

Clinical application of Nuss procedure for chest wall deformity in Poland syndrome

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Abstract

A 6-year-old girl with Poland syndrome was diagnosed with loss of hard and soft tissue on her affected chest. She required chest reconstruction to protect her thorax and improve her developmental delay. We applied a titanium device used for children with funnel chest to her, resulting in good shape. By using this device, we avoided sacrificing healthy rib and muscle; she could save the tissue for secondary surgery in puberty. The device made it possible to support the growth of her thoracic frame and has kept it in good shape. Also, 66 months after removing the device, her chest contour has been maintained. We evaluated the change of her chest frame by comparing the preoperative and postoperative CT index and found the result quite an improvement. We suggest that our technique would be easy, of low invasiveness, and safe for treatment of thoracic deformity in school age.

Key words: Poland syndrome, Nuss procedure, chest wall deformity.

Introduction

Poland syndrome is a rare congenital anomaly characterized by hypoplasia of the breast and nipple, insufficient subcutaneous tissue, absence of the costosternal portion of the pectoral major muscle, lack of the pectoral minor muscle, aplasia or deformity of the axillary and mammary region, and unilateral brachysyndactyly. This syndrome was reported by Alfred Poland in 1841. Some surgeons have demonstrated various methods of repair. Reconstruction and/or stabilization of the soft tissue defects (on the chest) usually may be achieved using muscle flaps or a prosthesis. However, in some cases of Poland syndrome, patients who have a severe chest wall deformity require repairs of hard tissue such as bone and cartilage. These reconstructions are generally invasive.

Streszczenie

U 6-letniej dziewczynki z zespołem Polanda zdiagnozowano ubytek tkanek twardych i miękkich klatki piersiowej. Pacjentka wymagała rekonstrukcji, aby ochronić jej klatkę piersiową i zmniejszyć opóźnienie rozwojowe. Zastosowanie urządzenia tytanowego używanego u dzieci z klatką piersiową lejkową zaowocowało prawidłowym kształtem klatki piersiowej. Wykorzystanie urządzenia pozwoliło też zapobiec pobraniu zdrowej tkanki żeber i mięśni i zachować tkanki na kolejną operację w okresie dojrzewania. Urządzenie umożliwiło wsparcie wzrostu klatki piersiowej pacjentki i utrzymanie jej prawidłowego kształtu. Po 66 miesiącach od usunięcia urządzenia kontur klatki piersiowej został utrzymany. Zmiany w kształcie klatki piersiowej oceniono za pomocą przedoperacyjnej i pooperacyjnej tomografii komputerowej, uwidaczniając wyraźną poprawę. Autorzy pracy oceniają, że opisana technika jest łatwą, małoinwazyjną, bezpieczną i uzasadnioną metodą leczenia deformacji klatki piersiowej u dzieci w wieku szkolnym.

Słowa kluczowe: zespół Polanda, operacja Nussa, deformacja ściany klatki piersiowej.

We achieved a modified Nuss procedure in the case of a girl suffering from Poland syndrome with severe chest deformity. Our method is simple, safe, easy and contributes to reduced hospitalization and cost. Moreover, patients can save their soft tissue for continuous treatment in puberty. We insist that this technique has provided satisfactory results with our aims in mind and should be considered as an initial treatment.

Case report

The patient was a 4-year-old girl with Poland syndrome without any other congenital anomalies or complications during delivery. Her descent was healthy. After her birth, she exhibited a loss of the complex of pectoral muscles on

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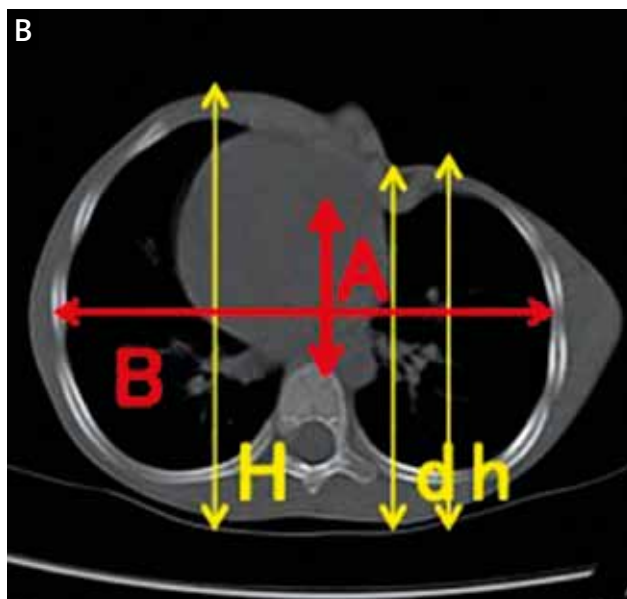


