Abstract:
An 8-month-old presented to a community hospital with respiratory distress, progressive abdominal distension, and emesis in the setting of fever and significant irritability. His illness course eventually progressed to requiring intubation and bedside exploratory laparotomy. He was subsequently diagnosed with abdominal compartment syndrome secondary to methicillin-resistant Staphylococcus aureus sepsis. The differential for abdominal distension in this age group is extensive, and the eventual diagnosis offers valuable teaching points in the management of the sick infant with abdominal distension.

Keywords:
abdominal compartment syndrome; respiratory distress; sepsis; abdominal distension

Respiratory Distress and Abdominal Distension in an 8-Month-Old ☆

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An 8-month-old immunized male infant, with no significant medical history, was transferred to a pediatric emergency department (ED) from a community hospital with the chief complaint of respiratory distress and fever. The patient's mother reported symptoms of increased work of breathing and fever with a maximum temperature of 102.3°F (39°C) that began 1 day prior to presentation. On the day of presentation, the patient was first taken to a community hospital approximately 20 miles from a children's hospital within a large metropolitan area. At the community ED, a chest radiograph was performed. The patient was diagnosed with an upper respiratory infection and discharged home without further intervention.

Twelve hours later, the patient returned to the same ED with continued fever; labored breathing; and 2 episodes of nonbloody, nonbilious emesis after feeding. Further workup was obtained during the second visit. A repeat chest radiograph was ordered and reported as normal. A 2-view radiograph of the abdomen revealed mild gaseous distention of the colon with prominent distal colonic stool. A complete blood count was remarkable for a white blood cell count of 3.1 K/μL, with an absolute neutrophil count of 2000 K/μL, hemoglobin of 11.8 g/dL, hematocrit of 34.6%, and platelets of 381 K/μL. A comprehensive metabolic panel was unremarkable. A lactic acid was reported to be 3.1 mEq/L. The patient was reported to have appeared lethargic with a weak cry. A mildly distended abdomen was noted, and the patient was reported to be in respiratory distress with grunting, tachypnea,

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and moderate retractions. Because of the patient's clinical examination, he was transferred via ground transport to the tertiary pediatric ED for further care. There were no complications noted during the transfer.

On presentation to the pediatric ED, the patient's vital signs were as follows: temperature 38.8°C, heart rate 190 beats/minute, respiratory rate 44 breaths/minute, oxygen saturation 95%, and blood pressure 113/65 mm Hg. The patient was ill appearing, pale, and tachypneic with moderate retractions. He had no rhinorrhea or nasal congestion, and his lungs were clear to auscultation bilaterally. The patient did have continued grunting throughout the examination. There was no rash or unusual bruising; however, the patient was noted to have diffuse pallor. The patient had a weak cry, responded to pain, moved all extremities equally; however, at rest, he would stay extremely still with minimal spontaneous movement. He was tachycardic without a murmur, and his capillary refill was 3 seconds. The abdomen was distended, tender to palpation, and with hyperactive bowel sounds. There was no cervical or axillary adenopathy appreciated. Mild inguinal adenopathy was noted.

Initially, the patient was fluid resuscitated with normal saline boluses, totaling 60 mL/kg over 60 minutes. A blood culture was obtained, and intravenous ceftriaxone and metronidazole were administered. A procalcitonin, point-of-care glucose, and point-of-care hemoglobin were obtained. A dose of 1 μg/kg of fentanyl was given intravenously for presumed abdominal pain. A urinary catheterization was performed; however, no urine was obtained. An abdominal ultrasonography was ordered to evaluate for intussusception and volvulus. Pediatric surgery was consulted prior to these results.

The point-of-care hemoglobin and glucose returned normal. The procalcitonin returned elevated at 15.6 ng/mL (0.00-0.05 ng/mL). The ultrasonography did not show evidence of intussusception; however, the possibility of malrotation or volvulus could not be assessed because of overlying bowel gas. The patient remained tachypneic and tachycardic, with grunting and nasal flaring. His abdomen became slightly more distended and firm. The child was further discussed with the pediatric surgeon who requested upper gastrointestinal tract radiography to further evaluate for abdominal pathology (Figure 1). This study demonstrated slowed gastric emptying possibly due to generalized bowel distention with no evidence of volvulus. A 2-view abdominal radiograph was repeated without evidence of free air or contrast in the peritoneum. The patient then began to have episodes of oxygen desaturation to 75%. He was placed on a nonrebreather mask to maintain oxygenation. His abdomen became increasingly distended and rigid. His extremities were cool, and his capillary refill time increased to 4 seconds. His temperature defervesced to 37.9°C; however, he remained tachycardic with a heart rate of 179 beats per minute. (Table 1.)

