Appendicitis – pitfalls and medicolegal implications

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Appendicitis, despite being the most common acute surgical condition of the abdomen in children, remains a diagnostic challenge.[1-6] It is estimated that up to 20% of children who present with acute abdominal pain will eventually be diagnosed with appendicitis.[7] Missed or delayed diagnosis of appendicitis is also among the top five most frequent malpractice claims against emergency department physicians.[1] Initial misdiagnosis by the first-contact physician approximates 30% in children 14 years or younger.[8] This fact is well described in the international literature and is due directly to the nonspecific symptoms and signs of early appendicitis. There is also no good accurate defining diagnostic test for early appendicitis and diagnosis based on physical examination is often impossible.[5] Furthermore delayed diagnosis caused by parents, physicians and surgeons further compromises the timely management of the disease.

The fundamental aspects of diagnosis in appendicitis in children are described below, with emphasis on how variations in anatomy and clinical presentation can make this difficult. Common causes of misdiagnosis are identified, and a strategy for reducing the rate of misdiagnosis is outlined in an attempt to expedite diagnosis and thereby improve outcomes and reduce reasons for litigation.

Red Cross Children's Hospital review

A recent review (2005) of 136 consecutive children with appendicitis at Red Cross War Memorial Children's Hospital has highlighted many of the difficulties involved in diagnosing appendicitis that may have clinical and medicolegal implications: (*i*) the peak incidence of appendicitis in prepubertal children occurred at 9 - 12 years of age with 3% < 3 years; (*ii*) symptoms of appendicitis had been present on average 2 - 3 days prior to admission (range 0 - 13); (*iii*) these findings were similar for both perforated and non-perforated appendicitis – whether this indicates that in fact there are two types of appendicitis, one that perforates and one that does not, is still a source of some debate; (*iv*) after a child was admitted to hospital with abdominal pain it could take surgical staff up to 67 hours to confirm the diagnosis for perforated and non-perforated appendicitis; and (*v*) life-threatening complications can result as a consequence of both delayed presentation and under-resuscitation. The overall complication rate was 10%.

Types of complications

Complicated appendicitis had a significantly longer hospital stay, 3 days for non-perforated compared with 5 days for the perforated group. The perforated group had, in some instances, stays for 3 - 8 weeks due to complications, with a significant increase in the number of operations needed to remedy the consequences of complications. These included enterocutaneous fistulas, severe iatrogenic ischaemic limb damage secondary to hypotension, multiple intra-abdominal abscesses, renal failure and prolonged intensive care stays.

The mortality rate was 0.7%. Although the disease can be far advanced and children admitted in extremis, death is unusual (0 - 2.4%).[1] During a reviewed 20-year period 6 children admitted to hospital with a subsequent clinical diagnosis of appendicitis died from the following causes: moribund on admission – 3; appendix stump leak following an appendectomy done for abdominal pain – 1; appendicectomy done in error in a child with acute rheumatic fever – 1; post-operative stress ulceration with fatal gastric haemorrhage – 1.

Medicolegal claims reviewed in the records of the Medical Protection Society (time unspecified) revealed 86 cases against doctors. Of these 50% related to postoperative complications, 20% to misdiagnosis and 7% to intra-operative mishap.

The postoperative complications were primarily due to retained collections, sepsis and bowel obstruction (63%). The next most common problems were systemic complications relating to cardiovascular compromise, e.g. pneumonia and in adults deep-vein thrombosis (21%). Lastly, retained instruments and foreign bodies such as needles, swabs and drains caused complications in 16%. Many of these relate to the late presentation of appendicitis and possible delay through patient or physician error.

Delayed diagnosis formed the basis of a significant proportion of claims laid against physicians; these included incorrect diagnosis, perforation, systemic sepsis and death. The difficulties and reasons for delay in diagnosis in paediatric cases are elucidated below.

Only a small percentage of complaints related to intra-operative mishap, most to difficult identification of the correct anatomy, e.g. removal of the ureter or incomplete removal

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of the appendix. Again complications were associated with advanced disease.

Anatomical considerations

Morphologically the appendix is the underdeveloped distal end of the caecum and is a long diverticulum projecting from the caecum at the confluence of the three taenia.[9]

The appendix can assume various anatomical positions, i.e. retrocaecal 74%, pelvic 21%, paracaecal 2%, subcaecal 1.5%, pre-ileal 1%, postileal 0.5%. In children, as opposed to adults, the caecum and the appendix assume a more abdominal than pelvic position. This diversity of position and relatively elevated position lead to varied presentations. The underdeveloped greater omentum is an important additional factor contributing to the difficulty in localising the inflammatory process to the right iliac fossa (RIF). As a consequence, young children are more likely to progress to generalised peritonitis. The pathological process from onset of appendicitis to perforation in adolescents and adults usually occurs within 24 - 36 hours. In young children this can be as little as 8 - 24 hours.[10]

Clinical presentation

The clinical presentation of appendicitis is related to the position of the appendix, the stage and progression of the disease, and the degree of peritoneal irritation. The natural clinical history can be divided into three phases. The first phase is largely nausea and vague abdominal pain without physical findings. Mild constitutional symptoms may be present. The next stage is progression to constant, localised pain in the RIF with systemic evidence of inflammation. The third phase is characterised by the inflammatory process progressing to perforation, abscess formation and peritonitis with concomitant worsening of abdominal signs, i.e. guarding preceding rigidity, marked abdominal distension and tenderness.