Fig. 1. A) Preoperative findings. **B)** Preoperative axial CT Measured values (cm): A = 4.61, B = 15.88, d = 10.88, H = 12.79, h = 10.95

the left chest and severe chest deformity caused by hypoplasia of ribs from the 3rd to 5th rib and paradoxical breathing (Fig. 1A). Computed tomography demonstrated warp of her thoracic cavity allowing the left lung to bulge toward the skin (Fig. 1B). Permission for surgical intervention was obtained by written informed consent from the patient and parents.

Under single right-lung ventilation, small incisions were made in the three areas of the chest along the deformity. Guided by thoracoscopy, a 9.44 inch-long titanium bar (Solve Co., Yokohama, Japan) was inserted beneath the lowest point of the front thorax and the intercostal space of ribs 4-5 (Fig. 2). We performed the whole method following the Nuss procedure, which is applied to the funnel chest. After the operation, the patient received adequate pain control with epidural anesthesia. No complication was observed during the perioperative period. The metal



Fig. 2. Intraoperative view. Insertion of the titanium bar for chest wall repair

bar removal was performed at 28 months postoperatively. At the latest clinical and radiologic follow-up examination 100 months after the operation, the chest had kept in good shape, with no regression of the deformity, and the patient did not report any impairment upon moving (Fig. 3A). Computed tomography demonstrated that the CT index improved from 0.29 to 0.85, the depression index from 0.85 to 0.92, and the symmetric index from 0.86 to 0.93 at 78 months after the surgery (Fig. 3B).

Discussion

Poland syndrome is a congenital progressive anomaly, causing chest wall deformities and atrophy of soft tissues [1]. Although various surgical techniques have been described for chest wall reconstruction depending on sex, age, and severity of the disease, muscle flaps and prostheses are the workhorses for chest augmentation. Especially for severe deformity cases, surgeons have to plan multiple operations. The deformity caused by hard tissue including rib, cartilage and sternum should be repaired in advance; subsequently augmentation of soft tissue should be carried out with flaps in puberty. To salvage the rib for donation, it is necessary to make a huge incision on the side of the healthy chest with a risk of mediastinal associated complications. The aim of this study was to determine whether the titanium plate that we have used in cases of funnel breast patients could be applied to Poland syndrome. In our experience, this technique effectively reduced the thoracic deformity of patients in school age; the metal device has applied continuous pressure on the patient's thorax from the inside, resulting in improvement not only of the dimple of the front chest but also the contortion of the side. Also, this use of our technique reduces the need for hospital admission and cost compared with the conventional treatments

