Comparison between operated muscular dystrophy and spinal muscular atrophy patients in terms of radiological, pulmonary and functional outcomes

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Comparison between operated muscular dystrophy and spinal muscular atrophy patients in terms of radiological, pulmonary and functional outcomes

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ABSTRACT

Comparison between operated muscular dystrophy and spinal muscular atrophy patients in terms of radiological, pulmonary and functional outcomes

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Study Design: Retrospective comparative study.
Purpose: To study and compare the surgical outcomes of muscular dystrophy (MD) and spinal muscle atrophy (SMA).
Overview of Literature: There are few reports that have evaluated and compared the surgical outcomes of MD and SMA patients.
Methods: The patients (n = 35) were divided into two groups: a MD group with 24 patients and a SMA group with 11patients. The average follow-up period was 21 months. All patients were operated for scoliosis correction using posterior instrumentation and fusion. In the immediate postoperative period, all efforts were made to reduce the pulmonary complications using non-invasive positive pressure ventilation and coughing assist devices. The patients were evaluated by radiograph in terms of the Cobb's angle, pelvic obliquity, T1 translation, thoracic kyphosis and lumbar lordosis. The pulmonary function and self-image satisfaction were also assessed.
Results: There was a lower correction rate in the MD group (41.5%) than in the SMA group (48.3%), even though the curves were smaller in the MD group. The correction in the pelvic obliquity was significantly better in the SMA group (p = 0.03).The predicted vital capacity showed a 4%

reduction in the MD group 1 year after surgery, while the SMA group showed a10% reduction. The peak cough flow and end tidal PCO_2 did not deteriorate and were well maintained. The average score for the improvement in self-image satisfaction postoperatively was 3.96 and 4.64 for the MD and SMA groups, respectively. The total complication rate was 45.7%; 14.3% of which were respiratory-related.

Conclusions: Surgical intervention for MD and SMA may be performed safely in patients with a very low forced vital capacity (< 30%) through aggressive preoperative and postoperative rehabilitation efforts.

Key words: muscular dystrophy, spinal muscular atrophy, neuromuscular scoliosis, surgical correction, pulmonary function

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I. INTRODUCTION

The progression of curves in patients with flaccid neuromuscular scoliosis, such as muscular dystrophy (MD) and spinal muscular atrophy (SMA), show a characteristically rapid progression, which can result in them being wheelchair-bound at an early age. Surgical intervention is often necessary before these curves become stiff. Early surgical correction has the definite advantage of better surgical outcomes with fewer complications[1,2]. The primary objective is to maintain the balance of the torso over a level pelvis in order to improve the functional independence and quality of life [1-9].Many studies reported that surgical intervention improves not only the sitting balance but also the quality of life[8,10-12]. However, whether surgical correction has the same beneficial effects on improving the pulmonary function is still controversial. A deterioration in the pulmonary function is a common complication in the postoperative period[4,5,13-16]. In patients with Duchenne muscular dystrophy (DMD) and SMA, the factor most closely associated with the morbidity and mortality of patients is the pulmonary function [17-19]. The probability of intraoperative and postoperative complications are increased in cases where the preoperative pulmonary function have already deteriorated, which can lead to a clinical dilemma as to whether surgical intervention should still be

contemplated. Many studies focused on the pulmonary function of either patients with DMD or SMA. However, there are no studies that evaluated and compared the outcomes of these two conditions. This study examined and compared the surgical outcomes of DMD and SMA to assess the differences in the improvement of the overall sitting balance, changes in pulmonary function, and overall functional status.

II. MATERIALS AND METHODS

Among the neuromuscular scoliosis patients treated with a surgical correction from 2002 to 2006 at our institution, only those patients with progressive flaccid neuromuscular scoliosis (MD and SMA) were enrolled in this study (n =35). These patients were divided into two groups: MD group (n = 24) and SMA group (n = 11). Within the MD group, there were 19 patients with DMD, 2 patients with the Fascio-Scapular-Humeral type, 1 patient with the Becker type and 2 patients who remained unclassified. The average follow up period was 21 months (range, 12 to 54 months).

