

# Comparison of baseline characteristics and postoperative complications in neuromuscular, syndromic and congenital scoliosis

Lorenzo R. Deveza<sup>a</sup>, Barkha N. Chhabra<sup>b</sup>, John Heydemann<sup>b</sup>, Chun Hung<sup>a</sup>, Dallas Vanorny<sup>a</sup>, Dion Birhiray<sup>b</sup> and Benny Dahl<sup>a,b,c</sup>

Nonidiopathic scoliosis encompasses a group of diagnoses, including neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis. The objective of this study was to compare the preoperative and postoperative clinical differences in pediatric nonidiopathic scoliosis patients with neuromuscular scoliosis vs. syndromic scoliosis/congenital scoliosis. This is a single-center retrospective review of all pediatric patients undergoing spinal instrumentation for nonidiopathic scoliosis during a 5-year period. Neuromuscular scoliosis patients ( $n=144$ ), syndromic scoliosis patients ( $n=44$ ) and congenital scoliosis patients ( $n=52$ ) were compared. Demographics, patient characteristics and outcomes were compared. Neuromuscular scoliosis patients had lower BMI z-scores and were more likely to have pulmonary disease, technology dependence and seizure disorder. Additionally, neuromuscular scoliosis patients underwent bigger procedures with more levels fused and a higher rate of pelvis fixation. By direct comparison, neuromuscular scoliosis patients tended to have more complications including deep surgical site infections, readmission in 30 days, return to operating room in 90 days and emergency care visits in 90 days. When controlling for the differences in their preexisting conditions and surgical procedure, we found that pelvic fixation

was a major confounding factor, whereas the others had no effect. We further subanalyzed cerebral palsy patients and found this group to exhibit no difference in complications compared to other neuromuscular scoliosis subtypes. Neuromuscular scoliosis patients have different characteristics and subsequent postoperative complications than those with syndromic scoliosis and congenital scoliosis. The difference in complication profile is mainly due to differences in surgical procedure and a higher rate of pelvic fixation. This should be considered when planning nonidiopathic scoliosis surgery among multidisciplinary teams. *J Pediatr Orthop B* 32: 350–356 Copyright © 2022 Wolters Kluwer Health, Inc. All rights reserved.

*Journal of Pediatric Orthopaedics B* 2023, 32:350–356

**Keywords:** nonidiopathic scoliosis, neuromuscular scoliosis, quality improvement

<sup>a</sup>Department of Orthopaedic Surgery, Baylor College of Medicine,

<sup>b</sup>Division of Orthopaedic Surgery, Texas Children's Hospital, Houston, TX, USA and <sup>c</sup>Spine Unit, Department of Orthopaedic Surgery, Rigshospitalet and University of Copenhagen, Copenhagen, Denmark

Correspondence to Barkha N. Chhabra, MD, MPH, Division of Orthopaedic Surgery, Texas Children's Hospital, Houston, TX 77030, USA  
Tel: +1 832 822 3100; fax: +1 832 825 9099;  
e-mail: bnchhab1@texaschildrens.org

Received 27 April 2021 Accepted 28 May 2022

## Introduction

Nonidiopathic scoliosis encompasses a group of diagnoses, including neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis [1]. Although not clearly defined, traditionally neuromuscular scoliosis includes cerebral palsy, muscular dystrophy, spinal muscular atrophy and other conditions affecting the neuromuscular system. Syndromic scoliosis includes Marfan's syndrome, Down syndrome, DiGeorge syndrome and osteogenesis imperfecta. Congenital scoliosis is defined by a malformation of one or more vertebral bodies. In many studies, these diagnoses are considered together [2–4]. However, clinically, surgeons recognize that there are differences in these populations affecting the treatment of each subgroup [3,5–8].

It is known that surgical treatment of children with neuromuscular scoliosis has an increased rate of complications compared to those with idiopathic scoliosis [8–10].

Further, studies have demonstrated differences in postoperative complication rate after posterior spinal instrumentation and fusion among children with syndromic scoliosis and idiopathic scoliosis [5], as well as with congenital scoliosis and idiopathic scoliosis [7]. However, the differences between neuromuscular scoliosis and those with syndromic scoliosis or congenital scoliosis have not been established. This becomes important when not only determining perioperative care but also institutionally when assessing quality measures for the hospital. For example, the US News and World Report, has until recently, included these diagnoses as a single group. This can unfairly demonstrate increased complications for institutions caring for more complex nonidiopathic scoliosis patients. Furthermore, with the push to institute safety protocols for adult and pediatric deformity cases, it is important to recognize the risk profile for each

diagnosis category to appropriately prepare the patient and families for possible complications [11].

The objective of this study was to compare the clinical differences in various types of pediatric nonidiopathic scoliosis, including neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis. Their baseline characteristics and co-morbidities are compared, followed by an assessment of their postoperative complication profiles.

