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# Wenlin procedure for asphyxiating thoracic dystrophy with severe pulmonary hypertension

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### Abstract

Surgery is the only effective method for the treatment of asphyxiating thoracic dystrophy, but it was considered that severe pulmonary hypertension was the contraindication. We report a 13-year-old male patient of asphyxiating thoracic dysplasia with severe pulmonary hypertension. We performed Wenlin procedure for him and achieved satisfactory results.

**Keywords:** Asphyxiating thoracic dystrophy, pulmonary hypertension, surgery

### Introduction

Asphyxiating thoracic dystrophy (ATD) is an extremely rare and dangerous autosomal recessive disease [1-4]. Due to the narrow and small thorax, most patients have serious respiratory dysfunction, and surgery is the only effective means to treat the disease [2-4]. However, severe pulmonary hypertension had been considered as a contraindication for surgery [5, 6]. We performed Wenlin procedure [2, 7] for a patient of ATD with severe pulmonary hypertension, and achieved satisfactory results.

### Case Report

The patient was a 13-year-old male. Immediately after birth, he developed severe hypoxia and cyanosis and had to be ventilated. After 15 days of treatment, his symptoms completely disappeared. Since then, he often had respiratory discomfort, which aggravated when crying and eating. After the age of 5, his condition gradually stabilized, but he couldn't exercise violently. One year before admission, his condition worsened and needed continuous oxygen inhalation. He finally admitted our hospital for surgical treatment.

Physical examination showed that his height was 141cm, and the chest circumference of nipple plane was 61cm. His thorax was narrow and small, and lateral chest walls were concave (Fig. 1A). Imaging examination showed obvious thoracic deformity, and both lungs were obviously compressed (Fig. 1B). The pulmonary artery systolic pressure was 79 mmHg. In order to reduce pulmonary artery pressure (PAP), Viagra was used briefly before operation, but the effect was not ideal. In order to eliminate symptoms quickly, the operation was carried out as scheduled.

Wenlin procedure was performed on both sides of the chest wall simultaneously [2, 7]. A longitudinal incision was completed at the axillary midline to expose the ribs and costal cartilages. Two tunnels were made in front of the sternum, and two arc-shaped steel bars were put into them respectively. The two sides of the steel bar were located directly in front of the depression of the lateral chest wall. The depression is lifted by steel wires and fixed on the steel bars (Fig. 1C and D). For some residual small depressions, the rib fixing bars were used for shaping. The operation was smooth without complications.

The patient underwent intubation ventilation for five days postoperatively, and weaned when his condition was stable. Then, oxygen was given by nasal catheter. The oxygen saturation was maintained above 92%, and the symptoms were significantly improved, but the pulmonary hypertension was not eliminated even Bosentan and Viagra were taken every day. His PAP was always above 75mmHg. Fortunately, this did not affect the improvement of the patient's condition. His chest circumference increased to 69cm, and the imaging examination showed that the shape of the thorax was significantly improved (Fig. 2A). He was discharged 49 days after operation.

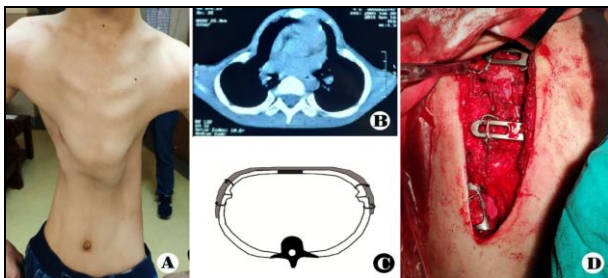
During follow-up, his symptoms did not completely disappear, but would worsen obviously after activity.

His pulmonary hypertension also existed all the time. Considering the possibility that the steel bars may compress the heart and lungs and affect the PAP, the steel bars were finally removed one year postoperatively (Fig. 2B). The patient's condition did not change significantly immediately after removal. One year after taking Bosentan and Viagra, his symptoms disappeared and PAP fell to normal finally.

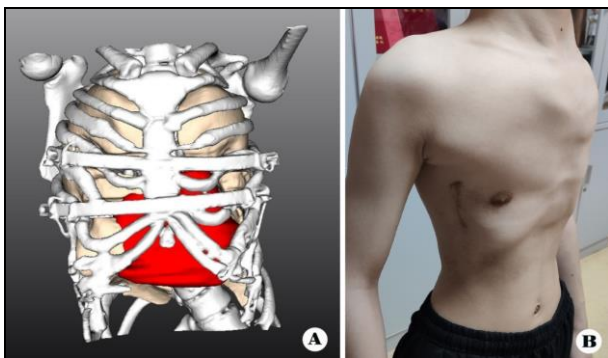
### Discussion

Operation for ATD is challenging, and severe pulmonary hypertension is regarded as a contraindication to ATD operation<sup>[5,6]</sup>. The mechanism of pulmonary hypertension is unclear, but we believe that the possible reason is related to the compression of the chest wall on the heart and lung. If compression can be eliminated, pulmonary hypertension may be eliminated possibly. Therefore, we did not regard pulmonary hypertension as contraindication.

Our patient had severe pulmonary hypertension, and the early postoperative effect was unsatisfactory, but the later results showed that the operation played a decisive role. Our procedure can directly increase the dimensions of the chest capacity while eliminating the depression, which laid the foundation for the final elimination of pulmonary hypertension.



**Fig 1:** Images before and related to the operation. Preoperative chest wall appearance (A); preoperative chest CT image (B); schematic diagram of operation (C); and operative image. (D).



**Fig 2:** Images after operation. Three-dimensional reconstruction image of thorax (A) and thorax appearance after steel bars removal.

### Conclusion

ATD with severe pulmonary hypertension will undoubtedly make surgery more challenging, but our experience shows that this situation should not be regarded as contraindication. If the correct surgical method can be used, satisfactory results would be obtained.

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