

Isolated Hypoplastic left Rib Anomaly

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Abstract

Presentation

A male infant was delivered at 31+3 weeks for reduced fetal movements and intra-uterine growth restriction. He required CPAP from birth and had a chest Xray at 4 hours of age.

Diagnosis

The Xray showed a hypoplastic left 4th rib, more gracile than adjacent ribs. There was no associated vertebral anomaly.

Treatment

All other investigations including echo, renal ultrasound, ophthalmology assessment and microarray were normal.

Discussion

Congenital rib abnormalities can be found in up to 2% of the general population. Hypoplastic ribs are described when the lateral margin of the ribs is 4mm medial to a tangent line drawn along the lateral margins of adjacent ribs and represent one of the least common congenital rib abnormalities. Typically, they are identified on incidental imaging and tend to occur in isolation. Follow up is required if associated with vertebral abnormalities, otherwise no treatment is required. This is relevant when counselling parents with a postnatal incidental diagnosis.

Introduction

Congenital rib abnormalities can be found in up to 2% of the general population¹. Hypoplastic or short ribs are described when the lateral margin of the ribs is 4mm medial to a tangent line drawn along the lateral margins of adjacent ribs and represent one of the least common congenital rib abnormalities. Classically the first rib is affected and occurs more frequently on the right, bilaterally and left in descending order and with a slight female tendency².

Case Report

A male infant was delivered at 31+3 weeks via emergency lower segment Caesarean section for reduced fetal movements and intra-uterine growth restriction in the setting of a non-reassuring cardiotocograph. His mother, a 34 year old primigravida, was a non-smoker with an unremarkable medical history and had a normal anomaly scan at 22 weeks' gestation. He required CPAP from birth and had a chest Xray at 4 hours of age (figure 1) for a rising oxygen requirement and received a dose of surfactant via the Intubation – Surfactant – Extubation (INSURE) technique. The chest Xray showed 12 ribs on the right and 11 on the left and a hypoplastic left 4th rib, more gracile than adjacent ribs. There was no associated vertebral anomaly. All other investigations including echo, renal ultrasound, ophthalmology assessment and microarray were normal.

Discussion

Typically, they are identified on incidental imaging and tend to occur in isolation, however, may be associated with other malformations. Hypoplastic or short ribs have been described in association with skeletal dysplasia's including syndromes such as thanatophoric dysplasia, achondroplasia and Ellis-van Creveld syndrome as well as Jeune syndrome and other short-rib polydactyly syndromes³. Short rib polydactyly syndromes are a collection of rare autosomal recessively inherited lethal skeletal dysplasias that are usually diagnosed antenatally on ultrasound, that in addition to short ribs have micromelia, pre and post axial polydactyly and hypoplastic thorax and may have anomalies in multiple major organs. In one single centre study of rib anomalies hypoplastic rib anomalies were the least commonly seen, less frequent than fusion anomaly and bifid ribs. Of all patients 72% were found to have a vertebral anomaly. Interestingly in those without an associated vertebral anomaly a combination of either rib and cardiac defect alone or rib and renal defect alone was seen in one third of cases⁴.

This case highlights the importance of appropriate investigation in such a presentation to establish that the defect is indeed an isolated one given the associations described. Longer term follow up is required if associated vertebral abnormalities given potential to develop scoliosis. Otherwise no follow up is required if all investigations are negative. This case report is relevant when counselling parents with a postnatal incidental diagnosis of a hypoplastic rib.

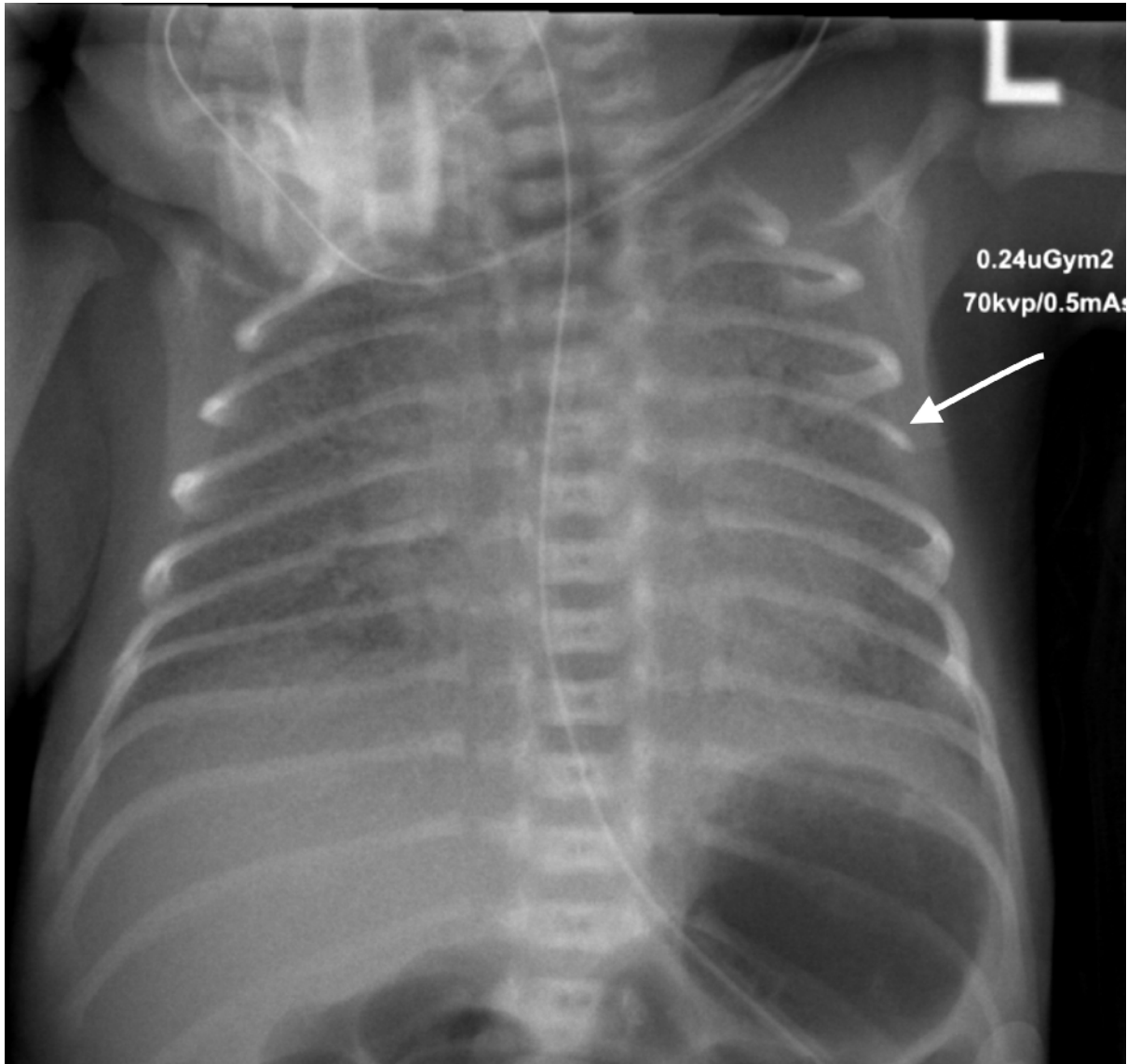


Image 1: Chest X-Ray of one day old 31 week with a hypoplastic left 4th rib indicated by white arrow.

Declarations of Conflict of Interest:

None declared.

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