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3 Late Preterm Infant with Respiratory Distress

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PRESENTATION

A 2.7-kg male neonate is delivered at 36 weeks' gestation by a 33-year-old gravida 1, para 1 woman via cesarean section. The cesarean delivery was indicated for nonreassuring fetal heart tones and arrest in the first stage of labor. The mother's pregnancy had been complicated by gestational diabetes (managed with metformin), chronic hypertension, and preeclampsia. The family history is not significant for any congenital cardiac defects or genetic syndromes. Fetal echocardiography suggested a coarctation of the aorta at 34 weeks of gestation. Fetal ultrasonography at 28 weeks' and 34 weeks' gestation did not show any anomalies.

After an uncomplicated delivery (Apgar scores of 8 and 9 at 1 and 5 minutes,

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respiratory distress, prompting reinitiation of CPAP. Subsequent imaging studies confirm the diagnosis (Fig 2).

DISCUSSION

Respiratory distress with bilious emesis in the neonate should prompt immediate evaluation and workup. Differential diagnosis in a 3-day-old late preterm neonate includes sepsis, respiratory distress syndrome, malrotation, volvulus, anatomic defects or variants including diaphragmatic hernias, and less commonly, necrotizing enterocolitis. For this neonate's presentation, chest radiography showed concern for loops of bowel in the left chest cavity (Fig 2). With subsequent chest ultrasonography and lateral table tilt chest radiography, the findings were most concerning for a congenital diaphragmatic hernia (CDH) (Fig 3).

Based on his prior studies and otherwise normal chest radiograph on the first day after birth, late-presenting CDH in the neonate was suspected. We suspected a hernia sac covered most of the defect, and gradual increasing abdominal pressures subsequently displaced portions of the confirmed bowels into the chest cavity.

AUTHOR DISCLOSURE Dr Rhine has disclosed that he owns stock in SonarMed, Nfant, and Novonate. Drs Kim and Arain have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

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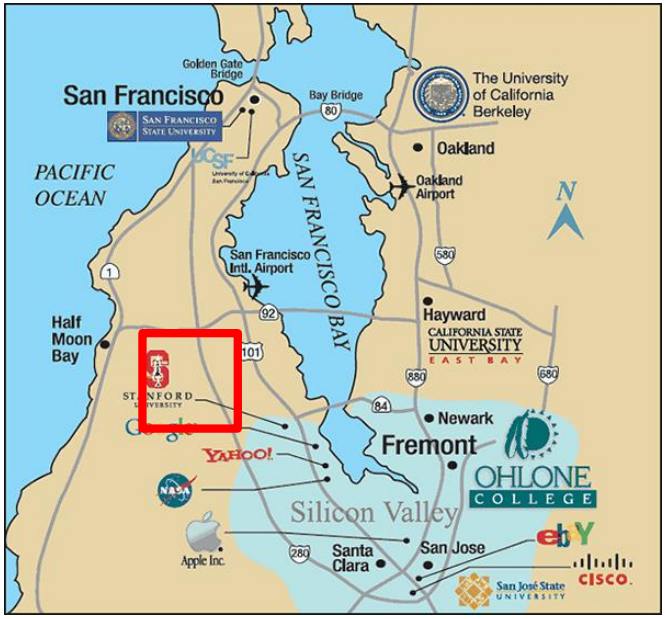

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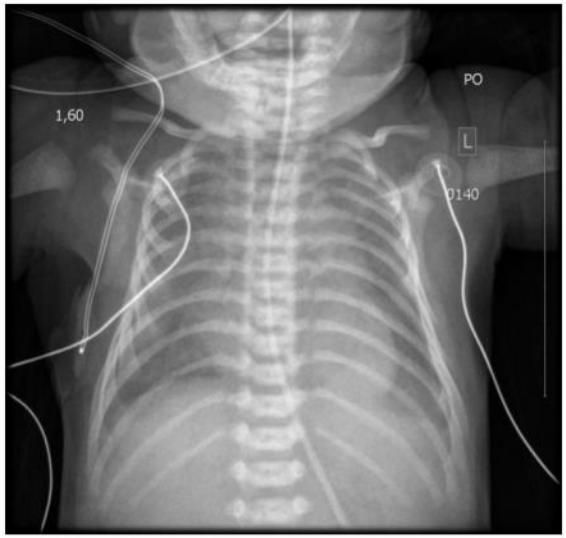


Figure 1. Normal anteroposterior chest radiograph first day after birth taken for respiratory distress.

