# Acute acalculous cholecystitis in children

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Acute acalculous cholecystitis (AAC)is a rare disease in African children and usually occurs as a complication of some other diseases, such as systemic infections. AAC can, however, be the primary pathology but, because of its low incidence, it is difficult to establish definite mechanisms and causes.

The case histories of six children (mean age seven years) with AAC are presented. They were treated in Queen Elizabeth Central Hospital, Blantyre, Malawi and in Gweru Provincial Hospital in Zimbabwe between 1985 and 1996. In four children, pre-operative ultrasonography showed the typical signs of AAC.

Cholecystectomy was performed on all seven children. One child died postoperatively from generalised sepsis and in two children a wound infection occurred.

In the tropics, where many children are seen with gastro-enteritis and other infectious diseases, the possibility of AAC must be borne in mind when a child is admitted with right upper abdominal tenderness and fever. The role of ultrasonography is emphasised. It is a reliable, non-invasive and quick investigation, which helps to establish the diagnosis before surgery.

Cholecystectomy is the treatment of choice,

because of the high incidence of necrosis of the gallbladder wall.

## Introduction

Acute cholecystitis is most commonly associated with gallstones. Acute acalculous cholecystitis in adults has been described after shock, extensive burns, in sepsis, as a postoperative complication and after receiving long-term parenteral feeding and this suggests a different pathogenesis.

AAC also has been described in children, although it is considered to be a rare condition. Ternberg<sup>1,13</sup>,in 1977, described 74 cases in children, of which 60 % occurred as a complication of other illnesses, such as gastro-enteritis and pneumonia.

Gallbladder disease is, however, rare in Africans. Onuigbo<sup>2</sup>, in Nigeria, found only 26 patients with gall bladder disease among 7,500 surgical specimens. In Uganda, Kawooya<sup>3</sup> found an incidence of 0.28% as detected by sonography and also Garrido<sup>4</sup> in Mozambique found a very low incidence of gall bladder disease in postmortem specimens in Maputo.

Biliary symptoms in the black child are often attributed to *Ascaris lumbricoides*, which may migrate into the common bile duct and cause transient obstructive jaundice. This will only occur in massive infestation of the small bowel with migration of the worms into the duodenum and common bile duct. Cholecystitis without jaundice and palpable masses in the bowel can not be

attributed to Ascaris infestations<sup>5</sup>.

In the tropics, the incidence of gastro-enteritis and other infectious diseases in children is very high and more cases of AAC would be expected, yet there are few published reports<sup>9,10</sup>.

The possibility of AAC must be borne in mind whenever a child is admitted with right upper abdominal tenderness and fever. Delay in establishing the diagnosis contributes to the high morbidity and mortality<sup>8,11,14</sup>.

### Patients and methods

Six cases of AAC in black children were seen in Queen Elizabeth Central Hospital in Blantyre, Malawi and in Gweru Provincial Hospital, Gweru, Zimbabwe between 1985 and 1996. There were four boys and two girls, with a mean age of seven years (range 3 to 11 years).

#### Case histories

- 1 A malnourished girl aged seven years presented with fever, abdominal pain, ascites, jaundice and vomiting.
  - Investigations: Hb 11.4g/dl, WBC 14.0 x 10<sup>9</sup>. Raised bilirubin but Widal negative. Ultrasonography not done.
  - She was treated by cholecystjejunostomy and histology showed a generalised *Schistosoma mansoni* infestation of the liver and gallbladder. Recovery was uneventful.
- 2 A five-year-old boy suffering from gastroenteritis was admitted with fever, pain in the right upper quadrant of his abdomen and ascites.
  - Investigations: Hb 9.4g/dl, WBC  $10.5 \times 10^9$ . Widal and malaria parasites negative. Ultrasonography confirmed a distended gallbladder with oedema of the gallbladder wall.
  - He underwent cholecystectomy and histology showed necrosis of the gallbladder wall. Recovery was complicated by wound infection.
- 3 A three-year-old boy with gastroenteritis and worm infestation was admitted with fever, jaundice, right upper quadrant pain and a palpable liver.

Investigations: Hb 10.9g/dl, WBC 29.5 x 10°, a raised bilirubin, negative Widal but positive malarial parasites. Ultrasonography showed a distended and thick-walled gallbladder.

He underwent cholecystectomy and histology showed non-specific cholecystitis. His recovery was complicated by a wound infection.

4 An 11-year-old girl was admitted with gastroenteritis complicated by fever, right upper quadrant abdominal pain and sepsis.

Investigations: Hb 10.3g/dl, WBC 6.8 x 10<sup>9</sup>, normal bilirubin and negative Widal and malarial parasites. Ultrasonography showed gross distension of the gallbladder.

She was treated by cholecystectomy and histology showed non-specific cholecystitis. Recovery was uneventful.

- 5 A five-year-old girl suffering from pneumonia, developed fever and abdominal pain with a palpable liver.

