CHOLEDOCHAL CYST-REPORT OF 4 CASES

Pages with reference to book, From 105 To 108

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Abstract

Four cases of Choledochal Cyst are presented. The preoperative diagnostic difficulty is re-emphasized. The pathology and surgical management is discussed. Choledochal Cysts are uncommon but correct management ensures symptom free future for the patient. Recent report advocating cyst excision, if technically possible, is also reviewed (JPMA 37:105 1987).

INTRODUCTION

Choledochal Cyst, an abnormal cystic dilatation of the biliary tree, was first reported by Vater[1] in 1923. Douglas[2] was the first to describe this anomaly accurately in 1852. The disease is rare and until 1926 only one case was encountered amongst 1938 Biliary operations performed at the Mayo Clinic[3]. Recently, with improved diagnostic methods, choledochal cysts are recognized with increasing frequency preoperatively.

CASE I: On 1st January 1984, two years six months old female child with a history of recurrent abdominal pain and bilious vomiting was admitted to the hospital. She was also not thriving. On examination a mass in the right hypochondrium was present. Laparotomy showed a large cystic dilatation of the common bile duct. Choledochocyst-duodenostomy was performed. Her post operative recovery was uneventful and the patient has been symptom free over the last 2 years.

CASE 2: On 25th February 1984, a 2 years old boy was admitted in the hospital with right upper quadrant mass. Exploratory laparotomy showed a large cystic dilatation of the common bile duct. Choledocho-cyst-duodenostomy was performed. His post operative recovery was uneventful and the child has remained symptom free for almost 2 years (Figure 1).

