An infant boy, the second child of healthy parents, was born at 35.5 weeks of gestation by cesarean delivery performed in emergency because of fetal bradycardia and polyhydramnios. At birth his weight was 2770 g (62nd percentile), length 48.3 cm (69th percentile), and head circumference 33.5 cm (64th percentile). Findings of a physical examination showed a broad forehead, a depressed nasal bridge, anteverted nares, a long and protruding philtrum, a high arched palate, retrognathia, joint contractures, and an umbilical hernia. The Apgar score was 6/8 at 1/5 minutes.

Because of progressive respiratory distress he required hospitalization and noninvasive support ventilation for the first 36 hours. At admission, a chest radiograph demonstrated a bell-shaped thorax with curved ribs and cupped anterior ends (Figure 1). These chest roentgenogram findings were further investigated by calculating the coat-hanger angle (CHA), which was 36° (Figure 2; available at www.jpeds.com). CHA refers to the average of the angles between the peak point of both sixth posterior ribs and the horizontal axis passing through their costovertebral articulations. If there is no peak point of the sixth posterior ribs, the center of the ribs is used instead. An upward angle is defined as +, and a downward angle as −. The horizontal axis is defined as a line passing through 2 points of both sixth costovertebral junctions.1,2 If this value exceeds 34° (CHA sign), in patients with suggestive dysmorphic features, a paternal uniparental disomy 14 syndrome, recently named Kagami-Ogata syndrome (KOS), should be suspected.2,3

KOS results in a unique phenotype characterized by facial abnormalities, abdominal wall defects, placentomegaly, polyhydramnios, and a small bell-shaped thorax with “coat-hanger” appearance of the ribs constituting its prominent pathognomonic feature.4,5 We performed a segregation study by microsatellite analysis polymorphisms using the leukocyte genomic DNA of the patient and parents, which confirmed our clinical suspicion.

There are several congenital disorders wherein thoracic hypoplasia is the sole radiological hallmark.6 The finding of a CHA sign is a well-characterized and simple feature that could suggest the diagnosis of KOS. This sign is described to be more severe than that seen in other genetic bone diseases and to persist from infancy through childhood/puberty. Because of a poor swallowing reflex, the patient required nasogastric feeding support until 3 weeks of age. He was discharged when he was 35 days of age. ■

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Figure 2. CHA calculation made via the formula: (angle A + angle B)/2. **Top,** Schematic overview of CHA calculation on a chest radiograph of a newborn without symptoms, followed by an ideogrammatic representation of the fourth to the seventh costovertebral joints (thoracic vertebrae and ribs); $\text{CHA} = 14^\circ$. **Bottom,** Schematic overview of CHA calculation on our patient’s chest radiograph followed by an ideogrammatic representation of the fourth to the seventh costovertebral joints (thoracic vertebrae and ribs); $\text{CHA} = 36^\circ$. 