

Ultrasound

Clinical case study

Managing pediatric patients with GI duplication cysts and pneumatosis intestinalis

Background

Gastrointestinal (GI) duplication cysts are a rare congenital abnormality. The cysts can occur anywhere along the GI tract but are most common in the small bowel. While they can be diagnosed at any age from antenatal to adult life, the cysts often present in children within the first two years of life. GI duplication cysts often require surgical intervention and usually have uneventful post-operative recovery.^{1,2}

The presence of gas or air within the bowel wall is termed pneumatosis intestinalis (PI). PI in a neonate can result in serious morbidity, especially in patients with complex congenital heart disease or prematurity. PI is often seen in inflammatory bowel conditions such as necrotizing enterocolitis (NEC), ileo-colic intussusception with bowel ischemia, or venous occlusion of the bowel. PI has been coined the "circle sign" in ultrasound, as the bubbles of gas within the perimeter of the bowel wall give the appearance of a continuous echogenic ring.^{3,4} On X-ray, PI is sometimes difficult to distinguish from gas originating from fecal matter. As such, ultrasound can play an important role in clarifying the diagnosis. Early detection of PI in the population at risk for NEC can lead to improved survival rates.^{4,5}

Patient history

Prenatal imaging of a fetus from a dichorionic triamniotic triplet pregnancy showed an abdominal cystic mass in the right hemi-abdomen. Fetal MRI and limited OB ultrasound performed at 30 weeks gestation at our institution allowed us to identify a nearly 4 cm cystic mass in the right hemi-abdomen without bowel obstruction.

High-resolution ultrasound evaluation with the eL18-4 PureWave linear array transducer allowed for identification of three distinct layers: an inner hyperechoic layer corresponding to mucosa, a middle hypoechoic layer corresponding to the muscularis layer, and an external hyperechoic layer corresponding to serosa, which when seen together constitute the "gut signature" sign. The diagnosis of intestinal duplication cyst was considered highly likely, given the stratified appearance of the cyst wall. The main differential diagnosis was a choledochal cyst based on the location of the mass.

Findings and protocol

Triplets were delivered at 35 weeks gestation. A post-natal abdomen sonogram was performed at 19 days of age using the mC12-3 and eL18-4 PureWave transducers. Imaging suggested bowel obstruction with a 4.5 cm unilocular cyst in the right abdomen that once again demonstrated a gut signature sign.

Laparotomy was performed at 19 days of age. A GI duplication cyst was identified in association with small bowel volvulus and obstruction. The duplication cyst was excised, the small bowel volvulus detorsed and the surgery was completed with resection of the affected segment and ileo-ileo anastomosis. The surgery successfully removed the GI duplication cyst.

GI duplication cyst conclusion

It is crucial to be aware of and make a definitive diagnosis of this rare abnormality, as it can cause substantial complications if left untreated. The use of the eL18-4 and mC12-3 PureWave transducers for obstetrical prenatal and pediatric imaging aids the ability to reach a definitive diagnosis.

Transverse fetal abdomen images demonstrate cystic mass and "gut signature" sign of a cyst in the right abdomen.



Transverse view at fetal stomach level

Magnified view of cystic mass

Sagittal view of right upper quadrant



Ultrasound with high-frequency transducers is a highly accurate, fast, cost-effective, reproducible and non-ionizing tool that provides high-quality images for managing pediatric patients.

Continued findings and protocol

Two weeks later, the infant began vomiting and an abdominal radiograph was suspicious for, but not definitive of, PI. We performed 2D imaging with the eL18-4 PureWave transducer using a bowel preset. All four quadrants were evaluated in transverse and sagittal planes with this high-frequency transducer. Special emphasis was given to the bowel wall, liver and peritoneum to evaluate for air in the bowel wall, portal venous gas in the liver, and free fluid or collection within the peritoneum. Transverse bowel images demonstrated air in the bowel wall continuous with the "circle" sign. This was a definitive ultrasound sign of PI. The infant was treated successfully with conservative management, including bowel rest, antibiotics and fluids. The patient continued to be monitored and was eventually released after 29 days.

PI conclusion

It is crucial to be aware of and make a definitive diagnosis of this rare abnormality as substantial morbidity and even mortality can occur if left untreated. Using high-frequency transducers such as the eL18-4 PureWave transducer for pediatric imaging aids the ability to identify and image PI.

2D images demonstrate portal venous gas in liver. Transverse bowel images demonstrate air in bowel wall continuous with the circle sign. Note the bubbles present along the entire bowel circumference, including the non-dependent bowel wall. In cases of intraluminal air (i.e., air within the bowel lumen, not wall), the air tends to accumulate only along the non-dependent wall.



Transverse views of the liver



Trans LUQ-LLQ

Transverse view of the left upper quadrant and left lower quadrant Rither Services Servi

Trans LLQ

Transverse view of the left lower quadrant



About the author

Monique Riemann, RMDS, RVT is an ultrasound technologist at Phoenix Children's Hospital with over 30 years of experience in the field. Her registries include Abdomen, OB/GYN, Vascular, Neuro and Pediatric. After many years in a lead sonographer position she transitioned to a research position where she has helped to create ultrasound exams that were once radiation-based by utilizing the "Think Ultrasound First" motto. In 2020 she became a published author with her article on sonography of magnetically controlled growing rods.

Technology perspective

Ultrasound offers imaging without the drawbacks of the need for sedation, as with MR, or exposure to ionizing radiation, as with CT.⁶ The Philips ultimate ultrasound solution for pediatric assessment aids clinicians in reaching a definitive diagnosis.

nSIGHT Imaging architecture



This imaging architecture features a multi-stage precision beamformer with massive parallel processing.

- Improves frame rates in all imaging modes
- Improves beam focusing throughout the image to visualize superficial vessels as well as deeper veins
- Provides better penetration with resolution at depth

eL18-4 PureWave transducer

The versatile ultra-broadband eL18-4 transducer is designed for pediatric, small parts, breast, vascular and OB imaging.

- Elevation focusing in conjunction with azimuthal focusing enables thin-slice imaging for exceptional detail resolution and tissue uniformity from near-to-far depth of field
- Extraordinary imaging and depth-of-field performance
- Superb border delineation in superficial imaging and elevated tissue conspicuity

mC12-3 PureWave transducer

Designed for pediatric patients, the mC12-3 transducer offers 30% more penetration than the C8-5 transducer, providing elevated tissue conspicuity and a unique ergonomic design and rounded edges for superb patient comfort.



Additional reading

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