

# Atypical Appendicitis in Children: Clinical Presentation, Imaging Features, and Management Plan

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**Abstract:** Atypical appendicitis is not uncommon in pediatrics. Factors that are usually seen in such presentations include: Age less than five years, developmental delay, ectopic anatomic location of the appendix, subacute or chronic appendicitis, and symptomatic treatment in addition to early inconclusive investigations. Children with Autistic spectrum disorder often suffer from communication problems, which might affect their ability to deliver their complaints effectively. It is usually parent's interpretation that physicians will rely on as they are the best to delineate minor changes of their children's behavior and body language signs, so that a correct diagnosis may be reached. Atypical appendicitis is usually a delayed diagnosis with increased morbidity and mortality. In this paper we illustrate the possible causes of atypical appendicitis in childhood, and review its clinical presentation, radiologic findings, and management plan as mentioned in medical literature. We also present a rare case of subacute appendicitis in a 5 years old autistic girl with atypical presentation, misleading imaging features, and re-admission after successful laparoscopy. Our aim is to present the importance of clinical assessment and suggest an approach to improve practice regarding childhood appendicitis which can be an endless trap for both patients and practitioners.

**Keywords:** Appendicitis, Atypical, Complicated, Subacute, Chronic Appendicitis, Imaging, Laparoscopy, Developmental Delay, Autism

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## 1. Introduction

### 1.1. Etiology

Acute appendicitis is the most common abdominal surgical emergency during childhood. 44% of pediatric appendicitis have atypical presentations with a relatively high perforation

rate up to 30% [1]. While ectopic anatomic location of appendix is usually the cause of atypical presentation of appendicitis in adults, many causes are possible in children. These causes include: age that is less than five years, medications, developmental delay, communication disorders, chronic and subacute appendicitis in addition to an anatomically ectopic appendix. These make the incidence of

atypical appendicitis relatively high in pediatrics. Ectopic location of the appendix is usually the result of developmental disorders, midgut malrotation or incomplete translocation during early fetal life [2, 3].

Location and length of the appendix may change with age due to many etiologies like intraperitoneal surgical interventions, trauma, inflammation, and adhesions. Surgical procedures on the abdomen or genitourinary systems may change the location of the appendix [3].

Ectopic inflamed appendix in the retro-caecal or retro-ileal positions is often shielded by the caecum and ileum, which can lead to a reduction in the pain severity and tenderness. In infants and children under 5 years old, appendicitis often lacks typical symptoms and may manifest as gastrointestinal-like symptoms with minimal abdominal tenderness. The use of analgesics, antipyretics, antiemetics, and other symptom-relieving medications can mask the abdominal pain, tenderness, and fever. This can potentially lead to misinterpretation of the symptoms. Therefore, it is advisable to avoid administering these medications in cases of uncertain abdominal pain diagnosis [4].

Antibiotics can change the course of disease and alter its presentation or duration. Many studies showed the successful use of antibiotics and analgesics to treat acute appendicitis without surgery [5]. Acute appendicitis is the most common abdominal emergency in developmentally delayed children. Autism Spectrum Disorder, Down Syndrome, Cerebral Palsy, and other Mental Disabilities usually add a diagnostic challenge to almost all abdominal emergencies, appendicitis namely, as these children are unable to communicate their symptoms properly to their caregivers or treating physicians.

### ***1.2. Clinical Presentation of Atypical Appendicitis***

The typical presentation of appendicitis as migratory abdominal pain from the epigastrium or periumbilical area to the right lower quadrant accompanied by anorexia, nausea and vomiting is usually absent in small children less than five years, which makes its diagnosis often difficult and delayed. It is also associated with a high perforation rate [6].