PRESENTATION

A 2.7-kg male neonate is delivered at 36 weeks' gestation by a 33-year-old gravida 1, para 1 woman via cesarean section. The cesarean delivery was indicated for nonreassuring fetal heart tones and arrest in the first stage of labor. The mother's pregnancy had been complicated by gestational diabetes (managed with metformin), chronic hypertension, and preeclampsia. The family history is not significant for any congenital cardiac defects or genetic syndromes. Fetal echocardiography suggested a coarctation of the aorta at 34 weeks of gestation. Fetal ultrasonography at 28 weeks' and 34 weeks' gestation did not show any anomalies.

After an uncomplicated delivery (Apgar scores of 8 and 9 at 1 and 5 minutes, respectively), the infant develops mild respiratory distress, which requires continuous positive airway pressure (CPAP). He is admitted to the NICU for anticipatory screening for coarctation and management of his respiratory distress. Chest radiography performed 1 day after birth shows no significant signs of respiratory distress syndrome and normal chest anatomy, with no defects (Fig 1). His respiratory distress subsides within 1 day after birth and feeding is initiated on the same day. Postnatal evaluation, including 4 extremity blood pressure measurements, pre- and postductal saturations, and physical findings, are reassuring within the first 72 hours, though cardiac imaging is yet to be performed. Four days after birth, the neonate develops bilious emesis and respiratory distress, prompting reinitiation of CPAP. Subsequent imaging studies confirm the diagnosis (Fig 2).

DISCUSSION

Respiratory distress with bilious emesis in the neonate should prompt immediate evaluation and workup. Differential diagnosis in a 3-day-old late preterm neonate includes sepsis, respiratory distress syndrome, malrotation, volvulus, anatomic defects or variants including diaphragmatic hernias, and less commonly, necrotizing enterocolitis. For this neonate's presentation, chest radiography showed concern for loops of bowel in the left chest cavity (Fig 2). With subsequent chest ultrasonography and lateral table tilt chest radiography, the findings were most concerning for a congenital diaphragmatic hernia (CDH) (Fig 3).

Based on his prior studies and otherwise normal chest radiograph on the first day after birth, late-presenting CDH in the neonate was suspected. We suspected a hernia sac covered most of the defect, and gradual increasing abdominal pressures subsequently displaced portions of the confirmed bowels into the chest cavity.

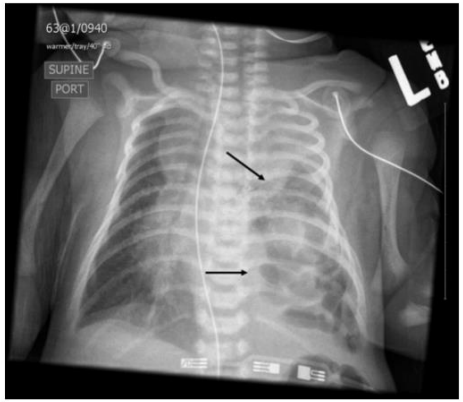


Figure 2. Anteroposterior chest radiograph demonstrating loops of bowel in the left pleural space (arrows).

