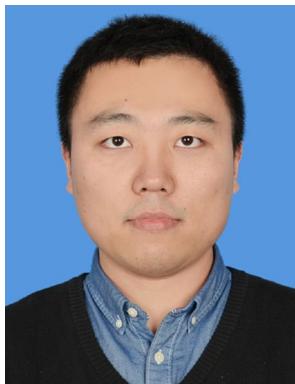


# Successful surgery for a neuromuscular scoliosis patient by pulmonary rehabilitation with forced vital capacity below 30%

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## Abstract



A rare case of a 15-year-old male patient with neuromuscular scoliosis with forced vital capacity (FVC) below 30%, who went through a successful surgery without any pulmonary complications, is reported herein. The patient had obvious asymmetric shoulders and poor exercise tolerance. The Cobb's angle of the main thoracic curve was 62.8°, and FVC in sitting position was 18% of predictive value. After skull traction and pulmonary rehabilitation, the FVC was still below 30%, and he finally went through surgery under this serious condition. By early pulmonary rehabilitation using home ventilator, he successfully recovered without any pulmonary complications. The patient had complete symptom remission and no deterioration of Cobb's angle was found during follow-up.

**Keywords** Neuromuscular scoliosis · Pulmonary rehabilitation · Forced vital capacity · Home ventilator

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## Introduction

Neuromuscular scoliosis (NMS), mainly caused by Duchenne muscular dystrophy (DMD) or spinal muscular atrophy (SMD), often causes respiratory failure due to weaknesses of the respiratory muscles. Thus, NMS can pose serious risks to anesthesia and is considered to be a relative contraindication to surgery [1]. In the past decades, scoliosis correction could be operated only in patients with forced vital capacity (FVC) above 30%; otherwise, the increased pulmonary complications could be fatal [1]. However, preoperative pulmonary rehabilitative support, such as home ventilator, can remarkably reduce surgical risks and postoperative respiratory complications [2]. In the past 10 years, five studies

of operations were reported in patients with neuromuscular disease with low FVC below 30% of predictive value according to our knowledge [3–7]. However, there are only isolated reports of successful operation without any postoperative respiratory complications in the previous studies [7].

In this study, we report on a patient with SMD that develops respiratory failure early in the course of his disease. This complicates the management of the spinal deformity. We present the result of scoliosis surgery in this patient with respiratory failure and on nocturnal ventilator support. There are very few reports of successful scoliosis correction with no respiratory complications achieved by active and comprehensive pulmonary rehabilitation in an SMD patient with FVC below 30% of predictive value.

## Case history

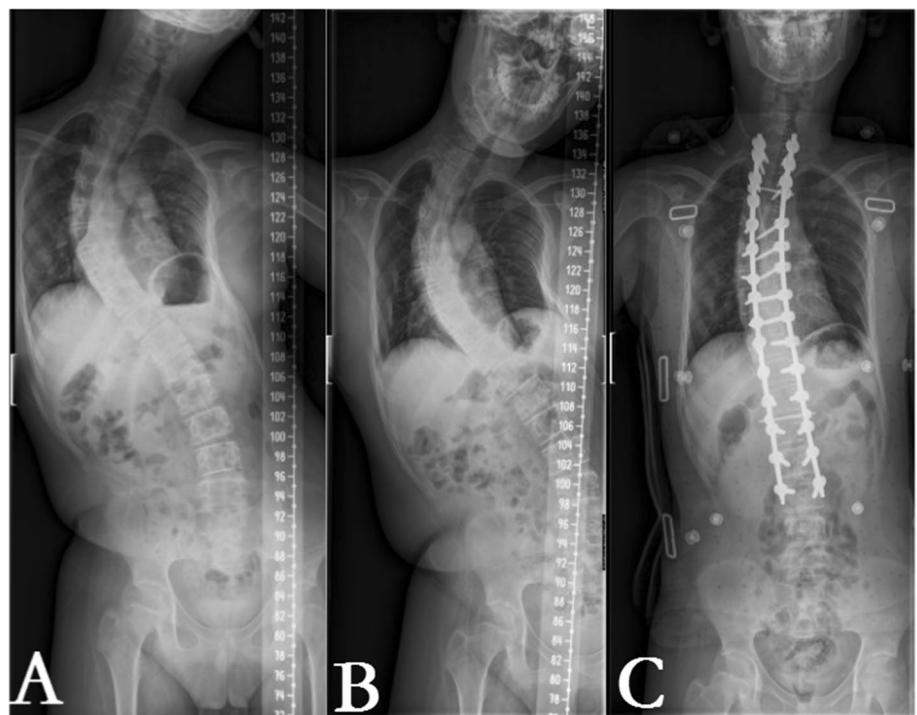
Herein, a 15-year-old male patient, who was found to have obvious asymmetric shoulders and poor exercise tolerance since 2 years ago, was taken to the First Affiliated Hospital, Sun Yat-Sen University by his father. He was very thin and weak, and the body weight was only 43 kg. He had no symptoms of dyspnea during daily life. However, when having jogging in the morning in school, he would suffer severe dyspnea and could not continue for even 1 min. This phenomenon was becoming more and more obvious during the last 2 years. The parents wanted to relieve the child's pain in appearance and improve quality of life, whatever the cost may be. The patient himself was also willing to take his

chance. Preoperative examination showed that Cobb's angle of the main thoracic curve was  $62.8^\circ$  (Fig. 1a). In the evaluation of pulmonary function, FVC in sitting position was 18% of predictive value, and pulmonary capacity was 1065.49 ml, which was significantly decreased than the normal value.

