

The Relationship between Preoperative Echocardiographic Evaluation and Spinal Deformity in Patients with Neuromuscular Scoliosis

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Abstract:

Introduction: Echocardiography is an important component of perioperative cardiac risk stratification in patients with neuromuscular scoliosis (NMS). However, there are little data regarding the relationship between preoperative echocardiographic findings and spinal deformity. We retrospectively reviewed preoperative echocardiographic data to investigate the relationship between echocardiographic evaluation and spinal deformity in NMS.

Methods: We reviewed 73 NMS patients (mean age: 13.3 years, male 66%) who underwent spinal correction surgery between 2008 and 2016. Echocardiographic data including ejection fraction (EF), valvar disease, and inferior vena cava diameter were collected from the preoperative exam. Demographic and radiographic data were also collected.

Results: Preoperative diagnoses included Duchenne muscular dystrophy, Fukuyama congenital muscular dystrophy, other dystrophy, spinal muscular atrophy, and congenital myopathies. Mean Body Mass Index (BMI) was 15.6 kg/m². Mean major Cobb angle before surgery was 86.6 ± 28.2°. Because of technical difficulty, complete echocardiographic data could only be collected and evaluated in 49.3% of patients. Neither right nor left sided cardiac evaluation could be completed in 20.5%. Patients in whom complete echocardiographic data could not be collected had significantly more extensive thoracic scoliosis with a more rigid curve and hypokyphosis, and were of lower weight and BMI than patients in whom complete data could be collected. Ten cases (13.7%) were diagnosed as having minor heart-related complications immediately after surgery, and they had higher right atrial pressures preoperatively.

Conclusions: Echocardiography can be technically difficult in NMS patients with extensive spinal deformities. We found that perioperative cardiac function could only be evaluated by echocardiogram in about half of NMS patients undergoing spinal correction surgery. The absence of an adequate preoperative cardiac evaluation could render these patients more susceptible to perioperative heart-related complications. Echocardiography may not be sufficient to evaluate cardiac conditions in children with extensive NMS.

Keywords:

neuromuscular scoliosis, echocardiography, perioperative complication, spinal correction

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Introduction

Spinal deformity in neuromuscular patients tends to be progressive, leading to deterioration in sitting-balance, general health, and quality of life¹⁾. Progressive disease in patients with neuromuscular scoliosis (NMS) can be difficult to manage with traditional braces and the majority of patients require surgical spinal correction²⁾. Unfortunately, spinal fusion and correction in NMS has a high rate of pe-

rioperative complications^{3,4)}. Pulmonary complications are one of the most frequently reported complications⁵⁾. NMS patients with compromised pulmonary function prior to surgery have a risk of pneumonia, atelectasis, and other respiratory complications in the post-operative setting^{6,7)}. Perioperative cardiac complications such as heart failure and cardiac arrest have also been reported^{8,9)}. Megna *et al.* reviewed over 2,000 cases of scoliosis and reported a three-fold higher risk of intraoperative cardiopulmonary arrest in patients with

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NMS¹⁰). Cardiac arrest was generally due to a cardiovascular cause in this patient population, making echocardiographic risk stratification an important part of the preoperative evaluation. However, there are no published studies examining the relationship between preoperative echocardiographic data and spine deformity in NMS patients. We retrospectively investigated echocardiographic data in NMS children just before surgery. This is the first cohort study to report a relationship between preoperative echocardiographic evaluation and spine deformity in NMS patients.