Table 1. Demographic and operative data

	Total	MD	SMA	p-value
Cases(n)	35	24	11	
Age(yr)	12.9±3.3	13.2±3.3	12.3±3.2	0.4397
Male: Female	26:9	19:5	7:4	0.3437
Height(cm)	145.8±16.6	147.5±18.3	141.9±11.8	0.3580
Weight(kg)	37.4±11.8	38.0±12.7	36.2±9.7	0.6711
Follow up(mo)	21.2±11.0	20.3±10.7	23.2±11.9	0.4710
Operation time(min)	715±226	688±228	773±222	0.3075
Blood loss(ml)	2,488±1,281	2,418±1,292	2,641±1,305	0.6096
Ventilator(day)	2.9±3.5	2.7±3.5	3.4±3.7	0.5951
ICU care(day)	3.7±4.0	3.5±3.5	4.4±5.0	0.5394
Complications(n)	16(45.7%)	11(45.8%)	5(45.5%)	0.9839

MD: Muscular dystrophy, SMA: Spinal muscle atrophy, ICU: Intensive care unit.

Table 1 shows the demographic and surgical data for both groups. Routine posterior instrumentation and fusion was performed using pedicle screws or sublaminar wires. Fusion up to the sacrum/pelvis was performed in 27 cases. Sacro-pelvic fixation using either iliac screws or Galveston was performed only for those cases with a sitting pelvic obliquity >10° using either iliac screws or a Galveston rod. In these cases, the anterior discectomy and fusion was performed prior to posterior procedures. After surgery, all patients were transferred to the intensive care unit. In the immediate postoperative period, all efforts were made to reduce the pulmonary complications using non-invasive positive pressure ventilation (NIPPV) and a coughing assist devices[17,20].

1. Radiological evaluation

Whole spine postero-anterior and lateral radiographs were taken in either the standing or sitting position preoperatively, immediately postoperatively, and every year thereafter. The radiographic coronal plane measurement parameters included the Cobb's angle, sitting pelvic obliquity, and T1vertebral (T1) translation. The T1 translation was measured as the horizontal offset between a vertical line drawn from the center of T1 to the central sacral vertical line [21]. The radiographic sagittal plane measurement parameters included the thoracic kyphosis and total lumbar lordosis measured using the Cobb's method. To avoid inter-observer and intra-observer variability, all radiological measurements were carried out twice by two orthopedic surgeons (HSC, JYK) with the mean of four sets of values used for statistical analysis.

2. Pulmonary function tests

In all patients, the pulmonary function tests were carried out immediately before and after surgery, at the one year follow-up, and every year thereafter. A respiratory rehabilitation specialist at the department of rehabilitation not associated with this study assessed the respiratory function. The pulmonary function in the sitting position was evaluated using the following parameters:

the vital capacity using a Micro Spirometer® (Micro medical Ltd, Rochester, UK), peak cough flow (PCF) (l/min) by ASSESS® (Health Scan Products Inc., Cedar Grove, NJ, USA), and the end tidal Pressure of CO_2 (ET PCO_2) content (mm Hg) using a CO_2partial pressure meter (BCI8200 Capnocheck Capnometer®, BCI International, Waukesha, WI, USA). The ability to expectorate was specifically noted and the PCF was measured because both can be useful predictors of postoperative respiratory complications. The ET PCO_2 was measured to monitor for hypercapnia because it was not feasible to obtain the arterial blood samples of the patients repeatedly. A section of the St. George's Respiratory Questionnaire (Division of Physiological Medicine, St. George's Hospital Medical School, London, UK) was used to assess the subjective changes in the pulmonary function (Table 2) [22,23].

Table 2. A part of St. George's Respiratory Questionnaire.

The answers below refer to activities that have typically caused you shortness of breath in the last few days
Grade 1: Sitting or lying still
Grade 2: Getting washed or dressed
Grade 3: Walking around the home
Grade 4: Walking outside on the level
Grade 5: Walking up a flight of stairs
Grade 6: Walking up hills
Grade 7: Playing sports or games

3. Functional evaluation and satisfaction

The patients' function was measured using the Swinyard et al. [24] functional grade scale. The level of patients' satisfaction and self-image was assessed via telephone interviews using question numbers 21, 22, 23, 24 and 30 of the SRS questionnaire ver. 30 (encompassing ver. 22 and 24,modified 11/12/03).SAS ver. 9.1 (SAS Inc., Cary, NC, USA) was used for statistical analysis. A student's t-test was used to compare the preoperative and postoperative data

between the two groups. A p-value < 0.05 considered significant.

