BILE PERITONITIS IN INFANCY

BY

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Extravasation of bile into the peritoneal cavity of infants and children, unassociated with trauma, has been described sufficiently rarely in the past to merit the reporting of two further cases. The term 'bile peritonitis' is misleading as a description of the clinical picture seen in these particular infants. The condition is essentially an escape of bile following rupture of some component of the extra-hepatic biliary system, forming a collection in the peritoneal cavity. Some of this is fluid drawn from the various peritoneal and intestinal tissues by the relatively high osmotic pressure of the escaping bile (Gross, 1953), but there is no acute peritonitis. This is, therefore, a bile ascites unaccompanied by disease of the liver or peritoneum.

Case Reports

Case 1. This child, the second of healthy parents, was admitted to hospital at the age of 6 weeks in December, 1949. He had been born normally, weighing 8 lb., after an uneventful pregnancy. The history was that his stools had been white since the passage of meconium ceased, that he had been jaundiced since the age of 2½ weeks, and that the intensity of the jaundice had not increased; two to three days before admission his abdomen had become distended, and bilateral inguinal herniae had appeared. On admission he was afebrile, slightly jaundiced, but not acutely ill; generalized wasting emphasized his gross abdominal distension and large herniae. He had in addition bilateral talipes equinovarus. The stools were cream coloured, but the urine contained neither bilirubin nor excess of urobilin. Both mother and child were Rh positive, and had negative Wassermann reactions. The abdominal distension increased slightly after admission and caused the infant some respiratory embarrassment. Paracentesis abdominis gave 50 ml. of heavily bile-stained fluid, sterile on culture, and in view of this laparotomy was performed. At operation, a large amount of bile-stained fluid was found in the peritoneal cavity. The gall bladder, cystic and hepatic ducts looked normal, as did the liver. The common bile duct was explored and a probe passed down into the duodenum. Some resistance was met at the sphincter of Oddi, but there was no genuine obstruction. A drain was left down to the duct and the abdomen closed, bile continuing to drain for some days. On the fifth post-operative day the child passed his first normal stools. The tube was removed on the ninth post-operative day, by which time the jaundice had disappeared. He has made good progress, and has been symptom free since.

Case 2. The first child of healthy parents, this boy was born naturally at term after a normal pregnancy and labour, weighing 7 lb. 7¼ oz. His mother thought that he had been slightly jaundiced from birth, and on leaving the maternity home she found he was passing white stools and dark urine. At the age of 1 month he developed a large left scrotal hernia, followed a week later by an equally large right-sided hernia. His progress otherwise was considered satisfactory, feeds were taken well, and he gained weight normally.

When first seen at the age of 8 weeks in March, 1952, he was an active, alert baby, not ill, weighing just over 10 lb. Apart from slight jaundice, the most striking features clinically were the large herniae, and a distended abdomen. Shifting dullness could be demonstrated, and the liver felt two fingerbreadths below the costal margin. The urine contained some bilirubin, but no urobilinogen, the faeces neither bilirubin nor stercobilin. The plasma bilirubin (5·4 mg.) and alkaline phosphatase (83 units) levels were raised. Both mother and child were Rh positive and had negative Wassermann reactions.

The similarity of this case to the first led to a diagnostic paracentesis abdominis on the day following admission, and a small amount of heavily bile-stained fluid was withdrawn, sterile on culture. An increasing abdominal circumference causing him some respiratory distress led to three more taps over the next eight days, 25 oz. of fluid being withdrawn in all. Fluid continued to accumulate, however, and as the infant's condition was deteriorating slightly, the decision to investigate further was made, and laparotomy performed three weeks after admission. On opening the peritoneal cavity, a further 12 oz. of bile-stained fluid was drained. There were many adhesions round the gall bladder area, and a small perforation was seen in the common bile duct just above the duodenum, beside which was lying a greenish black stone, 3 mm. in diameter. Probing showed that only the upper part of the common duct was patent, and that the lower part below the perforation was not. The opening in the common bile duct was enlarged slightly and was anastomosed to the duodenum over a short length of rubber tubing. The peritoneal cavity was drained by a tube to the region of the anastomosis. Culture of the bile was again sterile. Forty-eight hours after operation, the infant passed his first normal

yellow stool. Four days after operation the drain was removed, the wound was well healed after 10 days, and he started to gain weight satisfactorily. The child was discharged from hospital a few days later, jaundice having disappeared completely, the stools remaining normal in colour and consistency. Two months later liver function tests were as follows: Plasma bilirubin 0·2 mg. %, alkaline phosphatase 23 units, total proteins 6·2 g. % (albumin 4 g. %), and flocculation tests (thymol turbidity and colloidal gold) negative. The small rubber tube was passed via the bowel some three months after the operation. The baby has remained well and symptom free since, making excellent progress, and a recent repeat of the liver function tests has shown normal results.