A repeat complete blood count returned 2 hours later with a white blood cell count of 0.8 K/μL. Because of the patient's clinical decompensation, the decision was made to broaden antibiotics to include vancomycin. There was continued difficulty maintaining adequate oxygenation and ventilation, and the patient was intubated using rapid sequence intubation. He required positive end expiratory pressure of 15 cm H_2O to maintain adequate oxygenation. The patient was then transported to the pediatric intensive care unit (PICU) for further care.

In the PICU, the patient lost pulses in his bilateral lower extremities because of the extreme distension of the abdomen. He was deemed too unstable for the operating room, and a bedside exploratory laparotomy was performed. The bowel appeared diffusely edematous without evidence of ischemic injury, and there was no evidence of a volvulus, Meckel diverticulum, or intra-abdominal bands. A normal appendix was visualized, and there was no blood in the abdomen. Because of the diffuse bowel edema, the patient's abdomen was unable to be closed primarily, and a silo was placed over the intestines. The patient tolerated the procedure but remained in critical condition, on multiple vasopressors. The patient's ventilatory support was able to be quickly weaned to a positive end-expiratory pressure of 7 cm H_2O once the procedure was performed. Additionally, the patient's lower extremities regained some color, and a pulse was identified by Doppler ultrasonography. With this information, the suspected diagnosis was confirmed.

DIFFERENTIAL DIAGNOSIS

There is a broad differential for an infant with abdominal distension, vomiting, abdominal pain, fever, and increased work of breathing. In this patient, it is most important to identify and evaluate potential life-threatening intra-abdominal pathologies including malrotation with midgut volvulus, incarcerated inguinal hernia, adhesions with intestinal obstruction, and necrotizing enterocolitis. None of these diagnoses are associated with a primary symptom of fever, but because of the other symptoms and the severity of illness, these