Materials and Methods

This was a retrospective single center study. We analyzed 73 NMS patients (mean age: 13.3 years, male 66%) who underwent spinal correction surgery between 2008 and 2016. Preoperative echocardiography was routinely performed by qualified examiners to evaluate perioperative cardiac risk. Echocardiographic data including ejection fraction (EF), valvular disease, and inferior vena cava (IVC) diameter were collected from patients' records. Left heart function was evaluated by EF, and by aortic and mitral valve abnormalities. Right heart function was evaluated by IVC diameter at baseline and with respiration, and by pulmonary and tricuspid valve abnormalities. Based on the guidelines reported by Rudski *et al.*, we defined normal right atrial (RA) pressure as IVC diameter ≤ 2.1 cm with $>50\%$ collapse with sniff, and elevated RA pressure as IVC diameter >2.1 cm with $<50\%$ collapse with sniff. IVC diameter and collapse that did not fit these parameters was considered to represent intermediate RA pressure¹¹). We divided patients as follows: Group A: Transthoracic echocardiogram was adequately performed and all of the right and left heart parameters could be evaluated, Group B: Some of the left or right heart parameters could not be evaluated, indicating that the left or right echocardiographic assessment was not adequate, Group C: Neither left nor right heart parameters could be evaluated, indicating that the echocardiographic assessment was inadequate or failed. Demographic data including diagnosis, age, gender, height, weight, and Body Mass Index (BMI) were collected. The major curve Cobb angle and thoracic Cobb angle were measured using anterior-posterior radiographic imaging, and thoracic kyphosis (TK), thoracolumbar kyphosis (TLK), and lumbar lordosis (LL) were measured using whole spine radiography in the sitting position. Bending Cobb angle, TK, TLK, and LL were also measured in the supine position. We performed posterior instrumentation in all cases and collected clinical data for perioperative heart-related complications diagnosed by chest x-ray or unstable low blood pressure. We compared preoperative parameters, including age, height, body weight, BMI, preoperative major Cobb angle, EF, the rate of Groups A, B, and C in patients who did or did not have perioperative heart-related complications; furthermore, we compared the presence or absence of valvular disease.

Table 1. Patients' Demographic Data.

Patients [number]	73
Age [years] (mean \pm SD)	13.3 \pm 2.5
Gender	Male 48, Female 25
Diagnosis (number)	DMD (29) FCMD (11) BMD (1) UMD (4) Non-FCMD (4) SMA (17) Congenital myopathy (4) Noonan syndrome (1) Spinocerebellar degeneration (1) Multicore disease (1)
Height [cm] (mean \pm SD)	144.7 \pm 13.4
Weight [kg] (mean \pm SD)	33.6 \pm 14.8
BMI [kg/m ²] (mean \pm SD)	15.6 \pm 5.4

DMD: Duchenne muscular dystrophy

FCMD: Fukuyama congenital muscular dystrophy

BMD: Becker muscular dystrophy

UMD: Ullrich muscular dystrophy

SMA: Spinal muscular dystrophy

Statistical analysis

Statistical Package for Social Sciences (SPSS[®]) software, version 19.0 (IBM Japan Business Services Co., Ltd., Tokyo, Japan) was used for statistical analysis. Group comparisons were analyzed using independent-samples *t* test or a Chi-squared test. Significance was established at a value of $P < 0.05$.

Results

Patient Characteristics

Preoperative diagnoses included Duchenne muscular scoliosis (DMD) (29 patients), Fukuyama congenital muscular dystrophy (FCMD) (11 patients), Becker muscular dystrophy (BMD) (1 patient), Ullrich muscular dystrophy (UMD) (4 patients), Non-FCMD (4 patients), Spinal muscular atrophy (SMA) (17 patients), Congenital myopathy (4 patients), Noonan syndrome (1 patient), Spinocerebellar Degeneration (1 patient), and Multicore disease (1 patient). The mean age at the time of surgery was 13.3 years old (standard deviation (SD) 2.5 years). Forty-six patients (66%) were male and 27 (34%) were female. The mean height and weight were 144.7 cm (SD 13.4 cm) and 33.4 kg (SD 14.8 kg), respectively. The mean BMI was 15.6 kg/m² (SD 5.4 kg/m²). Patient demographic data are summarized in Table 1.

