulcers, which can be a tangible risk in such low BMIs, and the minimally invasive technique reduced blood loss and infection risk. The ability to contour the rods subfascially is especially helpful in small, rigid spines.

Despite the small patient size, we implanted 7.0 mm sacroiliac screws without navigation, guided by preoperative CT planning. This underscores the importance of precise surgical planning in these fragile patients.

Respiratory outcomes

The degree of preoperative pulmonary function abnormality is associated with thoracic dimensions as

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described by Canavese *et al.* [16]. In our population, as well, we described a correlation between thoracic width and forced expiratory volume. We interpret the *P* value, which is close to statistical signficance, as such due to the relatively small sample size. In our previous SMA type 2 series, we observed that MCGR did not significantly improve respiratory function but stabilized it over time [14]. We expect a similar outcome in our population of SMA type 1 patients treated with OWSER as maintaining thoracic growth appears critical in preserving pulmonary capacity. We acknowledge the lack of postoperative pulmonary testing as a limitation of our work, but at the same time, given the significant thoracic width improvement, we expect our patients not to decline in pulmonary function at long-term follow-up.

Emans *et al.* [17] presented a method to describe the potential maximum spine length and chest width in a population of idiopathic scoliotic patients, starting from pelvic width. We therefore tried to apply such a method to our cohort, but our results appeared not to be in line with the method described. We interpret this result as such, due to the very different patients' characteristics (idiopatic adolescents vs. SMA1 children), which probably makes it not suitable for our population.

Anesthesiological management

We are a referral center for patients with SMA1, who are managed with a lifelong program of regular respiratory, physiotherapeutic, cardiologic, and nutritional [18] checkups. We started administration of nusinersen and onasemnogene abeparvovec as soon as these drugs have made available [19]. Patients are usually ventilated in NIV way (in our cohort, one patient only used NIV only during upper airway infections and all the other ones used it regularly during nighttime).

We are therefore used to providing anesthesia to these patients for different procedures (e.g. placement of gastrostomies or fundoplication), but it is the first time we confront ourselves with spinal surgery at these early ages.

As these patients are known to present a potentially difficult airway, we choose to keep them spontaneously breathing during intubation. Patients are induced with growing doses of propofol and remifentanil and they receive oxygen through nasal cannulas while spontaneously breathing during videolaringoscopy, so that we are able to perform intubation without need to rush. We then place central venous lines for surgery as they are often patients with difficult peripheral venous access. Invasive blood pressure monitoring is essential during surgery, although we did not describe any hemodynamic compromise in our cohort, requiring vasopressor support. Regular use of NIV is helpful for the respiratory weaning after surgery as children are already used to it. It is important to avoid long fasting in these children as their glycogen storages are not adequate, therefore, we infuse intravenous balanced crystalloids with glucose from the night before surgery at a maintenance rate to avoid ketosis. Half of the patients required blood transfusion in our cohort, which seems reasonable given their relatively small blood volume.

We did not report any complications from anesthesia in our cohort, and we suggest these patients should be treated in expert hands with a multidisciplinary approach.

Limitations

We acknowledge our work presents some limitations. We present a small sample of patients with a relatively short follow-up period, which prompts us to present our results as encouraging but not definitive, particularly in view of the future trajectory of disease of these patients, which is still unknown to the scientific world. Still, these limitations could be also presented as strengths of our work as we first describe a new kind of population, with very particular characteristics (small age, low BMI), in whom we successfully used a minimally invasive system, with short ICU stay and short hospital stay.

Also, another limitation of the work is that we do not present postoperative pulmonary function tests. With longer follow-up times and a better knowledge of the evolution of these treated patients, we will be able to present results on the pulmonary function. Prospective studies will be necessary to further explore and confirm our findings and to provide a more comprehensive understanding of the clinical outcomes.

Conclusion

Spinal fusion remains the traditional approach for scoliosis in SMA, but is not suitable for early intervention. Fusionless systems like OWSER are safer and more appropriate in the early years when growth potential and respiratory development are paramount. As demonstrated in our first year with this system, it is feasible to surgically treat SMA type 1 scoliosis with promising biomechanical and clinical outcomes. It is important not to forget that good outcomes in this kind of very fragile patients depend on all the surgical and intensive care team, who is used to dealing with this very fragile patients.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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