Atypical appendicitis cases across all age groups often exhibit a similar pattern. A clinical study involving 270 patients under the age of 5, all diagnosed with appendicitis, revealed multiple atypical findings. Approximately one-third of the cases did not experience nausea or vomiting, while half did not have migratory pain. In 17% of cases, the pain persisted for more than 48 hours, and 16% experienced diarrhea. Pyrexia was absent in 82% of cases, while 55% showed no signs of guarding and 56% did not experience rebound pain. A third of the cases lacked tenderness, and a third experienced maximal pain in a location other than the right lower quadrant. Only 9.6% of cases had normal white blood cell count (WBC) [2].

Urinary frequency may indicate ureteric irritation, while minimal abdominal tenderness is typically observed without muscular rigidity. For instance, pain in the pelvic appendix is commonly localized to the lower abdomen, with limited tenderness detectable through rectal examination, which is

infrequently performed in pediatric cases. The occurrence of a left-side appendix is rare, approximately affecting 0.02% of the population, and is more likely in cases of gut malrotation or situs inversus. This condition typically manifests as left-sided abdominal pain concentrated in the left upper quadrant.

Maximal tenderness of the appendix, typically found in McBurney's point in the retrocecal region, can sometimes occur in atypical and uncommon locations. These locations include the left lower quadrant, near the lower edge of the liver close to the gallbladder, in contact with the duodenum, near the rectum, or even within a hernia in the scrotum. These anatomical variations can alter the presentation of atypical appendicitis and may resemble symptoms of acute diverticulitis, cholecystitis, duodenal ulcer, duodenitis, enteritis, or pathologies affecting the adnexa or testes [3, 4].

The medical history of this patient group relies on the impression and understanding of their caregivers. In severely affected children displaying hyperkinesia, hypertonia, and posture disorders, abdominal pain is typically minimal, making it challenging to detect tenderness or rigidity most of the time. Children with developmental delay need high clinical attention for the risk of complicated atypical appendicitis (7). Developmentally delayed children may exhibit manifestations of abdominal pain through sleep disturbances, sudden irritability, interrupted sleep, and aggressive behavior. Vomiting, fever, and tachycardia may also be present and more noticeable than the abdominal pain itself. Other symptoms that may be observed include profuse diarrhea (with or without blood), anorexia, decreased appetite, reduced wet nappies, and recent constipation [7].

### ***1.3. Radiological Findings in Atypical Appendicitis***

Abdominal ultrasound is usually the first recommended radiological tool in children with suspected appendicitis due to its non-invasive nature and high specificity (93%), although it has lower sensitivity (83%), this is probably due to the fact that it is operator dependent, and it is difficult to evaluate unprepared patients. (9) Avoiding unnecessary radiation is crucial, especially in children. Therefore, if ultrasound reveals inflammatory findings, there is no need for a contrast-enhanced CT scan [4]. Ultrasound results can reveal the presence of layering and thickening of the distal appendix >7mm in diameter, along with increased blood flow and moderate swelling of the tissues surrounding the appendix [10]. CT scan is the preferred imaging method with superior sensitivity (96%) and specificity (89%). [11] It is an excellent diagnostic radiology tool to assess inflamed appendix at any location. Appendiceal diameter 6.5mm or more, inflamed peri-appendiceal fat, and appendicolith are the typical appendicitis features by CT scan. However, it should be done only when ultrasound is inconclusive [5].

Computed tomography scanning findings of chronic and recurrent appendicitis are very similar to those of acute appendicitis [12]. stranding in the peri-caecal fat tissue is seen in 100% of cases, 88.9% show appendix dilatation, 66.7% focal thickening, 66.7% enlargement of abdominal lymph

nodes, and 50% of cases show a calcified appendicolith. Abscess formation or phlegmon collection may also be present [9, 10]. Magnetic resonance imaging should be an option when CT scan with contrast is contraindicated or not preferred especially in children and pregnant women. Appendix can be seen in T1-weighted and T2-weighted as hypo intensive thin-walled tubular structure [6].

