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The Journal of the American Osteopathic College of Radiology (JAOCR) is designed to provide practical up-to-date reviews of critical topics in radiology for practicing radiologists and radiology trainees. Each quarterly issue covers a particular radiology subspecialty and is composed of high quality review articles and case reports that highlight differential diagnoses and important teaching points.

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# Pediatric Radiology

Guest Editor: Bernard F. Laya, D.O.

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In This Issue

Bernard F. Laya, D.O.
Institute of Radiology, St. Luke’s Medical Center - Quezon City and Global City, Philippines

The field of medicine is constantly changing and as radiologists, it is very important to keep abreast with the current literature in order to be of better service to both our patients and referring clinicians. We are fortunate that every quarter for the last three years, we have been receiving a new issue of JAOCR to help us in this process. Since its introduction in 2012, our journal has gained tremendous support and following from the radiology community largely due to its education-focused format tackling pertinent, current, and must know topics in various subspecialties of radiology. Its open access, online format makes it even more appealing because it makes for a seamless search, appropriate for a busy radiology practice.

In this Pediatric Radiology issue, the authors present the current imaging trend on some of the more frequently encountered disease processes in children, ranging from intestinal obstruction, respiratory distress, and abdominal and intracranial masses. In the first review article titled, “Patterns of microcolon: imaging strategies for diagnosis of lower intestinal obstruction in neonates”, I collaborated with my colleagues from St. Luke’s Medical Center; Nathan Concepcion, Mariaem Andres, and Rafael Dizon to present a practical approach to diagnosis based on barium enema examination. In the second review article, “Imaging of neonatal lung disease”, Jonathan Wood and Linda Thomas skillfully present an important topic which some radiology residents often find challenging. Our colleagues from Dayton Children’s Hospital; Dawn Light, Francis Pianki, and Elizabeth Ey wrote a case report providing us a systematic approach to imaging diagnosis of abdominal masses in infants. The case report written by Matthew Minor and William O’Brien offers a detailed assessment in differentiating pineal region masses in children. The “Viewbox” articles on medulloblastoma by Valerie Hostetler and Claire C. Widule, as well as the article on posterior reversible leukoencephalopathy syndrome (PRES) written by Betsy Cheng and myself are classic examples of such intracranial abnormalities.

I would like to thank the JAOCR and Dr. William O’Brien for this honor and opportunity to serve the radiology community as a Guest Editor. Dr. O’Brien has been a very supportive mentor throughout the process and I am certain that JAOCR will continue to flourish under his watchful guidance. I would also like to thank all of the authors in this issue for their hard work and outstanding contributions. My sincere gratitude also goes to my colleagues, fellows, and residents at St. Luke’s Medical Center, Philippines for their unwavering trust and support. Not to forget, I would like to thank my family for their unconditional love, support, and understanding.

It is with great pride that I present to you the current pediatric imaging issue of JAOCR. I am certain that these articles will help you in your practice and I hope you enjoy reading them.

“Study while others are sleeping; work while others are loafing; prepare while others are playing; and dream while others are wishing.”

-William Arthur Ward
Introduction

Microcolon is a radiographic feature of low intestinal obstruction that results from intrauterine underutilization or what is termed “unused colon,” including entities in which meconium is not passed through the colon during in utero development. Prenatal and perinatal insults causing microcolon represent myriad of etiologies, which include hypoperfusion, as well as factors associated with dysmotility and stasis. Postnatal surgical procedures may also be responsible for this radiologic feature. Disease entities manifesting as microcolon include meconium ileus, small left colon syndrome, small intestinal and colonic atresia, and Hirschsprung disease.

Clinical and radiological features are important in the diagnosis of the disease but they are not pathognomonic. Radiography is the initial imaging study of choice for detection of low intestinal obstruction, which is non-specific; hence, there is a need for further evaluation through contrast enema study. In meconium ileus and small left colon syndrome, contrast enema is not only diagnostic but also therapeutic. In cases of Hirschsprung disease, rectal biopsy is required to determine the aganglionic segment responsible for the obstruction. Some patients with colonic atresia may also warrant rectal biopsy if contrast enema is equivocal. The goal of this article is to present a systematic radiologic approach to the diagnosis of microcolon, describe typical imaging characteristics, and discuss associated disease entities.

Background

A neonate presenting with distended abdomen requires prompt assessment by the clinician and systematic investigation by the radiologist. Clinically, neonates with abdominal distention may have accompanying symptoms of failure to pass meconium in the first 24-48 hours of life. This is highly presumptive of intestinal obstruction.

Microcolon, also termed as “unused colon,” is defined as a colon of abnormally small caliber but of normal length. There is no definite or absolute standard of measurement for this entity, although some authors state that a colonic segment with a caliber less than the interpedicular space of the L1 vertebra is considered microcolon. It has also been defined as a luminal diameter less than the height of an upper lumbar vertebral body. Microcolon is an important radiologic feature in neonates with bowel obstruction, particularly the distal portion of the bowel. This feature is best appreciated on fluoroscopic contrast enema studies. The colon is small because it is essentially unused.

There are variations in the radiologic pattern of microcolon, ranging from focal to long segment narrowing or even diffuse pattern. Whether the entire colon or a focal segment is affected, the distal colon is often involved. In light of its name, unused colon occurs because the intestinal secretions that make up the meconium in the fetal gastrointestinal tract do not reach the colon. It is due to obstruction in the low intestinal segments, anywhere from distal ileum to proximal colon or the entire colon itself. If the obstruction seats in the proximal intestinal tract, there is a chance for secretions to form and eventually reach the colon. In low gastrointestinal obstruction, when there is no transit of meconium into the colonic lumen, there is no stimulus for growth. Low gastrointestinal obstruction includes disorders such as meconium ileus, jejuno-ileal and/or colonic atresia, Hirschsprung, and small left neonatal colon or previously called “meconium plug” syndrome.

Radiographs of the abdomen and contrast enema have long been used in the investigation and accurate assessment of neonates with suspected lower intestinal obstruction. This article will discuss the
clinical presentation, imaging appearance, surgical correlation, and even treatment of various lower intestinal disease entities in the newborn with radiographic patterns of microcolon.

Radiologic Investigation

Radiographs

Plain radiograph of the abdomen, obtained in anteroposterior (AP) and lateral views is the initial imaging modality of choice in neonates presenting with abdominal distention to evaluate the possibility of obstruction. The timing when the radiographs are obtained is important, because taking it too soon after delivery may not allow enough time for air to make its way through the unobstructed portions of the intestine and could affect interpretation.

