

Patterns of Microcolon: Imaging Strategies for Diagnosis of Lower Intestinal Obstruction in Neonates

Bernard F. Laya, D.O., Mariaem M. Andres, M.D., Nathan David P. Concepcion, M.D., Rafael H. Dizon, M.D.

Institute of Radiology, St. Luke's Medical Center, Quezon City and Global City, Philippines

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.¹ 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.² It has also been defined as a luminal diameter less than the height of an upper lumbar vertebral body.³ 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.²⁻³ 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

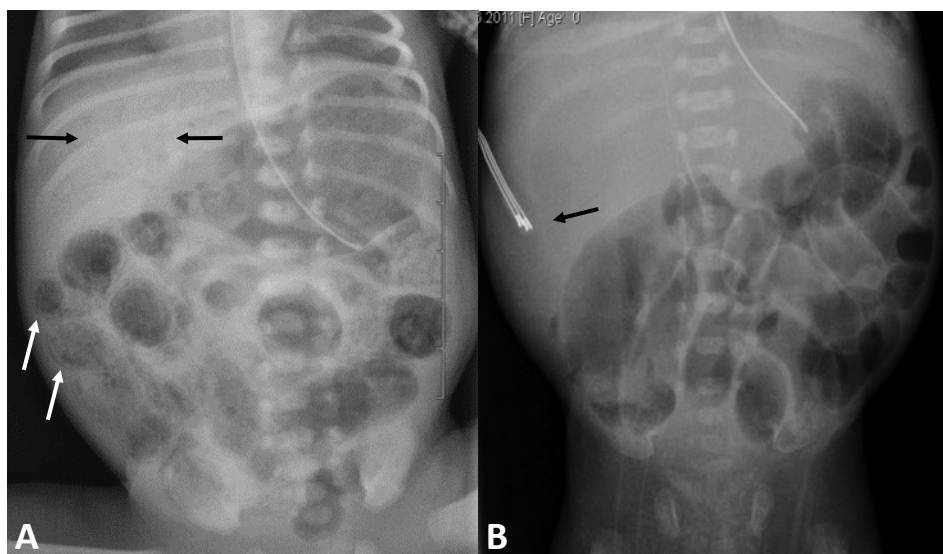


Figure 1.

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.

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).⁴ 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.⁴⁻⁵ 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

plain radiographic features of many (3 or more) dilated and air-filled small intestinal loops with a paucity of air within the colon and rectal regions (Fig. 2B).⁴⁻⁵ This distinction is important in deciding the next appropriate imaging step, whether to perform a fluoroscopic upper gastrointestinal (upper GI) study or a fluoroscopic lower intestinal study (contrast enema).

Fluoroscopic Single Contrast Enema

Low intestinal obstruction in neonates is one of the indications for a fluoroscopic single contrast enema study. It is the examination of choice to determine the

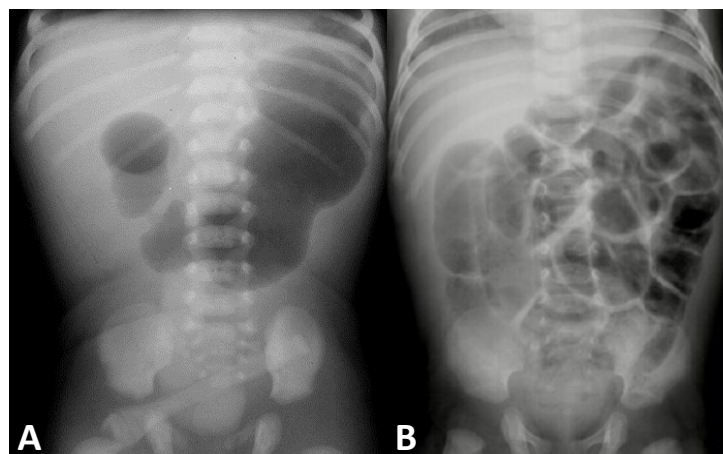


Figure 2.

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.