## 2. Case Presentation

A 5-year-old girl was brought to the pediatric emergency room due to weakness in her lower limbs and an unsteady gait that started the day before. She had also been experiencing nausea, vomiting, and diarrhea for a few days. Her mother, who is a specialist medical doctor, told that her child had developed severe diarrhea, fever, and vomiting eight days ago. She had visited her general practitioner at that time and was diagnosed with enterovirus-positive gastroenteritis. The GP prescribed symptomatic medications, and the child was discharged home. Two days later, mucoid blood was noticed in her stool, and she was started on oral metronidazole, ibuprofen, Hyoscine (Scopolamine) and acetaminophen for pain relief as needed. Over the course of three days, her symptoms gradually improved. She had been previously diagnosed with autism at the age of 2 years when she was presented with social and language delays.

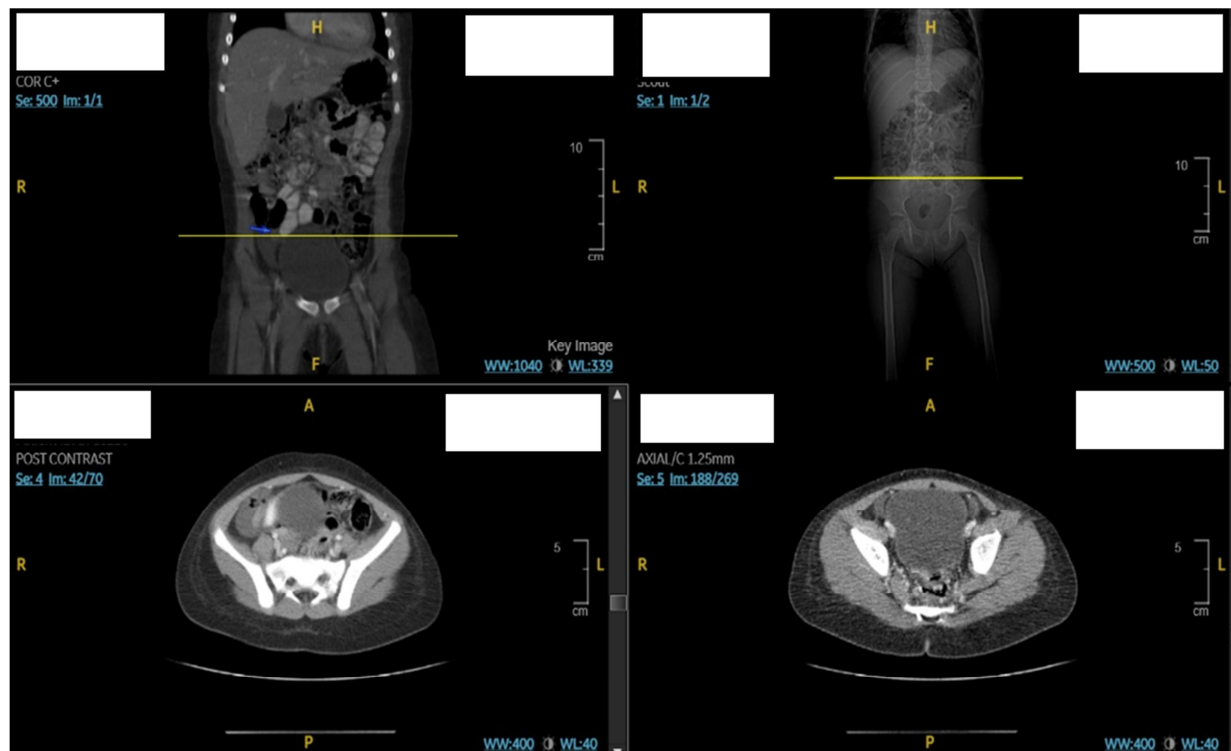
One day earlier, she exhibited signs of unhappiness, irritability, abdominal colic, nausea, anorexia, and

experienced diarrhea 2-3 times. There was no fever present. Today, since the morning, she began avoiding walking and could barely move her legs when pulled to stand. Due to the symptoms of gastroenteritis and weakness in the lower limbs, the child was admitted to the hospital from the emergency room for further investigation.