Upon initial inspection of the radiographs, it is important to rule out ominous signs which would require emergent surgical intervention. Such signs include pneumatosis intestinalis, portal venous gas, and pneumoperitoneum (Fig. 1). In neonates, the small and large bowel usually cannot be distinctly distinguished because the intestinal loops are featureless and sometimes do not lie in the predictable anatomical locations. However, the precise level of obstruction may be identified based upon the gas content and location of the air-filled bowel loops. A high intestinal obstruction pattern usually shows a few scattered air-filled loops in the upper abdomen (Fig. 2A). Low gastrointestinal obstruction generally has

Figure 1.
Ominous signs in abdominal radiographs. Frontal abdominal radiograph of a neonate (A) demonstrates pneumatosis intestinalis in the right hemiabdomen (white arrows) and portal venous gas in the liver (black arrows). Supine abdominal radiograph from another neonate (B) shows free air (black arrow). There is also air outlining both the external and luminal surface of the intestinal loops (“rigler sign”) indicative of free intraperitoneal air.

Figure 2.
Upper versus lower obstruction. Abdominal radiograph of a neonate (A) shows the typical “double bubble” sign of duodenal atresia, an example of upper intestinal obstruction. AP abdominal radiograph in a different patient (B) reveals multiple distended intestinal loops with paucity of air in the rectal region, indicative of lower intestinal obstruction.
Microcolon, Laya et al

Possible lower intestinal obstruction, it is recommended to use water-soluble contrast media, as there may be potential for bowel perforation or electrolyte imbalance.

In interpreting the contrast enema study, a pattern-based approach may be used in coming up with a differential diagnosis in a neonate with lower intestinal obstruction. There are four patterns that may be encountered in contrast enema: 1) Normal study; 2) Microcolon, where the luminal caliber of the entire colon is small and non-distensible; 3) Short microcolon, where the colon is small in caliber but terminates at any point before the cecum; and 4) Colonic caliber change, where there is a transition from small or normal-caliber colon distally and a distended colon proximally (Fig. 3). Each of these patterns offers a limited differential diagnosis, which accounts for about 98% of cases, and allows appropriate management decisions (Table 1).

![Figure 3. Four patterns on contrast enema for neonates suspected of lower intestinal obstruction.](image)

Normal (A), Microcolon (B), Short Microcolon (C), and Colonic caliber change (D).

Site of involvement if the level of obstruction is at the ileum or at any segment of the colon. In performing a contrast enema, a small soft catheter (8 French) is inserted into the rectum just above the anal verge without inflating the balloon tip. Not inserting it too high/deep and not inflating the balloon will aid in identification of a low transition point in Hirschsprung. The catheter is secured in place by taping it well at the perineum.

Infusion of contrast into the anus should be started with the patient in lateral position via gravity drip, minimizing the degree of pressure on the possibly diseased colon. Keen observation is important during the fluoroscopic evaluation, and selected images should be obtained as contrast is infused into the colon with special attention to the recto-sigmoid area and other regions with abnormal luminal caliber. If the colonic abnormality is immediately identified, no additional contrast filling proximal to the obstruction is recommended. If the entire colon is small in caliber, an attempt to reflux the contrast medium into the terminal ileum must be done. In neonates with possible lower intestinal obstruction, it is recommended to use water-soluble contrast media, as there may be potential for bowel perforation or electrolyte imbalance.

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<th>Patterns</th>
<th>Description</th>
<th>Differential Diagnosis</th>
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<td>Normal caliber and length of the colon.</td>
<td>1. No Obstruction</td>
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<td></td>
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<td>2. Hirschsprung Disease affecting the distal segment</td>
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<td></td>
<td></td>
<td>3. Total Colonic Aganglionosis (85%)</td>
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<td>Microcolon</td>
<td>Normal length but small luminal caliber of the colon, less than the interpedicular distance of lumbar vertebrae.</td>
<td>1. Meconium ileus</td>
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<td>2. Jejuno-ileal Atresia</td>
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<tr>
<td></td>
<td></td>
<td>3. Total Colonic Aganglionosis</td>
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<tr>
<td>Short Microcolon</td>
<td>Small luminal caliber of the colon that terminates at any point before the cecum.</td>
<td>1. Colonic Atresia</td>
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<tr>
<td>Colonic Caliber Change</td>
<td>Transition from small or normal-caliber colon distally and a distended colon proximally.</td>
<td>1. Small Left Colon Syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Hirschsprung Disease</td>
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Table 1. Contrast enema patterns, description and differential diagnosis.
Ultrasound

Post-natal ultrasound of the abdomen to rule out lower intestinal obstruction is not routinely performed, but it can be useful for meconium ileus and ileal atresia. Sonographic images of dilated bowel loops in meconium ileus are filled with echogenic material, while the loops in atresia are fluid-filled. It can also be used to assess other causes of lower intestinal obstruction and possible complications. Although ultrasound is mentioned as an imaging technique, this article focuses on the utility of radiographs and fluoroscopic contrast enema for the evaluation of microcolon.

Specific Disease Entities

Meconium Ileus

Meconium ileus (MI) is a functional low intestinal tract obstruction which involves the terminal ileum. MI is the earliest clinical manifestation of cystic fibrosis (CF), occurring in up to 15-20% of patients with CF. Conversely, greater than 95% of patients with simple MI have CF. Patients with cystic fibrosis have malfunctioning sodium-chloride pump which decreases the lubricating property of the intestines creating thick, viscid mucus. As a result, inspissated meconium blocks the distal ileum. MI makes up approximately 20% of low intestinal obstructions. The incidence in the United States is approximately 1 in 3000 live births per year. MI may be either simple or complicated, each with similar frequency. The simple form begins in utero where the thickened meconium obstructs the mid-ileum with resultant proximal dilatation, bowel wall thickening, and congestion. In complicated MI, the thick meconium obstruction leads to complications that include volvulus, atresia, necrosis, perforation, meconium peritonitis, and meconium pseudocyst formation (which may calcify).

Conventional abdominal radiographs show multiple dilated bowel loops with the meconium having a ground glass or soap-bubble appearance. There is absent to scant air-fluid levels, which is highly indicative of this type of low intestinal obstruction. However, the presence of air-fluid levels does not completely exclude this diagnosis. Dilated small intestines are identified on ultrasound with distinct echogenic material intraluminally, representing the thick meconium. Patients without a genetic predisposition are at low risk for MI, while those with genetic predisposition are at high risk for having MI. The fluoroscopic procedure of choice for its diagnosis is contrast enema in which reflux of contrast into the ileum is recommended to demonstrate the microcolon with a collapsed, meconium-filled, distal ileal segment (Fig. 4).

