PANCREATITIS—ACUTE AND RELAPSING—IN INFANCY AND CHILDHOOD

DAVID STEIN, M.B., F.R.C.S. (ENG.), *Department of Surgery, University of Cape Town*

Acute pancreatitis has been encountered in 11 children admitted to the Red Cross War Memorial Children's Hospital in Cape Town over the past 5 years. This article is based on these cases and gives a review of the literature concerning acute pancreatitis in children. It has been widely accepted that pancreatitis in childhood is a rare disease. Nevertheless, the disease occurs commonly enough to be considered in every case of abdominal pain, and it appears that the more conscious one is of the disease, the more commonly is the diagnosis made. Scanning the literature one finds only sporadic reports of pancreatitis in childhood under the age of 13 years.

In 1927 Schmieden and Sebening[1] studied 1,510 cases of acute pancreatitis, the youngest patient in their series being 13 years of age. In 1935, Dobbs[2] reported a 12-year-old child with acute pancreatitis, and he reviewed 14 additional cases from the literature. There have been subsequent articles by various authors reporting single cases of acute pancreatitis.[3-26]

In 1957 Blumenstock and his co-workers[3] reported 4 new cases and reviewed the literature up to 1957. To date there are reports in the literature of a total of 54 cases of pancreatitis occurring in children.[1-26] We have had 11 cases at the Red Cross War Memorial Children's Hospital, and none of these has previously been reported. The case reports of these 11 children are set out hereunder:

CASE REPORTS

Case 1

E.B., an 8-year-old girl, was well until 1 day before admission, when she complained of cramp-like abdominal pain followed by profuse vomiting and loss of appetite. There was no history of worms.

Examinations revealed an ill-looking child with a temperature of 101°F. and a pulse rate of 130 per minute. There was no jaundice. Her abdomen was tender in the peri-umbilical region and in the left loin, and rectal tenderness was noted. There was no rigidity. Bowel sounds were present. Her other systems were normal. Haemoglobin 13·5 G. per 100 ml.; white blood count 29,400 per c.mm.

Operation was performed the day after admission. The peritoneum contained free blood-stained fluid and there was considerable fat necrosis in the mesentery and omentum. The lesser sac contained free fluid and the pancreas showed marked evidence of haemorrhagic pancreatitis, the tail of the pancreas being more involved than the head. The abdomen was drained and the child made an uncomplicated recovery.

On the second postoperative day an ascaris worm was aspirated *via* the stomach tube. The drainage wound was dry by the 15th postoperative day. The serum-amylase level on the first postoperative day was 458 Somogyi units (normal 50-150), and this dropped to 400 a few days later. An intravenous cholangiogram was normal. The patient has been free of symptoms since discharge from hospital 3½ years ago.

Case 2

J.W., a 10-year-old girl, was well until 2 days before admission when, after eating some doughnuts, she vomited a great deal and collapsed with abdominal pain. She continued vomiting profusely and lost her appetite.

She was an ill, toxic child, showing 5% dehydration. Her blood pressure was 90/70 mm.Hg and her temperature was 99·4°F. The abdomen was not distended, but there was generalized tenderness, and it had a doughy feel. No bowel sounds were audible. Haemoglobin 13 G. per 100 ml.; white blood count 16,800 per c.mm. Electrolytes were normal. The serum-amylase level was 400 Somogyi units. The other systems were normal. Urine was normal.

Laparotomy was performed on the day of admission. The abdomen contained free blood-stained fluid, and fat necrosis was present throughout the abdominal cavity. The pancreas showed evidence of typical pancreatic necrosis. The gallbladder and liver were normal. No drain was inserted. Histology confirmed fat necrosis.

Postoperatively she passed 2 roundworms per rectum. She developed a raised temperature on the 4th day which persisted despite antibiotics, and a tender mass became palpable in the epigastrium on the 10th postoperative day.

A second operation was performed on the 21st postoperative day, when a typical pseudocyst of the pancreas was drained into the stomach by anastomosing the adherent anterior wall of the cyst to the posterior wall of the stomach. There was a dramatic drop in temperature and the patient made an uncomplicated recovery. Subsequent barium-meal examination failed to demonstrate the transgastric cystogastrostomy. Intravenous cholangiography was normal and she remains well 2 years after her discharge from hospital.

Case 3

S.M., a 4-year-old boy, was well until about 12 months before admission. During those 12 months he had had intermittent attacks of abdominal pain, vomiting and distension. Between the attacks his appetite had been good. He had had an intermittent dry cough and his mother thought that he might have had small haemoptyses. He had passed several roundworms per rectum.

