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ORIGINAL ARTICLE

Severe restrictive lung disease and vertebral surgery in a pediatric population

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Abstract The aim of this study is to describe the outcome of surgical treatment for pediatric patients with forced vital capacity (FVC) <40% and severe vertebral deformity. Few studies have examined surgical treatment in these patients, who are considered to be at a high risk because of their pulmonary disease, and in whom preoperative tracheostomy is sometimes recommended. Inclusion criteria include FVC <40%, age <19 years and diagnosis of scoliosis. The retrospective study of 24 patients with severe restrictive lung disease, who underwent spinal surgery. Variables studied were age and gender, pre- and postoperative spirometry (FVC, FEV1, FEV1/FVC), preoperative, postoperative and late use of non-invasive ventilation (BiPAP) or mechanical ventilation, associated multidisciplinary treatment, type and location of the curve, pre- and postoperative curve values, type of vertebral fusion, intra- and postoperative complications, duration of intensive care unit (ICU) stay and length of postoperative hospitalization. Mean age was 13 years (9–19) of which 13 were males and 11 females. Mean follow-up was 32 months (24–45). The etiology was neuromuscular in 17 patients and other etiologies in 7 patients. Mean preoperative FVC was 26% (13–39%). Eight patients had preoperative home BiPAP, 15 preoperative in-hospital BiPAP, and 2 preoperative mechanical ventilation. Nine patients had preoperative nutritional support. Preoperative curve value of the deformity was 88° (40°–129°). Nineteen patients with posterior fusion alone and 5 with anterior and posterior fusion were found. Mean duration of ICU stay was 5 days (1–21). Total postoperative hospital stay was 17 days (7–33). Ventilatory support in the immediate postoperative includes 16 patients requiring BiPAP and 2 volumetric ventilation. None of the patients required a tracheostomy. The intraoperative complications include one death due to acute heart failure; immediate postoperative, four respiratory failures (2 required ICU readmission) and one respiratory infection; and other minor complications occurred in six patients. Overall, 58% of patients had complications. Percentage of angle correction was 56%. After a follow-up of 30 months, FVC was 29% (13–50%). In conclusion, corrective scoliosis surgery in pediatric patients with severe restrictive lung disease is well tolerated, but the management of this population requires extensive experience with the vertebral surgery involved, and a multidisciplinary approach that includes pulmonologists, nutritionists and anesthesiologists. Currently, there is no indication for routine preoperative tracheostomy.

Keywords Neuromuscular scoliosis · Restrictive lung disease · Surgical treatment

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Introduction

The purpose of surgical correction of spinal deformity in pediatric patients with severe restrictive lung disease is to improve pulmonary function and quality of life, and increase the patients' life expectancy. The deterioration of pulmonary function in these patients is the result of the underlying disease. With progression of the vertebral deformity, the rib cage also becomes deformed and this leads to hypoxemia, hypercapnia, cor pulmonale, and pulmonary hypertension; hence, vertebral fusion is one of the keys to treating this condition [7, 9, 10].

The most frequent pulmonary alteration in these young patients is restrictive pulmonary disease, which is characterized by a decreased forced vital capacity (FVC), decreased forced expiratory volume in one second (FEV_1), and normal FEV_1/FVC ratio. Because of their respiratory impairment, patients have a high risk of postoperative complications, particularly when they undergo reconstructive vertebral surgery [13]. An FVC value less than 40% of the theoretical value is a sign of severe respiratory compromise [9].

Following surgery for spinal deformity, pulmonary function initially deteriorates and later recovers preoperative values; subsequently, deterioration continues with progression of the underlying disease [14]. Patients with severe restrictive lung disease associated with spinal deformity are a challenge for anesthesiologists and nutritional specialists, both in the preoperative preparation as well as the intraoperative and postoperative care. The related literature contains few reports on the management of these patients.

This study investigates the outcome of surgical treatment for pediatric patients with FVC less than 40% of the theoretical value and severe vertebral deformity, with particular emphasis on a multidisciplinary approach.

Materials and methods

A retrospective review was performed of 24 patients (13 males and 11 females) with severe restrictive lung disease requiring reconstructive vertebral surgery. The inclusion criteria were FVC less than 40% of the theoretical value, age less than 19 years, and a diagnosis of scoliosis. All the procedures were performed by the same group of surgeons.

The diagnoses included neuromuscular disease in 17 patients (10 myopathy, 6 spinal cord atrophy, and 1 postoperative paraplegia), and other etiologies in 7 patients (congenital scoliosis, Turner-Noonan syndrome, and idiopathic scoliosis). Some patients additionally presented severe concomitant conditions: previous pneumonectomy due to pulmonary atelectasis, pulmonary stenosis, pulmonary hypertension, a giant omphalocele, and psychomotor retardation.