III. RESULTS

The average number of segments fused were 15.8 ± 2.1(range, 11 to 19 segments) in the MD group and 16.1 ± 1.8(range, 12 to 19) in the SMA group. The preoperative Cobb's angle in the MD group was 67.6 ± 18.0°, which improved to 39.5 ± 24.3°. On the other hand, in the SMA group, the Cobb's angle improved from 80.7 ± 22.5° to41.7 ± 18.7° at the final follow-up. The MD group had a lower correction rate (41.5%) than the SMA group (48.3%) but the difference was not significant (p = 0.13). The preoperative pelvic obliquity in the MD group was 19.6 ± 10.3°, which improved to 10.5 ± 5.9°. The SMA group showed significantly better correction from 14.3 ± 7.9°to 7.3 ±3.2°(p = 0.03) (Table 3).

Table 3. Radiological parameters in coronal plane

		Cobb's angle			T1 translation			Pelvic obliquity		
	Cases	Preop	Postop	Last	Preop	Postop	Last	Preop	Postop	Last
Total	No	72.0±20.2	38.4±22.4	40.2±22.3	3.7±3.1	2.3±1.7	2.9±2.6	17.9±9.8	8.8±4.8	9.4±5.4
MD	24	67.6±18.0	38.1±24.1	39.5±24.3	4.4±3.6	2.6±1.9	3.3±3.0	19.6±10.3	9.3±4.9	10.5±5.9
SMA	11	80.7±22.5	39.0±19.6	41.7±18.7	2.4±1.3	1.7±1.3	2.2±1.4	14.3±7.9	7.8±4.7	7.3±3.2
p-value		0.0798	0.9168	0.7942	0.024	0.183	0.247	0.1457	0.4117	0.0299

Preop: Preoperative, Postop: Postoperative, Last: In last follow up, MD: Muscular dystrophy, SMA: Spinal muscle atrophy.

Table 4. Radiological parameters in sagittal plane

		T-K (T5-T12)			L-L(L1-L5)		
	Cases	Preop	Postop	Last	Preop	Postop	Last
Total	No.	32.8±21.5	26.2±15.8	28.3±18.8	13.2±23.3	34.7±16.0	36.2±16.0
MD	24	32.0±21.5	23.0±15.9	24.0±18.6	9.4±37.3	33.7±15.0	34.5±14.3
SMA	11	34.4±22.6	32.6±14.0	36.8±16.8	21.0±8.4	36.8±18.4	39.8±19.2
p-value		0.7630	0.0986	0.0635	0.0775	0.6081	0.3744

T-K: Thoracic kyphosis, L-L: Lumbar lordosis, Preop: Preoperative, Postop:

Postoperative, Last: In last follow up, MD: Muscular dystrophy, SMA: Spinal muscle atrophy.

Tables 3 and 4 list the other radiological results. Pulmonary function tests were performed on 27 and 10patients one and two years after surgery. The vital capacity at 1 year after surgery, which was measured as a percentage of the predicted normal value according to the age and trunk height of the patient, showed a 4% and 10% decrease in the MD and SMA groups, respectively. Although the SMA group showed a greater reduction in the predicted vital capacity, the difference in the absolute vital capacity changes between the two groups was not significant ($p > 0.05$) (Table 5).

Table 5. Pulmonary function tests

		Vital capacity				PCF		$ETCO_2$	
	Cases	Preop		Pod 1yr		Preop	Pod 1yr	Preop	Pod 1yr
	No.	ml	%a)	ml	%a)	l/min	l/min	mmHg	mmHg
MD	19	1342±592	48±16	1290±616	44±20	251±69	250±108	39±7	38±5
SMA	8	1029±610	40±23	835±458	30±14	179±42	200±37	35±3	33±5
p-value		0.2245	0.2965	0.0728	0.0787	0.0699	0.3847	0.1956	0.0264

PCF: Peak cough flow, $ETCO_2$: End tidal CO_2 content, Preop: Preoperative, Pod 1yr: At 1 year postoperative, MD: Muscular dystrophy, SMA: Spinal muscle atrophy. %a): The percentage of the predicted value of vital capacity considering the patients' height.

Table 6. The change of vital capacity at 2 years follow-up

	Cases	Preop		Pod 1yr		Pod 2yr	
	No.	ml	%a)	ml	%a)	ml	%a)
MD	5	1070	42.1	1104	38.7	1024	36.7
SMA	5	1280	48.1	964	34.1	904	31.1

Preop: Preoperative vital capacity, Pod 1yr: Vital capacity at postoperative 1 year, Pod 2yr: Vital capacity at postoperative 2 years, MD: Muscular dystrophy,

SMA: Spinal muscle atrophy. %a): The percentage of the predicted value of vital capacity considering the patients' height.