Examination revealed a well-nourished child. His temperature and pulse were normal. There was a large, firm nodular mass filling the epigastrium, extending under the costal margin on the left and reaching up to the right hypochondrium. The liver edge was just palpable and the spleen could be tipped below the left costal margin. Rectal examination was normal. Haemoglobin 12·5 G. per 100 ml.; white blood count 9,600 per c.mm. Differential count showed an eosinophilia of 9%. The erythrocyte sedimentation rate (ESR) was 40 mm. in the 1st hour (Westergren). Stools showed no fat globules or undigested fibres of meat. Trypsin was negative. Urine was normal.

Laparotomy was performed through an upper transverse incision. A large pseudocyst of the pancreas was present in the lesser sac and was adherent to the posterior surface of the stomach. A transgastric cystogastrostomy was performed. Several ascari were present throughout the gastro-intestinal tract.

The patient made an uncomplicated recovery and post-operative barium studies failed to show filling of the cyst. He was given an anthelminthic (piperazine citrate) before his discharge from hospital, with good effect, and there have been no episodes resembling pancreatitis in the 5 years since his discharge from hospital.

Case 4

D.A., an 8-year-old boy, fell out of a tree on to his abdomen 2 weeks before his admission. He complained of abdominal pain and vomited intermittently during the next week. These symptoms were associated with pyrexia, anorexia and progressive abdominal distension.

Examination revealed a malnourished child with a temperature of 100°F. There was diminished air entry and dullness at the left lung base. There was a soft doughy feel to the abdomen and a visible mass filled the epigastrium. This mass was smooth and soft with a well-defined inferior edge, but the edge was not well-defined laterally. The mass moved only slightly with respiration. The cardiovascular and central nervous systems were normal. Haemoglobin 10 G. per 100 ml.; white blood count 15,000 per c.mm. with a polymorphonuclear leucocytosis, but no eosinophilia. The ESR was 112 mm. in the 1st hour (Westergren). Urine was normal. X-rays showed a left-sided pleural effusion, and the left leaf of the diaphragm was slightly elevated. The stomach gas bubble was displaced forwards and upwards by a large mass occupying almost the entire upper half of the abdomen. There was an ununited fracture of the 12th right rib. The pleural effusion was aspirated and straw-coloured fluid containing polymorphs and lymphocytes was found; it was sterile on culture.

Operation was performed the day after admission. There was a turbid peritoneal exudate, and a large pseudocyst of the pancreas arising from the lesser sac and displacing and pressing on the stomach was found. A transgastric cystogastrostomy was performed. The patient made an uncomplicated recovery and has been symptom-free for the 3 years since the operation. Barium studies failed to show filling of the cyst.

Case 5

C.P., an 8-year-old girl, was well until 3 days before admission, when she vomited profusely and developed periumbilical cramp-like pains which radiated to the left of the abdomen. There was no distension, nor was there any history of worms.

Examination revealed an ill-looking child with a temperature of 101°F. Her tongue was furred. The abdomen felt tumid and there was guarding and tenderness in the upper half, especially in the left upper quadrant and loin. The other systems were normal. Haemoglobin 12 G. per 100 ml.; white blood count 21,000 per c.mm. Urine was normal.

Operation was performed the day after admission and bloodstained fluid and marked fat necrosis were found. The pancreas was enlarged and boggy. The gallbladder was normal, and several roundworms were seen throughout the gastro-intestinal tract. Nothing further was done and the abdomen was closed. The serum-amylase level on the 7th postoperative day was 320 Somogyi units. She was given an anthelminthic and has remained well for 1 year since the operation.

Case 6

G.K., a 6-year-old girl, developed epigastric pain associated with vomiting on the afternoon of admission. She had often passed roundworms in her stools and had a slight cough. For the past 2 years she had had repeated attacks of abdominal pain, usually lasting only 2 days.

Examination revealed a slightly dehydrated child with a temperature of 100·5°F. There was no jaundice. A vague mass filled the epigastrium, and it moved slightly with respiration. It was smooth and there was overlying tenderness and guarding. No hepatomegaly was noted. Haemoglobin 12 G. per 100 ml.; white blood count 17,000 per c.mm. Electrolytes were normal. Serum-bilirubin level 1·8 mg. per 100 ml. Serum-amylase level 1,200 Somogyi units. Biligrafin studies were unsatisfactory. Her other systems were normal. Urine was normal.

She was treated conservatively and the pyrexia and fullness in the epigastrium subsided by the third day. Subsequently the serum-amylase level fell to 133 Somogyi units. She was given a dose of piperazine citrate and passed a large number of roundworms. She has been well for the 3 years since her discharge from hospital.

Case 7

H.W., a 10-year-old boy, developed severe cramp-like abdominal pains and vomiting 2 days before admission. He was a well-nourished child with a normal temperature and pulse rate. There was no abdominal distension, but there was tenderness in the upper abdomen and left iliac fossa, associated with some guarding and rebound tenderness, but no rigidity. The other systems were normal. Haemoglobin 12 G. per 100 ml.; white blood count 14,000 per c.mm. Urine was normal. X-ray of the abdomen showed no abnormality.