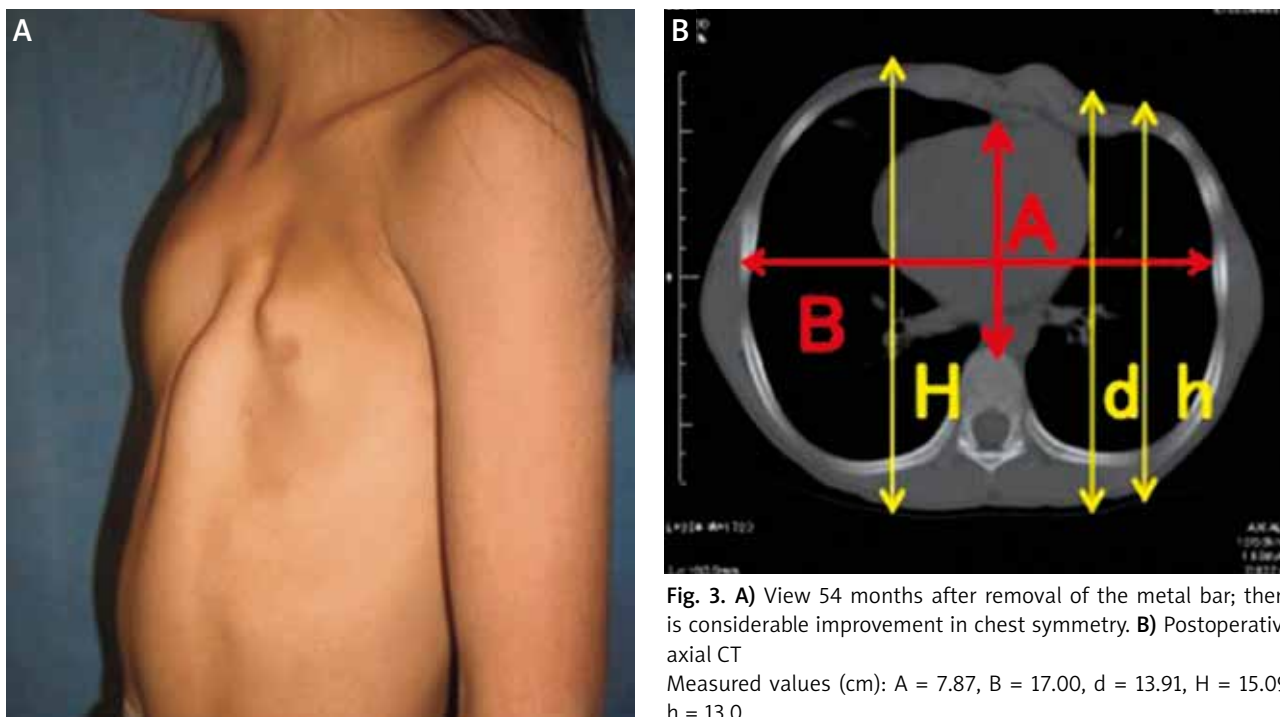


Fig. 3. A) View 54 months after removal of the metal bar; there is considerable improvement in chest symmetry. B) Postoperative axial CT
Measured values (cm): A = 7.87, B = 17.00, d = 13.91, H = 15.09, h = 13.0

which include rib osteotomy and soft tissue reconstruction. With careful handling the device guided by thoracoscopy during an operation, we have avoided much of the risk of this technique and performed it in a short time. When we remove the devices, the patient requires hospitalization again; the second operation is simple and easy.

Recently several reports [2-4], represented by Waldhausen *et al.* [5] and Berthet *et al.* [6], have suggested the usability of titanium devices on young patients for an initial reconstruction of hard tissue defects on the chest wall. In their comments, the authors referred to the handiness, safety, and stiff result of the device, confirming its effectiveness. However, these corrections were focused on the longitudinal expansion along the body trunk, so that did not produce a sufficient result in the deformity of the front chest. To overcome this disadvantage, Nishibayashi *et al.* [7] reported the case of a 4-year-old boy with Poland syndrome treated by Nuss method to repair the chest deformity. They succeeded in improving the over-projection of the sternum and the dent of the side chest without any complications. (Additionally they prevented the patient from sacrificing his healthy ribs.) Because the titanium devices are supposed to be removed three years after the surgery, as in funnel chest patients, it is unlikely that this technique would promote severe failure of the chest wall to thrive. In our case, we applied a symmetric titanium bar without transforming it to comply with the shape of the deformity.

Sixty-six months after removing the device, neither regression nor failure of chest growth has been noted. Although mild thorax deformity has remained, it is considered important to improve the form of hard tissue on the chest with a non-invasive method during the young period. We plan the second operation of soft tissue reconstruction,

but further follow-up is necessary to more accurately determine the outcome after placement of this device.

Conclusions

We have reported the case of a 4-year-old Poland syndrome patient who had congenital chest deformity. The patient was treated with a titanium bar device as used in funnel chest patients. Our technique is safe, reasonable, preserves the patient's healthy hard rib and soft tissue from an initial reconstruction, and promises to improve the chest deformity of the thoracic interior. We suggest that this method should be included in the initial treatment of chest abnormality syndrome.

Disclosure

Authors report no conflict of interest.

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