Echocardiography

Complete right and left heart examination could be completed (Group A) in 49.3% of patients (36/73 cases). Because of technical difficulty, left heart exam alone could be evaluated (Group B) in 30% (22/73 cases). Neither right nor

left heart exam could be completed (Group C) in 20.5% (15/73 cases) (Table 2). EF was evaluated in 95.9% (70/73) and EF \leq 50% was detected in 8.6% (6/70). The aortic valve could be evaluated in 80.8% (59/73 cases) and no abnormalities were observed. The mitral valve could be evaluated in 84.9% (62/73 cases) and mitral regurgitation (MR), including trivial valve regurgitation, was observed in 15.9% (11/62 cases). The IVC could be evaluated in 69.9% (51/73 cases). Based on IVC diameter and collapse with sniff, 54.9% (28/51 cases) were estimated to have normal right atrium (RA) pressure and 45.1% (23/51 cases) were estimated to have intermediate RA pressure. The tricuspid valve could be evaluated in 78.1% (57/73) and valve abnormality was detected in 47.4% (27/57). The pulmonary valve could be evaluated in 63.0% (46/73) and valve abnormality was detected in 28.3% (13/46) (Table 3).

Radiographic Measurements

The mean preoperative major Cobb angle and thoracic Cobb angle in the sitting position were 86.6° (SD 28.2°) and 39.4° (SD 30.3°), respectively. TK, TLK, and LL were 16.2° (SD 29.9°), 18.9° (SD 20.6°), and -8.3° (SD 47.7°), respectively. Bending Cobb angle, TK, TLK, and LL in the supine position were 57.1° (SD 27.4°), 13.1° (SD 16.6°), 3.4° (SD 18.9°), and 24.8° (SD 24.7°), respectively (Table 4).

Table 2. Patient Groups Based on Echocardiography Evaluation.

Group	Percentage (number)
A	48.3% (36/73)
B	30.0% (22/73)
C	20.5% (15/73)

Group Comparisons

We compared radiographic data, age, height, body weight, and BMI between Group A (N = 36) and Group C (N = 15). Compared to Group A, Group C had significantly larger thoracic Cobb angles and bending Cobb angles, less flexibility of the major Cobb angle, and less thoracic kyphosis ($p = 0.01, 0.011, 0.037, \text{ and } 0.045$). Weight and BMI were lower in Group C than Group A ($p = 0.049 \text{ and } 0.014$) (Table 5).

Heart-related complications

Although we did not experience any cases of severe heart-related complications such as cardiac arrest or acute heart failure perioperatively, ten cases (13.7%) were diagnosed as having minor heart-related complications immediately after surgery. Three cases were diagnosed by increased cardiothoracic ratios measured on chest x-rays and seven other cases by unstable blood pressure. They were successfully treated with vasopressor and/or diuretic drugs. We could not find any statistical difference in preoperative clinical

Table 4. Radiographic Data.

	mean \pm SD
Major Cobb [°]	86.6 \pm 28.2
Bending Cobb [°]	57.1 \pm 27.4
Thoracic Cobb [°]	39.4 \pm 30.3
TK [°]	16.2 \pm 29.9
TLK [°]	18.9 \pm 20.6
LL [°]	-8.3 \pm 47.7
Su TK [°]	13.1 \pm 16.6
Su TLK [°]	3.4 \pm 18.9
Su LL [°]	24.8 \pm 24.7

TK: Thoracic kyphosis
 TLK: Thoracolumbar kyphosis
 LL: Lumbar lordosis
 Su: in supine position

Table 3. Echocardiography Findings.