Brace of skull traction was used for 3 days before an acute coma occurred (Fig. 2), and the blood O<sub>2</sub> saturation (SaO<sub>2</sub>) dropped to 60%. He was transmitted to the intensive care unit (ICU) immediately. In the arterial blood gas analysis, partial pressure of carbon dioxide (PaCO<sub>2</sub>) was 48 mmHg and partial pressure of oxygen (PaO<sub>2</sub>) was 241 mmHg when the fraction of inspired O<sub>2</sub> (FiO<sub>2</sub>) was 40%. After 3 days' respirator-assisted ventilation in intensive care unit (ICU), the arterial blood analysis showed CO<sub>2</sub> retention, while the patient did not have any discomfort. Then, he was transferred to the normal ward. He received training for non-invasive intermittent positive pressure ventilation (NIPPV), which mode is assist-controlled ventilation (ACV). He was prescribed a home ventilator, and the ventilator settings were the mode of ACV, respiration rate of 18–20 times per minute, and support level of pressure during inspiration and expiration was 10 and 4 cmH<sub>2</sub>O, respectively. Based on the daytime monitoring of PaCO<sub>2</sub>, after about 4-h discontinuance of home ventilator, hypercapnia had redeveloped; he was, therefore, educated to apply the home ventilator while sleeping and additionally in the daytime. After application of home ventilator, the symptoms of hypercapnia during sleeping were resolved.

After being discharged, the patient had no complaints related to respiratory failure, but he suffered from the

**Fig. 1** Preoperative (**a, b**) and postoperative (**c**) whole spine radiograph show improved scoliosis. **a** Cobb's angle for the first time in hospital was  $62.8^\circ$ , **b** Cobb's angle increased to  $72.7^\circ$  after pulmonary rehabilitation at home, and **c** the postoperative angle was  $31.1^\circ$



**Fig. 2** Radiograph after skull traction for the first time in hospital



appearance, due to severe scoliosis: Cobb's angle of the main thoracic curve increased to  $72.7^\circ$  (Fig. 1b). After 4 months' Halo-gravity traction and pulmonary rehabilitation, he was admitted to the Department of Spine Surgery to receive surgery. The traction weight was as high as 20 kg for about 10 weeks. In the evaluation of pulmonary function, FVC in sitting position was 23% of predictive value, and pulmonary capacity was 1043.16 ml, which was significantly decreased than the normal value. He was considered high risk for scoliosis operation due to a very poor pulmonary function. After a serious case discussion in our department, we finally decided to give surgery for posterior fixation from the first thoracic vertebra to the third lumbar vertebra on December 14, 2015. During the operations, his vital signs were stable. From the time of admission, noninvasive PaCO<sub>2</sub> monitoring was conducted during sleep. The blood loss was 800 ml, and he had self-blood transfusion for about 400 ml. When surgery ended successfully, he was transferred to ICU, and NIPPV via home ventilator was applied right after intubation tube removal. Upon confirmation that there were no complications and hypercapnia, he was transferred to the general ward.

At 9 days after the surgery, intravenous antibiotics were injected due to hematuresis and odynuria. On January 4, 2016, 26 days after the operation, all stitches were removed and the wound healed very well. Cobb's angle was changed to  $31.1^\circ$  postoperatively (Fig. 1c). The shoulders were almost symmetric when standing. Then, he was discharged and continued to pulmonary rehabilitation using home ventilator. At postoperative 6 months, pulmonary function was reevaluated: the FVC was 19% of predicted value and pulmonary capacity was 1414.67 ml. He could have a normal daily life without home ventilator both in daytime and during sleep.

## Discussion

A high incidence of postoperative complications has been noticed in most studies that focused on patients with NMS surgery [8, 9]. The mean incidence of complications, including deep surgical-site infections, cardiopulmonary complications, and intraoperative neurologic complications, was 33% [9]. This has led patients with NMS to be high risk for surgery. They require a great deal of preoperative planning and preparation to prevent any complications.

Pulmonary complications, such as pneumonia, pneumothorax, and prolongation of trachea cannula, are most frequently occurring in patients with NMS [10]. According to Rawlins et al. [1], 19% of NMS patients and FVC below 40% may suffer respiratory complications after surgery. And in the past, scoliosis operation was contraindicated in patients with FVC below 30% of predictive value [11]. However, in studies in the past 10 years, there was no significant difference in surgical risk even if the FVC was below 30% of predictive value [3–7] (Table 1).

If respiratory muscles are weakening in patients with NMS, the tidal volume would decrease and the respiratory rate would increase subsequently. Accordingly, fatigue of the respiratory muscles should occur and induce retention of carbon dioxide. The patient in this case had a very low FVC below 30% of predictive value. Because of hypercapnia, he needed to be assisted by NIPPV, and normal ventilatory states were maintained during sleep. After the application of NIPPV for more than 4 months, it was possible to change from intubation to NIPPV immediately after a major operation without any difficulty.

In our patient, despite a very low FVC below 30% of normal predictive value, surgical scoliosis correction could

**Table 1** Studies involving NMS patients with preoperative FVC < 30%

Author	Number of patients	Preoperative pulmonary rehabilitation	Postoperative complications	Journal	Year
Gill [4]	8	Yes	Yes	<i>Spine</i>	2006
Takaso [3]	14	Yes	Yes	<i>Int Orthop</i>	2010
Modi [5]	18	No	Yes	<i>J Spinal Disord Tech</i>	2011
Chong [6]	18	Yes	Yes	<i>Spine</i>	2011
Lee [7]	1	Yes	No	<i>Ann Rehabil Med</i>	2013

be performed successfully without operation-related complications in an SMD patient with pulmonary rehabilitative support (NIPPV) being provided using a home ventilator.

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### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no competing interests.

**Informed consent** Informed consent was obtained from the patient mentioned in this article.

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