



Modified Nuss procedure for severe thoracic deformity associated with paternal uniparental disomy 14: Report of a case



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ABSTRACT

Paternal uniparental disomy 14 (pUPD14) is a rare condition, and only about 30 cases have been reported in Japan. It is characterized by facial abnormalities, a small chest, and ventral abdominal wall defects. An 11-year-old girl with pUPD14 had severe thoracic deformity with a projection on the right side and depression on the left side. On chest X-ray films and CT scans, her chest was narrow, the sternum showed clockwise rotation and depression, and the inferior parts of some ribs protruded anterior to the sternum. We performed a “modified Nuss procedure” to treat the patient’s thoracic deformity using a pectus bar. Costal cartilages were resected to increase thoracic mobility, and an anterior incision was made in the sternum down to the marrow to correct her thoracic deformity. Marked improvement was achieved postoperatively.

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Paternal uniparental disomy 14 (pUPD14) is rare condition and it is estimated that there have been about 30 cases in Japan [1]. Uniparental disomy (UPD) means that both copies of a pair of chromosomes are inherited from the same parent [2]. Several specific UPDs have been described, each of which is associated with a distinct clinical phenotype that depends on the parental origin of the chromosome. PUPD14 is characterized by facial abnormalities, respiratory problems associated with a small chest and so-called ‘coat-hanger’ ribs, and ventral abdominal wall defects in the neonatal period [3,4]. The respiratory symptoms show spontaneous improvement over time [1].

Here we describe a modified Nuss procedure, including costal cartilage resection and sternal incision to the bone marrow, which was performed to correct progressive severe thoracic deformity associated with pUPD14.

1. Case report

An 11-year-old girl presented with thoracic deformity. She had previously undergone abdominal wall repair for omphalocele and

pUPD14 had been diagnosed by chromosomal analysis. She had severe thoracic deformity as well as mild mental retardation. Her thoracic deformity had shown progression over the previous 12 months. The chest wall was asymmetrical, with prominence of the fifth, sixth, and seventh ribs on the right side, while there was a slight depression on the left side (Figs. 3A and 4).

The chest X-ray film revealed a small chest and a ‘coat-hanger’ appearance of the ribs (Fig. 1). Chest CT demonstrated clockwise rotation of the sternum, with the costal cartilages from the fifth cartilage protruding anterior to the sternum on the right side, while there was a depression on the left side centering around the sternum (Fig. 1C, D). The Haller index for pectus excavatum was 3.38 (Fig. 1D). In addition, the distal part of her sternum was recessed due to sternal curvature (Fig. 1B). The patient wished to have thoracic wall repair and there was a risk of future cardiorespiratory problems because her thoracic wall deformity was progressing. Therefore, we decided to perform surgical treatment of her thoracic deformity.

2. Operative technique

On preoperative CT scans, there was a thoracic wall bulge resembling pectus carinatum on the right and a depression like pectus excavatum on the left (Fig. 3A). Therefore, a surgical procedure that could correct this complex deformity was required. The following method was devised to treat both deformities (Figs. 2 and 3).

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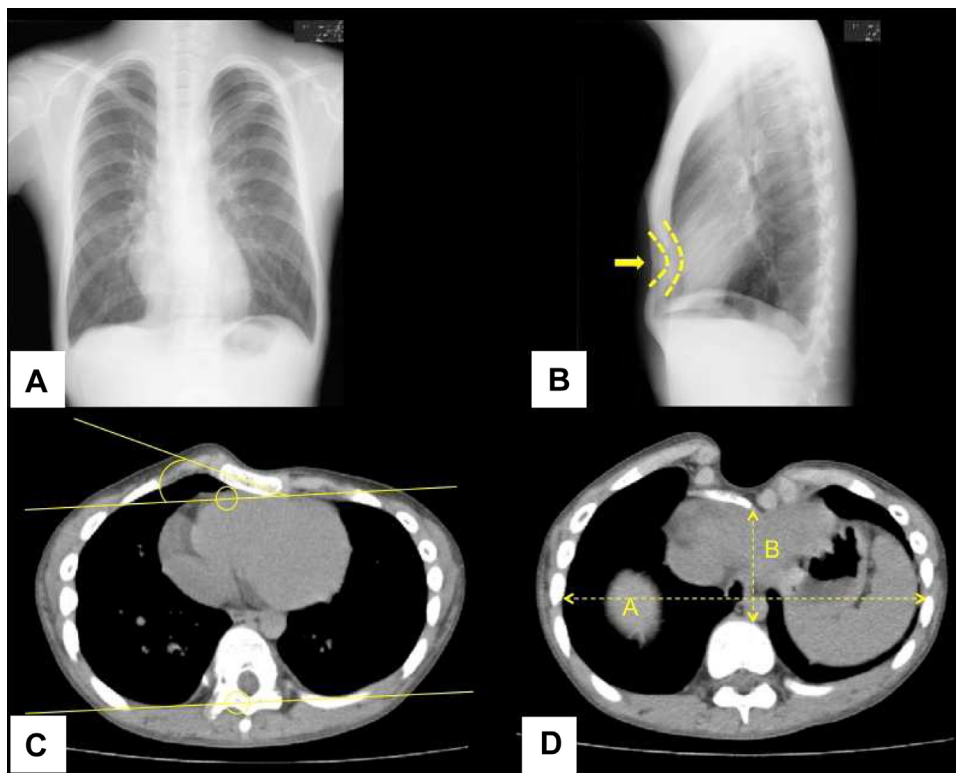