In the preoperative workup, all patients underwent meticulous assessment of their cardiorespiratory, nutritional, and orthopedic status. One patient required a prior gastrostomy for the nutritional supply and two others underwent orthopedic surgery of the lower limbs.

Evaluation of respiratory function included spirometry with the following variables: FVC expressed in liters and in percentage relative to the theoretical value, FEV_1 expressed in liters and percentage with respect to the theoretical value, and the FEV_1/FVC ratio, expressed in percentage.

Ten patients were receiving home ventilatory support before the intervention. Eight of them had a bilevel positive airway pressure (BiPAP) device and two had a volumetric ventilator (1 of them with a previous tracheostomy). In the preoperative preparation, BiPAP was used in 15 patients and volumetric ventilation in 2 patients.

None of the patients needed a tracheostomy prior to surgery. Nine had preoperative nutritional support, and various skeletal traction systems were used preoperatively in six patients as a coadjuvant to their respiratory preparation.

The following postoperative variables were recorded: days of intensive care unit (ICU) stay, days hospitalized, development of pulmonary complications (e.g., pneumothorax, atelectasis, pneumonia, respiratory failure), other surgery-related complications (e.g., neurological complications, wound infections), and mortality.

Study of the vertebral deformity consisted of radiographs in AP and lateral views, and while bending forward and in traction, to study the flexibility of the curve. Previous procedures to control the spinal deformity (growing rods) had been performed in two patients.

Posterior fusion was carried out in 19 patients, and anterior plus posterior fusion in 5 patients, 2 in one surgical stage and 3 in two stages, with a mean interval between the two procedures of 10, 12 and 9 days. The anterior approach consisted of a thoracotomy or thoracolumbotomy, depending on the vertebral levels to be fused. The mean number of fused levels was 5.7 (range 4–8). In the posterior approach, a mean of 15 vertebrae (range 12–16) were fused and the instrumentation was hybrid. Local autologous bone and bone from the tissue bank were used in all patients.

Following the procedure, postoperative immobilization was decided for patients who did not collaborate sufficiently and those who would be more comfortable with this measure.

Results

A total of 24 patients met the criteria for participation in the study: 13 males and 11 females, with a mean age at the time of surgery of 13 years (range 9–19 years). Mean follow-up was 32 months (24–45 months). All patients had undergone pulmonary testing to confirm the presence of severe restrictive pulmonary disease.

The mean preoperative angle of the main scoliotic curve was 88° (range 40°–129°). Following a mean radiographic follow-up of 25 months, a reduction from 88° to 37° was achieved (range 2°–80°), which represents a 56% decrease (range 5–90%).

Mean preoperative FVC was 0.76 l (range 0.28–1.9 l), that is, 26% of the theoretical value (range 13–39%),

preoperative FEV_1 was 0.71 l (range 0.28–1.9 l), 30% of the theoretical (range 16–41%), and the FEV_1/FVC ratio was 93% (range 62–100%).

The mean duration of ICU stay was 5 days (range 1–21) and postoperative duration of hospitalization was 17 days (range 7–33 days). During the immediate postoperative period, 16 patients required BiPAP and 2 volumetric ventilation.

The single intraoperative complication was the death due to acute heart failure of a patient with Duchenne disease and severe cardiomyopathy. Immediate postoperative complications occurred in 58% of patients. These included general complications: four cases of respiratory failure (2 requiring readmittance to the ICU for 3 days and 4 days, respectively), one respiratory infection, one recurrent atelectasis and one prolonged paralytic ileum; and surgery-related complications: three cases of surgical wound complications (2 bloody exudates and 1 dehiscence) and two cases of radicular pain that resolved with rehabilitation. One of the patients died 5 years after surgery as a consequence of the underlying condition (Duchenne disease).

After a mean follow-up of 30 months (range 20–33 months), spirometry values were: FVC 0.98 l (range 0.37–2.5 l), 29% of theoretical (range 13–50), FEV_1 0.81 l (range 0.37–1.58 l), 33% of theoretical (range 16–47%), and the FEV_1/FVC ratio was 95% (range 82–100%). In 3 patients, it was possible to withdraw prior ventilatory support following the procedure, 7 other patients who did not need prior ventilation continued without this treatment, and 12 patients who needed prior support continued with home ventilation.