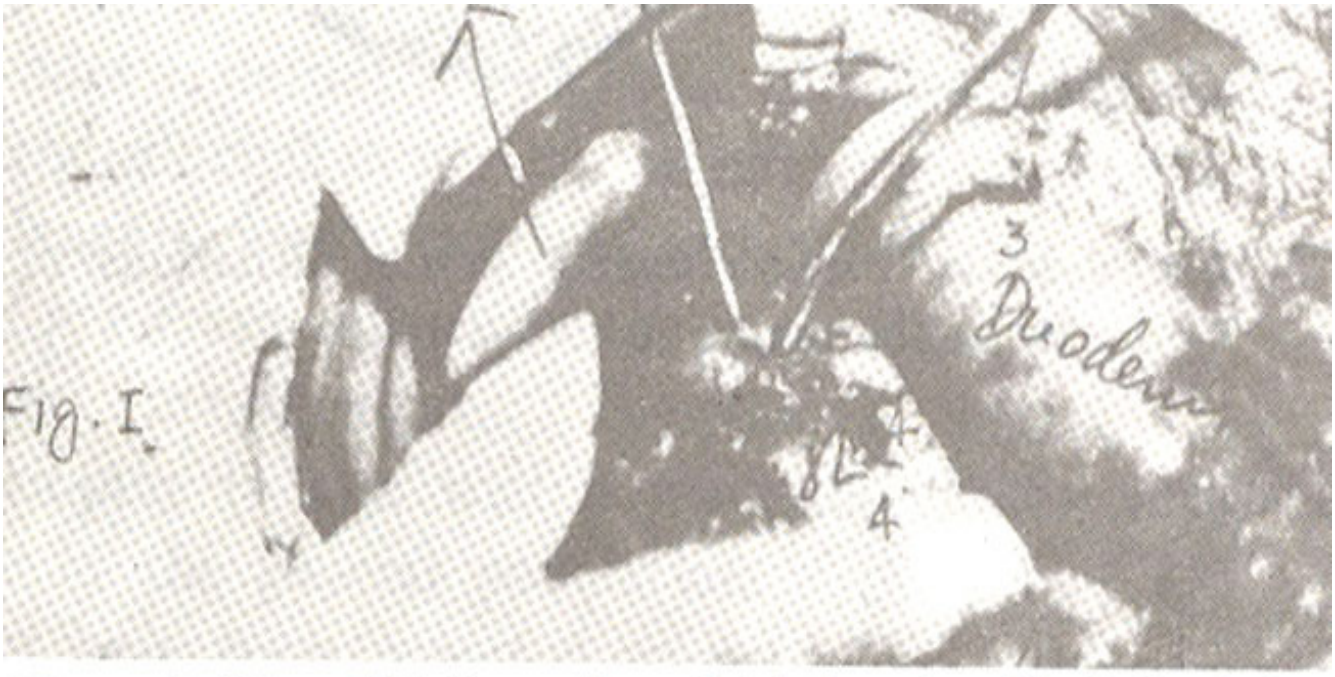


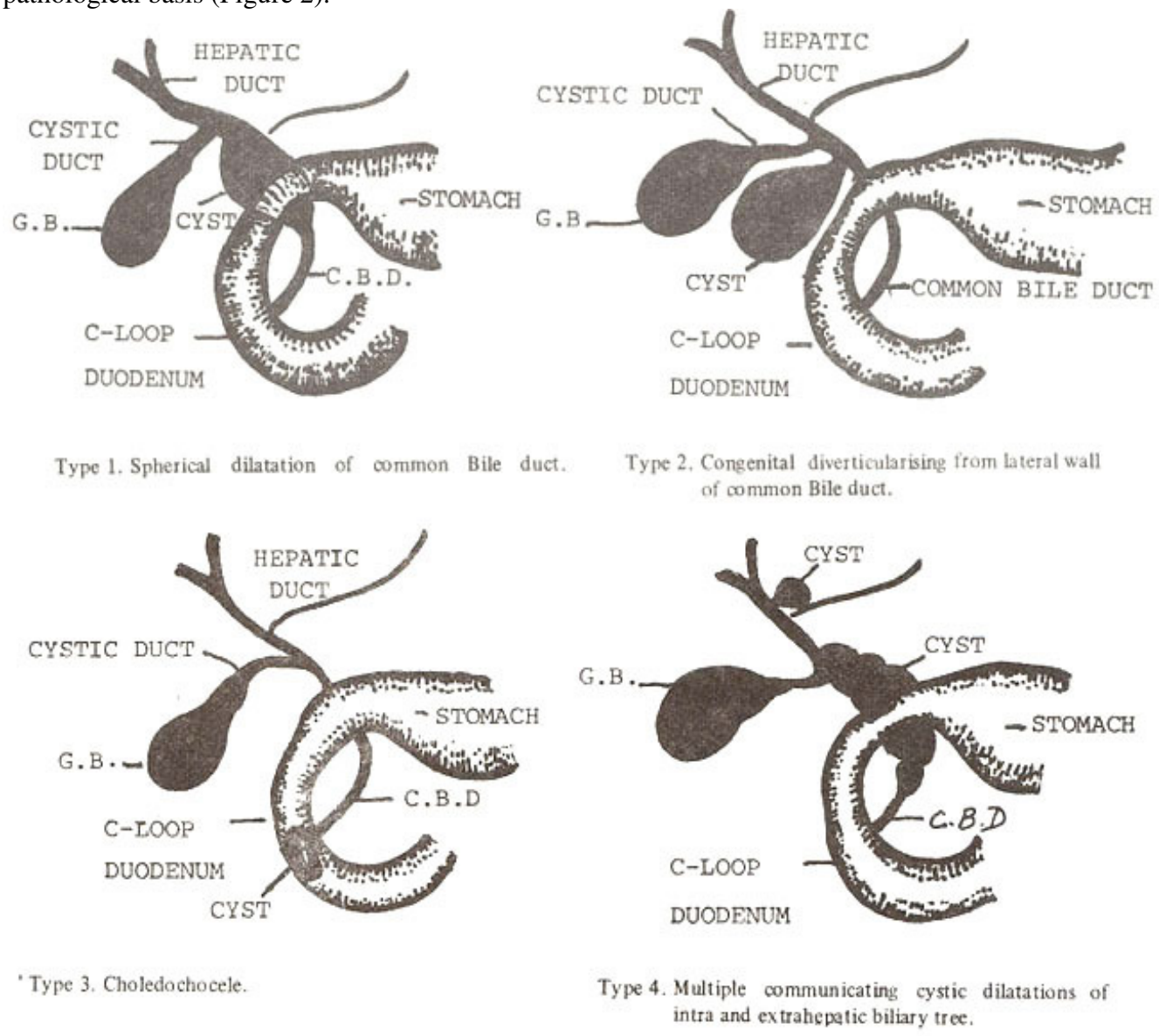
Figure 1. (Case 11) Operation field showing Choledochal Cyst, Gall Bladder, common bile duct and duodenum.

CASE 3: On 30th January 1985 a thirty years old female was admitted to the hospital with recurrent pain of nine years duration in the right hypochondrium. The pain had increased infrequency over the last 3 months. There was no history of jaundice. On clinical examination there was no palpable mass: Murphy's sign was positive. Oral cholecystogram showed non-functioning gall bladder. On laparotomy an inflamed gall bladder was found for which cholecystectomy was done. There was also associated cystic mass arising from the lateral wall of the common bile duct. Choledocho-cyst -duodenostomy was performed. Her post operative recovery was uneventful. She was discharged on the seventh day and has remained symptom free for over a year now. **CASE 4:** On 28th September 1985, a 15 years old female was admitted to the hospital with recurrent pain in the right hypochondrium associated with vomiting off and on for the last 4 years. On examination there was a cystic mass in the right hypochondrium. Intravenous cholangiogram did not outline the gallbladder,nor the cystic mass.
Laparotomy showed a large cystic dilatation of the common bile duct for which choledocho cystduodenostomy was performed. Her post-operative recovery was uneventful and the patient has remained symptom free since.