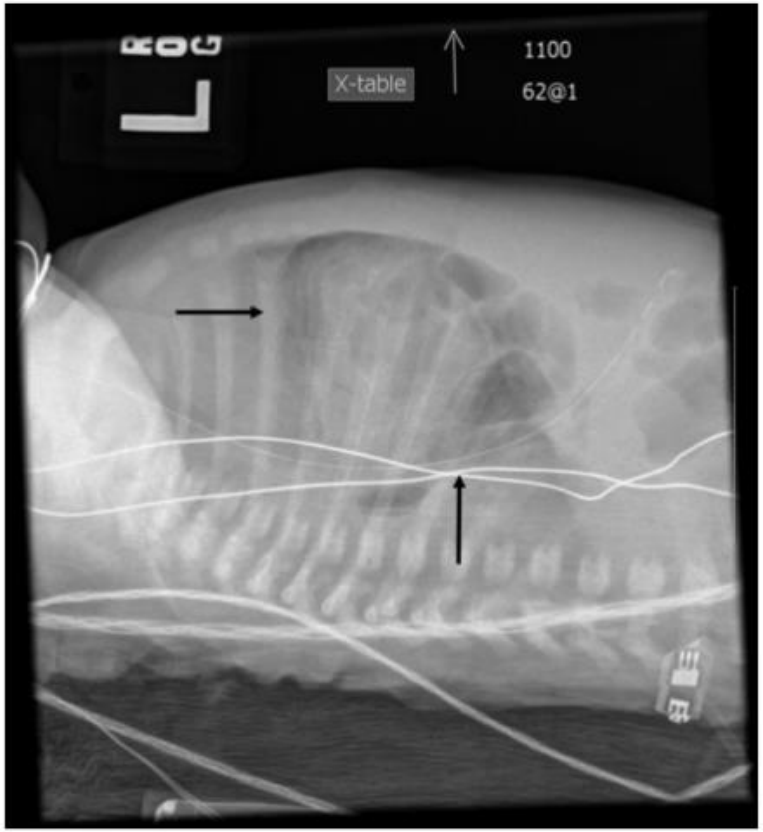
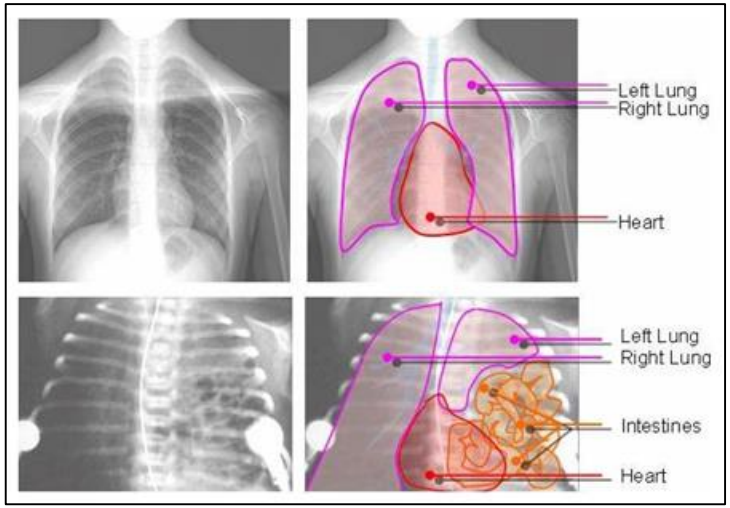


Figure 3. Lateral cross-table chest radiograph confirming loops of bowel in the left chest cavity (arrows).



CDH is an opening that forms a communication between the peritoneal and pleural cavities. CDH is one of the most common structural defects occurring in 1 to 4 in 10,000 births. (1)(2) If it occurs in utero, embryologic consequences, including lung hypoplasia and cardiac complications, can lead to poor postnatal outcomes, including death. Speculation on its pathogenesis is thought to be a failure in the embryologic development and closure of pleuroperitoneal folds early in gestation around 4 to 10 weeks. (3)(4)(5) There has not been a clear association with sex, but reports suggest that it occurs in boys more than in girls. (1) Approximately 2.5% to 25% of CDH cases present "late," which is defined as older than 30 days of age at detection. (5)(6)(7) Some retrospective studies speculate that the lower percentage is attributed to the improvement of prenatal screenings and treatment, while rural areas may report higher values because of limited resources. (7) Among these studies, data on the diagnosis of late CDH in the neonatal period are limited and a smaller fraction of the late-presentation groups may be included, because most of these cases may be asymptomatic. (6)(7)