Discussion

The literature has not established the limits for an indication of reconstructive vertebral surgery in patients with severe restrictive disease. Studies in pediatric patients with neuromuscular scoliosis have shown an inverse relationship between the vital capacity and the rate of postoperative complications [7]. Anderson et al. [1] reported a series of risk factors for the development of postoperative pulmonary complications in patients undergoing scoliosis surgery: scoliosis with a non-idiopathic etiology, obstructive lung disease, arterial hypoxemia, mental retardation, age less than 20 years, and anterior fusion. In our series, two of the criteria for inclusion were age under 19 and FVC less than 40% of the theoretical value. There was only one patient with psychomotor retardation, although all except one had scoliosis with a non-idiopathic etiology. These factors may have contributed to the development of postoperative lung complications in 25% of the series.

Spinal bracing using rigid jackets is an important stage in the management of scoliosis, particulary in children with muscle weakness. In these cases, the brace has most often considered a means of delaying the deformity progress and thus postponing surgical spinal fusion or as sitting support. Spinal bracing on respiratory function in neuromuscular disease results in a considerable reduction in FVC [2, 11]. Olafsson et al. [6] reviewed 90 patients with neuromuscular diseases and progressive spine deformity, treated with soft Boston orthosis. The mean pretreatment Cobb angle was 47°, with a mean brace-induced Cobb angle correction of 60%; after a follow-up of 3.1 years, the treatment was considered successful (<10° curve progression) in 23 patients. They concluded that brace treatment in neuromuscular scoliosis, except as a sitting support, can be questioned.

According to Wazeca et al. [13], one of the main risk factors for the development of postoperative lung complications is treatment with anterior fusion, and the risk increases if the procedure includes a thoracotomy. Five patients in our series were treated with a double approach, and respiratory complications occurred in the two patients who underwent the operation in a single stage. Among the 19 patients who were treated with posterior surgery alone, four (21%) experienced postoperative respiratory complications.

Pulmonary function is severely affected following scoliosis surgery [14]. In the immediate postoperative period, the functional respiratory test values decrease by up to 60% of the preoperative values, at 1 week, respiratory function is at 50%, and at 1 or 2 months, functional results return to the normal preoperative values, with no significant differences in the magnitude of the postoperative decrease according to the etiology of scoliosis (among 24 cases, 46% were idiopathic and 38% neuromuscular scoliosis), or the approach used in the surgical procedure. In a study including 298 scoliosis patients, Zhan et al. [15] reported the following rates of postoperative pulmonary complications as related to the preoperative FVC values: FVC 60–80% of theoretical, complications 2.72%; FVC 40–60%, complications 7.4%; and FVC less than 40%, complications 31.6%. In addition, the authors found statistically significant differences in relation to the surgical approach (complications were 18-fold more frequent in patients undergoing a transthoracic approach than in those treated by posterior fusion, $p < 0.05$). The potential impact of the etiology of scoliosis was not investigated.

In all the patients included in the present study, FVC was less than 40% of the theoretical value and in 71% the etiology was neuromuscular; the rate of postoperative pulmonary complications was 25%. In our opinion, the etiology of scoliosis is a risk factor for the development of postoperative pulmonary complications and for inability to

recover lung function. Whereas in idiopathic scoliosis the deformity itself is the cause of functional deterioration, in patients with neuromuscular disease the underlying disease persists even after the deformity is corrected. As to the status of lung function at long-term, some studies have provided evidence of improved pulmonary function at 1–2 years following surgery [5, 8], whereas in others this improvement was not observed [4, 10, 12]. In the present study, FVC was 26% (range 13–39%) of the theoretical value preoperatively. In Fig. 1, we can see the improving of the pulmonary function after the surgery: at 16 months (range 12–25 months) FVC was 27.4% (range 21–40.7%) of the theoretical value and 33% (range 16–47%) at a mean follow-up of 30 months (range 20–33 months); ventilatory support was discontinued in 3 patients, 7 continued without support, and 12 continued with ventilation. Figure 2 shows

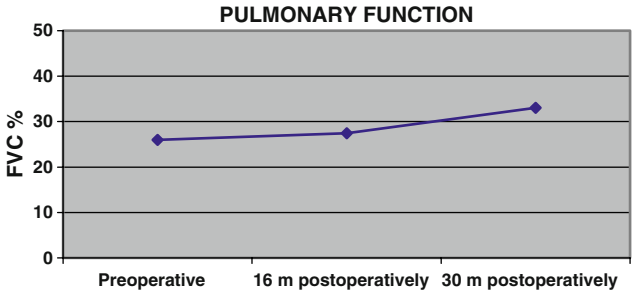

Fig. 1 Improving of the pulmonary function after the surgery (*m* months)