A laparotomy was performed on the day of admission and this revealed extensive fat necrosis in the omentum and mesentery. This was confirmed on histology. Several roundworms were noted in the small bowel. Nothing further was done. An intravenous cholecystogram was normal.

The patient was readmitted 4 months later with intestinal obstruction caused by a single adhesive band which was divided at operation. He did well, but a month later was again

admitted with epigastric pain and vomiting. There was no constipation. Examination revealed guarding and tenderness over the right half of his abdomen, maximal in the epigastrium. Bowel sound were audible but reduced. Haemoglobin 16 G. per 100 ml.; white blood count 8,000 per c.mm. X-ray of the abdomen showed no abnormality. His serum-amylase level was 320 Somogyi units, and a diagnosis of relapsing pancreatitis was made. With conservative treatment his pain disappeared and the serum-amylase level dropped to normal. He was treated with piperazine citrate and passed several roundworms. He is still well 4 years later.

Case 8

D.S., a 6-year-old boy, was quite well until 1 week before admission when he vomited his food. Soon afterwards a roundworm crawled out of his nose. He was given 'de-worming medicine' and subsequently passed 22 worms per rectum. Three days later he developed generalized abdominal pain which later became localized to his epigastrium. The pain was constant and he was most comfortable while sitting up and leaning forwards.

Examination revealed an ill-looking child with a furred tongue and a temperature of 103°F. There was no jaundice. The air entry was diminished at the right lung base. There was a large, smooth, cystic, tender mass filling the epigastrium; the mass did not move with respiration. No other masses were palpable and his cardiovascular and central nervous systems were normal. X-ray of the chest showed pleural thickening and consolidation of the lower lobe of the right lung. There was no effusion. X-ray of the abdomen showed an indefinite mass in the epigastrium, displacing the stomach to the left and the colon downwards. The serum-amylase level was 64 Street-Close units (normal 6 - 33). Bilirubin was within normal limits. Urine was normal.

He was treated by intravenous therapy, gastric suction, atropine and 'chloromycetin'. His temperature subsided on the third day. Thereafter the mass rapidly diminished in size and by the fifth day it was only just palpable. The serum-amylase level decreased from 64 Street-Close units on the day of admission to 43 units 2 days later, and 14·8 units at the time of his discharge from hospital, when the mass had disappeared entirely. His glucose-tolerance curve at this stage was normal. He has been free of symptoms for the 8 months since his discharge from hospital.

Case 9

R.A., a 6-year-old boy, was well until 1 hour before admission, when he developed central colicky abdominal pain associated with vomiting. There was no history of worms.

Examination revealed a well-nourished but ill child with a temperature of 100°F. There was slight abdominal tenderness and distension in the epigastrium. The other systems were normal and the urine clear. Haemoglobin 13 G. per 100 ml.; white cell count 21,000 per c.mm. There were numerous ova of *Ascaris lumbricoides* in the stool. The serum-amylase level was 2,280 Somogyi units.

A diagnosis of pancreatitis caused by worms was made and he was treated conservatively, with rapid improvement. He was given a course of piperazine citrate and has remained symptom-free for the 7 months since his discharge from hospital.

Case 10

D.J., a 6-year-old boy, developed abdominal pain following a kick in his abdomen. The next day he was vomiting profusely, and brought up a roundworm the day after that. The pain was dull and continuous.

He was an ill child with a pulse of 130 per minute and a temperature of 100·5°F. There were distension, tenderness and guarding over the whole abdomen, but there was no rigidity and bowel sounds were inaudible. Haemoglobin 15 G. per 100 ml.; white blood count 14,800 per c.mm. The serum-amylase level was 67 Street-Close units. The serum-bilirubin level was 0·8 mg. per 100 ml. X-ray showed a slight pleural reaction at the left lung base, and the posterior aspect of the left leaf of the diaphragm was slightly elevated. The abdomen showed gas in the small and large bowel in a pattern suggestive of ileus. Urine was normal.

Laparotomy was performed on the day of admission. Free clear fluid was present and the pancreas was grossly oedematous. The gallbladder and common bile duct were normal, but many roundworms were palpable in the small intestine. No drain was inserted.

The child made an uneventful recovery. The serum-amylase level on the 12th postoperative day was 48 Street-Close units. He has been free of pain for the 10 months since his discharge from hospital.

Case 11

M.v.B., an 11-year-old girl, fell on to the handlebars of her bicycle and injured her abdomen 6 weeks before admission. This was followed by a great deal of abdominal pain and some vomiting. Despite bed rest, the pain persisted, and after an interval of several days a mass was palpated in her epigastrium. A laparotomy was performed elsewhere and oedema and induration of the pancreas were found. The palpable mass was a large area of fat necrosis in the omentum. There were several other areas of fat necrosis, and one was biopsied. No further operative procedure was carried out, and her general condition improved, but after several days she again complained of abdominal pain and backache and she was transferred to our hospital where she also complained of pain in her elbows and knees.