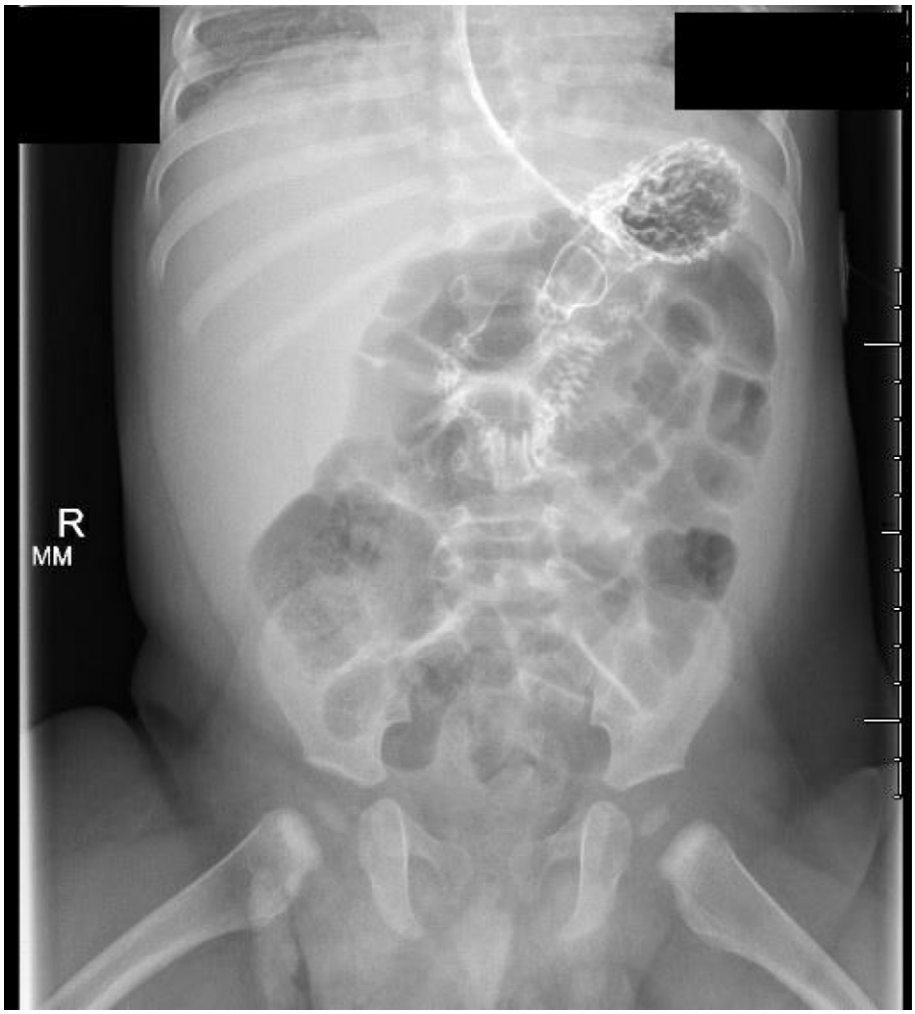

Figure 1. Abdominal radiograph after upper gastrointestinal tract study.

diagnoses were addressed first. The examination of the patient revealed no evidence of incarcerated inguinal hernia, which immediately eliminated this from consideration. Necrotizing enterocolitis was less likely because the development of this disorder in a previously healthy 8-month-old is rare. However, it may occur in setting of poor cardiac output from etiologies such as myocarditis and septic shock which can lead to intestinal necrosis.[1] Review of previous imaging obtained from the outside facility was reassuring that the abdominal radiograph had no evidence of pneumatosis intestinalis or free air. Malrotation usually presents in neonates less than 1 month of age, but older children who develop a volvulus usually have an acute onset of symptoms with bilious or nonbilious emesis.[2] Abdominal ultrasonography was unable to evaluate for malrotation or volvulus, and a subsequent upper gastrointestinal tract radiography ruled out this pathology.

The differential was then further expanded to include other common serious causes of this symptomatology that may not necessarily be life-threatening. These included the following: abdominal trauma, appendicitis, and intussusception.

TABLE 1. Differential of abdominal distension in the febrile infant.

Abdominal trauma	Appendicitis
Intussusception	Diabetic ketoacidosis
Malrotation with volvulus	Hirschsprung-associated enterocolitis
Incarcerated inguinal hernia	Abdominal compartment syndrome
Adhesions with intestinal obstruction	Hemophagocytic lymphohistiocytosis
Necrotizing enterocolitis due poor cardiac output	Pyelonephritis
Sepsis	Pneumonia

Nonaccidental abdominal trauma may have clinical manifestations including abdominal bruising, abdominal distension, or peritoneal signs.[3] Although the patient did not have signs of abdominal bruising, he did have significant abdominal distension and peritoneal signs. Intussusception was also considered because it most commonly presents with sudden, intermittent pain that is classically accompanied by inconsolable crying but can include emesis as the obstruction progresses.[4] It can also be associated with a lead point such as Meckel's diverticulum. Appendicitis was considered because, although it is rare in this age group, appendicitis can occur and practitioners must have a high suspicion in young children who have abdominal pain with vomiting, with or without fever.[5] However, the ultrasonography that was performed to screen for malrotation and volvulus visualized a normal appendix and yielded no evidence of intussusception.

Other uncommon life-threatening pathologies were taken into account including diabetic ketoacidosis, Hirschsprung disease resulting in Hirschsprung-associated enterocolitis, myocarditis, and abdominal compartment syndrome (ACS). Diabetic ketoacidosis usually presents with polyuria and polydipsia; however, it can also present with abdominal pain and vomiting especially in young children with a concurrent illness.[6] Knowing that the patient had a normal point-of-care glucose lowered this on the differential. Hirschsprung-associated enterocolitis, an uncommon complication of Hirschsprung disease was also considered. However, this will usually present with explosive diarrhea, fever, abdominal pain, and/or distension.[7] The patient's family reported no problems with stooling prior to this illness, and the patient had no reported diarrhea. Furthermore, although myocarditis can lead to decreased cardiac output resulting in necrotizing enterocolitis, as discussed above, it can also lead to abdominal pain due to passive hepatic congestion and referred pain caused by pericarditis.[8] The 2 chest radiographs from the outside facility did not demonstrate any cardiopulmonary abnormalities. An electrocardiogram was not obtained on this patient, although it may have been beneficial. Additionally, hemophagocytic lymphohistiocytosis, although extremely rare, can present in this age group with progressive organ dysfunction and abdominal distension in the setting of fevers.[9] The patient did not have elevated transaminases, and a ferritin was not obtained initially.