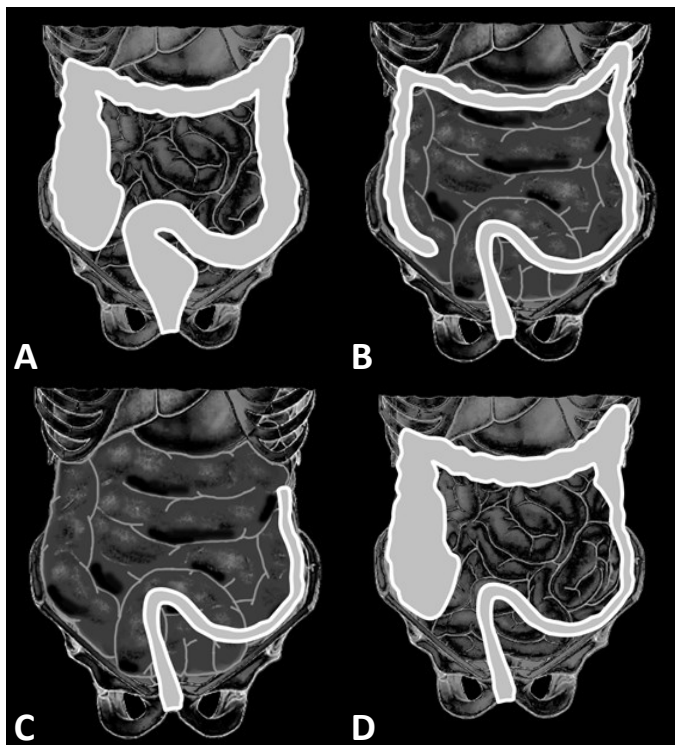


Figure 3. Four patterns on contrast enema for neonates suspected of lower intestinal obstruction.
 Normal (A), Microcolon (B), Short Microcolon (C), and Colonic caliber change (D).

site of involvement if the level of obstruction is at the ileum or at any segment of the colon.⁶ 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.

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**).³ Each of these patterns offers a limited differential diagnosis, which accounts for about 98% of cases, and allows appropriate management decisions (**Table 1**).³

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

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.^{5,7} It can also be used to assess other causes of lower intestinal obstruction and possible complications.⁷ 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.^{4,8-10} Conversely, greater than 95% of patients with simple MI have CF.¹⁰ Patients with cystic fibrosis have malfunctioning sodium-chloride pump which decreases the lubricating property of the intestines creating thick, viscid mucus.³ As a result, inspissated meconium blocks the distal ileum. MI makes up approximately 20% of low intestinal obstructions.¹¹ The incidence in the United States is approximately 1 in 3000 live births per year.¹⁰ 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).⁹⁻¹⁰

Conventional abdominal radiographs show multiple dilated bowel loops with the meconium having a ground glass or soap-bubble appearance.³ 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).¹¹

Water-soluble contrast is performed and serves a dual purpose, being both diagnostic and therapeutic, since the contrast loosens the obstructing concretions of meconium.⁹ The microcolon in most cases will eventually return to normal caliber. In certain cases,

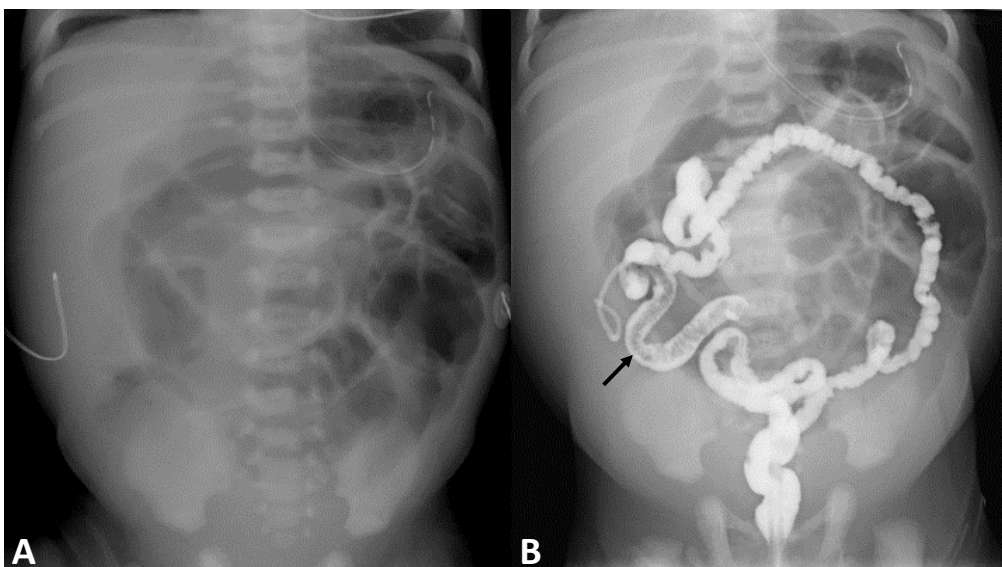


Figure 4.

Meconium ileus.

Frontal abdominal radiograph (A) reveals multiple dilated intestinal loops indicative of lower intestinal obstruction. Water-soluble contrast enema (B) reveals microcolon from the rectosigmoid area all the way to cecum. Multiple filling defects indicative of inspissated meconium (arrow) are seen in the terminal ileum.

