Bile Peritonitis in Infancy

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Spontaneous biliary peritonitis in infancy is rare. Yet the clinical picture in all reported cases is sufficiently similar for a recognizable syndrome to emerge. It is the purpose of this paper to add 2 further cases to the published material and to outline the views generally held about the condition.

Case Reports

Case 1. The patient was the third child of healthy parents, born at home following a normal pregnancy. She remained well until 20 months of age, when she was admitted to hospital, as a case of otitis media. The history was that she had become listless 2 days previously, had refused her food, and had vomited intermittently. Her bowels had been open regularly and were of normal colour. There had been no abdominal pain.

On examination, her temperature was 99° F. (37·2° C.), pulse 120, and respirations 40/min. There was no jaundice. The only abnormal physical finding was bulging of the left ear-drum. Hb 90%. WBC 17,600/c.mm. with a polymorph leucocytosis.

She was treated with oral penicillin. The following day her temperature was 101° F. (38·3° C.), an expiratory grunt had developed, and though no definite signs were found in the chest, the radiograph showed some mottling of the right mid-zone. Slight nuchal rigidity was also observed and a revised diagnosis of pneumonitis with meningism was made. CSF was normal.

One of us (W.L.) was asked to see her on the third day of admission because her abdomen had become distended. She was now apathetic with a flushed face. There was no jaundice. The left ear-drum showed some peripheral injection, but no bulging. Slight nuchal rigidity was still present. The abdomen was distended and showed yellow staining around the umbilicus and in the right flank and right iliac fossa. There was no rigidity, but tenderness and guarding were present in all quadrants. Serum amylase was normal.

It was thought that the child might have a traumatic biliary peritonitis, though injury was denied by the parents and no local bruising was found.

At laparotomy much clear, bile-stained fluid escaped; on culture this proved sterile. Exploration of the upper abdomen revealed a normal spleen, liver, and gall-bladder. A small perforation was found on the posterior wall of the

common bile-duct, immediately distal to the entry of the cystic duct. No free stones were found and none were palpable in the common bile-duct. Bouginage of the common duct, via the perforation, revealed no obstruction. The pancreas appeared normal. The small intestine was injected and the mesenteric nodes were enlarged, soft, and fleshy.

A small T-tube was placed in the common duct, through the perforation, and the abdomen was closed with a stab drain.

On the 20th post-operative day T-tube cholangiography showed easy filling of the common bile-duct and the rest of the biliary tree, with a free flow of dye into the duodenum. No abnormality was detected. The T-tube was removed and she made an uneventful recovery, and has remained well since.

Case 2. The patient was a female child, 4 weeks old, born after a normal delivery. When 2 weeks old, she began to vomit, but this lasted only 24 hours. The following day, however, the parents noticed that the stools were white, and they remained so until operation. A week later, mild jaundice was thought to be present by the parents, who noted also that her urine was dark in colour.

On admission the child was thin and undernourished. There was no obvious jaundice, but marked ascitic distension of the abdomen was present. No other abnormality was found on clinical examination apart from a slight umbilical discharge. Hb 79%. WBC 11,000/c.mm., normal differential. Serum bilirubin 6·6 mg./100 ml. (Other liver function tests, thymol turbidity and flocculation, zinc sulphate turbidity, colloidal gold, serum alkaline phosphatase, serum proteins, SGOT, SGPT normal.) WR negative. Electrolytes normal. Spectroscopic examination of faeces for stercobilinogen negative. Urinary chromatogram normal. Culture of the umbilical discharge revealed a scanty growth of coagulase-negative staphylococci and diphtheroid bacilli. Radiograph of chest was negative. Radiograph of abdomen suggested free fluid.

Ten days later her serum bilirubin was 2·0 mg./100 ml., and the alkaline phosphatase had risen from 11 to 48 units/100 ml. The remaining liver functions were still normal. Blood cultures were sterile.

During these investigations, the stools remained acholic, but the ascites increased steadily as judged by measurement of abdominal girth. There was no further vomiting and the child remained apyrexial throughout.

The problem, therefore, was of a 6-week-old child with persistent acholic stools, a progressively distending abdomen, and a mild fluctuating jaundice of obstructive type.