The last grouping in the differential was composed of infections not primarily in the gastrointestinal tract including urinary tract infections, pyelonephritis, pneumonia, and viral illnesses. Urinary tract infections can lead to acute kidney injury; renal failure; and, with seeding of bacteria in the blood, septic shock.[10] Urine was unable to be obtained in the patient. Pneumonia was considered as the patient may also appear to have abdominal pain, particularly with infiltrates in the lower lobes; however, the patient had a clear chest radiograph on presentation. These patients will typically present with associated symptoms such as fever, tachypnea, and cough.[11] Finally, viral illnesses such as parvovirus and enterovirus were entertained in the differential because they can lead to high fevers, ill appearance, and abdominal distension and pain.[12]

CASE PROGRESSION AND DIAGNOSIS

With the end-organ damage demonstrated by the patient's anuria and altered mental status, the loss of perfusion and pulses in the lower extremities, and the inability to close the abdomen primarily due to bowel edema, the diagnosis of abdominal compartment syndrome due to presumed sepsis was made. After the abdominal decompression was performed, the patient's positive end-expiratory pressure was weaned to 7 cm Hg, and he regained perfusion to his lower extremities.

The cause of the patient's initial decompensation continued to be explored. Ferritin, lactate dehydrogenase, and uric acid all returned within a normal range. A respiratory pathogens polymerase chain reaction (PCR) study returned negative. Parvovirus, HHV8, and enterovirus blood polymerase chain reaction studies also returned negative. An endotracheal tube aspirate was sent and returned with few white blood cells and some gram-positive cocci. At 17 hours after arrival to the ED, the initial blood culture grew gram-positive cocci. A BioFire Assay PCR was performed which identified the bacteria as methicillin-resistant *Staphylococcus aureus*. The patient's antibiotics were narrowed appropriately. On hospital day 4, the patient's abdomen was closed.

The patient's hospital course was complicated by 2 tension pneumothoraxes due to methicillin-resistant *S aureus* pneumonia, which grew from the patient's endotracheal tube aspirate, requiring bilateral chest tubes. Additionally, the patient developed edema of bilateral lower extremities, and Doppler ultrasonography revealed multiple deep venous thrombi with 1 occlusive thrombus. These thrombi were thought to be caused by the patient's initial hypercoagulable state and the multiple attempts that were initially made to gain central intravenous access. Additionally, on day 12 of hospitalization, the patient was noted to again have increased edema of the right leg. At that time, he was found to have significant osteomyelitis of his right lower extremity. This developed into

pyomyositis and osteonecrosis. The patient required multiple fasciotomies and debridement surgeries throughout his hospital stay.

The patient underwent an extensive immunologic workup, which all returned negative. After 30 days in the PICU, the patient was transferred to the general pediatric inpatient unit where he remained for another 28 days undergoing intensive inpatient rehabilitation. Two months after his initial presentation, he was discharged home. At his 6-month follow up, he was noted to be meeting all developmental milestones, tolerating a regular diet, and attending daycare without modifications.

DISCUSSION

Abdominal compartment syndrome was recognized as a distinct entity approximately 25 years ago; however, it has only been in the last 10 to 15 years that there have been formal attempts to standardize the terms for the disease and recommend treatments. ACS is the end point where either massive interstitial swelling or a space filling lesion leads to pathologically increased intra-abdominal pressure (IAP) causing intra-abdominal hypertension (IAH). The criterion standard for diagnosis of IAH/ACS is made indirectly by measuring bladder pressure through a Foley catheter. IAH is defined as a sustained or repeated elevation of IAP greater than or equal to 12 mm Hg, whereas *ACS* is defined as a sustained IAP greater than 20 mm Hg with new organ dysfunction or failure.[13]