Type	Percentage	Pathologic Description
I	23%	<ul style="list-style-type: none"> • Transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel. • The bowel is usually of normal length .
II	10%	<ul style="list-style-type: none"> • Involves two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact .
IIIA	15%	<ul style="list-style-type: none"> • Similar to type II, but with a mesenteric defect. • Bowel length may be foreshortened .
IIIB	11-22%	<ul style="list-style-type: none"> • 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%	<ul style="list-style-type: none"> • 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.

surgery may have to be performed with the goal of establishing intestinal continuity and preservation of maximal intestinal length.⁹ The prognosis for infants with both simple and complicated MI is excellent with reported survival rates approaching 100%.^{9,12}

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**.^{10,13-14} Structural obstruction due to atresia requires surgical management.

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.¹⁰ 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.¹⁴ The incidence of JIA is approximately 1 in 3000-5000 live births and affects both boys and girls equally.^{8,10,13-14} Approximately 1 in 3 infants is premature.^{10,13} Familial cases of intestinal atresias are rarely reported; most cases are sporadic.¹⁰

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.¹⁰ 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%.¹³

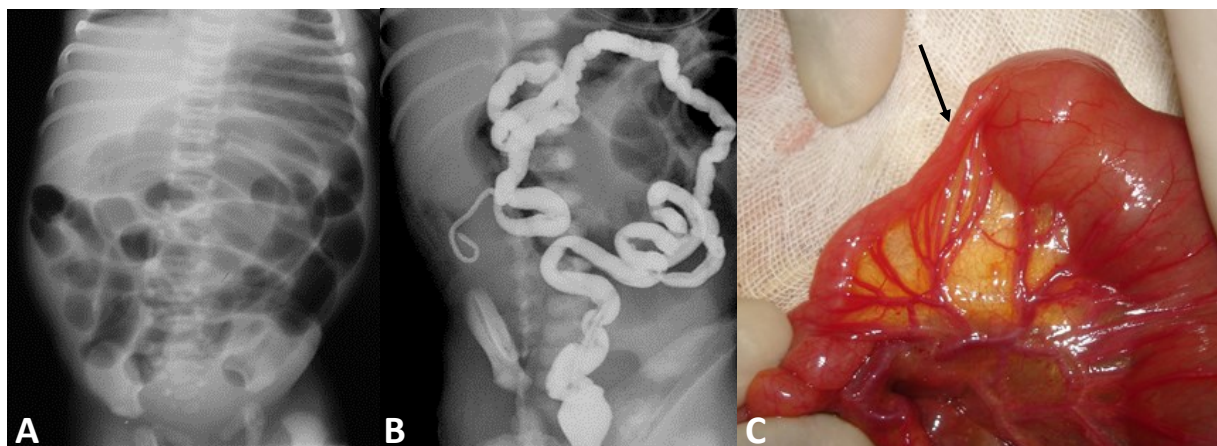


Figure 5. Ileal atresia.

Abdominal radiograph (A) demonstrates multiple dilated intestinal loops indicative of lower intestinal obstruction. Contrast enema (B) shows a microcolon but unable to reflux contrast into the terminal ileum. Gross pathologic picture (C) demonstrates atresia of the terminal ileum (arrow).

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.^{10,15} Mesenteric ischemic vascular insult remains the primary etiology. The classification of intestinal atresias also applies to colonic atresia.^{13, 15-16} Colonic atresia occurs in descending order of frequency at the sigmoid, splenic flexure, hepatic flexure, and ascending colon, respectively.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**).^{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**).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.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%.^{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.¹⁹

Hirschsprung Disease

Hirschsprung disease (HD) is a congenital bowel motility disorder that occurs in approximately 1 in 5000 live births.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.^{3-4,21} HD is common in boys (81.7%). In the majority of cases, recto-sigmoid involvement is seen.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.^{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