CDH screening occurs at 24 weeks' gestation with prenatal ultrasonography (at least 50% of cases diagnosed), or on rare occasion using fetal magnetic resonance imaging. (8) Postnatal signs and symptoms that should raise suspicion include a scaphoid abdomen in combination with respiratory distress. A CDH can be detected postnatally with a chest radiograph that displays herniation of abdominal contents into the chest cavity. (8)

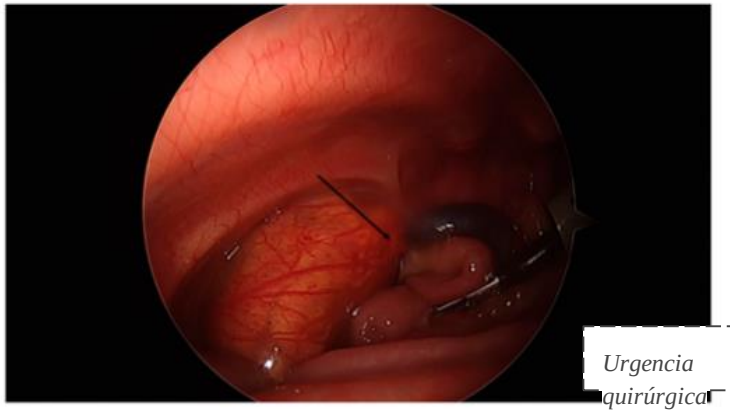


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Figure 4. Thorascopic view of the diaphgramatic hernia demonstrating small bowel loops (arrows) in the chest cavity.

A CDH is characterized based on the type of defect and its location. Left-sided hernias are more common, consisting of almost 85% of all reported CDH cases. Locations are labeled as posterolateral (Bochdalek), anterior (Morgagni), or central anterior (septum transversum of the Cantrell type). The defect can also vary within those respective locations and are identified by a letter classification system as described by Ackerman et al. (3) Posterolateral hernias are the most commonly reported type in literature. (4)

Management of CDH is a surgical urgency. Despite the good outcomes of CDH repair with delayed presentation in a neonate, missing the diagnosis could prove fatal. Herniation of the contents with fully developed lungs and vasculature can lead to respiratory distress and cardiac failure. CDH repairs involve thoracoscopic or minimally invasive surgery and the outcomes are usually favorable if detected early. Respiratory support in addition to bowel decompression is suggested to decrease intra-abdominal pressure and to limit the bowel dilation. Enteral nutritional support is withheld before operative repair, because the bowel location in the chest precludes enteral feeds. If prolonged medical resuscitation is anticipated, intravenous access and parenteral nutrition should be initiated.

Patient Course

The neonate underwent a thoracoscopic diaphragmatic hernia repair. Initial thoracoscopy confirmed a diaphragmatic hernia with the presence of small bowel loops and stomach in the thoracic cavity (Fig 4). Once gently reduced into the abdomen, a type A defect in the diaphragm was identified. In addition, there was a hernia sac encompassing the contents. After mobilization of the hernia sac, the diaphragmatic defect was repaired using interrupted permanent sutures. A pigtail catheter was left for chest drainage and the patient was taken back to the NICU for postoperative care. A repeat chest radiograph was obtained, which showed complete repair of the defect.

The neonate recovered well from the operation and he reached full feeds 12 days after birth and was placed on H2-blocker prophylaxis. He was discharged the same day and had no complications.

American Board of Pediatrics Neonatal-Perinatal Content Specifications

- Recognize the clinical features of extrapulmonary causes of respiratory distress.
- Recognize the imaging features of extrapulmonary causes of respiratory distress.
- Plan appropriate therapy for an infant with extrapulmonary causes of respiratory distress.

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