## Methods

A retrospective chart review was performed at Texas Children's Hospital (TCH) of all pediatric patients who underwent spinal surgery for any reason from July 2014 to June 2019. Inclusion criteria were patients less than 18 years of age at the time of surgery, including scoliosis cases and excluding those with a diagnosis of idiopathic scoliosis. These patients were then grouped by diagnostic category: neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis. We defined neuromuscular scoliosis as scoliosis associated with any neuromuscular conditions, including cerebral palsy, spinal muscular atrophy and muscular dystrophy. We defined syndromic scoliosis as scoliosis associated with known conditions that contribute to scoliosis, such as Marfan's syndrome, Down syndrome, osteogenesis imperfecta and DiGeorge syndrome. Congenital scoliosis was defined as a spinal deformity caused by any congenital vertebral anomaly. Using this cohort of patients, diagnostic categories were compared for their baseline characteristics and their operative complications.

Patient baseline characteristics included age, sex and BMI. Their baseline health characteristics were determined by assessing for the presence of cardiac disease, pulmonary disease, coagulopathy, seizure disorder and technology dependence. Patients were determined to have cardiac disease if they carried a diagnosis of congenital heart defect or cardiomyopathy. Patients were determined to have pulmonary disease if they carried a diagnosis of restrictive lung disease or obstructive sleep apnea. Coagulopathy was determined by having confirmed hematologic diagnoses such as Von Willebrand Factor deficiency. Technology dependence was determined by assessing whether the patient required bilevel positive airway pressure, continuous positive airway pressure or a tracheostomy tube to maintain oxygenation at any time in the perioperative care.

The surgical procedure was also included in the comparison. Specifically, we determined the number of spinal levels included in the construct, number of screws used, the screw density, whether the construct included fixation to pelvis and lastly whether the case concluded with plastics closure assistance.

The outcome parameters included mortality, surgical site infection, unplanned readmissions within 30 days,

re-operation within 90 days, emergency department visits within 90 days for any reason, revision surgery within 1 year and implant failure within 1 year. Additionally, pneumonia at 30 days and deep vein thrombosis/pulmonary embolus was assessed within the first 90 days after surgery.

Statistics were performed using analysis of variance (ANOVA) for all continuous variables and a logistical regression statistical test for all categorical data. A *P* value of less than 0.05 was considered statistically significant.

## Results

A total of 870 spinal instrumentation cases were identified at TCH from June 2015 to June 2019. Among these 638 cases were diagnosed with scoliosis, including idiopathic and nonidiopathic scoliosis. From this selection, 240 patients were categorized as having nonidiopathic scoliosis: 144 patients were categorized as neuromuscular scoliosis, 44 were syndromic scoliosis and 52 congenital scoliosis. The majority of neuromuscular scoliosis cases had cerebral palsy ( $n=52$ ), muscular dystrophy ( $n=18$ ), spinal muscular atrophy ( $n=11$ ), followed by post-traumatic neonatal spinal cord injury ( $n=5$ ) and other neuromuscular scoliosis ( $n=58$ , to include encephalopathy, hydrocephalus, spina bifida and syndromes when associated with severe global developmental delay, seizures, movement disorders). The average Gross Motor Function Classification System (GMFCS) score of patients with cerebral palsy was  $5 \pm 0.9$ . All patients with a GMFCS score of 5 had a fixation to the pelvis. The most frequent diagnosis for syndromic scoliosis was Marfan's syndrome ( $n=10$ ), followed by Ehlers-Danlos syndrome ( $n=3$ ), Neurofibromatosis ( $n=3$ ), DiGeorge Syndrome ( $n=3$ ) and other syndromic scoliosis ( $n=25$ , to include Down syndrome, CHARGE syndrome, Gaucher's Disease, mitochondrial diseases, Costello syndrome and Duane syndrome).

### Direct comparison between neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis

A direct comparison of patient demographics, baseline characteristics, and surgical data is presented in Table 1. Patients with neuromuscular scoliosis and syndromic scoliosis were older than those with congenital scoliosis ( $13.3 \pm 3.6$  neuromuscular scoliosis vs.  $13.0 \pm 2.5$  syndromic scoliosis vs.  $9.7 \pm 3.4$  congenital scoliosis;  $P < 0.001$ ) and had lower BMI-for-age Z-score ( $-0.3 \pm 1.9$  neuromuscular scoliosis vs.  $0.1 \pm 1.4$  syndromic scoliosis vs.  $0.7 \pm 1.3$ ;  $P = 0.001$ ). Similarly, the BMI percentile was lower for neuromuscular scoliosis patients. Regarding their preexisting conditions, patients with neuromuscular scoliosis had more pulmonary disease than those with syndromic scoliosis and congenital scoliosis (59.7% neuromuscular scoliosis vs. 36.8% syndromic scoliosis vs. 23.1% congenital scoliosis;  $P < 0.001$ ), technology dependence (37.9% neuromuscular scoliosis vs. 21.1% syndromic scoliosis

