Cholelithiasis in childhood and adolescence

A review

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Summary

Cholelithiasis and cholecystitis are uncommon in childhood and adolescence. Misconceptions as to the aetiology, natural history, diagnosis, and therapy of these conditions prompted us to review our experience. Nineteen patients ranging in age from 7 to 18 years underwent cholecystectomy for gallstones at the University of Illinois Hospital, Chicago, between June 1970 and June 1979. Seventeen patients were female and 2 male (ratio 8.5:1). The cause was unknown in 12 patients (63%) but in 7 patients (37%) haematological disease was the underlying disorder.

The most common presenting symptom (95%) was vague right upper quadrant or epigastric pain, often ignored or mistaken for peptic ulcer pain.

Oral cholecystography demonstrated gallstones in 15 of 16 patients. Ultrasonography, used more recently, was positive in 3 patients. Intra-operative cholangiography in 12 patients, and common duct exploration in 1 additional patient, failed to demonstrate calculi. No patients were readmitted for recurrent or retained biliary calculi after cholecystectomy.

Patients and methods

A retrospective study was undertaken of all patients under 18 years of age at the University of Illinois Hospital, Chicago, who had undergone cholecystectomy for cholelithiasis during a 10-year period, June 1970 to June 1979.

Patients were evaluated in terms of age, sex, underlying predisposing factors (including parity) and presenting clinical syndrome. The diagnostic work-up consisted of plain radiographs of the abdomen, oral cholecystography, haematological studies and, lately, abdominal ultrasound examination. The operative findings, results of operative cholangiography, and the presence or absence of common bile duct stones were similarly noted.

Results

Nineteen patients ranging in age from 7 to 18 years were operated on for cholelithiasis during the period under review. Thirteen patients were Black, 4 Hispanic, and 2 White. Seventeen patients were female, 2 male (8.5:1). Four patients had sickle cell anaemia and 1 patient each had sickle thalassaemia, sickle thalassaemia and sickle cell trait; a total of 7 patients with underlying haematological problems (36.8%). Two patients had previously had abdominal surgery (10.6%). The 2 males both had sickle cell disease associated with their gallstones.

Eighteen patients presented with a syndrome of vague right upper quadrant or epigastric pain (95%) and only 2 patients presented with clinical jaundice.

Recent weight loss and obesity were not prominent features in the group under study. Nine of the females were parous or on oral contraceptives. In only 2 patients (10.5%) were radiopaque calculi seen on plain abdominal radiographs. Sixteen of the 19 patients had oral cholecystograms, 15 of which outlined stones in the gallbladder. Three patients had ultrasound study of the gallbladder, all proved to have gallstones at operation. Therapy in all cases consisted of uncomplicated surgical ablation of the gallbladder. Twelve patients had intra-operative cholangiography and 1 patient underwent common bile duct exploration; no stones were found in the common duct in any of these 13 patients. No patient has returned with retained or recurrent common duct stones, although it should be borne in mind that the period of follow-up is short. Overall 1% of patients who underwent cholecystectomy were children.

Discussion

The assessment of the child or adolescent with abdominal pain presents a challenge of some magnitude to the clinician. Cholelithiasis and cholecystitis are uncommon in children and are often excluded in the differential diagnosis of abdominal pain in these patients. Autopsy studies have revealed that the incidence of cholelithiasis in children is less than 0.1%, although the clinical incidence varies from 0.13% to 1.5% in quoted series of all patients with biliary tract disease, and this would confirm our findings. Cholelithiasis occurs more commonly than acute cholecystitis and no patient in this series presented with the classic features of acute cholecystitis. The age range is variable, and calculi have been observed in stillborn fetuses. Females predominated in...
our series as they do in most other published reviews, although one study claims a male preponderance. The racial incidence is of importance when viewed in relation to the occurrence of haemolytic disorders. The racial variation also directly reflects the hospital population and the community it serves; it explains the greater number of Black children with cholelithiasis in our series. Similarly geographical, genetic, and dietary factors may also need to be assessed, as evidenced by the high incidence of biliary tract disease in Pima Indians in the USA.10

The causes of gallstones in children have been grouped by Brenner and Stewart1 into six categories: (i) infection and infections; (ii) anatomical — congenital and acquired, or external compression; (iii) constitutional, including obesity, recent weight loss, and hypertension; (iv) metabolic (hypercholesterolaemia) and hormonal; (v) hereditary — 20% of patients have a familial history in some series;11 in others, as in ours, this was absent;11 (vi) haemolytic.

Many of the literature reviews cited have various postulates on aetiology, some of which fall into the categories mentioned above. Blanket statements that ‘gallstones are commonly due to enough periods’14 in this series, the children with haemolytic disorders were all referred for elective cholecystectomy after presenting with vague abdominal and/or right upper quadrant pain.

The clinical presentation varies from florid acute cholecystitis to almost no symptoms at all. A history of fatty food intolerance was not recorded in any of the patients in this series, although 14 - 35% of patients in other studies volunteered this information.13,14,16 Younger children will rarely complain of or confirm fatty food intolerance.16 Sixty per cent complain of pain in the epigastrium and right upper quadrant,4 a finding which our series confirms. The specific symptoms of nausea and vomiting together with pain can present a confusing array of differential diagnoses and resulted in delayed recognition of up to 10 years in one series.11 The presence of jaundice should be carefully looked for. Most cases are haemolytic or inflammatory in origin, but may still be due to a common duct stone. Only 1 patient in this series had jaundice due to hepatitis, but in other series an incidence of 24-25% has been reported.1 A mass in the right upper quadrant is rare and found only in 10,7% of cases.1