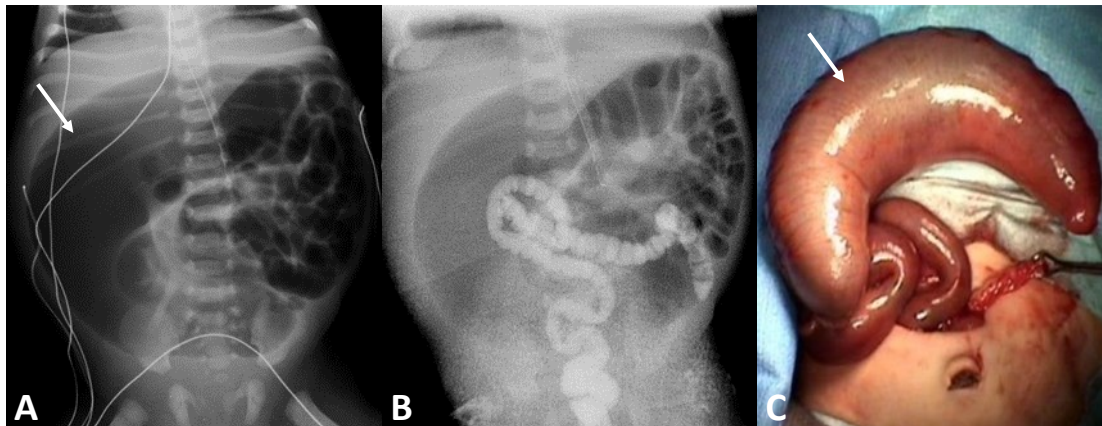


Figure 6. Colonic atresia. Abdominal radiograph (A) reveals multiple dilated intestinal loops with a single loop that is significantly dilated (arrow), raising the suspicion for an atresia. Contrast enema study (B) reveals a short microcolon pattern with a blind-ending loop, compatible with colonic atresia. Gross pathological specimen (C) demonstrates the dilated proximal blind-ended colon (arrow).

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.²²⁻²³ 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.²⁵ Contrast enema may be misleading if patients with HD also have meconium plug in colon.²⁰ The overall sensitivity and specificity of contrast enema study for the diagnosis of HD is 65-80% and 66-100%, respectively.²⁰ 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.²³

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.^{10,20,26}

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.^{4,25} TCA approaches an even distribution between boys and girls compared to short segment HD that has a male predilection.²⁷ 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.²⁷⁻²⁸ The classic question mark

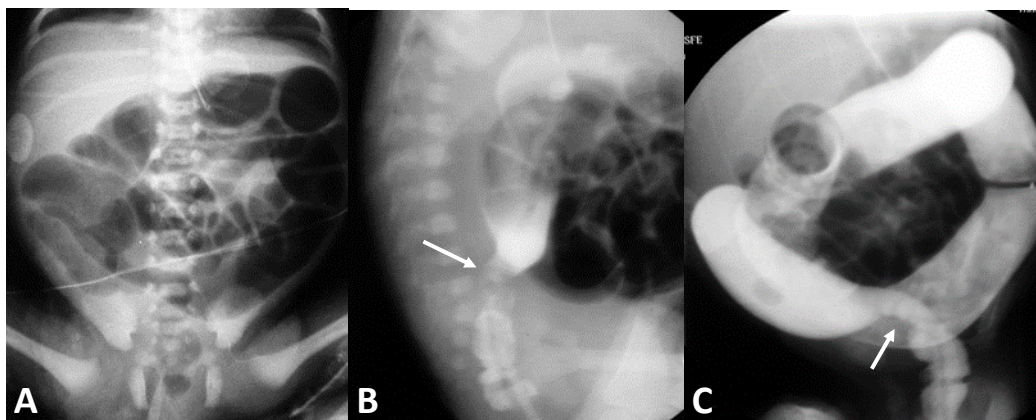


Figure 7. Hirschsprung disease. Abdominal radiograph (A) reveals multiple dilated intestinal loops indicative of lower intestinal obstruction. Lateral (B) and frontal (C) images following contrast enema demonstrate a small caliber rectum compared to the sigmoid. The radiographic transition zone (arrows) is persistent on both views.