On the other hand, both groups showed a similar 2-3% decrease in the vital capacity at the end of the second year (Table 6). However, the PCF and ET PCO_2 values did not deteriorate. Indeed, the PCF and ET PCO_2 values measured at the end of 1 year showed little deterioration and were almost comparable to their preoperative values (Table 5). The PCF values in the SMA group at the end of 1year even showed improvement from 179 ± 42 l/min to200 ± 37 l/min. Thirty-four patients, excluding one MD patient who died, were evaluated using the respiratory questionnaire both preoperatively and at the 1-year follow-up (Table 2). No significant differences were found between the two groups preoperatively and at 1-year follow-up, and significant improvement in respiratory grade was observed in the entire patient population at the final follow up ($p = 0.0191$). No significant differences in the Swinyard and Deaver[24] functional grades measured for the two groups were noted preoperatively and at the 1-year follow-up. There was significant improvement in the functional grade for the entire patient population ($p = 0.0314$).The "satisfaction with management" domain of the SRS-30 patient questionnaire was assessed by questions 21, 24and 30. The average score of this domain in the MD and SMA groups was 3.70 and 3.94, respectively, which was not statistically significant. The self-image satisfaction score was rated as 6.2/10 in the patients in the MD group and 6.9/10 in the SMA group (with an identical score of 4for both groups). However, in response to improvement in self-image postoperatively, the average score for the MD group was 3.96 while that for the SMA group was significantly better at 4.64 ($p = 0.01$).

1. Complications

In the MD group, the respiratory complications included2 cases of pneumonia and 1 case of atelectasis. The cardiovascular complications consisted of 1 case of cardiomyopathy, 1 case of pulmonary edema and 1 case of pericardial

effusion. The neurological complications consisted of 1case of seizure. The implant-related complications consisted of 1 case of iliac screw penetration into the pelvis. There were also 2 cases of wound infection. The gastro-intestinal (GI) complications included 1 case of ileus, 2 cases of superior mesenteric artery syndrome, and 1 case of upper GI bleeding. In the SMA group, the respiratory complications included1 case of atelectasis and 1 case of pneumonia. The cardiovascular complications consisted of 1 case of pulmonary congestion. There were no neurological or implant- related complications. There was 1 case of a seroma and 1 case of a wound infection. The GI complications consisted of 1 case of abdominal distention and 1 case of upper GI bleeding. Management of these complications involved removing the prominent hardware in the pelvic cavity and local debridement for the wound-related complications. GI complications were managed either conservatively, or by a gastro-jejunostomy, which was required in 1 case. The total complication rates for both groups are as follows: respiratory system complications (14.3%), cardiovascular complications (8.6%), GI symptoms, and wound infection (8.6%). A total of 16 out of 35 patients (45.7%) developed either surgical or medical complications. There was no difference between the two groups in terms of the complication rate (MD, 45.8%; SMA, 45.5%). There was no case of mortality in the immediate postoperative period. However, 1 case of DMD, who was followed up for 13months after surgery died due to a sudden deterioration in cardiac function at 14 months after surgery.

IV. DISCUSSION

One of the important goals of surgical intervention for flaccid neuromuscular scoliosis is to achieve a proper balance and sitting posture. In this series, although the scoliosis curves were smaller in the MD group, the correction rate in the MD group (41.5%) was lower than that in the SMA group (48.3%), but the difference was not statistically significant($p = 0.13$). Moreover, the correction in pelvic obliquity was significantly better in the SMA group.