Water-soluble contrast is performed and serves a dual purpose, being both diagnostic and therapeutic, since the contrast loosens the obstructing concretions of meconium. The microcolon in most cases will eventually return to normal caliber. In certain cases,
Jejuno-ileal Atresia

Just like other atresias of the intestinal tract, jejuno-ileal atresia (JIA) is thought to be caused by a prenatal vascular event resulting in ischemic obliteration of the intestinal lumen. Atresia of the jejunum and ileum are approximately equally distributed between the two anatomic regions. Loss of mesentery depends upon the length of the ischemic intestine and the non-viable intestine may disappear completely or may remain as a fibrous band. Multiple atresias resulting in segmentation may also be seen. The incidence of JIA is approximately 1 in 3000-5000 live births and affects both boys and girls equally. Approximately 1 in 3 infants is premature. Familial cases of intestinal atresias are rarely reported; most cases are sporadic.

Clinical presentation of JIA is variable, depending primarily on the anatomic location of the obstruction. A very proximal obstruction results in a scaphoid abdomen and bilious emesis, whereas a more distal ileal atresia can lead to massive abdominal distention which may be progressive. Failure to pass meconium is common. After birth, a neonate is unable to tolerate feeds and vomiting ensues, leading to rapid electrolyte derangement and dehydration.

Abdominal radiographs in JIA usually show multiple dilated, air-filled intestinal loops typical of low intestinal obstruction. Fluoroscopic contrast enema evaluation shows complete microcolon from the rectum all the way to the cecum. It is important to reflux the contrast media from the cecum into the terminal ileum to distinguish atresia from meconium ileus. Termination of the contrast in a blind-ending ileal loop (Fig. 5) is compatible with ileal atresia, in comparison to meconium ileus where the terminal ileum is filled with inspissated meconium.

Initial treatment for JIA consists of nasogastric decompression, fluid resuscitation, and broad-spectrum antibiotics. Operative repair is usually not emergent (in uncomplicated cases) but should proceed expeditiously. Surgical management is based on the location of the lesion, anatomic findings, associated conditions (malrotation, volvulus, or multiple atresias) noted at operation, and the length of the remaining intestine. The current survival rate is greater than 90%.

Atresia

Atresia is believed to be due to a mesenteric ischemic insult in utero resulting in a structural obstruction. Other proposed theories include failure of recanalization, intestinal perforation, drugs, and environmental factors. Contributing factors may include maternal smoking during pregnancy. Various types of intestinal atresia characterized by morphology are described on Table 2. Structural obstruction due to atresia requires surgical management.

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<tr>
<th>Type</th>
<th>Percentage</th>
<th>Pathologic Description</th>
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| I    | 23%        | • Transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel.  
            • The bowel is usually of normal length. |
| II   | 10%        | • Involves two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact. |
| IIIA | 15%        | • Similar to type II, but with a mesenteric defect.  
            • Bowel length may be foreshortened. |
| IIIB | 11-22%     | • Also known as “apple peel” deformity  
            • Consists of a proximal jejunal atresia, often with malrotation.  
            • Absence of most of the mesentery.  
            • Varying length of ileum surviving on perfusion from retrograde flow along a single arterial supply. |
| IV   | 25%        | • Multiple atresia of types I, II, and III  
            • Like a “string of sausages”  
            • Bowel length is always reduced  
            • Terminal ileum, as in type III, is usually spared. |

Table 2. Morphological types of intestinal atresia.
Colonic Atresia

Colonic atresia is a rare cause of intestinal obstruction with an incidence of 1 in 20,000 live births and comprises approximately 1.8-15% of intestinal atresias.\textsuperscript{10,15} Mesenteric ischemic vascular insult remains the primary etiology. The classification of intestinal atresias also applies to colonic atresia.\textsuperscript{13,15-16} Colonic atresia occurs in descending order of frequency at the sigmoid, splenic flexure, hepatic flexure, and ascending colon, respectively.\textsuperscript{17}

Newborns with colonic atresia usually present with progressive abdominal distension, bilious emesis, and failure to pass meconium. Abdominal radiographs demonstrate a distal bowel obstruction (multiple dilated bowel loops with air-fluid levels). A single markedly dilated loop with a large fluid level is often more indicative of atresia (Fig. 6A).\textsuperscript{13,15} However, due to the many variations of atresia, radiographic findings are diverse, and these findings are not absolute. Definitive diagnosis is suggested following a contrast enema which demonstrates a microcolon that terminates blindly at the point of colonic atresia (Fig. 6B and C).\textsuperscript{10}

Initial management of colonic atresia involves appropriate fluid resuscitation and close observation of fluid and electrolyte balance. Urgent surgical intervention is needed, because this anomaly has a higher risk of perforation (10% incidence) than seen in other intestinal atresias.\textsuperscript{14} Multiple atresias should always be excluded. A period of parenteral nutrition may be required until oral or enteral feeding is established. Most patients do well post-operatively with a survival rate of 90-95%.\textsuperscript{14,18} Rectal biopsy may be done if patients treated for colonic atresia manifest with delayed return of gut function, because of an established association of bowel atresia and Hirschsprung disease.\textsuperscript{19}

Hirschsprung Disease

Hirschsprung disease (HD) is a congenital bowel motility disorder that occurs in approximately 1 in 5000 live births.\textsuperscript{20} It is a form of functional intestinal obstruction characterized by failure of craniocaudal migration of ganglion cells to the submucosal (Meissner’s plexus) and intermuscular (Auerbach’s plexus) layers, resulting in upstream obstruction.\textsuperscript{3-4,21} HD is common in boys (81.7%). In the majority of cases, recto-sigmoid involvement is seen.\textsuperscript{22} The aganglionic segment shows failure to distend normally, resulting in a functional obstruction with proximal bowel dilatation and abnormal stool passage. Approximately 80% of patients with HD have short-segment distal aganglionosis; 10-15% have long segment involvement; and 5-13% have total colonic involvement.\textsuperscript{6,22-24} Children with HD are unable to pass meconium in the first 24 hours of life and show progressive abdominal distension.

Abdominal radiographs show signs of lower intestinal obstruction with variable abdominal bowel gas, bowel distention, and air-fluid levels. These radiographic findings are nonspecific; hence, barium enema must be performed. Characteristic radiologic
findings of HD on a contrast enema study include an abnormal rectosigmoid ratio of less than 1 (transverse diameter of the sigmoid is larger than the rectum on the lateral view) (Fig. 7), a transition zone of colonic narrowing, irregular contractions in the region of aganglionosis, and retained contrast material on delayed radiographs. It is important to note that the level of colonic caliber transition (radiographic transition) does not necessarily correspond to the surgical transition point. Additionally, delayed evacuation of contrast over 24 hours is not a specific sign of HD, and evacuation may even be normal. Contrast enema may be misleading if patients with HD also have meconium plug in colon. The overall sensitivity and specificity of contrast enema study for the diagnosis of HD is 65-80% and 66-100%, respectively. Thus, contrast enema maybe normal in some patients with HD and rectal biopsy is required in a neonate with clinical signs and symptoms suspicious for HD.