ACS can be caused by 2 distinct processes: first, by primary injury or disease of the abdomen and secondarily by interstitial swelling due to capillary leak. Abdominal trauma and ruptured abdominal aortic aneurysms both can lead to hemoperitoneum causing a space-filling lesion that increases IAP. This can also occur with ascites. Secondarily, ACS can occur because of aggressive fluid resuscitation in the setting of increased capillary permeability. This is most notable in the setting of both burns and sepsis. Furthermore, acute pancreatitis is a distinct entity as a cause for ACS because it is an inflammatory process associated with increased capillary leak and hypoalbuminemia. In the setting of acute pancreatitis, significant intraperitoneal visceral edema can develop with or without aggressive fluid resuscitation.[14]

Regardless of the initial cause of IAH/ACS, the end-organ effects of IAH are equivalent. The increasing intra-abdominal pressure leads to reduced ventricular compliance, reduced cardiac contractility, and reduced venous return within the heart. Additionally, with the expanding abdomen, the patient becomes much more difficult to ventilate because of the decreased chest wall compliance and decreased spontaneous tidal volumes. This leads to arterial hypoxemia and hypercarbia. Renal vein compression gives rise to impaired venous drainage, whereas decreased cardiac output causes renal artery vasoconstriction and worsening renal perfusion. Furthermore, blood flow to the mesentery, intestinal mucosa, and celiac/superior mesenteric artery is decreased. To compound this, IAH compresses the thin-walled mesenteric veins, impairing venous drainage and increasing intestinal edema. These events lead to worsening hypoperfusion, gut ischemia, decreased intramucosal pH, and lactic acidosis. Finally, intracranial pressure is increased and sustained while IAH exists, leading to a decrease in cerebral perfusion pressure and progressive cerebral ischemia. The physical signs of ACS include progressive abdominal distension, oliguria/anuria, hypotension, tachycardia, peripheral edema, and pulmonary decompensation. Evidence of hypoperfusion such as cool skin, obtundation, and restlessness can also be present.[13,15,16]

Management of IAH/ACS consists of supportive care while correcting the underlying etiology, with the primary goal being reducing intra-abdominal volume/pressure. This is accomplished by 5 main strategies: evacuating intraluminal contents, evacuating intra-abdominal space occupying lesions, improving abdominal wall compliance, optimizing fluid administration, and optimizing systemic and regional perfusion. In the ED, this may consist of inserting nasogastric and/or rectal tubes, elevating the head of the bed greater than 20°, and increasing abdominal wall compliance with pain control and/or sedation. If the patient is intubated, ventilatory support may be optimized with increased PEEP, decreased tidal volume, and permissive hypercapnia. Finally, although volume administration temporarily improved cardiac output, urine output, and visceral perfusion, it must be goal directed, especially in the setting of increased capillary permeability. There is no role for diuretic therapy in the acute setting.[15]

If IAP remains at greater than 20 mm Hg and/or new organ dysfunction/failure is present, the provider should strongly consider surgical abdominal decompression. This usually consists of a midline incision, with temporary closure of the abdomen usually incorporating of an abdominal silo, a negative pressure vacuum system, or a patch closure. The abdomen remains open while the underlying pathology can be treated, with most surgeons planning staged attempts at fascial closure every 48 hours. Abdominal decompression relieves intra-abdominal hypertension,

resulting in increased perfusion, the ability to decrease ventilator support, and improving venous return. Delays in progression to surgical decompression after the development of ACS can dramatically increase the risk of mortality in both primary and secondary causes of ACS.[15,17]

SUMMARY

The evaluation of a patient with abdominal distension in the setting of fever and respiratory distress can be attributable to many etiologies. In the setting of sepsis, increasing abdominal distension and clinical decompensation should trigger the thought of ACS as continued fluid resuscitation can worsen the clinical picture. ACS is caused by primary intra-abdominal pathologies, space-occupying lesions, as well as increased capillary permeability leading to interstitial edema. Regardless of etiology, sustained IAP will lead to progressive organ dysfunction and eventual organ failure. Once IAH/ACS is recognized, steps to decrease IAP should be used. These can include sedation, elevating the head of bed greater than 20°, and evacuation of intraluminal bowel contents. However, urgent surgical evaluation for surgical abdominal decompression is necessary while continuing to treat the underlying cause of the intra-abdominal hypertension. ✚

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