She was an obviously ill child. Her temperature was 101°F., and her pulse 158 per minute. There was evidence of recent weight loss, and there were several plum-coloured plaques on her legs, varying from 2 to 3 cm. in diameter, and raised about 1 mm. above the skin surface. These were almost identical to erythema nodosum in appearance and were warm and tender to the touch. The chest was normal, but a pericardial friction rub was heard over the apical area of the heart. Her abdomen was markedly distended with ascites. Haemoglobin 9·5 G. per 100 ml.; white blood count 14,750 per c.mm. The differential count was: 67% polymorphs, 16% lymphocytes, 2% monocytes and 15% eosinophils. Her ESR was 92 mm. in the 1st hour (Westergren). The urine was quite normal. The serum-amylase level was 8 Somogyi units.

A diagnosis of septicaemia was entertained, but repeated blood cultures were negative. The abdominal pain persisted and the patient vomited on several occasions. On the second day of admission the pain became very severe and radiated into her back. She now had more abdominal distension and tenderness and the bowel sounds became inaudible. She also complained of pain in her left knee and both elbows. Another red erythematous nodule, resembling erythema nodosum, was noted on her buttock.

Despite intensive antibiotic therapy the toxic state and rapid pulse persisted, and intravenous cortisone was commenced. By the next day there was a dramatic improvement, which was maintained. Three days later a skin biopsy of one of the erythematous nodules was performed and the histology was that of undoubted fat necrosis. The serum-amylase level was now repeated and was found to be 1,068 Somogyi units; the following day it was 3,000 units.

An abdominal paracentesis was performed, and 3,300 ml. of turbid, straw-coloured fluid was removed. The amylase content of this fluid was 53,000 Somogyi units. No increase in the serum lipids was noted. It was at this stage, i.e. 10 days after admission, that new areas of fat necrosis were noted in the skin overlying her calves, and 'probanthine' therapy was commenced.

The patient's condition gradually improved, the ascites resolved, the areas of fat necrosis of the skin cleared up and her pyrexia subsided. One month after admission to the Red Cross War Memorial Children's Hospital, her serum-amylase level was down to 267 Somogyi units.

She still complained of pain in the right forearm and there was now obvious bony swelling of the upper third of the ulna. There was also tenderness of the right humerus. X-ray of this humerus showed numerous small cystic translucencies with an overlying periosteal reaction. Similar changes of a lesser degree were present in her radius and ulna. The left humerus was also affected, but less so. No other bones appeared to be involved. The X-rays were repeated 2 weeks later, and marked progression was noted. There were numerous areas of bone destruction at the lower end of the right radius and to a lesser extent

on the left. There were multiple cystic areas in the upper ends of both bones on both sides.

The cortisone was discontinued 3 weeks before discharge from hospital and her condition continued to improve, the nodules in the skin completely resolved, and all her pain disappeared. The ascites did not re-collect. X-rays taken at later intervals showed a gradual re-ossification of the involved bones. No further attacks of pain have occurred.

This case, in view of its interesting features of disseminated fat necrosis in skin and bone, will be reported in greater detail by other members of our staff.[27]

DISCUSSION

Management of our cases was basically similar and was modified only because of difficulty in diagnosis. Of our 11 cases, 6 were identified only at operation for an 'acute abdomen'. When the pathological changes were detected, nothing further was done at operation, except in one patient in whom the lesser sac was drained. Three cases were recognized on admission; treatment in these was conservative and was the same as the usual postoperative regime given for the patients in this series. This consisted of gastric suction, intravenous fluid replacement, atropine intramuscularly to act as a vagal inhibitor, and antibiotics to combat any secondary infection of the pancreas. Pethidine was given if the pain persisted. In case 11 cortisone was given with a marked beneficial effect. In our later cases we added calcium gluconate to the intravenous therapy in view of the work done by Edmondson and Berne,[28] who showed that more than twice as much calcium as the total present in the circulating blood volume may be deposited in the areas of fat necrosis.

Conservative measures continued until all signs of the acute episode were over. An anthelminthic (piperazine citrate) was given to our patients if ascari were seen at operation or identified by vomiting, the passage of roundworms per rectum, or the recovery of ova in the stools. This was possible only several days after the acute episode had subsided.

Three methods of estimating the serum-amylase levels were used, i.e. Somogyi, Street-Close and Wohlgemuth. The method used in a particular case depended entirely on the one in use in the pathology laboratory at the time.

The serum-amylase level was highest at the onset of the disease and rapidly returned to normal. Continued high readings indicated activity of the disease or the formation of a pseudocyst.