	Evaluation Rate	Findings	Percent (number)
EF	95.9% (70/73)	\leq 50%	8.6% (6/70)
Aortic Valve	80.8% (59/73)	no abnormalities	
Mitral Valve	84.9% (62/73)	MR	15.9% (11/62)
Tricuspid Valve	78.1% (57/73)	TR	47.4% (27/57)
Pulmonary Valve	63.0% (46/73)	PR	28.3% (13/46)
Estimated RA Pressure			
IVC diameter	69.9% (51/73)	normal	54.9% (28/51)
		intermediate	45.1% (23/51)
		high	0.0% (0/51)

EF: Ejection fraction
 IVC: Inferior vena cava
 MR: Mitral valve regurgitation
 TR: Tricuspid valve regurgitation
 PR: Pulmonary valve regurgitation
 RA: Right atrium

Table 5. Comparison of Demographic and Radiographic Data between Group A and Group C.

Variable	Group A	Group C	p-value
Patients [number]	36	15	
Age [years] (mean±SD)	13±1.9	14.1±2.9	0.095
Height [cm] (mean±SD)	146.6±13.3	146.2±14.8	0.931
Weight [kg] (mean±SD)	37.4±15.0	28.7±11.5	0.049*
BMI [kg/m ²] (mean±SD)	17±5.5	13±3.7	0.014*
Cobb [°] (mean±SD)	79.1±24.6	95.5±33.1	0.057
Bending Cobb [°] (mean±SD)	49.2±22.9	69.5±29.5	0.011*
Flexibility [%]	39.2±16.7	28.4±15.6	0.037*
Thoracic Cobb [°] (mean±SD)	29.4±21.0	50.5±34.5	0.010*
TK [°] (mean±SD)	18.6±25.9	1.7±28.2	0.045*
TLK [°] (mean±SD)	-19.4±23.1	-16.8±19.2	0.707
LL [°] (mean±SD)	-4.3±41.6	-13.7±55.0	0.510
Su TK [°] (mean±SD)	14.3±14.7	7.4±9.1	0.103
Su TLK [°] (mean±SD)	-4.4±18.9	-0.3±22.3	0.506
Su LL [°] (mean±SD)	24±25.0	25.1±21.0	0.885

TK: Thoracic kyphosis

TLK: Thoracolumbar kyphosis

LL: Lumbar lordosis

Su: in supine position

* p<0.05

cal and echocardiographic parameters between patients who did and did not have perioperative heart-related complications, except for the existence of a difference in intermediate estimated RA pressure (p = 0.02).

Discussion

This study revealed that, because of technical difficulties, a complete right and left heart echocardiographic evaluation could be completed in only 49.3% of NMS patients before spinal surgery. In NMS patients, a large and rigid thoracic Cobb angle with hypokyphosis and lower weight and BMI correlated with technically difficult echocardiographic exams, resulting in a poor preoperative cardiac evaluation.

van Bockel *et al.* commented that scoliosis and poor echocardiographic acoustic windows in patients with DMD hamper accurate cardiac assessment¹². Chuah *et al.* reported a case in which transthoracic echocardiographic images were distorted due to the effect of kyphoscoliosis on cardiac orientation¹³. However, ours is the first study to demonstrate a correlation between spinal deformity and difficulty of echocardiography. Our study showed that only 49.3% of NMS patients could undergo adequately bilateral cardiac evaluation prior to surgery. We also found that echocardiography can be difficult in patients with larger and more rigid thoracic scoliosis and hypokyphosis. Yamamoto *et al.* have pointed out that adequate echocardiographic examination is often difficult in patients with scoliosis because of posture limitations resulting from physical deformities¹⁴. Progressed rigid spinal deformities with limb contractures make it more difficult for patients to remain appropriately positioned for echocardiographic examination, leading to poor echocar-

diographic acoustic windows and a low yield echocardiographic evaluation. Additionally, our results suggested that patients with lower weight and BMI had more difficulty with echocardiographic evaluation. It is possible that the uneven chest surface in these patients made it difficult to fit the echocardiography probe on the chest wall, leading to poor acoustic windows and difficult evaluation.