The current gold standard in the diagnostic confirmation of HD is histopathology based on rectal suction biopsy that shows absence of ganglion cells in the submucosa and increased acetylcholinesterase (AChE) activity in the lamina propria. The sensitivity and specificity of rectal suction biopsy are reported to be 97-100% and 99-100%, respectively.

Total colonic aganglionosis (TCA) is a rare form of HD affecting the total colon and distal 30-50 cm of the terminal ileum. Although short segment HD has no racial predilection, total colonic disease is more common in Caucasians; it is associated with trisomy 21, hydronephrosis, and dysplastic kidneys. TCA approaches an even distribution between boys and girls compared to short segment HD that has a male predilection. On contrast enema, TCA may have a normal appearance (85%), but may also demonstrate a microcolon (Fig. 8) or foreshortened “question mark” appearance of the colon. The classic question mark

Figure 7.
Hirschsprung disease.
Abdominal radiograph (A) reveals multiple dilated intestinal loops indicative of lower intestinal obstruction. Lateral (B) and frontal (C) images following contrast enema demonstrate a small caliber rectum compared to the sigmoid. The radiographic transition zone (arrows) is persistent on both views.
shape of the colon is observed in only 18% of children. Additionally, the transition zone and rectosigmoid index ratio are not reliable signs in TCA.

With timely diagnosis and recent advances in surgical management, most affected children can lead a normal and productive life. However, delayed diagnosis of HD beyond 1 week after birth significantly increases the risk of serious complications, which include Hirschsprung-associated enterocolitis, severe dehydration, sepsis, and even shock. These risks are higher in TCA compared to short segment HD.

### Small Left Colon Syndrome

Small left colon syndrome has been previously referred to as meconium plug syndrome, functional immaturity of the colon, and colon inertia of prematurity. This condition was first described as meconium plug syndrome in 1956 as “intestinal obstruction due to the inability of the colon to rid itself of the meconium residue in fetal life.” The location of the inspissated meconium in left colon is defined as the meconium plug. The exact etiology is unknown, but it tends to be self-limited and associated with immature myenteric plexus ganglia.
The incidence of meconium plug syndrome is estimated at approximately 1 in 500 live births. It is the most common cause of intestinal obstruction in offsprings of diabetic mothers, with maternal diabetes associated in 40-50% of the published cases. A minority of small left colon syndrome is associated with maternal magnesium sulfate administration for pre-eclampsia. It can be clinically difficult to distinguish this entity from a completely unrelated meconium ileus; despite prior nomenclature, small left colon syndrome has no association with cystic fibrosis. Meconium plugs found on contrast enema are associated with a 13% incidence of Hirschsprung disease.

Conventional radiographs of the abdomen show distal bowel obstruction. Air-fluid levels are typically absent in the first 48 hours and “soap-bubble” meconium may be seen in the collapsed left colon. Contrast enema shows a relatively normal rectum with small-caliber left colon containing multiple filling defects compatible with inspissated meconium. There is abrupt change of luminal caliber from a narrowed descending colon to the normal-sized splenic flexure and entire proximal colon (Fig. 9).

Management of this syndrome is largely supportive, since it typically improves following the contrast enema that is used to diagnose it. The clinical condition of most neonates improves rapidly with excellent outcomes following water-soluble enema.

Conclusion

Anomalies resulting in lower intestinal obstruction presenting as microcolon in neonates are not uncommon. The spectrum of abnormalities and symptoms is diverse, ranging from mild, self-limited conditions to complete intestinal obstruction requiring surgical intervention. Imaging evaluation plays an important role in the diagnosis and appropriate, timely intervention, which is aimed at preserving the child’s intestinal integrity and function. An understanding of proper selection of imaging modalities, use of optimal imaging techniques, and knowledge of characteristic imaging appearances of various causes of lower intestinal obstruction will enable an accurate diagnosis and optimize pediatric patient management.

References

Neonatal ICU chest radiographs are one of the most common pediatric radiology examinations performed. As modern medicine has advanced, the lower age limit of viability has continued to decrease. Nowadays, it is not uncommon for 23 week old infants to survive. Although there are many complications associated with prematurity, to include necrotizing enterocolitis, intracranial hemorrhage, and sepsis, the most common cause of neonatal morbidity and mortality remains lung disease. This article describes the pathology and radiographic findings of some of the most common lung disease encountered in neonates.

**Transient Tachypnea of the Newborn**

Transient tachypnea of the newborn (TTN), also referred to as retained fetal lung fluid, wet lung disease, or transient respiratory distress, is caused by prolonged clearance of fetal lung fluid. Fetal lung fluid is not the same as amniotic fluid; rather it represents an ultrafiltrate of the fetal plasma. Symptoms of TTN include mild to moderate respiratory distress which presents at birth but may be delayed up to 6 hours. The symptoms typically peak within 36 hours after delivery and resolve by 72 hours.

Common risk factors of TTN include precipitous deliveries and cesarean sections where it is thought that retained fluid is not fully expelled from the neonate's lungs as would occur during a normal vaginal delivery. Normally, 35% of fetal lung fluid is cleared in the first few days prior to birth secondary to increased gene expression for an epithelial sodium (Na+) channel. The rest is cleared by labor and postnatally during crying and breathing. Other risk factors include prematurity, maternal diabetes, hydrops and other forms of hypervolemia, maternal sedation, and a history of maternal smoking.

In vivo experiments demonstrate that the poor fluid absorption may be explained by a poorly developed epithelial Na+ transport protein. In utero, the fetal lung epithelium secretes chloride (Cl−) and fluid. Late in gestation, the lung epithelium develops Na+ proteins to absorb the fetal lung fluid by responding to increase catecholamines and glucocorticoids. It is thought that infants with TTN have mature surfactant and poorly developed respiratory epithelial transport proteins, as opposed to neonatal respiratory distress syndrome (NRDS) where both surfactant pathways and Na+ transport proteins are deficient.

Radiographic findings of TTN often include hyperinflated lungs and retained fluid within the alveoli and interstitium, to include pleural and fissural fluid, as well as increased perihilar interstitial markings (Figs. 1 and 2). Severe cases may show alveolar opacities from the retained fluid. Radiographic findings can be similar to heart failure, although without marked cardiac silhouette enlargement. Imaging findings of TTN typically improve within 24 hours as...
the excess fetal lung fluid is either absorbed or expelled.