Three patients were operated on for pseudocyst of the pancreas — one of them 3 weeks after laparotomy at the acute stage, when only fat necrosis and pancreatic oedema were seen. All 3 were treated by transgastric cystogastrostomy. This was done by opening the stomach and incising the posterior wall, which was adherent to the pseudocyst. The opening was anastomosed to the cyst wall with a continuous catgut suture, and the anterior incision in the stomach was then closed. These 3 patients have all done well and have remained asymptomatic. Postoperative barium studies failed to show any filling of the cysts.

I have been informed[29] of a 14-month-old child who presented with abdominal pain; an operation performed in Kuala-Lampur revealed an abdominal mass which was thought to be malignant. She was transferred to Great Ormond Street Hospital for Sick Children, where a pseudocyst of the pancreas was found and marsupialized to the anterior abdominal wall. Although the child made a satisfactory recovery, the fistula took one year to close.

In none of our cases was exploration of the ampulla of Vater or of the common bile duct done, and all our patients seem to have recovered satisfactorily. Unless some specific pathological change is seen at the initial attack of acute pancreatitis, exploration does not appear to be indicated, but in relapsing cases where hyperlipaemia is excluded, exploration may be called for (see later).

Aetiological Factors

Trauma

Trauma is a well-known aetiological factor in pancreatitis, and certainly a relatively common cause of pancreatitis occurring in childhood. At least 6 cases have been recorded in the literature, the injury usually being a severe blow to the abdomen.

Dobbs[2] reviewed 3 cases and pointed out that 1 of these developed symptoms of abdominal pain and vomiting only 2 days after being struck on the abdomen. Venable[4] published a case occurring in an 11-year-old girl, who developed pain and vomiting 5 days after receiving a blow to her abdomen. Adams[5] encountered a similar problem in a 7-year-old boy; he, too, only developed pain 3 days after being struck. All these patients were operated on and all recovered. Shallow and Wagner[6] reported an 8-year-old boy who developed pain immediately after he was struck on the abdomen. His serum-amylase level was markedly elevated and he was treated conservatively with complete recovery.

In our series there were 3 patients with pancreatitis caused by trauma, all developing pain immediately after being struck on the abdomen. One of them (case 4) presented 5 days after the onset of symptoms with a pseudocyst of the pancreas. These 3 patients were operated on and recovered completely, but case 11 had a stormy postoperative course and developed metastatic fat necrosis of the skin and bone marrow.

Mumps

None of our patients had a history of mumps infection or contact, but we did not do the complement-fixation test on any of them. Mumps pancreatitis is not infrequent, but seldom causes anything more than oedema of the pancreas and is hardly ever fatal.

Serum-amylase levels are of no value in making the diagnosis of mumps pancreatitis, because salivary-gland inflammation also causes a rise in the amylase level. Zelman[30] saw 600 cases of mumps during an epidemic, and of the cases he investigated, 73% had a raised serum-amylase level. He also showed that there was no correlation between the severity of the pancreatitis and the serum-amylase level. The diagnosis of mumps pancreatitis is therefore a clinical one, based on the presence of abdominal pain and tenderness. A patient suffering from mumps pancreatitis was operated on by Farnum,[31] who found a diffusely swollen pancreas and bloodstained peritoneal fluid. No fat necrosis was seen. Katz[29] reported a similar picture of pancreatic oedema in a patient operated on for abdominal pain during an attack of mumps.

Helminth Infestation

Ascaris lumbricoides is the commonest helminth found, usually in association with *Trichuris trichiura* and *Enterobius vermicularis*. Van Rensburg and Kark[32] claimed that 80% of the African and Indian hospital patients in the Durban area were infested with these parasites. A limited survey of children seen at the Red Cross War Memorial Children's Hospital has shown that 30% of them are infested with ascaris.[33]

The ascaris is one of the largest human parasites. The female is 25 - 35 cm. in length and 5 mm. in breadth. The male is smaller. The female lays up to 200,000 fertilized eggs per day in the intestine of man, the host. These are passed in the stool and deposited in the soil, where embryonation takes place over 9 - 13 days. There is no intermediate host and the ova are ingested with contaminated food. In the small bowel the covering membrane is digested and the larvae are set free. They burrow through the intestinal wall and reach the lungs *via* the lymphatics and venules, the liver and the right side of the heart. In the lung capillaries growth takes place up to 10 times the initial size of the parasite, and they then enter the bronchioles and migrate up the trachea to the epiglottis where they are swallowed. (It is thought that this migration may account for the cough occasionally present in children with worms.) By this time they are 2 - 3 mm. in length and take 2 months to develop into mature worms. Their lifespan is about one year. The adult ascaris may wriggle itself into any orifice; they may crawl up the nasopharynx into the eustachian tube, and if a perforated eardrum is present, escape out of the ear!

