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Surgical treatment of asphyxiating thoracic dystrophy with median thoracic expansion and Nuss procedure

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Abstract

Asphyxiating thoracic dystrophy can be divided into two types: type I is cylindrical with slight depressions on chest wall, and type II is non-cylindrical with serious depressions on the chest wall. The nature of the depressions determines the choice of operation. For type I patients, the relatively reasonable operation is median thoracic expansion. However, due to the existence of depressions, the effect is not ideal. In order to eliminate the effect of depression, we designed a operation with an additional Nuss procedure to median thoracic expansion. We applied this operation to a 3-year-old patient and achieved good results.

Keywords: Asphyxiating thoracic dystrophy, Median thoracic expansion, Nuss procedure

Introduction

Asphyxiating thoracic dystrophy (ATD), also known as Jenue syndrome, was first reported in 1955^[1, 2]. This disease is an autosomal recessive disease, which is extremely rare and dangerous. Its main damage comes from its small and narrow chest^[3, 4]. Due to the compression of two lungs, patients always have obvious respiratory insufficiency, and most of them die before puberty^[1]. It is generally believed that surgery is the only effective way to treat this disease. So far, several surgical methods have been reported^[1-6]. However, because the structural characteristics of the chest wall have not been fully considered, the results of these methods are not ideal. We received a 3-year-old female child with ATD who had a narrow chest and slight lateral chest depressions. We designed a special operation for her and achieved satisfactory results.

Case Report

A 3-year-old female ATD patient was referred to Guangdong Second Provincial General Hospital due to severe hypoxia. She had a full-term natural birth and was the only child of nonconsanguineous parents. After the patient was born, she received endotracheal intubation mechanical ventilation because of respiratory distress. She was weaned 9 days later, and was given oxygen with mask since then. The patient was asymptomatic when calm, but there was still hypoxia when eating and crying during the neonatal period. At the age of one year, she was hospitalized for pneumonia with hypoxia, and was discharged after using antibiotics and oxygen inhalation. Half a year before admission, she began to suffer from severe hypoxia and needed continuous oxygen inhalation. Finally, she admitted to our hospital for surgery. Physical examination showed that her height was 89cm, her chest circumference was 46cm, and the appearance of the chest was cylindrical (Fig. 1A). Imaging examination showed that her thorax was narrow and small, and there were slight depressions on both sides of the chest wall (Fig. 1B, C). She was finally diagnosed as ATD, and we performed surgical treatment for her. The operation was carried out under general anesthesia. Supine position was adopted, and the median sternotomy was made. After two sternal halves were stretched apart, four short steel bars were placed between the sternal halves and fixed with them from their posterior surface (Fig. 2A). Two short incisions were made on both sides of chest wall. An arc-shaped steel bar was inserted into the thorax and passed through the mediastinum, and then was turned over to prop up the depressions (Fig. 2A), thus an additional Nuss procedure was completed^[7, 8]. After all the bars were firmly fixed, the incisions were closed, and the total operation was completed (Fig. 2B). Mechanical ventilation was used continuously for 2 days postoperatively. After the condition was stable, the nasal catheter was used to give oxygen and stopped 9 days later.

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The oxygen saturation was maintained above 91%, and her chest circumference increased to 52cm. The patient was discharged 21 days postoperatively. At the early stage of follow-up, there was no hypoxia when she was calm, but there was still hypoxia when crying. The symptoms disappeared one year later, when X-ray examination showed that the shape of thorax was significantly improved (Fig. 2C).

Discussion

ATD is a very complicated thoracic deformity, and the operation is challenging. So far, several operations have been reported. According to the characteristics of operations, we divided them into three types, namely median thoracic expansion [2], lateral thoracic expansion [3, 4], and orthopedic surgery [5, 6]. These operations can increase the volume of the thorax and reduce the compression of lung tissue, so they all have a certain effect. However, there are some defects because the structural details of the chest wall are not fully considered.

By May 2022, we had completed operations for 33 ATD patients. Through the observation of these patients, we found that ATD could be roughly divided into two types: type I, the appearance of the thorax is cylindrical, and there are slight depressions on both sides of the chest wall; Type II, the appearance of the thorax is non-cylindrical, and there are obvious and deep depressions on both sides of the chest wall.

For type I ATD, because depressions of the chest wall are not serious, both median and lateral thoracic expansion can be used [2-4]. Since orthopedic surgery is designed for depression, and the depression of type I is not obvious, such surgery is not appropriate [5, 6]. For type II ATD, if median thoracic expansion is used, the depressions of the chest wall will not be eliminated [2]; if the lateral thoracic expansion is used, it will be extremely difficult to complete the corresponding operation due to the obvious depression on the chest wall [3, 4]. In contrast, the orthopedic surgery for this type is reasonable [5, 6].

Our patient's chest was cylindrical without serious depression, so she was a typical type I ATD patient and suitable to use median thoracic expansion. But we found that if the sternal halves were simply stretched apart, the depressions would be significantly deepened (Fig. 3A, B). This would not only affect the expansion of the thorax, but also counteract the total effect. Obviously, the depressions were the important defects affecting the surgical effect. In order to eliminate these defects, we add an additional Nuss procedure on the basis of median thoracic expansion. We used steel bar to support the depressions (Fig. 3C) [7, 8]. Because it could eliminate the depressions and ensure the good effect of median thoracic expansion simultaneously, we regarded that this was a reasonable method [8]. We applied this method to our patients and achieved satisfactory results.