Primary investigations revealed a hemoglobin level of 10, elevated white blood cell count (24,000 with 60% neutrophils), normal platelet count (300,000), high C-reactive protein (65), elevated ALT (75), elevated CPK (450 IU), and normal creatinine. Stool analysis showed positivity for Enterovirus and Blastocytosis Hominis. Physical examination indicated comfortable sleep, no fever, mild dehydration, soft abdomen, clear chest, and normal lower limb assessment. Analgesics were discontinued, and a full gait examination was planned.

After 6 hours, the child experienced intense pain in the chest, abdomen, right arm, and right leg. She vomited once and had watery stool without blood. Abdominal rigidity and tenderness were evident while she cried and displayed irritability but subsided when she calmed down. An abdominal ultrasound revealed a moderate collection of fluids in the right iliac fossa, with an inconclusive report that appendicitis could not be ruled out.

An abdominal CT scan with contrast was performed one hour later, which provided a definitive diagnosis of perforated complicated appendicitis with a fluid collection present.



**Figure 1.** Enhanced CT of the abdomen: acute perforated appendicitis (appendix thick wall enhancement surrounded with free fluid collection).

Based on the advice of the pediatric surgeon, it was recommended to continue the antibiotics ceftriaxone/metronidazole without the need for immediate

surgical intervention. After 12 hours, the child was found to be sleeping without pain and receiving intravenous fluids. To manage pain, the child was given perphalgan three times. The

vital signs recorded were a heart rate (HR) of 130, respiratory rate (RR) of 35, and blood pressure (BP) of 93/43mmHg. Repeat laboratory results showed a decline in overall health, with CBC revealing an Hb level of 9.8, WBC count of 16,000, CRP level of 260, and platelet count of 900,000. This deterioration in both clinical and laboratory findings prompted the consideration of other causes for intestinal perforation.

A Second opinion surgery referral was arranged to exclude other possible causes, collect samples and put a drainage. After 6 hours, Laparoscopy under general anesthesia disclosed simple unperforated appendicitis. Clear inflammatory fluid, which was accumulated around the cecum and in the pelvis, was cleared and appendectomy without drainage was performed. When the patient was seen in the following morning, she was symptom free. Liquid diet was commenced, with a plan to continue administering amoxiclav alone for 3 more days.

Later that evening, the mother expressed concerns about recurrence of diarrhea for 3times, accompanied by cramps, colic, and signs of irritability. Upon examination, the child appeared stable with no abnormal findings. The parents were reassured, and the child was discharged home. However, after another 12 hours, the mother sent a video via WhatsApp, depicting a stool container filled with 150 ml of bloody mucus collected from her daughter's stool. The mother was frightened, and the child appeared distressed and unwell while crying.



**Figure 2.** Bloody mucoid stool sample.

When the child was examined in the pediatric ER, she looked stable, not in pain, and alert. Her vital signs showed: heart rate (HR) of 90 bpm, respiratory rate (RR) of 25 cpm, blood pressure (BP) of 104/54 mmHg. Oxygen saturation (Sat) was at 97%, body temperature (T) was 37°C. Chest and abdomen appeared normal upon examination, with no rashes. The presence of occult blood in the stool was confirmed, with HGB level of 9.9, white blood cell count (WBC) was 12,000, and the C-reactive protein (CRP) level was 160. Additionally, (INR) was 1.2, and the partial thromboplastin time with kaolin (PTTK) was 45sc. Due to these findings, the patient was readmitted as a case of lower gastrointestinal bleeding.

Differential diagnosis of such a scenario is challenging and

must consider:

1. Bleeding from appendiceal invaginated stump.
2. Michel diverticulum.
3. Colon polyp or diverticulum.
4. Hepato-renal syndrome.
5. Vitamin K deficiency.
6. Henoch Schoenlein purpura.
7. Severe colitis caused by Clostridium.
8. Vasculitis with atypical presentation.
9. Ulcerative colitis with atypical presentation.

