Pediatric Surgery 3: 86,93, 1989

Current topics on the treatment of congenital biliary atresia and congenital biliary dilatation

Takeshi MIYANO, Koichiro IMURA, Toshiki OHYA
Department of Pediatric Surgery, Juntendo University, School of Medicine, Tokyo, Japan

A) Congenital biliary atresia (CBA)

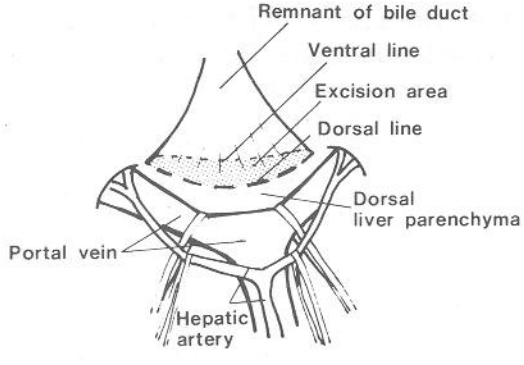
Since the hepatic portoenterostomy (so-called Kasai operation) was introduced, the prognosis of the biliary atresia (BA) has been greatly advanced and the operative result is still gradually improved by the early operation, the refinement of the operative technique and the postoperative care. Kasai mentioned that approximately 80% of BA would be curred by Kasai operation in near future [1].

Now generally speaking, however, only at most 30% of the patients with BA survive for a long time without jaundice, besides a majority of them have a portal hypertension or abnomal liver function. On the other hand, now the orthotopic liver transplantation is widely applied to the patients with BA and approximately 20% of mortality was obtained. Furthermore, in some institutes, the liver transplantation has been applied to the patients with BA as a primary procedure. Unfortunately, the liver transplantation is not available for various reasons in Japan. Nevertheless, it seems now that we are standing at the turning point concerning the treatment of BA.

a. Technical refinement of portoenterostomy

With regard to the technical aspect of portoenterostomy, firstly, the exposure and excision of the bile duct remnant at the portahepatis is the most important than any other operative technique in order to obtain a good bile flow postoperatively. Recently we passed the vessel tape and pulled the portal vein and the hepatic artery gently. Thus, the hepatic portal area behind the portal vein is exposed nicely, and then, cut the remnant on the dotted line (Fig. 1). This cutting (excision) is the most important and meticulous step,we use three times magnification throught this procedure. At this point we should not excise the liver parenchyma which causes the occulusion of the bile ductuli by the scar formation at the portal area. We should cut at the level where the extrahepatic bile duct just entered to the liver parenchyma. Particularly the excision of both sides of the inside of the portal area is important because the intrahepatic bile duct opened mainly in these areas.

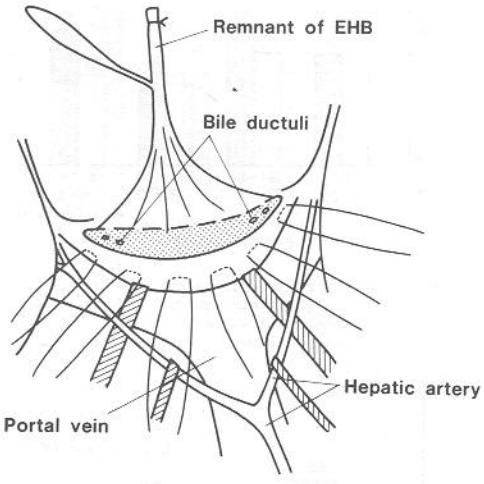
Anastomosis is also important especially at the posterior wall. The suture line should be far away from the excised area as much as possible in order to avoid the bile duct occulusion by the gran-



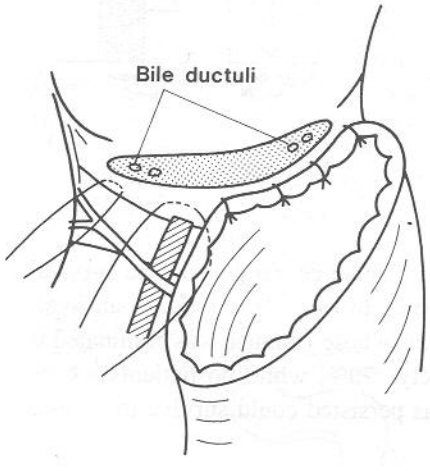
Exposure of Porta hepatis and Excision of the remnant of bile duct

Fig. 1

Address: Takeshi MIYANO MD Head of Pediatric Surgery, Juntendo University, School of Medicine 2-1-1, Hongo, Bunkyo-Ku, Tokyo, 113 Japan



Exposure of hepatic portal area
Fig. 2

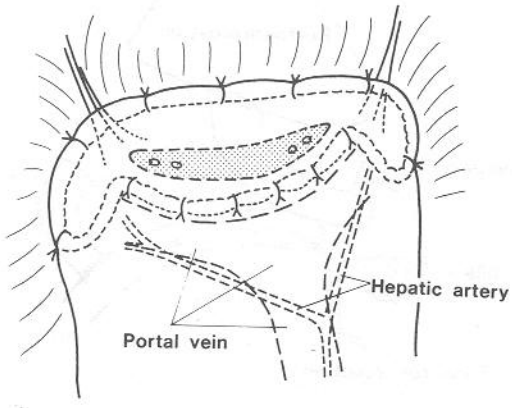


Hepatic Portoenterostomy
Fig. 3

ulation and the scar formation. From this aspect, a couple of 6-0 stitches are placed to the hepatic parenchyma just behind the portal area before the excision of the rudimentary bile duct. We use these stitches for the anastomosis of the posterior wall (Fig. 2,3,4).