In our series, the illness in 6 patients had a very definite relationship to worms, although the evidence incriminating them as the direct cause of the pancreatitis is tenuous. In one patient (case 8) the pancreatitis appeared to be related to the administration of 'de-worming medicine'. Patients 6 and 8 had clinical and laboratory evidence of pancreatitis with definite relationship to ascaris infestation. Both were treated conservatively and de-wormed with piperazine citrate after the acute episode. Like all our other patients, they recovered completely.

There need not necessarily be a worm in the pancreatic duct to cause pancreatitis, because a worm in the common duct may produce a similar picture. Cywes and Krige,[34] of the University of Cape Town Medical School, have shown that intravenous 'biligrafin' combined with tomography can demonstrate such worms in the common duct. In Vietnam, Tong-That-Tung *et al.*[35] have published their results in the treatment of 189 cases of pancreatitis in adults caused by ascaris infestation of the common bile duct. Unlike us, they advise immediate surgical removal of these ascari. Their mortality rate is 17%. In our series the disease was recognized in most cases only after laparotomy, but in direct contrast to Tong-That-Tung *et al.*[35] we have never opened the bile duct and our patients have all responded well to conservative measures.

Pancreatitis caused by ascaris has been regarded as a rarity, but in view of our findings and several reports in the literature, this is probably not so. Dobbs[2] reported 3 cases of pancreatitis in childhood associated with ascaris. The first child, aged 2 years, developed acute abdominal pain and vomiting and died without operation. Postmortem examination confirmed acute haemorrhagic pancreatitis, and although no cause was found, a roundworm was vomited shortly before death. The second patient, also a child of 2 years, developed abdominal pain and vomiting. Laparotomy showed acute pancreatitis. Shortly after the operation she vomited a roundworm and recovered completely. The third patient, a girl of 12 years, was originally reported by Novis,[7] and she presented with a 12-day history of abdominal pain and vomiting. She had an epigastric mass, and laparotomy confirmed the diagnosis of acute haemorrhagic pancreatitis. The pancreatic duct was opened from head to tail and 2 ascaris worms were removed from its lumen. The pancreas was repaired with interrupted catgut and she made an uneventful recovery.

Jaeger[8] reported a case where an ascaris worm was found in the pancreatic duct of a 2-year-old child. Duncan[9] reported the case of an 18-month-old boy with an abdominal mass which was diagnosed as an abscess and drained. Five days later the child died and at postmortem examination an ascaris was found in the remains of the head of the pancreas, which was the site of the abscess. Moore[10] recorded postmortem findings in a child where there was pancreatitis of the body and tail of the pancreas. Two ascari were found in the pancreatic duct of the body and an ascaris was found in a small necrotic cyst in the tail of the pancreas.

Steroids

In none of our cases was the disease caused by steroids. Baar and Wolff[11] reported 2 cases of pancreatitis, the first in a girl of 11 years with severe asthma, who was treated with cortisone. She died suddenly following a prolonged asthmatic attack and postmortem examination revealed acute haemorrhagic pancreatitis. The second patient was a boy of 3½ years, who for a year suffered from progressive dermatomyositis. He was treated with cortisone and was admitted one month before his death with abdominal pain and vomiting which responded to conservative measures, and just before he died he again had abdominal pain and vomiting. Postmortem examination revealed gross acute haemorrhagic pancreatitis.

Baar and Wolff suggested that cortisone is the aetiological factor in these cases, especially in view of the experimental work done by Stumpf and his colleagues,[36,37] who produced pancreatic lesions in cortisone-treated rabbits. Marczynska-Rabowska[12] reported a case of acute haemorrhagic pancreatitis in a boy of 5 years of age suffering from Still's disease. He was treated for 4 months with corticotrophin. He developed generalized abdominal pain and vomiting, deteriorated rapidly and died. Postmortem examination showed fat necrosis of the omentum and pancreas. The author suggested that the pancreatitis was due to the corticotrophin.

Common Bile Duct Pathology

Pathological changes in the common bile duct have been responsible for 4 cases of acute pancreatitis reported in the literature. In 3 of these the disease was of the relapsing type, and all have recovered.

Dobbs[2] reviewed a case published originally by Schmieden and Sebening.[1] This was a 13-year-old girl who was operated on for cholecystitis. The presence of pancreatitis with fat necrosis was noted. A stone was removed from the ampulla of Vater with complete relief.

Blumenstock *et al.*[3] reported a case of relapsing pancreatitis starting at the age of 15 months. Laparotomy at that stage revealed fat necrosis and a small perforation in the right hepatic duct, which was thought to be due to the pancreatitis. This was closed, and postoperatively the child had repeated attacks of pain, necessitating several admissions to hospital, each time with epigastric tenderness and a significant rise in the serum-amylase level. At the age of 5 years she had another operation and a grossly dilated common bile duct was found, with an obstruction just proximal to the ampulla of Vater. A probe could be passed *via* the ampulla into the pancreatic duct, but not into the common bile duct. A cholecyst-jejunostomy was performed and she has remained free of symptoms for 3 years since the operation.

