Congenital absence of multiple ribs

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***Background*: Congenital rib absence might be the absence of either single or several ribs or all ribs at one side. The sporadic cases were ever reported over years.**

***Methods*: A full-term Chinese infant was noticed that both shoulders were asymmetric significantly with left sagged shoulder at the age of 6 months. Her left hand could not rise over her shoulder and her gripping power was a little bit poor. She presented a good mental and motor development without other deformity. Imaging examination was given to the infant for diagnosis.**

***Results*: The absence of left three ribs was found on her chest X-ray film. The results of three-dimensional CT reconstruction image confirmed the absence of left 1st-4th ribs, the overlapping of 5th-7th ribs on the left side and 2nd-3rd ribs on the right side.**

***Conclusions*: Simple congenital absence of multiple ribs is confirmed based on her manifestation and imaging results.**

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***Key words*: infant;**
congenital rib absence;
three-dimensional CT reconstruction

Introduction

Congenital rib absence might be the absence of either all ribs at one side or single or several ribs. The incidence was undocumented and sporadic cases were reported over years.[1-4] This is a case report of a 17-month-old Chinese infant with the absence of left 1st-4th ribs, and the overlapping of 5th-7th ribs on the left side and 2nd-3rd ribs on the right side confirmed by three-dimensional CT reconstruction image.

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Case report

A 17-month-old girl, born as G1P1 at a gestational age of 39^{+4} weeks with a birth weight of 3.3 kg and no perinatal asphyxia, was noticed by her parents at the age of 6 months that the grip of her left hand was weaker than that of her right hand, in particular when she raised her hands, the left hand could not reach the level of the right hand. Her both shoulders were significantly asymmetric with narrow left shoulder compared to her right shoulder. The asymmetry of her shoulders tended to be more and more visible with her increasing month age (Figs. A and B). The absence of left three ribs was found on her chest X-ray film taken by other hospital one month ago (Fig. C). The results of three-dimensional CT reconstruction taken in our hospital showed the absence of left 1st-4th ribs, and the overlaping of 5th-7th ribs on the left side and 2nd-3rd ribs on the right side (Figs. D and E). The left scapula shifted internally with the inferior scapular angle moved to the vertebral column and the medial superior scapular angle moved upward over the clavicle. The little girl looked well generally with good appetite, defecation and sleeping. She had never had dyspnea since birth. She appeared to have a normal intelligence, could walk alone at 14-month-old age, and now could say simple syllables including "Baba, Mama", etc. There was nothing special occurring during her mother pregnancy and no similar family history was found.

Physical examination showed her good health condition with 9.8 kg body weight and 80 cm height. Her shoulders were significantly asymmetric owing to her left sagged shoulder. The development of her left chest was not very well. Her breath sounds bilaterally were normal and symmetric without the occurrence of inspiratory intercostal depression. The abdomen was soft in palpation without hepatosplenomegaly. She raised her left hand that can only reach her shoulder. However, the activity of her left hand was good and could hold and nip small things although her power of gripping was a little poor. The activities of her right hand and lower extremities were normal without brachydactylia and syndactyly of hands and feet. Nervous system examination also showed nothing abnormal.

The little girl was diagnosed as having "congenital absence of left 1st-4th ribs with overlapping of partial