Laparotomy was performed 12 days after admission (J.E.M.). On opening the abdomen through a right paramedian incision, a quantity of clear, bile-stained ascitic fluid escaped. The parietal peritoneum was stained green. Adhesions were present under the liver, and on dividing these, a thick bile-stained fibrinous area was encountered between the liver and the anterior surface of the stomach and duodenum. The gall-bladder appeared normal; darkly pigmented bile was aspirated from it. A ureteric catheter was tied into the gall-bladder with the intention of performing an operative cholangiogram. On syringing this through with saline, a small perforation was found in the anterior wall of the supraduodenal portion of the common bile-duct. The rest of the common bile-duct appeared normal and the mucosa was not inflamed. A probe could be passed through the perforation, easily into right and left hepatic ducts, but could not be passed downwards into the duodenum. The perforation in the common bile-duct was closed and cholecyst-duodenostomy performed. The abdomen was closed with a stab drain.

The child recovered and has remained well since.

Discussion

The cases described above characterized the two modes of presentation of spontaneous biliary peritonitis in infancy.

The acute case: Simon and Schorr (1958) reported a case in many respects similar to Case 1. This was a 4-week-old female infant in whom vomiting, abdominal distension, and a finger-wide icteric strip from umbilicus to xiphisternum had been noted some days before admission. There was no pyrexia, but a leucocytosis was present. The stools varied from light yellow to white on admission, and there was no jaundice.

Hindmarsh (1947) reported a case of a 22-month-old infant with a 36-hour history of listlessness, vomiting, and anorexia, but no abdominal pain. Some diarrhoea was present, but the stools were of normal colour. She was pyrexial and there was a leucocytosis. On examination, her general condition was poor and the abdomen was slightly distended and generally tender. She was thought to have peritonitis, secondary to appendicitis. Byrne and Bottomley (1953) reported the case of a 3-week-old female infant with a 36-hour history of restlessness, crying, drawing the legs over the abdomen, and anorexia, but no vomiting. The following day, the child cried almost continuously, pyrexia and leucocytosis were present, and she appeared acutely ill. The abdomen showed moderate distension, there was generalized tenderness and

guarding and a diagnosis of appendicitis was also made. Pettersson's (1955) third case (Ehrenpreis) was a female infant, 2 weeks old, with a history of intermittent vomiting, but normal stools and no fever. Abdominal distension and a raised serum bilirubin were noted at the second week and a diagnosis of peritonitis was made.

In Case 1, the disease pursued a rather milder course over several days with listlessness, anorexia, and vomiting. Pyrexia and leucocytosis were present, but it was not until the fifth day that abdominal distension, with generalized tenderness and guarding, and flank and umbilical staining, were noted. At laparotomy, in each of these 5 cases, free bile was found in the peritoneal cavity.

In Simon and Schorr's case, and also in Pettersson's case, a perforation covered by fibrin was found in the anterior wall of the common bile-duct. In Hindmarsh's case retroperitoneal oedema and flecks of lymph were noted in the region of the duodenum, but the site of perforation was not determined, while in Byrne and Bottomley's case one of several congenital cysts at the junction of cystic and common bile-duct was found to have ruptured. Simple drainage led to recovery in 4 of these 5 cases. In Case 1, the perforation of the common bile-duct was on the posterior wall at its junction with the cystic duct, and T-tube drainage led to recovery. It is difficult to see how, in the acute case, a pre-operative diagnosis can be made.

The chronic case: 14 cases of biliary peritonitis, presenting in a chronic manner, have been recorded, and their clinical features, together with our Case 2, are shown in the Table.

The clinical picture is fairly constant. Usually within the first month of life, jaundice and acholic stools are noted. The jaundice fluctuates, is usually slight, and may disappear for a time. The acholic stools persist throughout the illness. Some variation in colour may occur, and in 2 instances the stools were reported to be normal. Urobilinogen is absent from the urine in most cases. Abdominal distension is next observed, on the average at about the sixth week, when free fluid is clinically and radiologically detectable. If paracentesis abdominis is performed at this stage, sterile bile-stained fluid will be aspirated. At about the same time, scrotal swellings appear. These, described in some reports as 'herniae', result from distension of the tunica vaginalis by the bilious ascites. In some instances, the 'herniae' was the only reason for the parents seeking a medical opinion, thus serving to emphasize the mild course of the illness.

The disease appears to be painless as a rule, though attacks of screaming have been noted. The infants