  Investigations: Hb 8.7g/dl, WBC 7.0 x 10°, normal bilirubin and both pegative Widal and
  - normal bilirubin and both negative Widal and malaria parasites. Ultrasonography showed a thick-walled gallbladder.
  - Histology of the excised gallbladder showed necrotising cholecystitis. The patient died of generalised sepsis.
- 6 A 10-year-old girl without previous history of illness developed fever, abdominal pain and vomiting.
  Investigations: Hb 13.8g/dl, WBC 8.7 x 10<sup>9</sup> with normal bilirubin and negative malarial parasites.

Ultrasonography was not done.

Cholecystectomy was done and histology showed necrotising cholecystitis. She recovered without complication.

## **Results**

Mild fever, nausea, vomiting and abdominal pains were the common symptoms. Sometimes a mass was palpable in the right upper quadrant of the abdomen. Jaundice was not a consistent feature and was found in only two of the six patients. In general the symptoms were those of an infection

and the abdominal symptoms were often overshadowed by the concomitant illness.

Laboratory tests were not really helpful and only showed that an infection was present. The Widal test is not specific. The diagnosis was reached preoperatively in four cases from ultrasonography.

Five of our patients underwent conventional cholecystectomy and, in three, necrosis of the gallbladder wall was found. In the child with extensive Schistosomiasis, it was technically impossible to remove the gallbladder, so a cholecystojejunostomy was performed.

The average hospital stay was 18.7 days. The average pre-operative illness was 6 days and the pre-operative delay was three days.

One child died due to generalised sepsis and two children had wound infections necessitating drainage and a prolonged hospital stay.

#### Discussion

AAC in children is a recognised but rare disease<sup>1,8,11,12,13,14,15,16</sup>. Ternberg and Keating described 74 cases of AAC in children and found that, in 60 %, the AAC occurred as a complication of other illnesses, most frequently upper respiratory tract infections and gastro-enteritis<sup>13</sup>. They conclude that biliary stasis and superimposed infection caused by dehydration, fever, prolonged fasting and/or prolonged ileus are the mechanisms causing AAC.

According to Glenn and Becker, however, it is not biliary stasis but vascular change in the gallbladder wall which is the main mechanism<sup>14</sup>. Severe trauma, sepsis and massive blood transfusions are conditions in which a clotting cascade may occur. Activation of the Hageman factor (factor Xll) in the clotting cascade may lead to thrombosis of the blood vessels in the gallbladder wall.

Goris, however, when describing AAC in adults, believes that biliary stasis associated with prolonged parenteral feeding and bacterial colonisation may be contributing factors only. He believes that, in acutely ill patients, systemic hypoperfusion and/or hypoxia cause decreased blood flow to the gallbladder. Torsion and narrowing of the bile ducts

have been found in these cases and are believed to have contributed. In all the cases of AAC described by Goris, positive cultures were made from the bile<sup>7</sup>. Iudin<sup>16</sup> also found organic disorders (strangulation, long cystic duct and poor evacuation) in all 18 reported cases of acalculous cholecystitis in children .

In this group of six children, no obvious gallbladder disorders were found in five cases but, in the child with schistosomiasis, there were many adhesions which may have contributed to the cholecystitis. None of the children had worm infestations or haemolytic anaemia. In one child, a bile culture was positive. Three children were admitted with gastro-enteritis and subsequently developed AAC, while one child had severe pneumonia on admission. These children could fit either causal hypothesis but the numbers are too small to reach conclusions. Most likely a combination of events causes AAC.

Ultrasonography has proved to be a very valuable and reliable non-invasive diagnostic tool in cholecystitis. It permits a rapid and accurate evaluation of the right upper abdomen. Particularly in children, this non-invasive method has great advantages. Greenberg et al<sup>6</sup> recognised three important signs of gallbladder disease:

- 1 a marked thickened sonolucent gallbladder wall
- 2 a thickened hyper-reflective irregular wall
- 3 cholelithiasis

Ultrasonography is particularly reliable in detecting calculi with an accuracy of over 90%. In AAC, no calculi are found but the thickened wall is a very typical and persistent sign and this was found consistently in our patients, who had ultrasonography.

Most authors recommend cholecystectomy because, in many series, necrosis of the gallbladder wall has been described with a high incidence of perforation. Ternberg and Keating feel that there is a place for cholecystotomy which is easier to perform in a severely ill child. It has been used frequently in the treatment of AAC and Eggermont has shown that ultrasound-guided, percutaneous transhepatic cholecystotomy can be a safe initial

## treatment in severely ill patients<sup>17</sup>

Roentgenographic studies in children who had a cholecystotomy have demonstrated a perfectly normal oral cholecystogram postoperatively. Recurrent cholecystitis with the need for a later cholecystectomy however has also been reported. Recently, Holcomb has described laparoscopic cholecystectomy in children with good results<sup>18</sup>.

We believe that, in the African setting, cholecystectomy is the treatment of choice and that cholecystotomy could be reserved for the very ill patient.

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