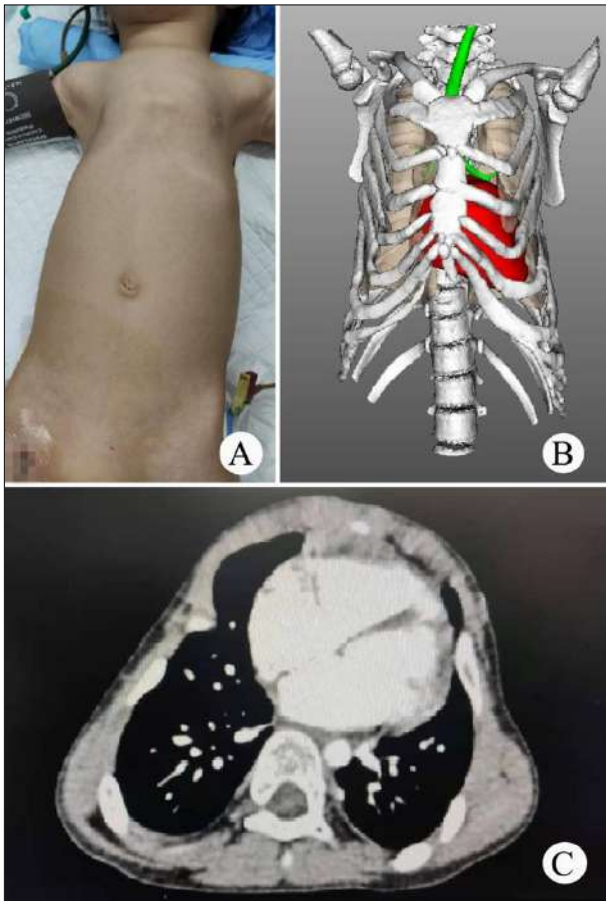


Fig 1: Preoperative photograph. A, The appearance of the thorax before operation; B, The three-dimensional reconstruction image of the thorax shows that the thorax is narrow and small, and the appearance is cylindrical; C, CT scan showed slight depression on lateral chest wall.

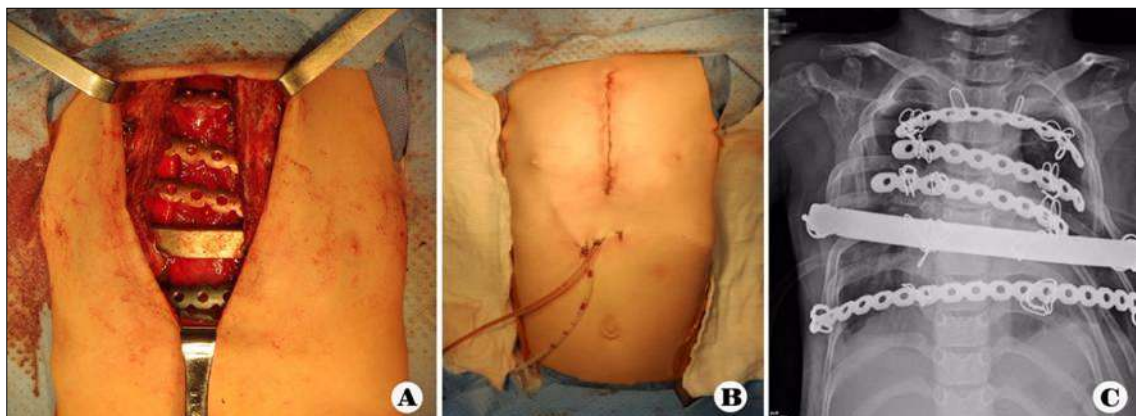


Fig 2: Perioperative and postoperative photograph. A, Steel bars are placed between the two halves of the sternum to stretched them apart; B, Postoperative appearance of thorax; C, X-ray examination one year after operation.

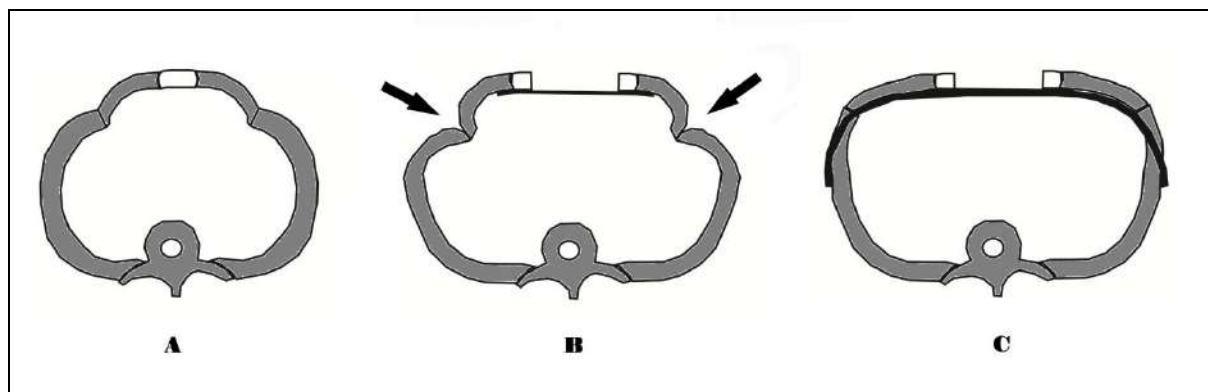


Fig 3: Operation schematic diagram. A, Chest diagram of type I ATD; B, The depressions of lateral chest wall will deepen after simple median thoracic expansion; C, The depressions of the lateral chest wall will be eliminated after the additional steel bar was used for supporting.

Conclusion

Our method can eliminate the defects of median thoracic expansion and obtain better surgical results. Therefore, it is a relatively reasonable operation. However, due to limited experience, its actual effect needs to be further confirmed.

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