Discussion

Caulfield (1936), stating that severe trauma had been responsible for any previously reported cases that he could find, put on record the first two cases with a different aetiology. These were both full-term male infants, born after normal deliveries, and the first symptoms in each case were noted at the age of 3 weeks. The first infant, in whom slight jaundice and the passage of white stools were quickly followed by increasing abdominal distension and scrotal swelling, was subjected to laparotomy at the age of 6 weeks, as paracentesis producing heavily bile-stained fluid had been followed by fairly rapid accumulation with some respiratory distress. At operation it was found impossible to distinguish the anatomy of the extra-hepatic ducts, and simple drainage to this region was carried out, with complete recovery. The second infant did not develop abdominal and scrotal swelling or jaundice until three weeks after the appearance of white stools. He was not operated on, and died at 11 weeks. Necropsy showed stenosis of the common bile duct, rupture of the duct just above the stenosis, and a large bile-filled sac in the upper abdomen which had ruptured and filled the peritoneal cavity. Death was presumed to be due to inanition.

Hindmarsh (1947) described the case of a 22-month-old girl, previously healthy, who had presented after a 36-hour illness with abdominal distension, generalized abdominal tenderness, fever and shock. She was found at operation to have a large quantity of bile-stained fluid in the abdomen. Simple drainage was performed, and she made a good recovery. The bile was sterile on culture. A cholecystogram done several months later was normal.

Byrne and Bottomley (1953) reported a fourth case, that of a 3-week-old girl, thriving until she presented with a 36-hour illness, showing abdominal distension, generalized abdominal tenderness, fever and shock. She was found at operation to have a large quantity of bile-stained fluid in the abdomen, a completely solid gall bladder embedded in the right lobe of the liver, and several small cystic masses at the junction of the cystic and common ducts, one of which was ruptured and presumably the site of extravasation. Simple drainage again gave a complete recovery, though follow-up is being continued.

The combination of stools without bile pigment, indicating a complete obstruction to the flow of bile, with slight jaundice and slight or absent bilirubinuria, suggests that providing it is being normally formed, the bilirubin is not being reabsorbed into the blood stream but is escaping elsewhere. The presence of abdominal distension and shifting dullness in the two cases now recorded, with these other findings, thus made the diagnosis almost certain. It is interesting to note that in both of Caulfield's cases, as in ours, the rise in intra-abdominal pressure was sufficient to cause bilateral inguinal herniae, and it is a reflection on the mildness of the illness that in three of the four cases it was the presence of the herniae alone that worried the parents sufficiently to seek medical advice. Diagnosis in Hindmarsh's and Byrne's cases, on the other hand, was impossible pre-operatively, and the histories of both conform much more to the picture seen in adults, when extravasation of bile presents as an acute abdominal emergency with considerable associated shock. The reason for these two contrasting modes of presentation is obscure. A gradual leakage of bile over a longer period as opposed to a sudden large extravasation may be the answer, but it is at least possible that the formation of large scrotal herniae as intra-abdominal pressure rose in the four male infants acted as a kind of safety valve, thereby altering the clinical picture.

The aetiology of Case 1 is uncertain. At operation, apart from slight resistance to the probe at the sphincter of Oddi, the extra-hepatic biliary system appeared normal in all respects, and the actual site of rupture was not found. A tempting theory is that the probing bougie dislodged a plug of inspissated bile, possibly at the site of resistance, so releasing the obstruction. The infant had a leucocytosis (as did the cases of Hindmarsh and Byrne), but he was afebrile, the bile-containing ascitic fluid was sterile on culture, and it is difficult to correlate the rather insidious history with an acute infection. In Case 2 a definite abnormality, stenosis or failure of development of the lower end of the common bile duct, was present, with the actual perforation visible above it. The minute gall stone found may have been instrumental in causing the rupture, though it may equally well have been formed

after the extravasation had occurred. A similar anomaly, also with rupture above the stenosis, was responsible for Caulfield's second case, and rupture of a congenital cyst in the region of the cystic duct gave rise to symptoms in Byrne's and Bottomley's case. Caulfield's first case is more difficult to explain: perhaps again a plug of bile, or even a gall stone could be postulated, but proof of the former anyway would be impossible and it must remain an academic point. Hindmarsh's patient is also puzzling, and one wonders if, in view of her age, she could, without her parents' knowledge, have sustained some trauma before the onset of symptoms.

This condition, as Caulfield stressed, is an indication for surgical exploration, though it is probably justifiable to try the effect of paracentesis one or more times first. Neither of the infants now described presented as abdominal emergencies, which meant that adequate preparation for operation, including the administration of vitamin K, could be undertaken. Simple drainage after probing the extrahepatic ducts in the first case, and anastomosis of the common duct above the stenosis to the duodenum in the second, were successful. Simple drainage alone proved successful in the three other surviving cases previously reported.

The prognosis of this condition in infancy and childhood when surgically treated appears to be good. Perhaps the very fact that the bile is released means that back pressure on the liver and therefore subsequent damage to it is avoided.

Summary

Two cases of so-called bile peritonitis in infancy, unassociated with trauma are reported, and the available literature described.

The diagnosis and aetiology and treatment are discussed.

It is stressed that the condition is an indication for surgical treatment.

Both these children were admitted under the care of Dr. Smallpeice, and we should like to thank her for permission to describe the cases, and for much encouragement and help in the preparation of this article.

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