**Table 1 Patient baseline characteristics and surgical procedural data**

	NMS	SS	CS	P value
Patient demographics				
Number of cases	144	44	52	
Age in years (Ave; SD)	13.3±3.6	13.0±2.5	9.7±3.4	<0.001
Gender (% female)	41.0%	69.9%	65.8%	
BMI (Ave; SD)	19.7±5.3	20.3±4.7	21.1±7.1	0.277
BMI z-score (Ave; SD)	-0.3±1.9	0.1±1.4	0.7±1.3	0.001
BMI percentile (Ave; SD)	47.8±35.3	49.4±35.6	69.6±28.8	0.001
Cardiac disease (%)	10.5	31.6	17.9	0.243
Pulmonary disease (%)	59.7	36.8	23.1	<0.001
Technology Dependent (%)	37.9	21.1	17.9	0.020
Seizure Disorder (%)	33.1	7.9	5.1	0.003
Coagulopathy (%)	7.3	5.3	0.0	0.992
Surgical data				
Number of levels (Ave; SD)	15.7±3.8	13.8±2.0	9.6±4.7	<0.001
Number of screws (Ave; SD)	22.8±7.9	20.6±5.2	12.9±5.3	<0.001
Screw density (%)	73±18.1	76.2±24.1	72.3±17.0	0.6311
Pelvis fixation (%)	60.5	9.5	14.6	<0.001
Plastics closure (%)	1.4	0.0	3.8	0.118

CS, congenital scoliosis; NMS, neuromuscular scoliosis; SS, syndromic scoliosis.

**Table 2 Direct comparison of complications between neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis**

	NMS	SS	CS	P value
Postop complications				
Deaths in 30 days (%)	1.4	0.0	0.0	0.997
Deaths in 1 year (%)	2.8	0.0	0.0	0.996
SSI (deep) (%)	10.4	0.0	1.9	0.085
SSI (superficial) (%)	11.1	9.3	9.6	0.735
Readmission in 30 days (%)	14.0	9.3	3.8	0.061
Return to operating room in 90 days (%)	13.3	4.7	5.8	0.145
Emergency care visits in 90 days (%)	21.7	14.0	11.5	0.106
Revision surgery in 1 year (%)	15.3	9.3	9.6	0.295
Instrument failure in 1 year (%)	4.2	7.0	0.0	0.991
Pneumonia in 30 days (%)	5.6	7.0	1.9	0.297
DVT/PE (%)	2.1	0.0	0.0	0.996

CS, congenital scoliosis; DVT, deep vein thrombosis; NMS, neuromuscular scoliosis; PE, pulmonary embolus; SS, syndromic scoliosis; SSI, surgical site infections.

vs. 17.9% congenital scoliosis, 0.02) and seizure disorder (33.1% neuromuscular scoliosis vs. 7.9% syndromic scoliosis vs. 5.1% congenital scoliosis;  $P=0.003$ ). There was a trend towards higher cardiac disease in syndromic scoliosis patients, including cardiomyopathy and congenital heart defects, but the difference was NS.

There were many differences in surgical procedures among the diagnosis groups, where neuromuscular scoliosis patients underwent more involved spinal procedures (Table 1). This was evident as neuromuscular scoliosis patients had a higher average number of fused spinal levels ( $15.7 \pm 3.8$  levels neuromuscular scoliosis vs.  $13.8 \pm 2.0$  syndromic scoliosis vs.  $9.6 \pm 4.7$  levels congenital scoliosis;  $P<0.001$ ) and subsequently a higher number of screws used. There was no difference in screw density per procedure for any group. Notably, the majority of neuromuscular scoliosis patients have pelvis fixation, which was not as frequent for syndromic scoliosis and congenital scoliosis patients (71.6% neuromuscular scoliosis vs. 9.5% syndromic scoliosis vs. 14.6% congenital scoliosis;  $P<0.001$ ).

We did not find differences in the rate of plastic closure at our institution.