Treatment of TTN is typically supportive with oxygen and maintenance of body temperature. As the fluid clears, the lung parenchyma may develop a reticulonodular appearance on radiographs. The fluid typically clears superiorly to inferiorly and peripherally to centrally, although this may be variable given patient positioning.

**Meconium Aspiration Syndrome**

Meconium aspiration syndrome (MAS) is the most common cause of respiratory distress in the term or post-term neonate. Meconium is the earliest stool in an infant, and its components include epithelial cells, mucus, amniotic fluid, bile, blood, and lipids. Meconium was once thought to be sterile, although research has shown that approximately half of meconium is populated by *Escherichia coli* and the remaining half by lactic acid producing bacteria, such as *Lactobacillus*.³

Most cases of meconium aspiration occur during the distress of labor; however, in utero meconium passage and aspiration can occur secondary to fetal stressors, such as hypoxia and sepsis. These stressful events cause a vagal response by the fetus, leading to defecation.

Clinically, infants may present with cyanosis, tachypnea, and tachycardia. Signs of respiratory distress are invariably present, including intercostal retractions and nasal flaring. If the neonate defecated in utero, ingestion of meconium within the amniotic fluid can result in a yellow or greenish appearance to the skin, nails, and urine.

Aspiration of meconium results in airway obstruction with a ball-valve mechanism, chemical pneumonitis, and inactivation of surfactant by the bile salts, causing secondary surfactant deficiency.⁴ This complex pathophysiology results in a wide range of radiographic manifestations of the disease.

The most common radiographic finding is pulmonary hyperinflation secondary to the ball-valve mechanism of air trapping. Air trapping, in combination with the chemical pneumonitis, results in barotrauma which may cause pneumothoraces, pneumomediastinum, and pulmonary interstitial emphysema (*Fig. 3*). Other radiographic findings include perihilar ropey opacities and interspersed areas of atelectasis (*Fig. 4*). Pleural effusions can be seen but are uncommon.
gestations, oligohydramnios, and maternal diabetes. Maternal diabetes is thought to cause fetal hyperinsulinemia which interferes with surfactant biosynthesis, leading to NRDS. Boys and Caucasian babies are also at increased risk, for unknown reasons.

Surfactant is produced in the endoplasmic reticulum of type II pneumocytes, which are found in the alveolar walls. The surfactant is transported to the surface of the pneumocyte where it is combined with surfactant apoproteins on the surface to form a lipid monolayer. The surfactant layer reduces the surface tension and allows the alveoli to more easily expand. If the type II pneumocytes are not mature at the time of birth, surfactant deficiency occurs. The collapsed alveoli result in decreased oxygenation, causing an increase in the pulmonary vascular resistance. This in turn increases right to left shunting through a patent ductus arteriosus, which usually does not close in the setting of prematurity and low blood oxygenation. The increased shunting through the PDA exacerbates the infant’s hypoxia.\(^6\)

The term hyaline membrane disease is derived from the appearance of hyaline membranes in the bronchiole walls. The hyaline membranes, which contain fibrin, mucin, and necrotic alveolar cells, are a byproduct of prolonged alveolar collapse. The lecithin to sphingomyelin ratio in the amniotic fluid is

Meconium aspiration syndrome can result in persistent pulmonary hypertension of the newborn. Treatment includes endotracheal intubation to facilitate suctioning below the vocal cords, administration of surfactant to replace the surfactant inactivated by bile salts, and prophylactic antibiotics. The radiographic manifestations usually resolve by 48 hours, although may take weeks if the meconium has a lower water content.

Meconium aspiration results in significant morbidity and requires extensive treatment. Singh, et al. studied 7,518 neonates with the diagnosis of meconium aspiration syndrome. 9% of the patients required ICU admission, 2.4% required transfer to another NICU for convalescent care, and 1.2% died.\(^5\)

**Neonatal Respiratory Distress Syndrome**

Neonatal respiratory distress syndrome (NRDS) is the clinical term used to describe surfactant deficiency. It is also referred to as lung disease of prematurity. The term hyaline membrane disease is a histologic term and describes a byproduct of the disease.

The incidence of NRDS is approximately 6 in 1000 births.\(^6\) Risk factors include prematurity, multiple

![Figure 4. Meconium aspiration with perihilar opacities. Portable chest radiograph in a newborn infant with history of meconium aspiration shows rope-like perihilar opacities.](image)

![Figure 5. Classic appearance of NRDS. Portable chest radiograph demonstrate decreased lung volumes and granular opacities which are more confluent centrally, resulting in air bronchograms and effacement of the pulmonary vasculature. Patient is intubated with the ETT tip at the carina.](image)
frequently used as a marker of fetal lung maturity. Fetal lung fluid flows into the amniotic fluid throughout gestation. At approximately 32 to 33 weeks of gestational age the lecithin content rapidly increases, indicating maturing fetal lungs and production of surfactant by type II pneumocytes.

Radiographic findings in the setting of NRDS include stigmata of prematurity, to include a bell-shaped thorax and absence of humeral head ossification centers. The classic pattern of NRDS includes bilateral and symmetric granular opacities, air bronchograms, effacement of the pulmonary vasculature, and decreased lung volumes (Fig. 5). The classic appearance of NRDS is less commonly seen given the early administration of surfactant, frequently before baseline imaging is obtained, and tendency for early intubation. This results in appearances that can mimic meconium aspiration syndrome or neonatal pneumonia with increased lung volumes and focal areas of consolidation. Cystic lucencies from expanded parenchyma and asymmetric aeration can resemble PIE. If only a single lung receives surfactant, it may asymmetrically expand and cause mediastinal shift.

Dinger, et al. reported radiographic findings in 110 neonates after treatment with surfactant. Uniform improvement was shown in 38%, asymmetric improvement in 35%, and no improvement in 10%. They found that the asymmetric improvement was most pronounced in the middle and upper right lung fields. 7

Symptoms, which usually present in the first hours of life, include expiratory grunting and nasal flaring with possible cyanosis. Treatment includes exogenous surfactant administration and mechanical ventilation. If these therapies are not sufficient, extracorporeal membrane oxygenation (ECMO) can be used to allow the lungs to mature. Complications include barotrauma from mechanical ventilation and oxygen toxicity of the pulmonary parenchyma, as well as hemorrhage from surfactant therapy.