Many authors have advocated surgical intervention in DMD cases when the forced vital capacity (FVC) remains >35% of the normal predicted value. However some have reported that deformity correction can be performed safely using NIPPV in patients with a FVC as low as 20-30%[20,25]. Bach and Sabharwal[17] reported that pulmonary complications in patients with muscular disorders or respiratory failure can be reduced using NIPPV. In the present study, 9 patients (6 cases in MD group, 3 cases in SMA group) had a preoperative vital capacity < 30% of the predicted value (range, 14.9 to 29.8%). These patients underwent an aggressive preoperative pulmonary rehabilitation program in conjunction with the respiration rehabilitation team at the department of rehabilitation medicine. Immediate postoperative care focused on reducing the period of mechanical ventilation through endotracheal intubation. In23 out of 35 patients, the total period of ventilator use was less than 2 days. The patients were then subjected to early pulmonary rehabilitation using NIPPV and coughing assist devices. The authors suggested that an intensive respiratory rehabilitation played a key role in maintaining the relatively low incidence of respiratory complications (14.3%) in this series despite the very low FVC preoperative values. Another important goal of surgical intervention in cases of flaccid neuromuscular scoliosis is to reduce the rate of decline in pulmonary function and possibly increase the life expectancy of these patients. Kurz et al.[13] reported that as the scoliosis angle increases by 10°, the vital capacity decreases by 4% in DMD patients. Chng et al.[11] reported a decrease in the rate of FVC declined from 7.7% to 3.8%in SMA patients through a surgical correction of the spinal deformity. Robinson et al.[14] reported that the scoliosis angle of SMA patients was closely related to the pulmonary capacity, and that the PCF was improved by surgical correction. In addition, the surgically-induced lengthening of the thoracic cavity and the improved posture could stabilize the pulmonary capacity or even improve it in the long run[14,15,26]. In the present study, the SMA group showed a greater decrease in the FVC (%) at the end of the first year,

which is likely because the predicted vital capacity was affected by the increased truncal height. The SMA group showed a larger increase in truncal height due to the greater degree of correction of scoliosis and the predicted FVC showed a steeper inclination. The effect of the change in truncal height was not a concern at the end of the second year and both groups showed a similar decrease (2% in the MD group and 3% in the SMA group), which is consistent with other reports[4,16]. Interestingly, unlike the vital capacity, the PCF and $ETPCO_2$ did not show any deterioration. The PCF and $ETPCO_2$ values measured at the end of 1 year showed very little deterioration and were comparable to their preoperative values. In addition, the respiratory questionnaires showed significant improvement in the respiratory grade of patients in both groups despite the decrease in vital capacity. The relatively well-maintained PCF and ET PCO_2 values, which are important determinants of symptomatology, might have induced these results. Therefore, it is believed that the aggressive preoperative and postoperative respiratory rehabilitation played a beneficial role in maintaining the PCF and ET PCO_2.High complications rates, ranging from 24-75%, have been reported in neuromuscular scoliosis patients treated surgically. Mohamad et al.[18] reported that the overall complication rate was approximately 33.1% and that the complications usually occur in the respiratory system (19.4%). In the current study, the post-surgical complications were approximately 45.7%, with a relatively lower respiratory system rate (14.3%).In many studies on flaccid neuromuscular scoliosis, most patients and their guardians claimed that surgical intervention improved their function, appearance, sitting balance and overall quality of life[26]. Aprin et al.[27] reported that surgical intervention resulted in a high satisfaction level (86%) in SMA patients and their guardians. In this study, both groups showed high levels of satisfaction post-operatively with no significant difference between them. The mean score for the improvement in the post-operative self-image in the MD group was 3.96 while that in the SMA group was significantly better at 4.64. It

is believed that this lower average score for self-image in the MD group might be due to the lower correction rates in the scoliosis curve. However, this requires further investigation with a longer follow-up. One of the major limitations of this study was the disparity in the size of the two study groups, which made a precise statistical comparison between the two groups difficult. In addition, the follow-up period was short for some of the patients. However, these patients had rare, life-threatening conditions and such disparities were unavoidable. Moreover, a direct comparison of two different diseases was not possible. Nevertheless, this study may have clinical significance in the terms of the assessment and comparison in the two representative conditions of flaccid neuromuscular scoliosis, namely MD and SMA.

V. CONCLUSION

Surgical intervention resulted in good outcomes in both groups in all aspects. There was no significant difference between the two groups in terms of the pulmonary function, satisfaction or functional grade. Patients with a very low FVC (< 30%) could be operated on safely. Moreover, aggressive preoperative and postoperative rehabilitation efforts can help maintain the PCF and ET PCO_2 values in these patients.

REFERENCES

1. Cervellati S, Bettini N, Moscato M, Gusella A, Dema E, Maresi R. Surgical treatment of spinal deformities in Duchenne muscular dystrophy: a long term follow-up study. Eur Spine J 2004;13:441-8.