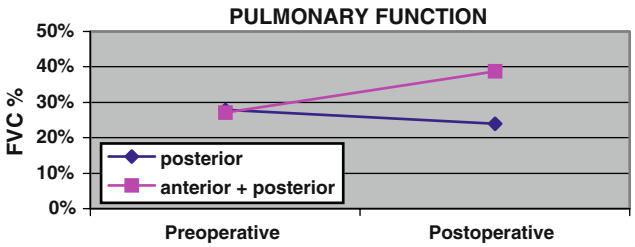

Fig. 2 Preoperative and postoperative pulmonary function (FVC) in posterior fusion and anterior and posterior fusion

Table 1 Summarization of results of several series

Variables	Wazeka et al. [13]	Rawlins et al. [9]	Gill et al. [3]	Our group
N	21	32	8	24
Follow-up (months)	47.3	58	48	32
Age (years)	11.5	13	12	13
Double approach	18	19		5
Posterior approach alone	3	13		19
Anterior levels fused	6			6
Posterior levels fused	11	13		15
Preoperative Cobb angle	82°	87°	70°	88°
Postoperative Cobb angle	35°		32°	37°
Preoperative spirometry	FVC 30.5%	VC 31%	FVC 20%	FVC 26%
Postoperative spirometry			VC 18%	FVC 29%
ICU (days)		6	3	5
Duration postoperative hospitalization (days)	18	27	14	17
Complications	0 Deaths	3 Postoperative tracheostomy	0 Deep infection	1 Death
	0 Neurological complication	2 Pneumonia	0 Pulmonary complication	2 Radicular pain
	1 Implant failure	2 Reintubation	0 Tracheostomies	2 Bloody exudates
	2 Deep infection	1 Pseudoarthrosis		1 Dehiscence
	2 Pancreatitis	1 Preoperative tracheostomy		4 respiratory Insufficiency
	1 Postoperative tracheostomy			1 Atelectasis
	2 Pneumonia			1 Pneumonia
	1 Preoperative tracheostomy			1 Prolonged paralytic ileum
	2 Pneumothorax			
	4 Atelectasis			

the evolution of the pulmonary function of two subgroups of patients: posterior fusion and anterior and posterior fusion. It shows that in anterior and posterior group, the postoperatory FVC is higher than the preoperative results; we think that this result can be explained because patients in double approach were not neuromuscular patients, so when you get the fusion, they suffer an improve in their pulmonary function.

There were only three articles in the literature whose results could be compared with those of the present study, although the criteria for inclusion differed. Wazeka et al. [13] performed a retrospective study in patients with FVC less than 45%, age under 21, and severe thoracic scoliosis (21 cases). Rawlins et al. [9] reviewed patients with restrictive lung disease and FVC less than 40%, age under 18 and vertebral deformity (32 patients). Gill et al. [3] prospectively studied patients with progressive scoliosis, myopathy and respiratory failure (8 patients). The findings from our group of patients and the results from these studies are summarized in Table 1.

What we wish to emphasize from these data is the low rate of major postoperative complications in our group of patients. Table 2 shows the complications of posterior fusion and anterior and posterior fusion. We can see that 9 of 19 of posterior fusion has no complications, but 1 of 5 in the anterior and posterior fusion. In this last group, two major respiratory complications happened. We explain it because in the anterior approach (thotacotomy and thoracolumbotomy) you need a selective intubation in one lung, and the recovery of the pulmonary function is worst.

We believe that this low rate of major complications is because multidisciplinary treatment was provided preoperatively and postoperatively, and few patients underwent corrective surgery involving a double approach. Many of our patients were admitted to the hospital some days before surgery to undergo extensive nutritional and respiratory preparation, together with methods for orthopedic skeletal traction, which make management of the deformity at the time of surgery less complex.

Table 2 Complications in posterior fusion and in anterior and posterior fusion

	Posterior fusion ($n = 19$)	Anterior + posterior fusion ($n = 5$)
Minor complications	2 Bloody exudates	2 Radicular pain
	1 Prolonged paralytic ileum	
	1 Dehiscence	
Major complications	3 Respiratory insufficiency	1 Respiratory insufficiency
	1 Pneumonia	1 Atelectasis
	1 Intraoperatory death	
No complications	9	1

Conclusions

Major reconstructive vertebral surgery is well tolerated in patients with restrictive lung disease. Often these patients are excluded as candidates for a corrective intervention because of their deteriorated pulmonary function. Correct preoperative assessment is important and should be carried out by several specialists working in the pediatric age group: anesthesiologists, pulmonologists, cardiologists, and professionals in nutrition. Extensive familiarity with spinal surgery is needed to decrease the operative time and bleeding, and to avert complications, and postoperative follow-up should be supported by the specialists who participated in the preoperative assessment. At present, there are no indications for tracheostomy, but several preoperative interventions are known to improve the patients' nutritional and respiratory status, and make the treatment for vertebral deformity less complex at the time of surgery.

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