

A rare case of congenital absence of ribs in a newborn baby

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ABSTRACT

Congenital absence of ribs is a rare condition. The defect is generally unilateral, and the first rib is usually unaffected. We report a case of congenital absence of ribs in an early-term (37 + 1 week), female baby. The 5th and 6th ribs on the right side were absent. The baby had mild respiratory distress which resolved within 1 day and had no other congenital anomalies. Rarely, it can result in severe lung restriction and cor pulmonale.

Key words: Congenital anomalies, Congenitally missing, Infant, Ribs

Congenital absence of ribs is a rare condition. The defect is generally unilateral, and the first rib is usually unaffected [1]. The complete absence of ribs is less frequent than the arrest in the development of ribs. The arrest of development is generally manifested at the costal cartilage and the anterior parts of the ribs. It does not occur at any constant level [2]. Isolated defects of the highest and lowest ribs have minimal clinical pulmonary consequences. Missing midthoracic ribs may be associated with the absence of the pectoralis muscle (Poland syndrome), and lung function can become compromised. Associated kyphoscoliosis and hemivertebrae may accompany this defect [3].

Here, we report an early-term female newborn baby with congenital absence of the 5th and 6th ribs on the right side.


CASE REPORT

An early-term (37 + 1 week), female baby was born through cesarean section to a 33-year-old mother with gestational diabetes and pre-eclampsia. There was no history of consanguinity. Antenatal anomaly scan was said to be normal. The baby had a birth weight of 1975 g, a length of 43.5 cm, and the head circumference of 31 cm with a ponderal index of 2.39. The baby was found to have symmetric intrauterine growth retardation. The baby cried immediately after birth but was found to have respiratory distress soon after. The baby was shifted to the neonatal intensive care unit and kept under hood oxygen. A clinical diagnosis of transient tachypnea of the newborn was made.

Routine physical examination showed a heart rate of 132 beats/min, respiratory rate of 68 cycles/min with mild retractions, saturation of 93%, and mean arterial pressure of 46 mmHg. Chest examination showed no obvious asymmetry of the chest anteriorly though mild hollowing of the chest was noted in the right infra-axillary region (Fig. 1). On palpation on the right side of the chest, a midthoracic bony defect was noticed with palpable bony prominence above and below the defect. On auscultation, bilateral air entry was equal and normal vesicular breath sounds were heard. The cardiac examination showed regular heart rhythm and normal heart sounds with no murmurs. The abdomen was soft on palpation. The vertebral column and limbs had no obvious deformity.

Chest X-ray showed normal lung fields with the absence of the 5th and 6th ribs on the right side (Fig. 2). Contrast-enhanced computed tomography (CECT) of thorax showed complete absence of the right 5th rib and part of the 6th rib, with expansion and fusion of the right 3rd and 4th ribs, 7th and part of the 6th rib, and crowding of 9th, 8th, 7th, and 6th ribs along with the expansion of sternocostal joints while the rest of the ribs had no deformities with normal lung fields (Fig. 3). To rule out other associated anomalies, neurosonogram and ultrasonogram of the abdomen and pelvis were done which were normal. 2D echo showed small ostium secundum-atrial septal defect 3 mm.

Respiratory distress settled within 2 h but the baby continued to have minimal oxygen requirement for 3 days. Direct breastfeeds were started once the baby was off oxygen and shifted to the mother's side on day 5 of life.

Access this article online	
Received - 13 July 2023 Initial Review - 25 July 2023 Accepted - 30 September 2023	Quick Response code 
DOI: 10.32677/ijcr.v9i10.4168	

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Figure 1: No obvious asymmetry of the chest anteriorly though mild hollowing of chest was noted in the right infra-axillary region



Figure 2: Chest X-ray showed normal lung fields with absence of 5th and 6th ribs on the right side

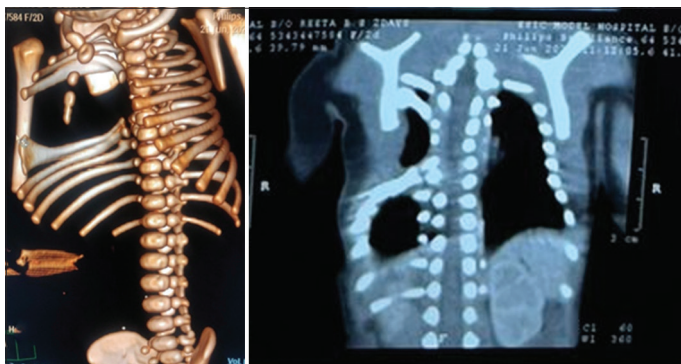


Figure 3: Contrast-enhanced computed tomography of thorax showed complete absence of the right 5th rib and part of 6th rib, with expansion and fusion of right 3rd and 4th rib, 7th and part of 6th rib, and crowding of 9, 8, 7, and 6th ribs with expansion of sternocostal joints while the rest of the ribs had no deformities with normal lung fields

DISCUSSION

Congenital absence of ribs is a rare condition without certain etiology. It may be related to impaired blood supply during the embryonic period [1]. Congenital absent ribs can present alone or

as part of many congenital anomalies like in “Poland Syndrome”. Poland syndrome is a rare congenital disorder associated with various anomalies such as ipsilateral syndactyly, brachydactyly, dextrocardia, herniation of the lung, underdevelopment of upper ribs, aplasia, or hypoplasia of the breast [3]. Our baby did not have any of these abnormalities.

Absence of 2nd-5th ribs anteriorly can be associated with lung herniation and significant abnormal respiration. The lung was soft and non-tender and may be easily reducible on examination. CECT in our case showed normal lungs with no herniation or defects [4].

Chest radiographs demonstrate the deformation and absence of ribs with secondary scoliosis. Most rib abnormalities are discovered as incidental findings on a chest film. In general, neonatologists would be detecting the superior rib deformity at the time of birth, inferior rib deformity can be overlooked as inferior rib deformity is difficult to detect at the time of birth, most of the time discovered as an incidental finding [5,6]. In our case, the bony defect was detected on clinical examination and confirmed by X-ray and CECT. The absence of the 5th and 6th ribs in our case did not affect the respiratory and cardiovascular functions.

There was a similar case report where a full-term male newborn with absent 5th – 8th ribs on the left side presented with transient tachypnea [7]. Another article reported a full-term male newborn with absent superior anterior 2nd-4th ribs on the left side without respiratory distress [5].

Complicating sequelae include severe lung restriction (secondary to scoliosis), cor pulmonale, and congestive heart failure. Symptoms are often minimal but can cause dyspnea. Respiratory distress is rare in infancy [8]. In case of clinical compromise or significant lung herniation, homologous rib grafting can be performed. Rib-expanding procedures can also be undertaken. To correct associated chest wall anomalies with rib abnormalities, a modified Nuss procedure can also be performed. Cosmetic breast surgery might be required in adolescent girls with rib anomalies [9].

CONCLUSION

Congenital absence of ribs is a rare condition and the defect is generally unilateral. In severe cases, it can result in severe lung restriction and cor pulmonale.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Hollin S, Manjunath MA, Karaippa P, Prathap N. A rare case of congenital absence of ribs in a newborn baby. *Indian J Case Reports*. 2023;9(10):315-317.