2. Shapiro F, Bresnan MJ. Orthopaedic management of childhood neuromuscular disease. Part 1: Spinal muscular atrophy. J Bone Joint Surg Am 1982;64:785-9.

3. Galasko CS, Williamson JB, Delaney CM. Lung function in Duchenne muscular dystrophy. Eur Spine J 1995;4:263-7.

4. Kennedy JD, Staples AJ, Brook PD, et al. Effect of spinal surgery on lung function in Duchenne muscular dystrophy. Thorax 1995;50:1173-8.

5. Ramirez N, Richards BS, Warren PD, Williams GR. Complications after posterior spinal fusion in Duchenne's muscular dystrophy. J Pediatr Orthop 1997;17:109-14.

6. Galasko CS, Delaney C, Morris P. Spinal stabilization in Duchenne muscular dystrophy. J Bone Joint Surg Br1992;74:210-4.

7. Karol LA. Scoliosis in patients with Duchenne muscular dystrophy. J Bone Joint Surg Am 2007;89 Suppl 1:155-62

8. Larsson EL, Aaro S, Normelli H, Oberg B. Weight distribution in the sitting position in patients with paralytic scoliosis: pre- and postoperative evaluation. Eur Spine J2002;11:94-9.

9. Smith RM, Emans JB. Sitting balance in spinal deformity. Spine (Phila Pa 1976) 1992;17:1103-9.

10. Berven S, Bradford DS. Neuromuscular scoliosis: causes of deformity and principles for evaluation and management. Semin Neurol 2002;22:167-78.

11. Chng SY, Wong YQ, Hui JH, Wong HK, Ong HT, GohDY. Pulmonary function and scoliosis in children with spinal muscular atrophy types II and III. J Paediatr Child Health 2003;39:673-6.

12. Mercado E, Alman B, Wright JG. Does spinal fusion influence quality of life in neuromuscular scoliosis? Spine (Phila Pa 1976) 2007;32(19 Suppl):S120-5.

13. Kurz LT, Mubarak SJ, Schultz P, Park SM, Leach J. Correlation of scoliosis and pulmonary function in Duchenne muscular dystrophy. J Pediatr Orthop 1983;3:347-53.

14. Robinson D, Galasko CS, Delaney C, Williamson JB, Barrie JL. Scoliosis and lung function in spinal muscular atrophy. Eur Spine J 1995;4:268-73.

15. Sucato DJ. Spine deformity in spinal muscular atrophy. J Bone Joint Surg Am 2007;89Suppl 1:148-54.

16. Velasco MV, Colin AA, Zurakowski D, Darras BT, Shapiro F. Posterior spinal fusion for scoliosis in Duchenne muscular dystrophy diminishes the rate

of respiratory decline. Spine (Phila Pa 1976) 2007;32:459-65.

17. Bach JR, Sabharwal S. High pulmonary risk scoliosis surgery: role of noninvasive ventilation and related techniques. J Spinal Disord Tech 2005;18:527-30.

18. Mohamad F, Parent S, Pawelek J, et al. Perioperative complications after surgical correction in neuromuscular scoliosis. J Pediatr Orthop 2007;27:392-7.

19. 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;26:216-20.

20. Gill I, Eagle M, Mehta JS, Gibson MJ, Bushby K, Bullock R. Correction of neuromuscular scoliosis in patients with preexisting respiratory failure. Spine (Phila Pa 1976)2006;31:2478-83.

21. Gupta MC, Wijesekera S, Sossan A, et al. Reliability of radiographic parameters in neuromuscular scoliosis. Spine (Phila Pa 1976) 2007;32:691-5.

22. Kim HS, Moon ES, Park JO, et al. A change of pulmonary function in the patients with severe non-idiopathic scoliosis following reconstructive spine surgery. J Korean Soc Spine Surg 2006;13:106-13.

23. Jones PW, Quirk FH, Baveystock CM. The St George's respiratory questionnaire. Respir Med 1991;85Suppl B:25-31.

24. Swinyard CA, Deaver GG, Greenspan L. Gradients of functional ability of importance in rehabilitation of patients with progressive muscular dystrophy and neuromuscular diseases. Arch Phys Med Rehabil 1957;38:574-9.

25. Marsh A, Edge G, Lehovsky J. Spinal fusion in patients with Duchenne's muscular dystrophy and a low forced vital capacity. Eur Spine J 2003;12:507-12.