Comparing their complication profiles, we found trends toward higher complications in neuromuscular scoliosis patients, but these did not reach significance (Table 2). Notably, there was a higher rate of postoperative deaths in patients with neuromuscular scoliosis at 30 days (1.4% neuromuscular scoliosis vs. 0% syndromic scoliosis/congenital scoliosis) and at 1 year (2.8% neuromuscular scoliosis vs. 0% syndromic scoliosis/congenital scoliosis), but the analysis did not show statistical significance. There was a borderline statistically significant increase in the rate of deep surgical site infections (SSI) in patients with neuromuscular scoliosis (10.4% neuromuscular scoliosis vs. 0% syndromic scoliosis vs. 1.9% congenital scoliosis;  $P=0.085$ ). Similarly, neuromuscular scoliosis patients had trends towards increased rates of readmissions at 30 days (14.0% neuromuscular scoliosis vs. 9.3% syndromic scoliosis vs. 3.8% congenital scoliosis;  $P=0.061$ ), return to operating room at 90 days (13.3% neuromuscular scoliosis vs. 4.7% syndromic scoliosis vs. 5.8% congenital scoliosis;  $P=0.145$ ), emergency center visits in 90 days (21.7% neuromuscular scoliosis vs. 14.0% syndromic scoliosis vs. 11.5% congenital scoliosis;  $P=0.106$ ) and need for revision surgery in 1 year for any reason (15.3% neuromuscular scoliosis vs. 9.3% syndromic scoliosis vs. 9.6%;  $P=0.295$ ). However, these did not reach statistical significance. The rates of instrument failure in 1 year, perioperative pneumonia and perioperative deep vein thrombosis/pulmonary embolus were not significantly different.

### Controlling for confounding variables

Because of the differences in baseline characteristics and surgical procedure among the diagnosis groups, we performed a controlled variable analysis. First, a univariate analysis was performed for each diagnosis and the different complications (Table 3). In this analysis, we found that a diagnosis of neuromuscular scoliosis alone was a significant risk factor for SSI ( $P=0.021$ ) and return to the operating room in 90 days ( $P=0.049$ ), while it was a borderline significant risk factor for readmission in 30 days ( $P=0.066$ ) and emergency care visits in 90 days (0.073). Neither syndromic scoliosis nor congenital scoliosis reached significance as a risk factor for any of the complications analyzed.

The differences in baseline characteristics and surgical procedure were BMI, pulmonary disease, technology dependence, seizure disorder, number of surgical levels and pelvis fixation. We therefore controlled for each of these variables to assess their effect on neuromuscular scoliosis diagnosis as a risk factor for complications (Table 3). We found that controlling for BMI did not change the effect of neuromuscular scoliosis alone as a risk factor for any of the complications while controlling for pulmonary disease, technology dependence and

seizure disorder had little effect on SSI, readmission in 30 days and return to the operating room in 90 days. The comorbid conditions did seem to be a subtle confounder for emergency care visits in 90 days and revision surgery in 1 year. Notably, the surgical procedure was an important confounding variable for SSI. Specifically, pelvis fixation was the major confounding variable for each of the complications (Table 3), which effectively increases the *P* value of neuromuscular scoliosis to insignificant levels.

#### Comparing cerebral palsy patients to all other neuromuscular scoliosis patients

Given that cerebral palsy patients are often thought as a distinct group and presumably have higher rates of fixation to the pelvis, we performed a subgroup analysis on this group (Table 4). cerebral palsy patients did have lower BMI and had a higher rate of technology dependence and seizure disorder when compared to neuromuscular scoliosis patients with spinal muscular atrophy, muscular dystrophy, post-traumatic neonatal spinal cord injury, and other neuromuscular scoliosis (to include encephalopathy, hydrocephalus, spina bifida, syndromes when associated with severe global developmental delay, seizures, movement disorders). However, these were determined to not be confounding factors. As expected, cerebral palsy patients did have a higher rate of fixation to the pelvis (81.2% cerebral palsy vs. 48.1% neuromuscular scoliosis other;  $P < 0.001$ ). Despite the higher rate of pelvis fixation in cerebral palsy patients, there were no statistically significant differences in complication rates in cerebral palsy patients (Table 4). However, the difference in mortality in 1 year approached significance with a higher rate in cerebral palsy patients (5.8% cerebral palsy vs. 1.1% neuromuscular scoliosis other;  $P = 0.142$ ).

#### Comparing effect of pelvis fixation on complications based on diagnosis

Because pelvic fixation was found in our analysis to be an independent risk factor for complications, we sought to determine its relative association with postoperative complications (Table 5). When controlling for diagnosis, pelvic fixation was a statistically significant risk factor for SSI [odds ratio (OR) = 2.8 (1.1–7.6);  $P = 0.031$ ] and readmission in 30 days [OR = 4.2 (1.3–18.8);  $P = 0.029$ ].

It did approach (but did not reach) significance for return to operating room in 90 days [OR = 2.8 (1.0–10.5);  $P = 0.079$ ] and emergency care visits in 90 days [OR = 2.0 (0.8–5.3);  $P = 0.129$ ]. When accounting for patient diagnosis, we found that neuromuscular scoliosis increased the OR for SSI (OR = 4.2), readmission in 30 days (OR = 10.8), return to operating room in 90 days (OR = 4.8), emergency care visits in 90 days (OR = 2.8) and revision surgery in 1 year (OR = 2.5) (Table 5). A diagnosis of syndromic scoliosis or congenital scoliosis had minimal effect on the OR for pelvis fixation alone. Interestingly, readmission in 30 days increased nearly four-fold with a diagnosis of syndromic scoliosis most commonly for pneumonia.