**Bronchopulmonary Dysplasia**

Bronchopulmonary dysplasia (BPD), also known as chronic lung disease of infancy, is a disease of unclear etiology, although it is likely multifactorial. The disease was originally thought to be caused from NRDS and its treatment. The etiology of BPD is now less clear given the advancements in neonatal treatment for NRDS, such as surfactant, steroids, and low pressure algorithms of positive pressure ventilation. Infectious organisms, such as *Ureaplasma urealyticum*, which is the most common contaminant in amniotic fluid, have also been implicated as potential causes of BPD. It has been postulated that inflammatory cytokines associated with *Ureaplasma* infection injures the respiratory epithelium, which is then further damaged by oxygen toxicity or barotrauma. 6,8,9

The radiographic findings of BPD vary and have evolved over time from continuing advances in medical therapy. Northway, et al. originally described four radiographic stages of BPD: stage 1 (2-3 days after birth) resulted in the typically granular opacities of NRDS; stage 2 (4-10 days) demonstrated granular opacities and with superimposed complete pulmonary opacification in more severe cases (Fig. 6); stage 3 (10-30 days) revealed small cystic lucencies alternating with small focal opacities; and stage 4 (greater than one month) showing a “bubbly” appearance due to enlargement of cystic lucencies and linear or ropy opacities. 10

**Figure 6. Bronchopulomonary dysplasia (BPD).** Portable chest radiograph of a neonate with bronchopulmonary dysplasia demonstrates diffuse granular opacities with more focal consolidation in the right lung.
Interstitial gas may extend along numerous potential spaces in the interstitium toward the peripheral lung. Subpleural blebs can form and rupture, resulting in a pneumothorax. Interstitial gas may also extend centrally, resulting in pneumomediastinum. The radiographic appearance of PIE can occasionally be confused with aspiration pneumonia, pulmonary edema, and neonatal respiratory distress syndrome.

There are two types of PIE, acute and persistent. Acute PIE appears radiographically as “bizarre tubular and cystic lucencies” which may be focal or diffuse (Fig. 7). The term persistent PIE is reserved for PIE that lasts longer than 1 week. As with the acute form, it may be focal or diffuse. The cysts of persistent PIE have been described as being lined with multinucleated giant cells. Persistent PIE may be confused with other types of cystic thoracic chest masses in the infant. However, PIE can usually be distinguished from other cystic lesions, since it arises occurs and progresses in a ventilated patient.

Management of PIE varies and includes high-frequency ventilation, placing the affected side of the chest down if the PIE is unilateral, and selective bronchial intubation to help spare the affected lung. Persistent PIE is managed conservatively, although focal cases can be resected if they are severe. If successfully treated, radiographic findings improve or resolve, depending upon the degree of improvement and underlying parenchymal injuries (Fig. 8).

Summary

Neonatal lung disease remains one of the most common causes of morbidity and mortality in this patient population, especially in the setting of prematurity. Neonatal chest radiographs play a critical role in the diagnosis, categorization, and management of the myriad of underlying neonatal lung diseases. Therefore, it is critical that radiologists involved in interpreting neonatal chest radiographs be familiar with the imaging manifestations of common neonatal lung pathologies. This will allow for prompt and accurate characterization, as well as expedite and guide treatment.
References


Figure 8. Development and resolution of PIE.
Portable chest radiograph in a premature newborn infant (A) demonstrates an endotracheal tube tip near the carina, diffuse granular opacities, and no interstitial air. Examination performed two days later (B) shows interval development of PIE within the left pulmonary interstitium. Three days later, the right lung develops PIE (C). By two weeks from the initial radiograph, the pulmonary interstitial emphysema has resolved (D).


Pineal Region Mass

Matthew Minor, M.D.,1 William T. O’Brien, Sr., D.O.2,3

1Department of Radiology, Wilford Hall Ambulatory Surgical Center, Joint Base San Antonio-Lackland, TX
2Department of Radiology, David Grant USAF Medical Center, Travis Air Force base, CA
3Department of Radiology, Uniformed Services University of the Health Sciences, Bethesda, MD

Case Presentation

A 15-year-old boy presented with acute exacerbation of chronic headache, as well as new onset nausea, vomiting, and visual changes. Past medical history and review of systems were noncontributory. Physical examination revealed mild papilledema and paralysis of upward gaze. The patient was subsequently referred for an emergent head CT, which was initially performed without contrast but then with contrast based upon the preliminary findings, followed by an MRI of the brain with and without contrast (Fig.).

Figure. Axial contrast-enhanced CT image (A) reveals an enhancing pineal mass with central regions of calcification. There is dilatation of the visualized portions of the third and lateral ventricles, as well as mild transependymal flow of CSF along the margins of the lateral ventricles. Sagittal T1 weighted MR image (B) with contrast demonstrates a lobulated, enhancing pineal region mass with compression of the underlying tectal plate and cerebral aqueduct. There is dilatation of the third ventricle and a normal-sized fourth ventricle.
Case Report, Minor et al.

Key Imaging Finding

Pineal region mass

Differential Diagnoses

- Pineal cyst
- Pineal germ cell tumor
- Pineal cell tumor
- Tectal plate glioma
- Meningioma

Discussion

Pineal region masses include those that originate from the pineal gland, as well as those that arise from adjacent structures. Masses of the pineal region range from simple, benign cysts to high-grade neoplasms. Imaging plays an important role in establishing the appropriate diagnosis or a reasonable list of differential considerations, as well as for identifying underlying complications.

Tumors originating from the pineal gland represent 3-8% of all pediatric intracranial neoplasms and 0.4-1.0% of all adult intracranial neoplasms. Germ cell tumors are most common, particularly germinoma, followed by pineal cell tumors, to include pineocytomas and pineoblastomas.

The pineal gland ranges in size from 10-14 mm and is located within the midline above the tentorium and superior colliculi and below the splenium of the corpus callosum and vein of Galen. It develops as a diverticulum in the diencephalon roof of the 3rd ventricle during the second month of gestation. The gland itself is attached to the posterior aspect of the 3rd ventricle by the pineal stalk. The mature gland secretes melatonin, an endocrine hormone involved in multiple pathways, but most commonly known for its association with circadian rhythms. The pineal gland is composed of 95% pineocytes (specialized neurons related to retinal rods and cones) and 5% astrocytes. Unlike most intracranial structures, the gland is outside of the blood-brain barrier.

Small pineal masses may be asymptomatic, but as lesions increase in size, they compress adjacent structures and may become symptomatic. Poppen and Marino initially suggested 3 clinical phases to pineal region masses: 1) headaches with nausea and vomiting; 2) blurred vision, diplopia, changes in mental states, drowsiness, papillary changes, ataxia or dizziness, and paralysis of the extra-ocular muscles; 3) papilledema, weakness, and spasticity. Two common syndromes associated with pineal region masses include the Sylvian aqueduct syndrome and Parinaud syndrome. These syndromes are similar and result from compression of the mesencephalon. Typical clinical findings include paralysis of upward gaze, abnormalities of the pupil, and nystagmus retractorius. One dreaded complication of a pineal region mass is pineal apoplexy, which is a sequelae of hemorrhage into a pineal cyst or tumor with sudden decrease in consciousness and headache.