Variations to the typical scenario

The inflamed appendix is in a 'hidden position' unable to be detected by the palpating hand in 15% of patients with simple appendicitis and in 30 - 68% of patients with gangrenous or perforated appendicitis (pelvic, retrocolic, retro-ileo-colic), thereby changing the clinical manifestations of appendicitis.[3,11] When the appendix is in a 'hidden position', patients will present with less abdominal pain, less local tenderness in the RIF and a longer duration of symptoms before the diagnosis is established. A higher perforation and sepsis rate occurs and the differential diagnosis is more varied. This atypical presentation with relatively unimpressive physical findings may cause the patient to delay seeking medical attention and may dissuade a physician from considering appendicitis as the cause of the patient's symptoms.

An inflamed appendix touching the rectum or in the retro-ileal position may mimic diarrhoea, which can be substantial in watery volume and mimic gastroenteritis closely.[12] This symptom is common enough to potentially confuse the definitive diagnosis. In the retrocolic position it may cause renal angle tenderness and in the pelvis on the bladder wall, dysuria and even leucocytes in the urine.

Diagnosis

Despite technological advances, the key to successful diagnosis of appendicitis is a careful history, a thorough satisfactorily carried out, and if indicated serial, physical examination, a high index of suspicion and supporting selected laboratory studies. If more than one of these aspects indicate appendicitis, further evaluation is usually unnecessary. Only in confusing cases or atypical presentations, should further diagnostic studies be ordered.

A rectal examination to aid the diagnosis is rarely done in children. In a large study with 100 consecutive children with appendicitis, rectal examination only contributed to the diagnosis in 3% of cases.[13] It is a very distressing and traumatising event for children and not encouraged at Red Cross Children's Hospital.

Ancillary diagnostic studies done in elucidation of the cause of acute abdominal pain in children are used to supplement clinical evaluation, but they do not appreciably reduce the frequency of missed appendicitis or negative appendectomy.[6,14] A moderate polymorphocyte leucocytosis is normally present, but the count may be normal in cases with perforated appendicitis and can also be raised with other causes of abdominal pain. Use of the leucocyte count alone may result in missed diagnosis or unnecessary surgery.

Plain abdominal radiographs usually contribute little to the diagnosis of appendicitis and are not routinely recommended. However, if they are ordered, certain findings may be found such as the presence of a faecalith, abnormal and atypical lower quadrant gas patterns, localised ileus, lumbar scoliosis and obliteration of the psoas shadow. Plain radiographs are more useful in identifying other causes than confirming the diagnosis of appendicitis. Chest radiographs may be particularly helpful[15] to exclude an occult pneumonic process which may mimic appendicitis.

In experienced hands ultrasound may be helpful (accuracy 85%), but this is not universally available.[15] Ultrasound, however, can provide a non-invasive method to review the entire abdomen and pelvic cavities.

Focused computed tomography (CT) with bowel contrast to diagnose appendicitis has a positive yield of 95%.[16] However, it exposes patients to significant radiation and has not reduced the negative appendectomy rate, again emphasising the fact that history and physical examination by an experienced physician is as accurate as CT in diagnosing appendicitis.[17]

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In an effort to improve diagnosis, laparoscopy both for evaluation and treatment can be an important adjunct, especially in peri-pubertal girls and children with equivocal findings. It is, however, an invasive procedure requiring general anaesthesia. The role of laparoscopic appendicectomy is still under debate with some trials showing an improved outcome[18] while others show no benefit.[19] The era of laparoscopy has, however, caused a dramatic increase in the number of medicolegal complications; how this will affect the diagnosis and management of appendicitis has yet to be elucidated.

Surgical consultation is necessary for children in whom appendicitis is suspected. There are no standard guidelines laid down for surgical referral by first-contact physicians when appendicitis is suspected, or when its classic features are absent. However, it is common practice to observe patients with acute abdominal pain of uncertain cause to allow the pathology to declare itself, thereby avoiding an unnecessary laparotomy. The intention is to resuscitate if needed, repeat abdominal examinations, and do further diagnostic studies. When used in this setting, observation should not lead to a higher rate of complicated appendicitis.

Differential diagnosis

Differential diagnosis of acute appendicitis varies depending on the age and sex of the patient and is particularly difficult in the young non-verbal child. In paediatric populations, the diseases most commonly mistaken for appendicitis are acute nonspecific abdominal pain of unknown origin, gastroenteritis, urinary tract infections, viral enterocolitis, intestinal parasitosis, primary peritonitis, dysfunctional voiding, pneumonia and uncommonly Meckel's diverticulitis.