#### Discussion

In this study, we have attempted to clarify the clinical differences between nonidiopathic scoliosis subtypes, including those with neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis. The differences are important when determining a patient's risk profile and management, which is best performed on a case-by-case basis. As we have shown, the diagnosis sub-type can be a risk factor for downstream complications. However, it is important to recognize that differences in complication rates are tied to differing preoperative characteristics. This carries additional significance as these subtypes are often categorized together when comparing hospital quality measures across institutions. In the era of value-based medicine, it is relevant to understand the expected rates of complications for more complex populations. Overall, we have shown that neuromuscular scoliosis patients are more likely to be underweight, have higher rates of comorbid pulmonary disease with subsequent technology dependence, and have higher rates of seizure disorder than other nonidiopathic scoliosis subtypes. Seizure disorders have been associated with longer ICU stays and more respiratory complications [12]. Additionally, these patients are more likely to require larger fusion constructs with fixation to the pelvis. With more preexisting conditions and larger procedures, neuromuscular scoliosis patients tend to have more complications. Still, we found only pelvic fixation to be an independent risk factor for complications.

**Table 3 Univariate analysis of each diagnosis subgroup on complications demonstrates neuromuscular scoliosis alone is associated with multiple complications. Multivariate analysis demonstrates the effect of confounding variables on neuromuscular scoliosis *P* value. Pelvis fixation is the confounding factor that removes the association of neuromuscular scoliosis with the listed complications**

	NMS <i>P</i> value by controlling for confounder								
	NMS	SS	CS	BMI z-score	Pulmonary disease	Technology dependence	Seizure disorder	Number of levels	Pelvis fixation
SSI (deep)	0.021	0.992	0.155	0.015	0.042	0.037	0.021	0.989	0.993
Readmission in 30 days	0.066	0.714	0.084	0.065	0.058	0.075	0.061	0.034	0.377
Return to OR in 90 days	0.049	0.209	0.256	0.026	0.113	0.089	0.048	0.01	0.252
Emergency care visits in 90 days	0.073	0.448	0.176	0.046	0.221	0.214	0.123	0.087	0.259
Revision surgery in 1 year	0.186	0.438	0.425	0.089	0.259	0.214	0.112	0.004	0.577

CS, congenital scoliosis; NMS, neuromuscular scoliosis; OR, odds ratio; SS, syndromic scoliosis; SSI, surgical site infections.



**Table 4 Comparison of cerebral palsy and all other neuromuscular scoliosis subtypes on patient preoperative characteristics, surgical procedure and subsequent postoperative complications**

	CP	NMS – other	P value
<b>Patient demographics</b>			
Number of cases	52	92	
Age in years (Ave; SD)	14±3.5	12.9±3.7	0.091
Gender (% female)	40.4	51.1	0.217
BMI (Ave; SD)	17.8±3.2	20.8±6.0	<0.001
BMI z-score (Ave; SD)	-1.1±2.1	0.1±1.7	0.001
BMI percentile (Ave; SD)	34.4±29.9	55.6±36	<0.001
Cardiac disease (%)	9.6	11.1	0.789
Pulmonary disease (%)	50.0	66.7	0.063
Technology dependent (%)	26.9	45.8	0.034
Seizure disorder (%)	65.4	9.7	<0.001
Coagulopathy (%)	7.7	6.9	0.874
<b>Surgical data</b>			
Number of levels (Ave; SD)	16.4±3.8	15.2±3.8	0.099
Number of screws (Ave; SD)	24.5±7.6	21.6±7.9	0.055
Screw density (%)	75.7±18.2	71.1±17.8	0.192
Pelvis fixation (%)	81.2	48.1	<0.001
Plastics closure (%)	1.9	1.1	0.685
<b>Postop complications</b>			
Deaths in 30 days (%)	1.9	1.1	0.685
Deaths in 1 year (%)	5.8	1.1	0.142
SSI (deep) (%)	9.6	10.9	0.813
SSI (superficial) (%)	13.5	9.8	0.502
Readmission in 30 days (%)	11.8	15.2	0.570
Return to operating room in 90 days (%)	9.8	15.2	0.365
Emergency care visits in 90 days (%)	23.5	20.7	0.690
Revision surgery in 1 year (%)	3.8	4.3	0.885
Instrument failure in 1 year (%)	7.7	4.3	0.406
Pneumonia in 30 days (%)	3.8	1.1	0.296

CP, cerebral palsy; NMS, neuromuscular scoliosis; SSI, surgical site infections.