After a discussion with the pediatric gastroenterologist and pediatric surgeon, an abdominal U/S was performed to exclude intraperitoneal bleeding, 10mg vitamin K was given, and IV fluid increased to 1.5 maintenance. The patient was kept NPO with close observation with vital signs being monitored every hour. Order to repeat the laboratory tests after 4 hours was put with a recommendation to prepare the patient for a colonoscopy within 24 hours or sooner if her condition worsened. The following tests were requested for the next blood extraction: blood film, ANCA, PANCA, coagulopathy profile study, creatinine, albumin, bilirubin, ALT, and GGT.

Over the next 24 hours, the child had a single instance of passing a small amount of watery bloody stool. However, there was no deterioration in their vital signs, which remained stable. All laboratory results came back normal, and the INR returned to a normal range. Both renal and hepatic function were also within normal limits.

The following morning, this piece of underwear was seen in the patient's room. The mother had thoroughly washed it with water and soap to ensure proper cleaning.



**Figure 3.** Pigment on the child's underwear.

The pigment observed was not blood but had a blood-like appearance. After reviewing her dietary history, it became evident that the substance resembling mucoid blood in the stool container was, in fact, a combination of cherry jelly, juice, and ice cream. The child, who is autistic, consumed these items in copious quantities, and the presence of weak stomach acidity contributed to the unusual appearance.

### 3. Discussion

Childhood autism, small age, subacute appendicitis, treatment with antibiotics and excessive symptomatic

medications all were causes of atypical appendicitis in this patient at the same time.

Presentation started as gastroenteritis symptoms with minimal abdominal tenderness. Analgesia, antipyretics, antiemetic and other symptomatic medications might have minimized abdominal pain, tenderness, and fever. Vomiting, disturbed sleep and Anorexia also improved with treatment and that caused misinterpretation of findings. Appendicitis in developmentally delayed children more often presents late with a local abscess or spreading peritonitis. A pelvic mass or intestinal obstruction from the small bowel adhering to the wall of an appendiceal abscess is common. This leads to increased morbidity and mortality [8]. These children also have higher rates of postoperative emergency room visits and hospital readmissions after undergoing laparoscopic appendectomy compared with normal children [8]. Computed tomography might be ordered instead of ultrasound to increase diagnostic yield in this group [8]. In autistic children decreased amount of wet nappies, and recent constipation might be seen usually. Sleep disturbances, sudden irritability, interrupted sleep, and aggressive behavior can be manifestations of abdominal pain in developmental delay children.

Rigidity and tenderness also might be hidden by analgesia, or become intermittent, with very soft abdomen in between [3]. In childhood appendicitis, the time window between inflammation and perforation is typically narrow, emphasizing the importance of early clinical suspicion for optimal outcomes. The presence of an antalgic gait or lower limb pain, accompanied by symptoms resembling gastroenteritis, should raise concerns and increase doubts regarding appendicitis. [11]. It could be the result of direct psoas muscle irritation and even inflammation or a prophylactic maneuver by the child to decrease peritoneal pain [11]. It seems mandatory to arrange urgent abdominal ultrasound in this case; keeping in mind that perforation in imaging study is not necessarily a definite diagnosis as seen in this patient.

Chronic appendicitis can sometimes be radiologically tricky even by CT scan [13, 14]. "Misty mesentery" on imaging is a term used to describe the radiological presentation of mesenteric fat that has been invaded by inflammatory cells, fluid, tumors, and/or fibrosis can be seen in other cases like Pancreatitis, diverticulitis, inflammatory bowel disease, TB, lymphoma, and mesenteric panniculitis which should be also kept in the differential diagnoses [15, 16]. In this particular case, both ultrasound and CT scan results unfortunately led to a misdiagnosis, as they indicated signs of perforation and the presence of a phlegmon collection. Regrettably, this confusion resulted in a further delay in performing laparoscopy, as advised by the pediatric surgeon. The patient's conservative management plan was reevaluated due to clinical deterioration presented as tachycardia and deterioration in blood pressure in the absence of fever, dehydration, or pain in addition to worsening acute inflammatory markers.