Factors contributing to delays in diagnosing appendicitis

A variety of factors have been cited as causing delays in the diagnosis of appendicitis.[6,15,20,21] The ability to diagnose appendicitis is related to the patient's age, the history obtained and the diligence with which the physical examination is performed. In children under 3 years of age, appendicitis is virtually never diagnosed before perforation and should be considered in children presenting with a triad of abdominal pain, tenderness and vomiting.[2] Early diagnosis is further compromised by the child's inability to verbalise symptoms and endure physical examination, concern among caretakers about other more possible diagnoses, especially gastroenteritis or small-bowel obstruction, and failure by one or more physicians (67%) to diagnose appendicitis early.[5]

In a study to identify differences between correctly diagnosed appendicitis and misdiagnosed cases that resulted in litigation between 1982 and 1989, missed cases appeared less acutely ill, had fewer complaints of right lower quadrant pain, required significant analgesia for undiagnosed abdominal pain or symptoms and, more often, received an emergency room discharge diagnosis of gastroenteritis (50%) or nonspecific abdominal or urinary tract infection. Most of the patients were incorrectly sent home without counselling or instruction. After inappropriate discharge, the average time to correct diagnosis was 39 hours. Delayed diagnosis was associated with a 91% incidence of ruptured appendicitis, and required more surgery with more postoperative complications. In addition, the complication rate was higher in patients who did not receive appropriate discharge or follow-up instructions and in those whose parents did not return for repeat visits.

Diagnosis is often delayed if there is pre-admission uncertainty regarding the diagnosis or when patients/parents and physicians consider an alternative diagnosis.

In several studies[5,22] information regarding patient and physician delay was available in 93% of cases. Patient delay ranged from half a day to 45 days with a prolonged delay period of more than 3 days, more frequently seen in the misdiagnosed group. Physician-related delay ranged from 2 to 430 hours, and of interest to note, the mean physician delay was similar in acute uncomplicated appendicitis and advanced cases.[11] Another study records that 45% of their patients with ruptured appendices had previously been seen by their family physician who failed to make the correct diagnosis.[23]

Removing a 'lily white' appendix

Removing a 'lily white' appendix is primarily due to the uncertainty of diagnosing appendicitis. A normal appendix is removed in between 6% and 18% of patients. The current rate of negative appendectomies in the USA is approximately 20%.[1,21] Consensus is that this rate should be < 10%.[24] Morbidity associated with the removal of a non-infected appendix is in the order of 6 - 17%, which is significant.

Conclusions

Mistakes in diagnosis do occur and shortening the interval from symptom onset to surgery is definitely desirable. There are, however, common and universal factors/reasons for the diagnostic dilemma in acute appendicitis. All these factors are well described.

1. The typical signs and symptoms of acute appendicitis are nonspecific early on and may be varied, i.e. atypical presentation. These symptoms and signs are often under-valued and ignored.

2. It is often impossible to diagnose appendicitis on the basis of a single physical examination: indeed most errors occur after only a single examination. Repeated abdominal examination is crucial even if the child has to be admitted for observation.

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3. Absence of the classic signs and/or symptoms of acute appendicitis, pain but no nausea or vomiting, can confuse the physician.

4. Administration of narcotic pain medication followed by discharge can mask symptoms, and if required should warrant admission or further investigation.

5. Patient and parents are often sent home with only a vague understanding of signs to look for or inadequate instructions to follow.

6. Parental factors include delay in contacting health professionals, not recognising the symptoms as being significant and failure to follow instructions. When parents were encouraged to observe their sick child at home, a higher incidence of perforation occurred (65%).[4]

7. Misdiagnosis is common with treatment of a different diagnosis, i.e. urinary tract infections, gastroenteritis, pneumonia.

8. Obvious signs and symptoms are overlooked.

9. Children with appendicitis will have a higher incidence of vomiting, diarrhoea, dysuria and respiratory symptoms contributing to misdiagnosis of the condition.

10. Although appendicitis under the age of 3 years is infrequent, every effort should be made to diagnose the disease early, to reduce complications if perforation has occurred. The rate of perforation in this age group is high.

11. Red Cross Children's Hospital experience has shown that because of uncertainty about the diagnosis or symptoms mimicking other conditions, the final diagnosis and surgery were often delayed before the correct diagnosis was established. These findings again indicate the difficulty in making a diagnosis even if children have been admitted to a teaching hospital. Delay in diagnosis does not necessarily result in perforation.

Recommendations

To improve the diagnosis of acute appendicitis, shorten the clinical pre-appendectomy period, reduce associated morbidity and decrease medicolegal implications, we would like to put forward the following recommendations.

1. Always suspect the possibility of acute appendicitis in a child with acute onset of abdominal pain even if the symptoms and clinical findings are atypical. Always ask: is this, or is this not, appendicitis?

2. When in doubt, it is advisable to submit the child to active observation by repeated planned examinations[25] or to

give specific discharge instructions to the family/guardian regarding their return if the condition does not improve. Earlier follow-up should also be arranged.

3. A patient who presents with acute abdominal pain migrating to the RIF and with local signs, falls into a group that has a diagnostic accuracy of 95% for acute appendicitis. Only when the clinical presentation is equivocal or a mass is present in the RIF, should imaging modalities be utilised to establish the diagnosis.

4. We believe that this strategy would reduce time delay and lead to timely surgical intervention.

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