**Table 5 Multivariate analysis showing the odds ratio and P values for the association of pelvis fixation alone with multiple complications**

		Pelvis fixation +		
	Pelvis fixation	NMS	SS	CS
SSI (deep)	OR=2.8 (1.1–7.6), <i>P</i> =0.031	4.2	2.8	2.8
Readmission in 30 days	OR=4.2 (1.3–18.8), <i>P</i> =0.029	10.8	18.1	4.2
Return to operating room in 90 days	OR=2.8 (1.0–10.5), <i>P</i> =0.079	4.8	2.8	2.8
Emergency care visits in 90 days	OR=2.0 (0.8–5.3), <i>P</i> =0.129	2.8	2.5	2.0
Revision surgery in 1 year	OR=1.9 (0.7–5.8), <i>P</i> =0.202	2.5	1.4	1.9

Odds ratios are shown for each diagnostic category when combined with pelvis fixation.

CS, congenital scoliosis; NMS, neuromuscular scoliosis; SS, syndromic scoliosis; SSI, surgical site infections.

In many ways, these findings are expected because neuromuscular scoliosis includes a group of patients with high baseline health-care needs, such as cerebral palsy, muscular dystrophy and spinal muscular atrophy. Many require full-time care to address their activities of daily living. The findings of lower BMI, higher prevalence of pulmonary disease (obstructive sleep apnea or restrictive lung disease), technology dependence and a higher incidence of seizures, are consistent with prior reports [13–15]. These conditions have been shown to predispose neuromuscular scoliosis patients to a higher incidence of postoperative pulmonary issues and pneumonia, with

prior reports ranging from 22 to 50% postoperative pulmonary complications [14–16]. In our study, we did not find higher rates of pneumonia in neuromuscular scoliosis patients compared to syndromic scoliosis and congenital scoliosis patients. However, it has also been shown that patients with syndromic scoliosis [5] and congenital scoliosis [17,18] carry an increased risk of pulmonary complications. As with neuromuscular scoliosis, the presence of preoperative pulmonary disease increases the risk of postoperative pulmonary complications in patients with congenital scoliosis [17,18]. Ultimately, there was a higher mortality rate in neuromuscular scoliosis patients. Despite not reaching statistical significance, each mortality was due to respiratory failure. It is important to recognize that the differences may not have shown significance due to this study being underpowered and future inquiries with larger cohorts may demonstrate a significant difference.

Nevertheless, the differences in BMI and preexisting conditions between neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis were not associated with statistically significant differences in complication profiles. However, neuromuscular scoliosis patients did trend towards higher rates of SSI, in addition to other complications, such as readmission in 30 days, return to operating room in 90 days, emergency care visits in 90 days and revision surgery in 1 year. The rate of SSI in neuromuscular scoliosis patients has been reported to range from 6 to 24% [15,19], while the rates of SSI for idiopathic scoliosis patients are on the order of 1–2% [20]. The rate of SSI in neuromuscular scoliosis patients in this study was consistent with prior reports (10.4%), while the rates in syndromic scoliosis and congenital scoliosis patients, 0% and 1.9%, respectively, were comparable to idiopathic scoliosis patients. This is a similar finding to a large nationwide database comparing syndromic scoliosis and idiopathic scoliosis, which reported 0.4% SSI in each subgroup [5]. This suggests that, although syndromic scoliosis and congenital scoliosis patients are likely to have a higher risk profile than idiopathic patients, they may be closer in risk profile to idiopathic scoliosis than neuromuscular scoliosis subtypes. One possible explanation for the higher infection rate in neuromuscular scoliosis patients includes longer fixation constructs, more often involving instrumentation to the pelvis, which was found in this study. Another explanation is that neuromuscular scoliosis patients are generally immobile, risking pressure ulcers.

When we controlled for potential confounding variables, we found only the surgical procedure to be an independent risk factor for complications. Despite neuromuscular scoliosis patients having lower BMI and higher rates of pulmonary disease, technology dependence and seizure, these characteristics were not found to be independent risk factors for postoperative complications. With regards

to BMI, a prior study also found no differences in postop complications for underweight neuromuscular scoliosis patients, but rather a higher complication rate in obese patients [21]. In our study, we found that surgical procedure was an independent risk factor. Specifically, we found that when controlling for pelvic fixation, the differences in complication profile borne out by diagnosis were eliminated. This suggests that the main risk factor for increased rates of complications in neuromuscular scoliosis patients was pelvic fixation, which has been previously reported [22]. An increased rate of complications after pelvic fixation is likely multifactorial, including that this procedure is typically indicated for nonambulatory patients without trunk control. However, despite pelvic fixation typically being indicated for GMFCS 5 cerebral palsy patients, we did not demonstrate statistically significant increases in any complications in this group when comparing against other neuromuscular scoliosis patients; even though they did have a higher rate of pelvic fixation in our study. At TCH a multidisciplinary pathway has been in effect since 2014 and was developed for all nonidiopathic scoliosis patients before surgery. The first step entails an indications committee review where the Spine Surgeon Committee agrees that surgery is indicated. The preoperative plan including proposed levels of fusion, special equipment needs, need for preoperative halo traction as well as intraoperative traction are discussed and approved by multiple members of the Spine Surgeon Committee. Our current practice is to obtain a group consensus on the proposed fusion levels. Consideration for pelvic fixation is made for all nonambulatory patients who do not exhibit trunk control, GMFCS 5.