The pre-operative rate of misdiagnosis is said to be as high as 18-20%, hence the need for accurate pre-operative work-up. The plain film of the abdomen will demonstrate only 10% of calculi and, as confirmed in our study, had limited value. One study, however, claims that it is of diagnostic importance.17

Oral cholecystography is confirmatory in 75-80% of cases, with a 95% positivity rate in our series. The technique for children is similar to that for adults, except for infants in the first few months of life.4 Abdominal ultrasonography is a non-invasive technique for evaluating the biliary tree, and will accurately indicate the presence of gallstones in 90% of patients.18,19 It will demonstrate the presence or absence of dilated extra- and intrahepatic ducts. In 2 of the 3 patients in this study who underwent ultrasonography it was the only pre-operative investigation and was completely successful. It is an easy, cheap outpatient study and has an important role in the diagnosis of abdominal pain in children and in those in whom the results of oral cholecystography are questionable. For children in our institution it has become the first-line diagnostic procedure in the work-up of cholelithiasis. Recently, radionuclide imaging of the biliary tract has come into use with the development of 99mTc-labelled biliary tracers.20 Other routine laboratory assays are of importance, notably that of the alkaline phosphatase level to assess hepatic excretory function.

Surgery is the only form of therapy for cholelithiasis if morbidity and/or mortality is to be avoided.1,12 In our series there were no deaths or complications; this compares with a quoted operative mortality rate of 0.9%.4 Common duct stones are rare in children, occurring in only 6-10% of reported cases,14 and the role of intra-operative cholangiography is therefore controversial.17 In our series, 13 patients had intra-operative cholangiography and 1 patient underwent common duct exploration because of the presence of jaundice. All procedures failed to show the presence of common duct stones, despite the presence of small stones in the gallbladder. We no longer perform operative cholangiography routinely, or common duct exploration, unless dilated ducts are seen pre-operatively on ultrasonography or are found in the operating room, or there is a history of pancreatitis or jaundice in the absence of haematological disease.

In children with haemolytic anaemia concomitant splenectomy has been frowned upon,6 but with modern techniques the procedure is safe;6 where necessary we remove the spleen.

Conclusions

On the basis of our findings and a review of the literature to date we may make the following points:

1. Cholelithiasis and cholecystitis are uncommon in childhood.
2. Clinical signs and symptoms are nonspecific, the main symptom being epigastric pain.
3. The cause is unknown in the majority of patients, and related to haematological disease only one-third of cases.
4. In the absence of jaundice, oral cholecystography is reliable for demonstrating stones in the gallbladder.
5. Ultrasound is an important diagnostic tool for demonstrating stones in the gallbladder as well as the presence of dilated intra- or extrahepatic bile ducts.
6. Operative cholangiography should not be routine in children and should be reserved only for patients in whom dilatation of the intra- or extrahepatic bile ducts is demonstrable either by ultrasonography (pre-operatively) or at the time of cholecystectomy.

REFERENCES

Fasting plasma glucose and glycosylated haemoglobin levels in the assessment of diabetic control in non-insulin-dependent diabetes in the young

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Summary

Response to diet and drug therapy was assessed in a group of 85 Indian patients with non-insulin-dependent diabetes in the young (NIDDY). There was a significant decrease in fasting plasma glucose (FPG) values on therapy (pretreatment 13.3 ± 0.5 mmol/l; post-treatment 9.7 ± 0.4 mmol/l) (P<0.001). Prior to therapy the majority of patients had either moderate (40%) or severe (59%) diabetes; on therapy, the majority had either mild (21%) or moderate (62%) diabetes. Estimation of glycosylated haemoglobin (Hb A1) levels revealed that control was excellent (Hb A1 <10%) in 47% of patients and excellent or adequate (Hb A1 ≤12%) in 78%. Hb A1 levels correlated significantly with the FPG value (r=0.78; P<0.001). In 8 patients with iron deficiency anaemia the Hb A1 level did not fall within the correlation norms between Hb A1 and FPG. Treatment of the anaemia restored the correlation norms. Hb A1 levels were significantly higher in patients with microvascular complications (12.1 ± 0.8%) than in those without any vascular complications (10.3 ± 0.9%) (P<0.01).

Reports on non-insulin-dependent diabetes in the young (NIDDY) in its most classic form with autosomal dominant inheritance (MODY) would suggest that it is a mild non-progressive disorder bordering on chemical diabetes and that good control is usually achieved by diet and the use of oral hypoglycaemic agents.

Campbell also obtained good control with oral hypoglycaemic agents (mainly chlorpropamide) and dietary restriction in Natal Indians with NIDDY. However, Cosnett found that oral therapy resulted in improvement in less than 20% of his group of Indians with NIDDY. It was therefore of interest to study the control achieved by diet and oral hypoglycaemic agents in the much larger group of Indians with NIDDY which we have been studying, using more rigorous criteria in assessing the control achieved.

Patients and methods

The 85 Indians with NIDDY studied have been reported on previously and full clinical details have been given. All were advised to adhere to a low-carbohydrate diet by a dietitian and issued with a diet sheet.

Since euglycaemia could be achieved in only 1 patient by diet alone (she had a normal fasting plasma glucose level), therapy with the oral hypoglycaemic agents was instituted in the remaining 84 patients. The drugs used and the maximum doses offered are given in Table I. Chlorpropamide or glibenclamide, usually in combination with phenformin, was most commonly used. All patients were followed up for a minimum period of 1 year on therapy. The patients were seen monthly, and in addition to a clinical assessment and examination of the urine for glucose, ketones and albumin, blood samples were withdrawn for plasma glucose and glycosylated haemoglobin (Hb A1) estimation. Hb A1 levels were determined on the same day that the samples were taken, by cation exchange chromatography at 22°C.

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Date received: 12 February 1982.