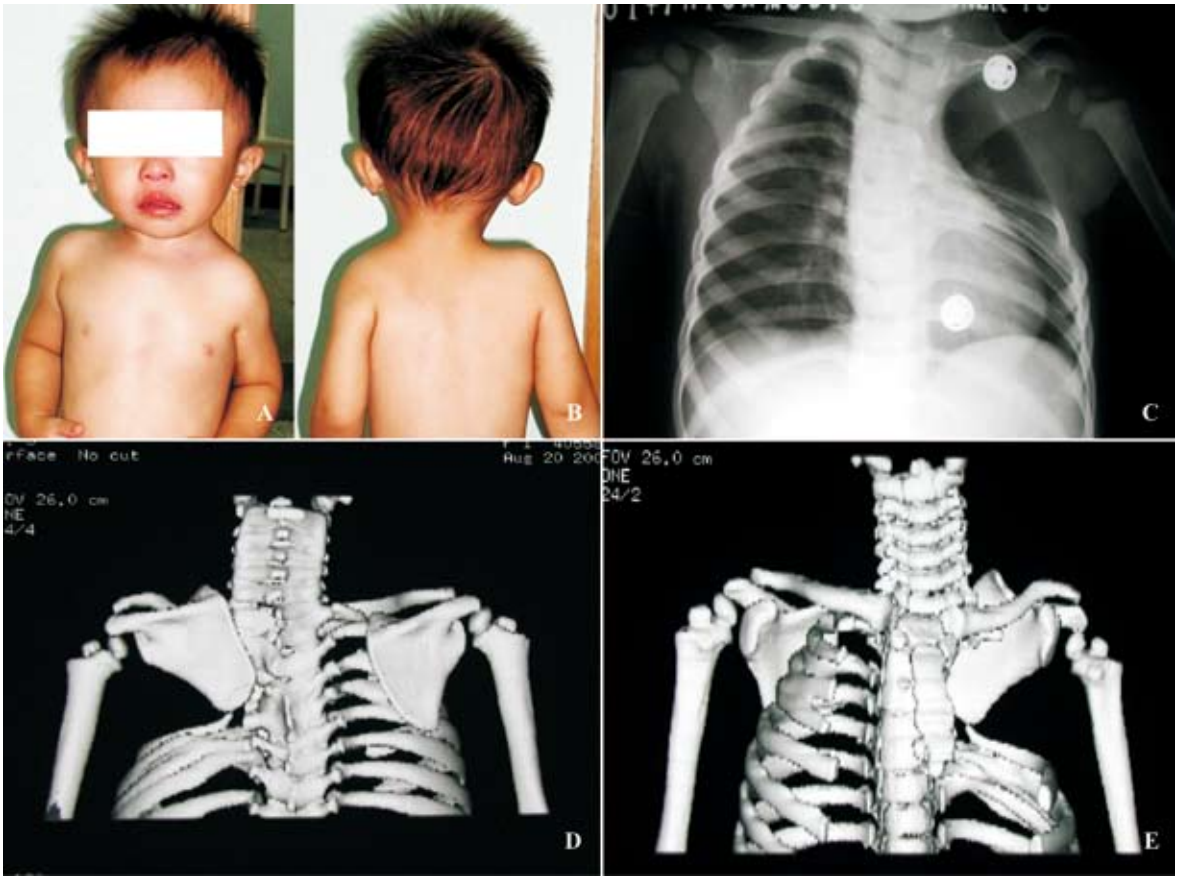

Fig. A & B: The little girl's portraits in front and in back showing her asymmetric shoulders with left sagged shoulder; **C:** Chest X-ray showing the absence of left 1st-3rd ribs. **D & E:** Three-dimensional CT reconstruction image on the back and in front showing the absence of left 1st-4th ribs, and the overlapping of 5th-7th ribs on the left side and 2nd-3rd ribs on the right side. The left scapula shifted internally with the inferior scapular angle moved to the vertebral column and the medial superior scapular angle moved upward over clavicle.

ribs in both sides" based on her clinical manifestations and imaging diagnosis.

Discussion

There are twelve pairs of ribs in the normal human body. 1st-5th ribs are joined with the sternum, 6th-10th ribs are joined with the margin of rib cartilage, and 11th-12th ribs are free ribs, respectively. The etiology of rib absence remains unclear. It was surmised that a local blood-supply insufficiency might result in rib dysplasia during the embryonic period.[5-7] The absence of parasternal 2nd-5th ribs is common and usually accompanied with dysplasias of mammary glands, local subcutaneous tissue, and ectopectoralis and entopectoralis partially or entirely. The absence of underthoracic 6th-12th ribs is rare and usually accompanied with hemicone, meningomyelocele, diaphragmatic eventration, congenital heart disease, duplication of the intestine or anomaly of the rectum, etc. It was reported that an infant with trisomy 13 had bilateral presence of cervical ribs and the bilateral absence of 12th thoracic ribs.[8] It is not difficult to distinguish simple congenital rib absence from cases of trisomy 13. The latter usually appears to have a special face and other malformations. Primary signs of confirmed diagnosis for the patient with Poland syndrome include the absence of pectoralis and the developmental deformity of fingers except for the absence of ribs.[9,10] There is also report about congenital rib defects with the Pierre Robin syndrome.[11]

Usually, single rib absence is not managed necessarily if there is no significant clinical symptom. When several ribs are absent, however, paradoxical breathing could be induced owing to the lack of support for the thorax and malacia of the chest wall. The chest malactic zone is depressed during inspiration and prominent during expiration, thus the pendular movement of the mediastinum can be induced. Such infants usually manifest dyspnea and cyanosis soon after birth, which may be aggravated

with feeding and blubbering. Emergency operation is necessary when infants have absence of multiple ribs with respiratory distress. If the range of rib absence is small, local compression bandaging is used to control paradoxical breathing and pendular movement of the mediastinum. However, immediate operation should be performed for infants with repeated infection in the respiratory tract. A few options are given to the repair of defective chest wall, including broad fascia transplantation for small-range rib absence, adjacent self-rib transplantation, and coverage by the skin flaps of either latissimus dorsi or musculus obliquus externus abdominis, marlex polyethylene mesh,[12] and sternum-reversal operation for large-range rib absence. The symptoms of untreated patients can be improved with increasing age, but severely deformed spinal column and thorax may be inevitable.

The present case should be a simple congenital rib absence based on the manifestations. The etiology remaineds to be confirmed. The infant had no symptom of dyspnea or infection. Her intrathoracic organs were not affected despite the absence of four ribs on one side. However, as growing up, clinical complications may occur with negative impact on her psychological development owing to her asymmetric shoulders. Whether the repair operation is necessary was discussed by pediatricians, surgeons and orthopedists at this hospital. Once the repair operation is undertaken, frequent modification operations will be continued to adapt her enlarged thorax during growing-up. It was determined at last that the orthopedic repair of thorax would not be undertaken at present. The girl is kept to follow up now, and the operative repair would be made in an appropriate time later on if no complications occur. Particular attention should be paid to protect the unsubstantial thorax of the little girl for the avoidance of accident intrathoracic-organ injury. No indications are available for the management of the overlapping of ribs since her respiratory function and thorax are not influenced significantly.

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