DISCUSSION

The incidence of choledochal cyst is unknown. Large series quote an incidence of approximately I in 13000 births. Sixty percent recorded cases are from Japan[4].
The high incidence of choledochal cyst in the Japanese is well known. The disease has\ been reported in Japanese literature since 1905. By 1936,55 cases had been reported[5]. This high incidence of choledochal cyst found in the Japanese has not been observed in other Asian races[6]. The prevalence of this condition in this part of the world is not recorded. The clustering of these 4 cases in a course of 2 years could just be a chance occurrence since our hospital records show no previous cases.

A ratio of 74% females to 26% males[7] has been reported. In the present series there are 3 females (75%) and one male (25%). It is predominantly found in children and young people though no age is exempt: the oldest patient recorded being a 78 years old woman[8],fifty five percent of cases occur in the first 10 years of life and 35% between the ages of 10—40 years, the remaining 10% are over the age of 40 years[6]. Our report. of 2 cases (50%), less than 10 years of age and 2 cases (50%) 10-40 years age range is consistent with reported age distribution. Alonsoleg, Rener and Passango in l959[9] postulated that the majority of choledochal cyst are congenital. This is supported by the associated high incidence anomalies of biliary tract with this condition[10]
Longmire, Mandiola and Gordon in 197111 described 4 types of choledochal cysts on clinico pathological basis (Figure 2).



Type 1. Spherical dilatation of common Bile duct.

Type 2. Congenital diverticularising from lateral wall of common Bile duct.

Type 3. Choledochocele.

Type 4. Multiple communicating cystic dilatations of intra and extrahepatic biliary tree.

Figure 2. Types of Choledochal Cyst (Longmire et al., 1971).

1) Spherical dilatation of common bile duct comprise majority of cases. Our cases 1 ,2 and 4 belong to this group.
2) Congenital diverticulum arising from lateral wall of common bile duct. This variety is rare, only 14 cases are documented to date[12]. Our case No.3 belongs to this group.

3) Choledochocele: Dilatation of intraduodenal portion of common bile duct: 9 cases have been reported to date[13]
4) Multiple communicating cystic dilatation of intra and extra hepatic biliary tree was originally described by Mcwhorter[14] attributed to Caroli, Soupault and Kossakiwski[15] and designated as type IV chole dochal cyst by Longmire et al[11]. A total of 45 cases have been reported to date.
The typical Macroscopic and Microscopic features of choledochal cyst have been extensively reviewed[5,7,16,17] The cyst may arise from any part of the duct, can increase to any size and the duct proximal to the cyst may be normal or minimally dilated. The gallbladder is not usually dilated.
The preoperative diagnosis of the cyst has proved to be an elusive goal in most cases due to rarity of the condition and the variability of clinical manifestations. The classical triad of vague intermittent upper abdominal pain, intermittent jaundice and right upper abdominal mass is rarely seen. None of our patients had these features together. Plain x-ray films of abdomen were not helpful. Barium studies were not done in any of the cases. Oral cholecystogram, which was done in case 3 was not helpful. Intravenous cholangiogram in the jaundiced patients may outline the cyst. It was done in case 4 but did not help in diagnosis. Liver function tests done ,in all cases were normal. Grey scale ultrasound a very effective diagnostic tool and recently being widely used, was not available to us. Operative cholan-giogram at exploratory laparotomy most often provides a definitive diagnosis. The complication of choledochal cyst such as cirrhosis, ascending cholangitis, perforation and carcinoma in the cyst wall are grave and stress the importance of early correct management. The risk of carcinoma developing in cyst wall is markedly higher than that in the general population. A total of 63 cases have been reported to date; an incidence of 47%1 8 The overall risk of biliary tract carcinoma varies from 0.007%-0.04% where as it appears to be 2% in association with choledochal cyst[19] The treatment of choledochal cyst is by operation. Conservative treatment is attended by mortality reaching l00%[20]. The main controversy concerns the type of surgical management. Choledocho-cyst-duodenostomy has been the procedure adopted in all the 4 cases. This was first described by Bakes in 1907[21] and is still frequently employed. A side to side anastomosis is performed between the most dependent part of the choledochal cyst and the second part of the duodenum. A dependent drainage is easily established [22].The operation is technically very easy and is more physiological. It has beer, found a most satisfactory procedure and,m our cases, has not been attended by any complication so far. The frequent argument against this procedure is that it is associated with high incidence of ascending cholangitis; this has not occuned in any of our 4 cases.