TABLE
Details of 15 Cases of Chronic

Case No.	Date	Author	Sex	Age on Admission* (wk.)	Time of Onset of Symptomatology (wk.)				Vomiting
					Jaundice	Distension	Acholic Stools	Scrotal Swellings	
1	1936	Caulfield	M	6 (7)	3	5	6	4	Yes
2			M	11 (12†)	7	4	3	8	Yes
3	1952	Snyder, Chaffin, and Oettinger	F	6 (7)	5	5	2	—	Yes
4	1954	Krebs	M	9 (15†)	13	8	Reported normal Since birth	8	Yes
5	1955	Coffey and Oweida	M	3 (6)	Since birth	5-6	Reported normal Since birth	NR	NR
6	1955	Davies and Elliot-Smith	M	6 (7)	2½	5	1	5	NR
7			M	8 (11)	Since birth	8	1	4-5	NR
8			F	3½ (8)	4	Tumour felt, 7	3	—	Yes
9	1955	Pettersson	M	6 (7)	4	5	4	6	No
10			M	5 (8)	3½	8	3½	8 (R) only	No
11	1959	Talalak	?	7 (7)	1	7	Reported normal	—	NR
12	1959	Gertz	F	9 (22)	5	9	9	9	NR
13	1961	Johnston	F	3 (6)	3	6	2	6	Yes
14	1964	Colver	M	7 (7)	Since birth	6	Since birth	6	NR
15	1965	This paper (Case 2)	F	4 (6)	3	4	2	—	Yes

NR, not recorded, or no comment.

GB, gall-bladder.

CBD, common bile-duct.

* Age at operation is given in parentheses.

may appear well, but more usually are apathetic, refusing food, and failing to thrive. Vomiting is common. Fever and leucocytosis are usually absent. The degree of abdominal distension ultimately becomes gross, with engorged abdominal wall veins and pouting umbilicus. This may lead to respiratory distress with rapid deterioration in the general condition.

Laparotomy was most commonly proceeded with in the fifth week after the onset of symptoms, when the child was not relieved by repeated paracenteses or when the general condition worsened. At laparotomy, in all cases but one (where the biliary leak had remained encapsulated), free bile-stained fluid escaped from the incision. Peritoneal hyperaemia, small gut distension, and enlargement of the mesenteric lymph nodes, were commonly found. The findings on further exploration were variable; either the anatomy was too obscure to determine the site of the biliary leak, or further dissection revealed a small perforation of the anterior wall of the common bile-duct, usually at the junction of the cystic duct with the common hepatic duct, or immediately below this. In 2 cases, the perforation was at the junction of the gall-bladder with cystic duct. The gall-

bladder and bile-ducts were not distended and looked normal, though occasionally inflamed. Bile had either escaped freely from the perforation, or an attempt at encapsulation had been made (denoted by 'bile sac' formation in the Table), bile being loculated by a fibrous layer overlying the region of the hepato-duodenal ligament. Exploration through this layer would reveal the perforation in the common bile-duct. Although such 'sacs' were found in 9 cases, in only 1 had the 'sac' remained intact, enlarging to reach the left iliac fossa, so that the case presented as an abdominal tumour.

Operative management varied. In the absence of distal stenosis of the common bile-duct, simple drainage in the region of the perforation, catheter drainage of the common bile-duct via the perforation, or simple suture of the perforation, with or without cholecystostomy, was performed. The presence of marked distal common bile-duct stenosis led to the adoption of short-circuiting procedures. No deaths resulted from this simple operative management, except for one case where the bile sac was thought to be a choledochal cyst and was anastomosed to the duodenum.

The aetiology of the condition has been obscure.

Bile Peritonitis in Infants

Pyrexia	Leucocytosis	Urine for Urobilinogen	Paracentesis†	Operative Findings*	Management	Outcome
No	Yes	- ve	+ ve	Anatomy obscure; leakage from region of bile-ducts	Simple drainage	Recovery
No	No	- ve		No op.; at necropsy perf. CBD; bile sac; distal incomplete stenosis§	Non-operative	Death
Yes	No	- ve	+ ve	Perf. junction cystic duct with GB bile sac; CBD stones	CBD drainage + simple drainage	Recovery
NR	Yes	- ve		No op.; perf. CBD with small stone	Non-operative	Death
NR	NR	NR	+ ve	Perf. CBD	Choledocho-jejunostomy	Sudden death 3 mth. later; no necropsy
NR	NR	- ve	+ ve	No perf. found; on exploration CBD slight resist. at sphincter of Oddi	Simple drainage	Recovery
NR	NR	- ve	+ ve	Perf. CBD with distal stenosis and small stone	Choledocho-duodenostomy + simple drainage	Recovery
NR	NR	NR		Bile sac extending to left lower abdomen; no free bile; perf. CBD; no stenosis	Closure of perf.	Recovery
No	NR	NR		Perf. CBD; bile sac; no stenosis	CBD drainage + simple drainage	Recovery
No	NR	NR		Perf. CBD; bile sac; stenosis ampulla Vater (mild)§	Dilatation of ampulla; closure of perf.; simple drainage	Recovery
NR	NR	NR		Perf.; bile sac; at necropsy perf. CBD found; two stones in GB and one in peritoneal cavity	Bile sac thought to be choledochal cyst and anastomosed to duodenum	Death
NR	NR	- ve	+ ve	Perf. CBD; bile sac; distal stenosis	Simple drainage	Recovery
NR	NR	+ ve		Perf. CBD; bile sac; no stenosis	Closure of perf. + cholecystostomy	Recovery
No	Yes	- ve	+ ve	Perf. junction cystic duct with GB; inspissated bile; no stenosis	Irrigation CBD; closure of perf. + cholecystostomy	Recovery
No	No	- ve		Perf. CBD; bile sac; distal stenosis	Closure of perf. + cholecyst-duodenostomy, simple drainage	Recovery