Plechas[13] reported a patient who had a history of recurrent attacks of abdominal pain since the age of 2 years. Laparotomy at the age of 10 years showed no abnormality. On his fourth admission to hospital for abdominal pain, another laparotomy was performed and this showed acute pancreatitis with fat necrosis. Recurrent attacks of pain persisted, and at 17 years of age yet another laparotomy revealed a scarred ampulla with a common bile duct twice its normal diameter, showing clinical evidence of inflammation. The pancreatic duct was 10 times its normal size. After sphincterotomy there was a smooth postoperative course and the patient has been symptom-free for over a year.

Gibson and Haller[14] reported a 4-year-old Negro child with a 7-day history of abdominal pain and vomiting. The serum-amylase level was 1,105 Somogyi units. Conservative therapy was tried, but jaundice occurred on the third day and a laparotomy revealed a choledochal cyst with marked pancreatic oedema and induration. A choledocho-duodenostomy was performed and the child has been well since then. In none of our cases of choledochal cysts in children has there been any evidence of pancreatitis.

Among 67 reported cases of pancreatitis in childhood, only four had surgically correctable lesions of the common bile duct or ampulla, i.e. 6%. However, there were only 10 cases of relapsing pancreatitis, 3 of which were due to surgically correctable common duct pathology, i.e. 33%.

Hyperlipaemia

In 3 reported cases hyperlipaemia is cited as the cause of pancreatitis in childhood. In all of them the disease was of the relapsing type.

Lipaemia in children is usually associated with diabetes mellitus and nephrosis, but may occur with liver disease and hypothyroidism. Anaemia, leukaemia and the lipoidoses, such as von Gierke's disease, Niemann-Pick disease and the xanthomatoses, are associated with an increase in blood lipids. Thannhauser[38] published 10 cases of pancreatitis associated with hyperlipaemia, but none were in children. Whether hyperlipaemia is the cause or effect of pancreatitis is still uncertain. The experimental work of Wang *et al.*[39] has shown a definite rise in lipids in all cases of induced pancreatitis. However, they do accept that familial hyperlipaemia may cause pancreatitis. The exact mechanism is uncertain, but there may be embolization of agglutinated serum-lipid particles to the pancreatic vessels.

Collett and Kennedy[15] reported the case of a child with known relapsing pancreatitis associated with hyperlipaemia, and Poulsen[16] reported a similar case. Both had recurrent attacks of abdominal pain and vomiting associated with hyperlipaemia, manifested by chylous serum and elevated phospholipids and cholesterol. Both had siblings with hyperlipaemia, and one developed xanthomatous lesions of the skin. Jaffe *et al.*[17] reported a 6-month-old Indian child with acute pancreatitis caused by familial hyperlipaemia. This infant had the usual picture of abdominal distension, tenderness, and a temperature of 100·2°F. Pancreatitis was found at laparotomy, but at the time it was noted that when the vessels were cut, a salmon-pink fluid escaped. The peritoneal fluid contained fat globules. The serum-amylase level was 40 Wohlgemuth units (normal 3 - 10) and the total lipids were 190 (normal 16 - 30). The serum-lipid level of the father was 33 and of the mother, 18. The child has subsequently had repeated attacks of abdominal pain resembling attacks of acute pancreatitis.

Relapsing Pancreatitis

This is much rarer than acute pancreatitis in childhood, and besides our 1 case, there are only 10 cases recorded in the literature. The diagnosis is often difficult because the serum-amylase level may be only slightly elevated, if at all.

Warwich and Leavitt[18] reported a patient with relapsing pancreatitis who had 3 attacks over 2 years. The diagnosis of pancreatitis was made by laparotomy at the first admission. Subsequent investigations failed to show any abnormality of the biliary tree, nor was there any calcification of the pancreatic region. The patient was well between the episodes of pain and her serum-amylase level returned to normal.

Blumenstock *et al.*[3] reported the case of a girl with attacks at 15, 19 and 42 months. The diagnosis was made by operation during the first attack of pain, and subsequent attacks were characterized by a rise in the serum-amylase level. At the age of 5 years she again had a laparotomy, and a grossly dilated common bile duct was found, although the pancreatic duct did not appear obstructed. A cholecyst-jejunostomy was performed and she remained free of symptoms after the operation.