Pineal cyst

Pineal cysts are very common and have an outer wall composed of 3 layers: inner gliotic tissue, middle pineal parenchymal tissue, and outer connective tissue. Pineal gland cysts are usually asymptomatic and range in size from 2-15 mm. If the cyst reaches 15 mm or larger, however, it may become symptomatic secondary to compression of the tectum. The vast majority of cysts are simple, unilocular, and follow CSF density (CT) and signal intensity (MRI), although the cyst may not completely suppress on FLAIR sequences secondary to proteinaceous content. Peripheral enhancement may be seen but is usually incomplete. Complex cysts may be multilocular with variable signal intensity but do not have solid enhancing components. Atypical cysts are followed on imaging and clinically to ensure stability and benignity.

Germ cell tumor

Germinomas are the most common intracranial germ cell tumor (GCT) and represent 1-2% of all intracranial neoplasms. They represent the most common pineal malignancy. On CT, germinomas are typically hyperdense due to high cellularity and tend to engulf pineal calcifications centrally within the mass. On MRI, germinomas are typically iso- to hyperintense to gray matter on T1 and T2 sequences and demonstrate avid, homogenous enhancement. They may also have cystic components. Germinomas are
prone to dissemination throughout the cerebrospinal fluid (CSF); therefore, imaging of the remainder of the neuroaxis should be performed. Pathologically, germinomas are very cellular tumors and highly responsive to radiation therapy. In the presence of a germinoma, hCG will often be elevated within the CSF.³ Teratomas are less common GCTs composed of elements from all three germ layers and typically have cystic and solid components with macroscopic fat.

**Pineal cell tumor**

Pineal parenchymal tumors range from low-grade pineocytomas (WHO grade I) to high-grade pineoblastomas (WHO grade IV). Pineocytomas are typically well demarcated, hypo- to isointense on T1, and hyperintense on T2 with avid enhancement. The lower grade masses may have cystic change with solid nodular components. More aggressive lesion are often large, lobulated, and highly cellular. Regions of increased cellularity are hyperdense on CT and may demonstrate restricted diffusion. Pineal calcifications may be seen along the periphery in an “exploded” pattern. Pineoblastomas have heterogeneous MR signal intensity and avid but more heterogeneous enhancement. Obstructive hydrocephalus due to compression of the cerebral aqueduct is common. Aggressive pineal cell tumors are prone to CSF dissemination; therefore, imaging of the entire neuroaxis should be performed.⁴

**Meningioma**

Meningiomas are benign extra-axial tumors (WHO grade I) of the meninges which result from over-proliferation of arachnoidal cap cells. They are the most common extra-axial tumors and comprise 13-20% of intracranial tumors in adults.⁴ Most lesions are asymptomatic unless there is compression of adjacent structures. Meningiomas are typically iso- to hypointense to gray matter on T1 and variable in signal intensity on T2 weighted sequences, depending upon their cellularity and presence of calcifications. A CSF cleft and broad dural base confirms an extra-axial location. Lesions enhance homogeneously and typically have a dural tail, which is nonspecific, but highly suggestive of a meningioma. On CT, meningiomas present as well-circumscribed dural-based masses which are iso- to hyperdense compared to brain parenchyma with intense, homogenous enhancement. Calcifications and adjacent bony hyperostosis are characteristic.⁴

**Tectal plate glioma**

A tectal plate glioma is a subset of midbrain gliomas that are typically indolent in nature and low-grade. The most common associated finding will be hydrocephalus as a sequelae of tectal plate enlargement and compression of the Sylvian aqueduct. On MRI, the tumor is most commonly well-circumscribed, iso- to hypointense on T1, and hyperintense on T2 weighted sequences. The glioma will demonstrate variable enhancement. Given that progression of tectal plate gliomas is rare, the overall prognosis is favorable with symptomatic treatment for hydrocephalus.⁵
Diagnosis

Germ cell tumor (germinoma)

Summary

A variety of pathological entities may involve the pineal region, ranging from benign cysts to malignant neoplasms. Patients may be asymptomatic or present with symptoms most often related to compression of adjacent structures. Cross-sectional imaging, especially MRI, is essential in the work-up and management of pineal region masses. Simple, uncomplicated pineal cysts are benign and do not need further imaging. When presented with an enhancing mass originating from the pineal gland, on the other hand, it is important to image the entire neuroaxis, as aggressive lesions in this region are prone to CSF dissemination.

References


The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
Case Presentation

A previously healthy 8-month-old girl was brought to the pediatrician with increasing abdominal girth. On physical exam, the patient was tachypneic, and her abdomen was distended. Constipation was suspected and the patient was referred for a conventional radiograph of the abdomen, which demonstrated a large abdominal mass (Fig. 1A). An abdominal ultrasound (not shown) and contrast-enhanced CT of the abdomen and pelvis (Fig. 1B and C) was performed to further characterize the mass. Laboratory evaluation revealed an elevated serum alpha-fetoprotein of 394 ng/ml (reference range 0-99 ng/ml). Serum CEA and b-HCG were normal. The serum LDH was low at 185 U/L (normal range 208-274 U/L). The patient underwent laparotomy with subsequent definitive surgical management at a tertiary pediatric treatment facility.

Figure. AP supine view of the abdomen (A) demonstrates diffuse soft tissue density filling the right flank and midline of the abdomen. Air-filled bowel loops are displaced into the left upper quadrant. Axial (B) coronal (C) contrast-enhanced CT images show a solid and cystic mass distorting the inferior margin of the right hepatic lobe.
Key Imaging Finding

Large solid and cystic right upper quadrant mass in an infant

Differential Diagnoses

Renal origin – 55%
- Hydronephrosis
- Polycystic kidney disease
- Mesoblastic nephroma
- Nephroblastomatosis – Wilms tumor spectrum
- Renal vein thrombosis
- Ectopic kidney

Gastrointestinal origin – 15%
- Duplication cyst
- Mesenteric cyst
- Omental cyst
- Meconium pseudocyst

Pelvic origin extending into abdomen – 15%
- Ovarian cyst
- Hematocolpos
- Sacrococcygeal teratoma

Adrenal origin – 10%
- Adrenal hemorrhage
- Neuroblastoma
- Teratoma

Hepatobiliary origin – 5%
- Benign
  - Hepatic Cyst
  - Choledochal cyst
- Infantile hemangioendothelioma
- Mesenchymal hamartoma
- Infantile hemangioma
- Malignant
  - Hepatoblastoma
  - Undifferentiated embryonal sarcoma
  - Undifferentiated rhabdomyosarcoma of the biliary tree

Discussion

The differential diagnosis of infantile abdominal masses is broad and based on age, location, and structure of the lesion. In infants, there are 3 primary locations for masses: renal (55%), gastrointestinal (GI) tract (15%), and extension of a pelvic mass (15%). Ultrasound is often the first imaging modality used in the evaluation of an abdominal mass since it is widely available, does not expose the infant to radiation, and can usually be performed without sedation. The origin of the tumor can frequently be determined by ultrasound. The lesion can be classified as cystic, solid, or mixed. Vascularity of the lesion and evidence of vascular invasion should be evaluated prior to biopsy or resection.