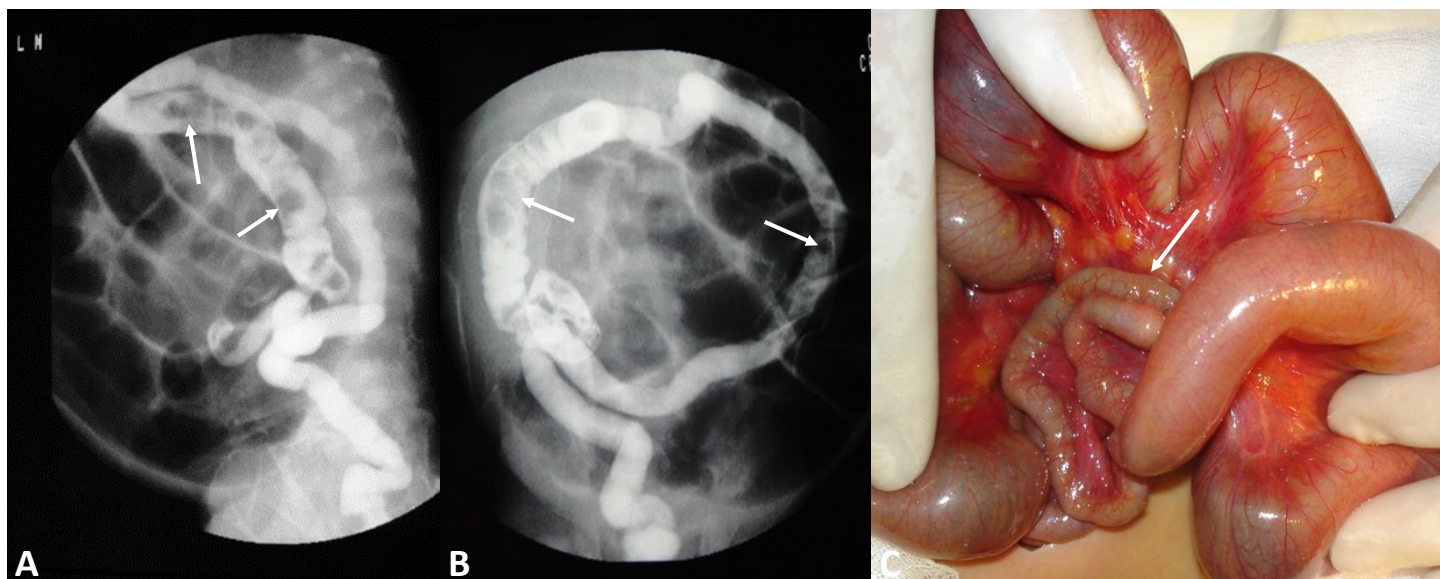


Figure 8. Total colonic aganglionosis.

Lateral (A) and frontal (B) fluoroscopic spot images demonstrate a microcolon involving the entire colon. Inspissated meconium (arrows) is seen throughout the small-caliber colon (Case courtesy of Pedro A. Daltro, MD, Rio de Janeiro, Brazil). Intra-operative image (C) from another patient shows luminal caliber discrepancy between the microcolon (arrow) and significantly dilated small intestines.

shape of the colon is observed in only 18% of children.²⁴ Additionally, the transition zone and rectosigmoid index ratio are not reliable signs in TCA.²⁸

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.²⁰ These risks are higher in TCA compared to short segment HD.^{20,24}

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.^{6,29} 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.”³⁰ 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.^{6,29}

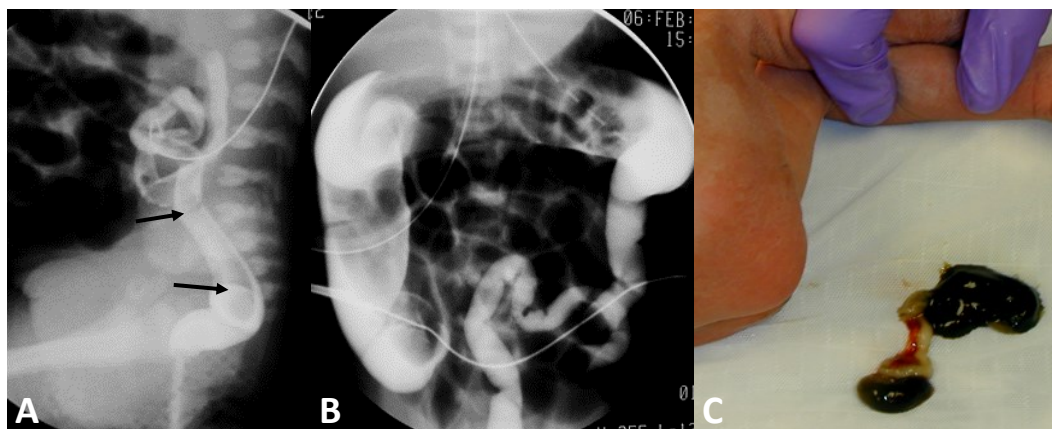


Figure 9. Small left colon syndrome.

Lateral fluoroscopic spot image from a contrast enema (A) reveals a small caliber descending colon with filling defects (black arrows) compatible with inspissated meconium. Frontal view (B) shows a caliber change from a small left colon to a dilated transverse colon. Following the contrast enema, the patient passed a thick inspissated meconium (C).

The incidence of meconium plug syndrome is estimated at approximately 1 in 500 live births.¹⁰ 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.^{4,25} A minority of small left colon syndrome is associated with maternal magnesium sulfate administration for pre-eclampsia.^{4,25} 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.³¹ Meconium plugs found on contrast enema are associated with a 13% incidence of Hirschsprung disease.²⁹

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**).³²

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.^{10,31}

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.

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