Davis and Kelsey[19] published a case of relapsing pancreatitis in an 8-year-old boy. He had a 3-week history of abdominal pain, distension and vomiting. Abdominal paracentesis was performed 3 times; each time reddish fluid was obtained. At laparotomy nodules of fat necrosis were found and the omentum was adherent to the anterior abdominal wall above the umbilicus. For 6 months postoperatively he had intermittent upper-abdominal pain and a raised serum-amylase level, but after that the pain disappeared. Whether this was a genuine case of relapsing pancreatitis seems doubtful. The picture might have been due to intra-abdominal rupture of a pseudocyst of the pancreas which took a long time to heal.

Comfort *et al.*[20] reported 2 cases, the first an 11-year-old boy who had a hypoplastic pancreas and hereditary diabetes. He had severe intermittent epigastric pain associated with nausea and vomiting. Hypoproteinaemia, oedema, hypocalcaemia and tetany developed after 2 severe attacks of pancreatitis. In less than 2 years he died of profound pancreatic insufficiency and malnutrition. The second patient was a 12-year-old boy with recurrent attacks of abdominal pain. He had multiple intrapancreatic calculi. He recovered after removal of most of these calculi and a short course of X-ray therapy to his pancreas.

Collett and Kennedy,[15] Poulsen,[16] and Jaffe *et al.*[17] each reported a child with chronic relapsing pancreatitis associated with hyperlipaemia. Each child had recurrent attacks of abdominal pain and vomiting. Hyperlipaemia was manifested by chylous serum and elevated serum lipids. One developed xanthomatous lesions of the skin. Two had siblings with hyperlipaemia.

Wolman[21] reported 3 cases of pancreatitis in children, one being of the acute relapsing type. This was in a 10-year-old boy with repeated attacks of abdominal pain. Appendicectomy was performed, but the appendix was normal. His serum-amylase level was 80 Wohlgemuth units, and all other investigations were negative. Subsequent attacks of pain over the next 18 months were associated with a rise in the serum-amylase level up to 160 Wohlgemuth units. A laparotomy was ultimately performed, and although no pathological changes could be detected, a sphincterotomy was done and the child has been free of pain since then.

Plechas[13] described an interesting case of a child who presented at the age of 10 years with a history of recurrent bouts of abdominal pain since the age of 2 years. Laparotomy was performed on his first admission (at 10 years of age) and no abnormality was found. On his fourth admission to hospital a second laparotomy showed pancreatitis, and on his fifth admission (now 17 years of age) a third laparotomy showed a scarred ampulla of Vater. The pancreatic duct was 10 times its normal size and the common bile duct appeared to be clinically inflamed and was twice its normal diameter. Sphincterotomy was performed and he has been free of pain ever since. An important feature in this case is that at no time was his serum-amylase level raised.

In our one case of relapsing pancreatitis we found no cause for the recurrence except that roundworms were still present when the child had his second attack of pain. He was treated conservatively, and since he was 'dewormed' he has had no subsequent episodes of pancreatitis.

Hereditary Pancreatitis

Five kindreds of hereditary pancreatitis have been reported.[40] Hereditary pancreatitis resembles non-hereditary relapsing pancreatitis except for the striking familial incidence and its early onset and absence of precipitating factors. It is inherited *via* a dominant autosome, and although the defect is not clear, many of the patients excrete excess lysine and cystine. No anomalous biliary systems have been demonstrated.

This disease must not be confused with cystic fibrosis of the pancreas, which is transmitted by a recessive trait and presents from early childhood with wasting, steatorrhoea, recurrent respiratory infections, meconium ileus, and increased sodium chloride in the sweat test. The age onset of hereditary pancreatitis is usually in the teens, but one

case presented at 3 years of age. None of our cases appeared to fall into this category.

SUMMARY

Eleven cases of pancreatitis in childhood have been presented. Of these, one was of the relapsing type. There were no deaths and all have recovered completely. At least 54 cases of pancreatitis in childhood have been reported previously in the literature and, of these, 10 were of the relapsing type. There were 24 deaths among these 54. The disease is probably not as rare as has been believed and many surgeons can recall isolated cases they have treated. Many cases remain undiagnosed, while others are not published. It therefore appears that, although pancreatitis may be uncommon in childhood, it should always be considered in the differential diagnosis of abdominal pain in children.

Our series brings the total number of reported cases to 65. Among these no aetiological factor was found in 29. Worm infestation was associated with 12. In 4 of these ascari were actually demonstrated in the pancreatic duct. Trauma accounted for 9 cases with no mortality, and the interesting feature here is that pain may only develop several days after the initial trauma. Three cases were associated with the administration of steroids and all these children died.

Acute relapsing pancreatitis in children is rare. Altogether 10 cases have been reported. Three of these were due to idiopathic familial hyperlipaemia and 1 to recurrent worm infestation, while 3 had surgically correctable common bile duct pathological conditions.

Pseudocysts of the pancreas were found in 3 of our cases and all were satisfactorily treated by transgastric cystogastrostomy.

One of our patients developed disseminated fat necrosis in skin and bone.

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