Age at death. ‡ A blank indicates that paracentesis was not performed.

§ Inspissated bile.

We are in agreement with Johnston (1961) that the relative constancy of the site of perforation would argue a developmental *locus minoris resistentiae* which might rupture as a result of raised intraduct pressure. Duct stenosis was reported in 6 cases (though only of a degree sufficient to warrant bypass in 2 of them). In 1 of them stenosis was found at necropsy, no operation having been performed. In 3 others, stones were found either in the common bile-duct or lying alongside the perforation, and in one case inspissated bile was found in the common bile-duct in the absence of distal stenosis. Thus obstructive features were found in 10 of the 15 cases tabled. In 4, stenosis was reported not to be present, but in the 5 cases in which the presence or absence of stenosis was undetermined, stones were found in the common bile-duct of 3 of them. It is conceivable that inspissated bile may cause temporary obstruction, which may resolve or lead to raised intraductal pressure with subsequent perforation. Once perforation has occurred, inspissation might no longer be detectable. Although Brent (1962) was of the opinion that biliary inspissation was not the cause of jaundice in infancy, yet Rickham and Lee (1964) reported neonatal cases

where at exploration of obstructive jaundice operative cholangiography alone cleared an intrinsic obstruction into the duodenum and resulted in cure. We consider that obstruction, whether from stenosis, stone, or inspissated bile, plays a dominant role in the pathogenesis, and this is further supported by the fact that jaundice and/or acholic stools were always observed before perforation (as evidenced by distension) occurred, though not in the case reported by Krebs (1954).

Spontaneous perforation of the common bile-duct may be related to the formation of choledochal cysts. Such an association was suggested by Caulfield (1936) and by Johnston (1961). Burnell and Markey (1965) report a case of choledochal cyst associated with spontaneous perforation of the common bile-duct and argue that probably most cysts arise from this cause. It will be noted from the Table that 'bile sac' formation occurred in 9 cases. If such a 'sac' did not rupture, it would probably remain undiagnosed in infancy, and further organization of its wall could lead to the appearances seen in later life.

A distinction must be drawn between the biliary ascites in the cases described above, and bile

peritonitis as generally recognized in the adult. In the latter, the release of biliary pus is a painful and shocking process. In the former, the leak of sterile bile is comparatively painless and appears to be well tolerated in the peritoneal cavity of infants, since on the average it was 2 weeks from the onset of distension to operation. Noetzel (1909) incised the gall-bladders of rabbits and closed the abdomen without closing the gall-bladder. He drained the abdominal cavity, and the animals all survived. However, when he repeated the experiment, and introduced micro-organisms, the animals died. It is not the bile itself, but the concomitant infection that earns bile peritonitis in the adult its poor prognosis.

The difference in clinical presentation of the acute and chronic case in infancy may be accounted for if it is postulated that in the former, the extravasation of bile is sudden and massive, while in the latter, the initial breach in the duct wall is pinpoint, allowing localization for a time, or a gradual accumulation of bile in the peritoneal cavity over a longer period.

This disease, though rare, should be easily recognized as Davies and Elliot-Smith (1955) have pointed out, for the combination of mild fluctuant jaundice with acholic stools and the absence of urobilinogen in the urine means that in the presence of normal bile production, bilirubin is neither reaching the gut nor the blood-stream in quantity, while the abdominal distension with shifting dullness, and scrotal swellings containing fluid, point to where the bile has escaped. Once a diagnosis is made, operation is mandatory. The operative procedure adopted will depend on the findings, but short-circuit procedures should be reserved for cases with distal common bile-duct stenosis. Otherwise simple drainage will suffice.

Summary

Two cases of biliary peritonitis in infancy, one acute and the other chronic, have been described.

Tabulation from the published reports of the symptomatology of the chronic group results in the emergence of a syndrome, characterized by acholic stools, mild fluctuating jaundice, ascites, and fluid 'herniae'. Operative findings and management have been outlined.

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