Renal masses comprise the majority (55%) of abdominal masses in infants. Hydronephrosis is most common with other etiologies including polycystic kidney disease, mesoblastic nephroma, nephroblastomatosis - Wilms tumor spectrum, renal vein thrombosis, and ectopic kidney. GI tract masses account for about 15% of abnormalities. Common causes include duplication cysts and mesenteric or omental cysts, as well as meconium pseudocysts. Pelvic masses extending into the abdomen make up another 15% and include ovarian cyst, hematocolpos, and sacrococcygeal teratoma. Non-renal flank masses make up 10% of cases and include adrenal hemorrhage (most common in neonates), neuroblastoma, and teratoma.

Hepatobiliary masses are least common, comprising only 5% of abdominal masses in infants. The most common benign hepatic tumors of infancy are infantile hemangioendothelioma, mesenchymal hamartoma, and infantile hemangioma. Hepatoblastoma is the most common primary malignant hepatic tumor in this age group. Other malignant liver tumors in infants include undifferentiated embryonal sarcoma and embryonal rhabdomyosarcoma of the biliary tree.

This mass was surgically proven to be a mesenchymal hamartoma, which is the second most common benign liver tumor in children. It was successfully resected without complication or need for additional therapy. This entity usually presents as a multicystic mass in the liver in a child younger than 2 years of age. The stroma is usually cystic or mixed but can contain angiomatosus components and be
multifocal. There are reports that patients may have increased occurrence of a balanced translocation at the long arms of chromosomes 11 and 19.

CT images are notable for large feeding vessels. With contrast administration, there is enhancement of the septae and solid (stromal) elements. Pathologically, mesenchymal hamartoma is composed of myxomatous mesenchyme and malformed bile ducts which may bulge from the liver or may even appear pedunculated. Rapid expansion can occur with accumulation of fluid in the cystic spaces. Complications include ascites, jaundice, and rarely congestive heart failure.

If the tumor is detected prenatally, there is increased risk of preterm delivery. In utero percutaneous decompression of the larger cysts improves prognosis. This tumor is often very large at initial detection, making surgical excision complicated. Although rare, cases of malignant transformation into undifferentiated embryonal sarcoma are reported. Although the tumor grows rapidly in young children, over time it may shrink. It may be followed with ultrasound in asymptomatic patients.

Diagnosis

Hepatic mesenchymal hamartoma

Summary

When an abdominal mass is discovered in an infant, conventional radiography is often performed first followed by ultrasonography. The most common etiologies of an abdominal mass in an infant include renal, GI, and pelvic masses, followed by adrenal and hepatic lesions. By determining the organ of origin, internal characteristics, and vascularity of the lesion, the extensive differential diagnosis can be narrowed. Additional imaging with CT or MRI can be of benefit if surgical excision is contemplated.

References

Valerie C. Hostetler, M.D., Claire C. Widule, MS-4

1Department of Diagnostic Radiology, David Grant USAF Medical Center, Travis Air Force Base, CA
2F. Edward Hébert School of Medicine, Uniformed Services University of the Health Sciences, Bethesda, MD

Medulloblastoma.

This nine-year-old boy presented with acute onset headaches, emesis, and truncal ataxia. CT (not shown) was performed initially, showing a posterior fossa mass with hydrocephalus and transependymal flow of CSF. Axial MR images demonstrated a large midline posterior fossa mass with cystic components. Solid portion of the mass were hyperintense to white matter and isointense to gray matter on T2 (A), hypointense on T1 (B), and heterogenously enhancing (C). Increased signal was present within the mass on diffusion-weighted imaging (D).

Medulloblastoma is a WHO grade IV tumor, most commonly affecting children in the first two decades of life; however, lesions may also present in young adults. The tumor typically arises from the cerebellar vermis at the roof of the 4th ventricle; cerebellar hemisphere involvement is far less common and occurs more frequently in older patients. Patients commonly present with rapid onset symptoms of increased intracranial pressure due to obstructive hydrocephalus. CT typically demonstrates a well-circumscribed midline posterior fossa mass, hyperattenuating to brain parenchyma with cystic features in roughly half of cases and calcifications in approximately 10-20% of cases. On MR, T1 sequences show iso- to hypointensity of the mass; T2 signal intensity is more variable. Prominent, heterogeneous enhancement is typical on both CT and MR. DWI characteristically shows restricted diffusion.

Treatment includes surgical resection with chemotherapy and/or radiation. Due to frequency of CSF drop metastasis, imaging the entire neuroaxis is vital prior to treatment.

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Posterior Reversible Encephalopathy Syndrome (PRES).

An 11-year-old boy presented with hypertension, seizure, and acute onset of blurred vision. An MRI was performed, showing bilateral asymmetric FLAIR hyperintensity in the cortical and subcortical regions of the occipital and parietal lobes (A and B). No abnormal enhancement was noted following gadolinium administration (C).

Posterior reversible encephalopathy syndrome (PRES) is a complex disorder that occurs in patients with a variety of complex systemic and metabolic conditions. The pathogenesis remains unclear but is believed to be a result of autoregulation dysfunction and vascular injury. It may occur at any age from 4 to 90 years, many of whom have co-morbidities such as hypertension, impaired renal function, and autoimmune disease. The most common presenting symptoms are seizures, mental status change, visual abnormalities, headache, nausea or vomiting, aphasia, and even shock.

MRI depicts the characteristic T2 and FLAIR high signal edema, typically bilateral, involving the cortical and subcortical white matter regions of the parietal and occipital lobes. Frontal and temporal lobe involvement may also be seen. Basic imaging pattern follows the watershed zones with three pattern variants depicting the lateral, medial, and posterior blood supply. The cerebellum, basal ganglia, and brainstem are less commonly affected. Complications include ischemia from conversion to cytotoxic edema and hemorrhage from oxidative stress and reperfusion injury. Management is aimed at identifying and ameliorating the underlying cause. The prognosis in majority of patients is favorable as most cases completely resolve; however, some develop frank ischemia with progression to infarction and occasionally death.