The US News and World Report consider all nonidiopathic scoliosis subtypes as a single entity. Among the questions asked by their ranking system that was assessed here, neuromuscular scoliosis patients trended towards a higher rate of readmission in 30 days, return to operating room in 90 days and return to the emergency center in 90 days. Based on our analysis, this may be related to their higher rates of pelvic fixation. Considering the differences, institutions that treat a higher percentage of neuromuscular scoliosis patients in comparison to syndromic scoliosis and congenital scoliosis patients might be unfairly deemed to have a higher rate of complications in their nonidiopathic scoliosis patients. Moreover, there are likely differences among neuromuscular scoliosis sub-diagnoses that are not detailed here but are nevertheless important, such as the higher risk of pulmonary complications that children with Rett syndrome have as compared to those with cerebral palsy [23]. Cohen *et al.* [23] found that despite better preoperative motor function, lower GMFCS levels, as well as shorter anesthesia and surgery durations, patients with Rett Syndrome experienced more respiratory failure, prolonged positive pressure ventilation use, and longer ICU stays after spinal instrumentation and fusion than did children with cerebral palsy [23]. Additionally, Jain *et al.* [8]

demonstrated increased blood loss in patients with cerebral palsy as compared to other neuromuscular scoliosis subtypes, as well as in comparison to syndromic scoliosis. Overall, these differences should be recognized when comparing institutions based on their complication profile for nonidiopathic scoliosis.

The present study has several limitations. First, this is a retrospective study at a single institution, and the findings may therefore not be broadly applicable to other practice types and patient populations. Second, neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis are broad diagnostic definitions, which could be further subdivided into more specific diagnoses to detail their presurgical differences, as well as their subsequent complications. For example, dural ectasia size has been found to increase in size in scoliosis patients with Marfan syndrome and may lead to a higher procedural risk [24]. However, aside from cerebral palsy, the relative sample sizes of each of these sub-diagnoses would be too small to allow a relevant analysis. Lastly, while we have made effort to clarify confounding variables, there are likely more that are not presented here. For example, while we have found pelvic fixation to be an independent risk factor for the complication, this could be because pelvic fixation is reserved for more severe patients who do not exhibit trunk control. Although we attempt to include predisposing factors such as co-morbidities and BMI in the analysis, there are other potential contributing factors not analyzed here, such as preoperative mobility or independence. Still, this does not rule out the possibility that pelvic fixation alone is an independent risk factor even in patients that do not have other predisposing characteristics.

## Conclusion

In conclusion, this study clarifies the differences in baseline characteristics and postoperative complications in neuromuscular scoliosis, syndromic scoliosis and congenital scoliosis. Overall, we found neuromuscular scoliosis patients have a lower BMI, have higher rates of pulmonary disease, technology dependence and seizure disorder; in addition, they are more likely to have larger surgical constructs including pelvic fixation. While neuromuscular scoliosis patients trend towards having higher rates of postoperative complications, such as SSI, this is more likely to be related to their higher rates of pelvic fixation in our study. These differences in risk profile are important when quality measures are assessed in nonidiopathic scoliosis subtypes.

## Acknowledgements

The authors would like to acknowledge the Texas Children's Spine Study Group. Benny Dahl received financial support from the Alfred Benzon Foundation.

## Conflicts of interest

Benny Dahl, consultancy with Stryker. Lorenzo R. Deveza has stock ownership with Lento Medical, Inc. For the remaining authors, there are no conflicts of interest.