Fig. 1. Chest X-ray films (A, B) and chest computed tomography scans (C, D). (A) The thorax wall is narrow and elongated. (B) There is a depression in the lower sternum. (C) The sternum shows about 30° of clockwise rotation. (D) Costal cartilages project anterior to the sternum. There is compression of the heart by the sternum.

The procedure involved simultaneous elevation of the depression of the distal sternum and the left thoracic wall, while flattening the rib cage on the right side by positioning a pectus bar in the pleural cavity (Fig. 2). In addition, we resected several costal

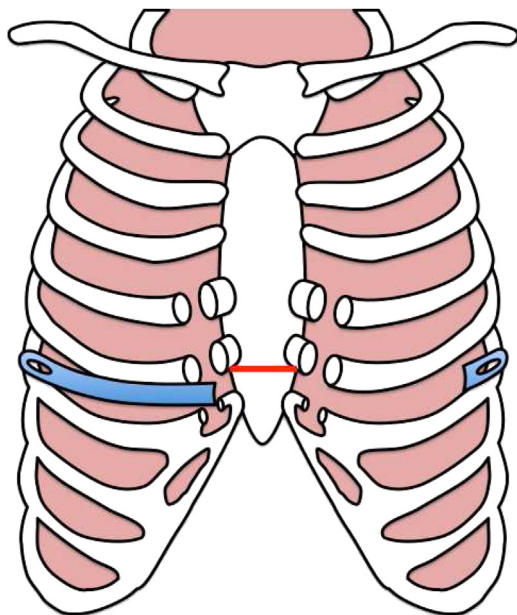


Fig. 2. The pectus bar was inserted from the right sixth intercostal space at the right sternal border to the left sixth intercostal space at the anterior axillary line. Incision in the sternum (-). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

cartilages to increase the mobility of the rib cage, and incised the anterior sternum to correct her sternal recession.

After a 4-cm arc-shaped incision was made in the anterior thoracic wall, the attachment of pectoralis major was incised to expose the costal cartilages.

The perichondrium of the bilateral fifth, sixth, and seventh costal cartilages was incised and dissected, after which these cartilages were resected at a site 1 cm from the sternum. Then the perichondrium was repaired with absorbable sutures. Next, a V-shaped incision was made in the sternum down to the marrow and hemostasis was achieved with bone wax.

Subsequently, a pectus bar was shaped so that the left and right thoracic walls would become symmetrical. The bar had a larger curve on the right side and a slightly smaller curve on the left side, with a small recess at the center (Fig. 3B).

Bilateral 3-cm incisions were made at the sixth intercostal level in the midaxillary line and subcutaneous tunnels were created to insert the pectus bar. Then, a 5 mm 30° thoracoscope was inserted from the right seventh intercostal space for thoracoscopic guidance.

To compress the projecting costal cartilages, the pectus bar was positioned subcutaneously from the sixth intercostal space near the right sternal border as the right hinge point. Then the left fifth and sixth ribs and the recessed distal sternum were elevated on the left side, using the left sixth intercostal space near the midclavicular line as the left hinge point (Figs. 2 and 3B). Finally, the pectus bar was fixed to the sixth and seventh ribs bilaterally using No. 5 non-absorbable sutures at the midaxillary line and wound closure was done without inserting a drain tube. The operating time was 165 min and the blood loss was 37 ml.

After surgery, her thoracic wall deformity was markedly improved (Fig. 4). She had a subcutaneous hematoma and atelectasis of the right lung postoperatively, but both improved with

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