During laparoscopy, huge amount of clear inflammatory fluids was suctioned from the pelvis and ileocecal area.

Surprisingly, the inflamed appendix was intact, accessible with no adhesions, further histopathology demonstrated nonspecific subacute inflammatory changes in the appendix layers.

The optimal approach for treating complicated or perforated appendicitis in pediatrics remains a topic of debate. However, it is agreed that laparoscopy offers superior outcomes compared to a conservative plan with supportive care, allowing for exploration of the affected area, drainage placement, and subsequent administration of appropriate antibiotics [17, 18]. Tazocin is the preferred treatment for peritonitis, while a combination of Cefepime and metronidazole is recommended for peritonitis following perforated appendicitis [18, 19, 20].

For childhood appendicitis, laparoscopic appendectomy is the recommended treatment approach, regardless of whether the condition is simple or complicated [19]. When perforation is suspected, it is also advisable in pediatrics to explore the area, put drainage, and potentially perform an appendectomy if applicable [20]. Laparoscopy may also be considered an accurate diagnostic tool for other possible rare causes like perforated gut or diverticulum, Crohn's disease, Tuberculosis, lymphomas, or congenital anomalies [20]. Lower Gastrointestinal bleeding after appendectomy is extremely rare in children. However, it is a critical situation that requires a multidisciplinary team approach to recognize the etiology and provide best evaluation and treatment. Pediatric surgeon, gastroenterologist, hematologist should be involved early in the management, which is usually better to be performed in a pediatric intensive care unit. It is important to note that, although rare, appendectomy may be complicated by bleeding, making it an unpredictable factor that should be considered in the differential diagnosis.

## 4. Conclusion

In children, appendicitis can manifest with atypical symptoms and signs in up to 44% of cases. The anatomical location of the appendix, as well as surgical interventions, chronic inflammation, trauma, and age-related changes, all contribute to this conundrum. Subacute or chronic appendicitis can also lead to a longer, less severe clinical course, adding to the bewilderment. Special attention should be paid to patients with developmental delay, autism, and cerebral palsy, as these conditions can obscure clinical findings.

Administering antibiotics and symptomatic medications can further modify the disease course and clinical presentation, complicating matters even more. It has been observed that when a patient's parent is a doctor, atypical and delayed presentation of appendicitis can occur. Clinicians should be aware that migratory abdominal pain, abdominal rigidity, and rebound tenderness are not commonly seen in childhood atypical appendicitis.

If a child with gastrointestinal-like symptoms presents with lower limb pain or antalgic gait, urgent investigation is required to rule out appendicitis. When ultrasound findings

confirm appendicitis, no further radiological studies are necessary. However, in cases with inconclusive ultrasound findings, a contrast-enhanced CT scan should be undertaken, as it is an effective method for identifying uncommon appendix locations.

The accuracy of ultrasound and CT scan diagnoses for complicated perforated appendicitis can be limited. Therefore, a low threshold for laparoscopy should be maintained in both typical and atypical appendicitis in children, regardless of symptom duration. Paediatricians and surgeons should be mindful of the high incidence of atypical appendicitis in children and consider it even when unexpected.

Treatment approaches for complicated or perforated appendicitis are subject to debate, making it more confusing. However, laparoscopy, which involves exploration of the area, drainage placement, and antibiotic treatment, is generally preferred over a conservative plan with supportive care and delayed intervention if necessary. Tazocin is recommended as the primary treatment for peritonitis, while a combination of Cefepime and metronidazole is preferred for peritonitis following perforated appendicitis. When the patient's parent is a doctor, atypical appendicitis should be expected.

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