## References

- Williams BA, Matsumoto H, McCalla DJ, Akbarnia BA, Blakemore LC, Betz RR, et al. Development and initial validation of the Classification of Early-Onset Scoliosis (C-EOS). *J Bone Joint Surg Am* 2014; **96**:1359–1367.
- Divecha HM, Siddique I, Breakwell LM, Millner PA; British Scoliosis Society Members. Complications in spinal deformity surgery in the United Kingdom: 5-year results of the annual British Scoliosis Society National Audit of Morbidity and Mortality. *Eur Spine J* 2014; **23**(Suppl 1):S55–S60.
- Qiu Y, Zhu F, Wang B, Yu Y, Zhu ZZ, Qian BP, Zhu LH. Clinical etiological classification of scoliosis: report of 1289 cases. *Orthop Surg* 2009; **1**:12–16.
- Salsgiver E, Crotty J, LaRussa SJ, Bainton NM, Matsumoto H, Demmer RT, et al. Surgical site infections following spine surgery for non-idiopathic scoliosis. *J Pediatr Orthop* 2017; **37**:e476–e483.
- Chung AS, Renfree S, Lockwood DB, Karlen J, Belthur M. Syndromic scoliosis: national trends in surgical management and inpatient hospital outcomes: a 12-year analysis. *Spine (Phila Pa 1976)* 2019; **44**:1564–1570.
- Gjolaj JP, Sponseller PD, Shah SA, Newton PO, Flynn JM, Neubauer PR, et al. Spinal deformity correction in Marfan syndrome versus adolescent idiopathic scoliosis: learning from the differences. *Spine (Phila Pa 1976)* 2012; **37**:1558–1565.
- Poorman GW, Jalai CM, Diebo B, Vira S, Buza J, Baker J, et al. Congenital etiology is an independent risk factor for complications in adolescents undergoing corrective scoliosis surgery: comparison of in-hospital comorbidities using Nationwide KID's Inpatient Database. *J Pediatr Orthop* 2019; **39**:406–410.
- Jain A, Njoku DB, Sponseller PD. Does patient diagnosis predict blood loss during posterior spinal fusion in children? *Spine (Phila Pa 1976)* 2012; **37**:1683–1687.
- Kannan S, Meert KL, Mooney JF, Hillman-Wiseman C, Warrier I. Bleeding and coagulation changes during spinal fusion surgery: a comparison of neuromuscular and idiopathic scoliosis patients. *Pediatr Crit Care Med* 2002; **3**:364–369.
- Murphy NA, Firth S, Jorgensen T, Young PC. Spinal surgery in children with idiopathic and neuromuscular scoliosis. What's the difference? *J Pediatr Orthop* 2006; **26**:216–220.
- Berven S. Patient safety in spinal deformity surgery: the development of standard work protocols, moral hazard in decision making, and the need for prospective validation and protocol adoption. *Spine Deform* 2019; **7**:653–654.
- Abousamra O, Sullivan BT, Shah SA, Yaszay B, Samdani AF, Cahill PJ, et al. Do seizures compromise correction maintenance after spinal fusion in cerebral palsy scoliosis? *J Pediatr Orthop B* 2020; **29**:538–541.
- Persson-Bunke M, Häggglund G, Lauge-Pedersen H, Wagner P, Westbom L. Scoliosis in a total population of children with cerebral palsy. *Spine (Phila Pa 1976)* 2012; **37**:E708–E713.
- Rumalla K, Yarbrough CK, Pugely AJ, Koester L, Dorward IG. Spinal fusion for pediatric neuromuscular scoliosis: national trends, complications, and in-hospital outcomes. *J Neurosurg Spine* 2016; **25**:500–508.
- 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.
- Kang GR, Suh SW, Lee IO. Preoperative predictors of postoperative pulmonary complications in neuromuscular scoliosis. *J Orthop Sci* 2011; **16**:139–147.
- Wu L, Zhang XN, Wang YS, Liu YZ, Hai Y. Risk factors for pulmonary complications after posterior spinal instrumentation and fusion in the treatment of congenital scoliosis: a case-control study. *BMC Musculoskelet Disord* 2019; **20**:331.
- Yin S, Tao H, Du H, Feng C, Yang Y, Yang W, Duan C. Postoperative pulmonary complications following posterior spinal instrumentation and fusion for congenital scoliosis. *PLoS One* 2018; **13**:e0207657.
- Ramo BA, Roberts DW, Tuason D, McClung A, Paraison LE, Moore HG 4<sup>th</sup>, Sucato DJ. Surgical site infections after posterior spinal fusion for neuromuscular scoliosis: a thirty-year experience at a single institution. *J Bone Joint Surg Am* 2014; **96**:2038–2048.
- Mackenzie WG, Matsumoto H, Williams BA, Corona J, Lee C, Cody SR, et al. Surgical site infection following spinal instrumentation for scoliosis: a multicenter analysis of rates, risk factors, and pathogens. *J Bone Joint Surg Am* 2013; **95**:800–6, S1.
- Malik AT, Tamer R, Yu E, Kim J, Khan SN. The impact of Body Mass Index (BMI) on 30-day outcomes following posterior spinal fusion in neuromuscular scoliosis. *Spine (Phila Pa 1976)* 2019; **44**:1348–1355.
- Anari JB, Spiegel DA, Baldwin KD. Neuromuscular scoliosis and pelvic fixation in 2015: Where do we stand? *World J Orthop* 2015; **6**:564–566.
- Cohen JL, Klyce W, Kudchadkar SR, Kotian RN, Sponseller PD. Respiratory complications after posterior spinal fusion for neuromuscular scoliosis: children with Rett syndrome at greater risk than those with cerebral palsy. *Spine (Phila Pa 1976)* 2019; **44**:1396–1402.
- Ito K, Kawakami N, Ohara T, Saito T, Tauchi R. Increase in dural ectasia size in scoliosis patients with Marfan syndrome. *J Pediatr Orthop B* 2021; **30**:235–238.