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REFERENCES

1. Vater, A. Dissertatioo Inaugualis Medica. Proes. Diss. qua Scirrhis Vicerum Disseret C.S. Ezerlus, 4 Wittembergaes Pamphlets, Vol. 881, p. 22. (Another copy H22, 70, 19. Edinburagh,University Library, l723;p.l.
2. Douglas, A.H. Choledochal cyst. Med. Sd. (London), 1852; 14:97.
3. Judd, E.S. and Greene, E.I. Choledochus cyst. Surg. Gynecol. Obstet., 1928; 46:3 17.
4. Jones, P.G., Smith, ED., Clake, A.M. and Kent, M. Choledochal cysts; experience with radical excision.J. Pediatr. Surg., 1971;6:112.

5. Yotsuyanagi, S. Contributions to aetiology and pathogenicity of idiopathic cystic dilatation of common bile duct with report of 3 cases; new aetiological theory based on supposed unequal epithelial proliferation at stage of physiological epithelial occlusion of primitive choledochus. Gann, 1936; 30:601.
6. Lee, S.S., Mm, P.C., Kim, G.S. and Hong, P.W. Choledochal cyst; a report of nine cases and review of the literature. Arch. Surg., 1969;99:19.
7. Tsardakas, E. and Robentt, A.H. Congenital cystic dilatation of the common bile duct; report of three cases, analysis of 57 cases and review of the literature. Arch. Surg., 1956; 72:311.
8. Madding, G.F. Congenital cystic dilatation of the common bile duct. Ann. Surg., 1961; 154:288.
9. Alonso-Lej, F., Rever, W.B. Jr. and Passango, DJ. Congenital choledochal cyst with a report of 2 and an analysis of 94 cases. Int. Abstr. Surg., 1959; 108:1.
10. Komi, N. Uclaka, H., Iheda, N. and Khsiwazi, Y. Congenital dilatation of biiary tract; new classification and study with particular reference to anomalous arrangement of the pancreatiobiiary ducts. Jpn. J. Gastroenterology, 1977; 12:293.
11. Longmire, W.P. Jr., Mandiola, S.A. and Gordon, H.E. Congenital cystic disease of the liver and biiarysystem. Ann. Surg., 1971; 174:711.
12. Murphy, B., Waldron, R., Drumm, J. and McCarthy, C.F. Choledochal cysts, report of 3 cases and review. Postgrad. Med. J., 1984; 60: 397.
13. Oldbourne, N.A. Choledochal cyst. A review of the cystic anomalies of the biliary tree. Ann. R. Coil. Surg. England, 1975;56:26.
14. McWhorter, G.L. Congenital cystic dilatation of the common bile duct. Report of a case with cure. Arch. Surg., 1924; 8:604.
15. Caroli. J., Saupaulat, R., Kossakowski, J., Plaker, L. and Pahadowska, M.La dilatation polykstique congenitale des voires biliares intrahepatiques. Sem. Hop. Paris, 1958; 34:488.
16. Fonkaisrud, E.W. and Boles, E.T. Jr. Choledochal cysts in infancy and childhood. Surg. Gynecol. Obstet., 1965; 121:733.
17. Mahour, G.H. and Lynn, H.B. Choledochal cyst in children. Surgery, 1969; 65:967.
18. Todani, T., Tabuchi, K., Watanabe, Y. and Kobayashi, T. Carcinoma arising in the wall of congenital bile duct cysts. Cancer, 1979; 44: 1134.
19. Longmire, W.P. Jr., McArthur,M.S.,Bastounis, E.A. and Hiatt, J. Carcinomas of extrahepatic biiary tract.Ann. Surg., 1973;178:333.
20. Attar, S. and Obeid, S. Congenial cyst of the common bile duct; a review of the literature and a report of 2 cases. Ann. Surg., 1955; 142:289.
21. Bakes, J. ,Offizielles Protokoil der K.K. Gesellschaft der Arzte in Wien. Wissenschaftliche Litzung (contribution to discussion). Wien. Klin. Wochenschr., 1907;20:298.
22. Ravitch, H.M. and Snyder, G.B. Congenital cystic dilatation of the common bile duct: special reference to radiographic studies. Surgery, 1958; 44:752.