26. Bridwell KH, Baldus C, Iffrig TM, Lenke LG, Blanke K. Process measures and patient/parent evaluation of surgical management of spinal deformities in patients with progressive flaccid neuromuscular scoliosis (Duchenne's muscular dystrophy and spinal muscular atrophy). Spine (Phila Pa1976) 1999;24:1300-9.

27. Aprin H, Bowen JR, Mac Ewen GD, Hall JE. Spine fusion in patients with

spinal muscular atrophy. J Bone Joint SurgAm 1982;64:1179-87.

APPENDICES

MD: muscular dystrophy

SMA: spinal muscular atrophy

DMD: Duchenne muscular dystrophy

ICU: intensive care unit

NIPPV: non-invasive positive pressure ventilation

PCF: peak cough flow

ET PCO_2: the end tidal Pressure of CO_2

Preop: preoperative

Postop: postoperative

Last: in last follow up,

T-K: thoracic kyphosis

L-L: lumbar lordosis

PCF: peak cough flow

Pod 1yr: at 1 year postoperative

%a): the percentage of the predicted value of vital capacity considering the patients' height

GI: gastrointestinal

FVC: forced vital capacity

ABSTRACT (IN KOREAN)

근이영양증과 척수성 근위축증으로 인한 척추 측만증 환자의 수술 후 방사선학적, 호흡기계 및 기능적 결과의 비교

<지도교수 김학선>

연세대학교 대학원 의학과

정 현 수

연구 계획: 후향적 연구

연구 목적: 근이영증 환자와 척수성 근위축증 환자에서 발생한 척추 측만증의 수술적 결과를 연구하여 비교하여 보고자 한다.

연구 배경: 기존의 연구 중 근이영양증 환자와 척수성 근위축증 환자에서 발생한 척추 측만증에 대해 수술적 치료 결과를 비교한 것은 거의 없다.

대상 및 방법: 근이영양증으로 인해 척추 측만증이 발생하여 수술한 환자는 24명이었으며 척수성 근위축증으로 인한 환자는 11명이었고, 이들 환자들의 평균 추적 기간은 21개월 이었다. 모든 환자는 신경근육병성 척추 측만증에 대해서 후방 교정술 및 유합술을 받았으며, 수술 직후 기간 동안 호흡기계 합병의 발생을 막기 위해서 비침습적 양압 인공호흡기 (noninvasive positive pressure ventilation, NIPPV) 및 기침 보조기구 사용하였다. 모든 환자는

방사선 사진상 콥스각, 골반 경사각, 흉추 번의 전위, 흉추 후만 및 요추 전만 각도를 측정 하였으며, 폐 기능 검사 및 자아상에 대한 만족도 또한 조사하였다.

결과: 수술 전 측만각도는 근이영양증 환자군에서 좀더 작게 측정되었으나 수술 후 교정 정도는 척수성 근위축증 환자군에서 좀더 높게 측정 되었고, 골반 경사각 또한 척수성 근위축증 환자군에서 통계학적 의미 있게 높은 교정을 얻을 수 있었다. 수술 후 1년째 추시 한 폐기능 검사상 폐활량의 기대치는 근이영양증 환자군에서 4%의 감소를 보였고 척수성 근위축증 환자군에서 10%의 감소를 보였으나 최고 기침유량과 최종 배출 이산화탄소 농도 (End tidal PCO_2)는 잘 유지 되는 것을 확인하였다. 수술 후 자아상에 대한 만족도는 근이영양증과 척수성 근위축증 환자군에서 각각 3.96과 4.64로 측정되었으며, 전체적인 합병증의 발생은 45.7%이었으며 이중 14.3%가 호흡기계와 관련된 합병증이었다.

결론: 근이영양증과 척수성 근위축증 환자 중 폐활량이 30%이하로 심각하게 감소된 환자라도 수술 전후에 적극적인 호흡 재할을 시행한다면 수술적 치료를 비교적 안전하게 시행할 수 있다.

핵심되는 말: 근이영양증, 척수성 근위축증, 신경근육병성 척추측만증, 수술적 교정, 폐기능

PUBLICATION LIST

내용

1. Chong HS, Moon ES, Kim HS, Ankur N, Park JO,Kim JS, et al. Comparison between operated muscular dystrophy and spinal muscular atrophy patients in terms of radiological, pulmonary and functional outcomes. Asian Spine J 2010;4(2):82-8.