Several cases of cardiac arrest and acute heart failure have been reported during neuromuscular scoliosis surgery. The authors of these reports indicate that accurate prediction of perioperative cardiac risk was difficult^{8,9,15}. Our results imply that a higher RA pressure could be a risk factor for perioperative heart-related complications. However, because of the limited sample size in the current study, it is hard to conclude that a higher RA pressure can clearly predict the perioperative heart-related complications. So, as the previous authors indicated, we think detection of obvious perioperative heart-related events is still difficult. In addition, based on our findings, we hypothesize that a difficult and inaccurate preoperative echocardiographic evaluation of NMS patients is one reason for unexpected severe perioperative cardiac complications. In patients with advanced NMS, particularly in those with large and rigid thoracic deformities and low BMI, we would recommend that clinicians evaluate the risk of perioperative heart-related events with additional cardiac assessments, such as brain natriuretic peptide, creatine kinase¹⁶, multigated cardiac radionuclide ventriculography¹⁷, and cardiac magnetic resonance imaging^{18,19}.

In this study, several valvular abnormalities (including trivial valve regurgitation) were detected. The prevalence of MR was 15.9%, TR 47.4%, and PR 28.3%. In adolescent idiopathic scoliosis (AIS), the prevalence of cardiac valve abnormalities has been reported as 13% to 28%^{20,21}. Our study suggests that NMS patients have a higher prevalence of valvular abnormalities than AIS patients. It has been reported that DMD and BMD patients have high rates of echocardiographic abnormalities due to degenerative changes of the papillary muscles or the ventricular myocardium²²⁻²⁴. Our study included 49 patients (67.1%) with muscular dystrophy, which may have biased our findings. Furthermore, it has been reported that patients with severe scoliosis have compromised right ventricular function²⁵. In the current study, RA pressure was estimated by IVC diameter and we did not detect any signs of right heart dysfunction. Given the high prevalence of heart abnormalities in NMS patients, accurate preoperative cardiac evaluation is an important component of risk stratification prior to spinal correction surgery.

One limitation of this study is that the results of echocardiography could be influenced by the experience or technique of the examiners. In the future, a study using the results of standardized echocardiographic examinations may be needed.

Echocardiography can be technically difficult in children with advanced spinal deformities. We found that only about half of NMS patients could be adequately evaluated by pre-

operative echocardiography. The lack of adequate cardiac evaluation prior to surgery could lead to unexpected and serious heart-related perioperative complications. Echocardiography could be insufficient to evaluate cardiac conditions in children with advanced NMS and additional examinations may be needed.

Conflicts of Interest: The authors declare that there are no conflicts of interest.

References

- Gibson DA, Koreska J, Robertson D, et al. The management of spinal deformity in Duchenne's muscular dystrophy. *Orthop Clin North Am.* 1978; 9: 437-50.
- Sussman MD. Treatment of scoliosis in Duchenne muscular dystrophy. *Dev Med Child Neurol.* 1985; 27: 522-4.
- Ramirez N, Richards BS, Warren PD, et al. Complications after posterior spinal fusion in Duchenne's muscular dystrophy. *J Pediatr Orthop.* 1997; 17: 109-14.
- Modi HN, Suh SW, Yang JH, et al. Surgical complications in neuromuscular scoliosis operated with posterior- only approach using pedicle screw fixation. *Scoliosis.* 2009; 4: 11.
- Beckmann K, Lange T, Gosheger G, et al. Surgical correction of scoliosis in patients with severe cerebral palsy. *Eur Spine J.* 2016; 25: 506-16.
- Kang GR, Suh SW, Lee IO. Preoperative predictors of postoperative pulmonary complications in neuromuscular scoliosis. *J Orthop Sci.* 2011; 16: 139-47.
- Harper CM, Ambler G, Edge G. The prognostic value of preoperative predicted forced vital capacity in corrective spinal surgery for Duchenne's muscular dystrophy. *Anaesthesia.* 2004; 59: 1160-2.
- Schmidt GN, Burmeister MA, Lilje C, et al. Acute heart failure during spinal surgery in a boy with Duchenne muscular dystrophy. *Br J Anaesth.* 2003; 90: 800-4.
- Irwin MG, Henderson M. Cardiac arrest during major spinal scoliosis surgery in a patient with Duchenne's muscular dystrophy undergoing intravenous anaesthesia. *Anaesth Intensive Care.* 1995; 23: 626-9.
- Menga EN, Hirschfeld C, Jain A, et al. Intraoperative Cardiopulmonary Arrest in Children Undergoing Spinal Deformity Correction: Causes and Associated Factors. *Spine (Phila Pa 1976).* 2015; 40: 1757-62.
- Rudski LG, Lai WW, Afilalo J, et al. Guidelines for the echocardiographic assessment of the right heart in adults: a report from the American Society of Echocardiography endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. *J Am Soc Echocardiogr.* 2010; 23: 685-713.
- van Bockel EA, Lind JS, Zijlstra JG, et al. Cardiac assessment of patients with late stage Duchenne muscular dystrophy. *Neth Heart J.* 2009; 17: 232-7.
- Chuah SS, Al-Mohammad A. Large Eustachian valve and kyphoscoliosis. *Heart.* 2005; 91: e17.
- Yamamoto T, Kawano S, Sugiyama D, et al. Predicting scores for left ventricular dysfunction in Duchenne muscular dystrophy. *Pediatr Int.* 2012; 54: 388-92.
- Reid JM, Appleton PJ. A case of ventricular fibrillation in the prone position during back stabilisation surgery in a boy with Duchenne's muscular dystrophy. *Anaesthesia.* 1999; 54: 364-7.
- Yamamoto T, Kawano S, Sugiyama D, et al. Predicting scores for left ventricular dysfunction in Duchenne muscular dystrophy. *Pediatr Int.* 2012; 54: 388-92.
- van Bockel EA, Lind JS, Zijlstra JG, et al. Cardiac assessment of patients with late stage Duchenne muscular dystrophy. *Neth Heart J.* 2009; 17: 232-7.
- Silva MC, Meira ZM, Gurgel Giannetti J, et al. Myocardial delayed enhancement by magnetic resonance imaging in patients with muscular dystrophy. *J Am Coll Cardiol.* 2007; 49: 1874-9.
- Puchalski MD, Williams RV, Askovich B, et al. Late gadolinium enhancement: precursor to cardiomyopathy in Duchenne muscular dystrophy? *Int J Cardiovasc Imaging.* 2009; 25: 57-63.
- Dhuper S, Ehlers KH, Fatica NS, et al. Incidence and risk factors for mitral valve prolapse in severe adolescent idiopathic scoliosis. *Pediatr Cardiol.* 1997; 18: 425-8.
- Hirschfeld SS, Rudner C, Nash CL Jr, et al. Incidence of mitral valve prolapse in adolescent scoliosis and thoracic hypokyphosis. *Pediatrics.* 1982; 70: 451-4.
- D'Orsogna L, O'Shea JP, Miller G. Cardiomyopathy of Duchenne muscular dystrophy. *Pediatr Cardiol.* 1988; 9: 205-13.
- Reeves WC, Griggs R, Nanda NC, et al. Echocardiographic evaluation of cardiac abnormalities in Duchenne's dystrophy and myotonic muscular dystrophy. *Arch Neurol.* 1980; 37: 273-7.
- Sanyal SK, Leung RK, Tierney RC, et al. Mitral valve prolapse syndrome in children with Duchenne's progressive muscular dystrophy. *Pediatrics.* 1979; 63: 116-23.
- Li S, Yang J, Li Y, et al. Right ventricular function impaired in children and adolescents with severe idiopathic scoliosis. *Scoliosis.* 2013; 8: 1.

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