b. The opeartive result of Suruga II modification and analysis of prognostic factors

With regard to the preventation of the ascending cholangitis which is the most serious postoperative complication, various operative techniques



Hepatic Portoenterostomy
Fig. 4

Operative Procedure for Prevention of Ascending cholangitis

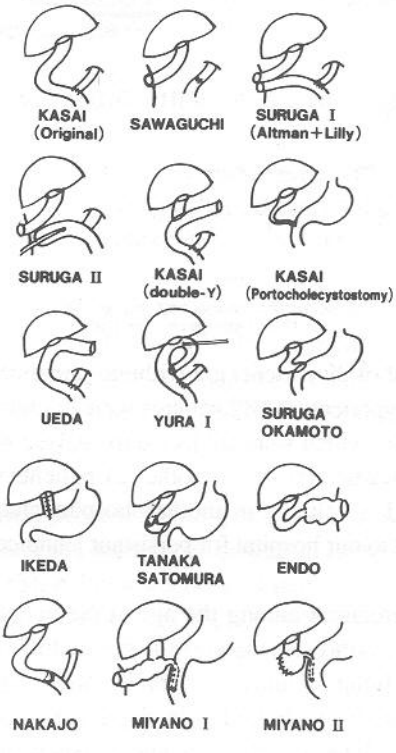
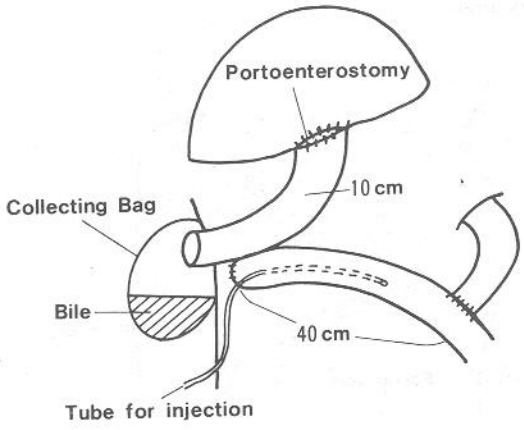


Fig. 5

were reported (Fig. 5).
Since 1977 we have been utilized so-called Suruga II enterostomy (Suruga II modification) in 103 patients (Fig. 6). Suruga II modification made it easy to manage a postoperative care and is most widely used in Japan today.

From 1977 to 1986, in the Department of Pediatric Surgery, Juntendo University Hospital, one hunred and three patients with BA were treated by Suruga II modification. 29 patients are alive, whi



Suruga II type enterostomy

Fig. 6

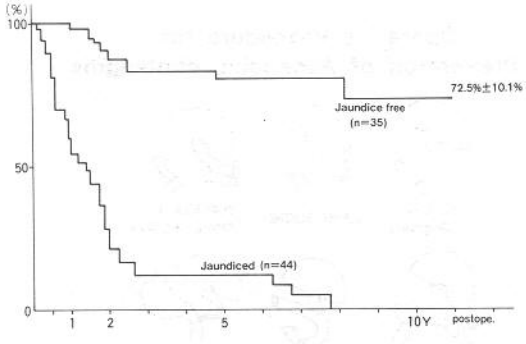


Fig. 7. 10 years survival rate (Kaplan-Meier method)

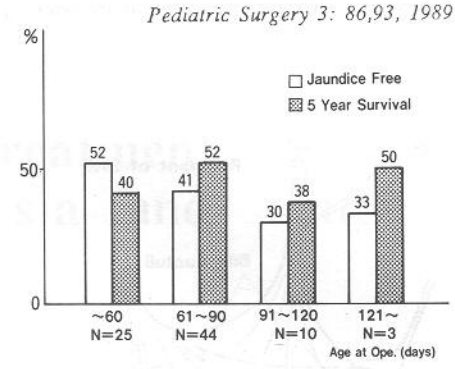


Fig. 8. Correlation among the age at operation, jaundice free and 5 years survival

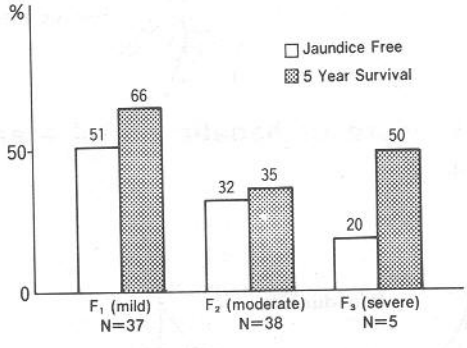


Fig. 9. Correlation among the degree of liver fibrosis, jaundice free and 5 years survival.

le 16 out of 29 patients are without jaundice and portal hypertention. 82 patients were operated on primarily, which were subjected to analyse various prognostic factors. The other 21 patients were operated on initially in another hospital and then referred to our hospital for persistant jaundice.

The correlation among the age at the operation, the liver fibrosis, the size of the ductuli in the fibrotic biliary remnant at porta hepatis and prognosis has been studied. We chose these factors because all these factors are able to be evaluated at the operation. The prognosis was judged by using the five years survial's rate (SR) and the elimination rate of the jaundice (ER). The survival's rate was calculated by using the Kaplan-Meier method.

I) The survival rate

82 patients were divided into two groups according to the elimination of jaundice. Group 1 consisted of 38 patients whose jaundice was eliminated after the operation, and so did Group 2 45 patients whose jaundice remained. The survival curve is shown in (Fig. 7). 10 years survival rate of patients whose jaundice was eliminated is approximately 70%, while no patients whose jaundice was persisted could survive more than 10 years.

II) The age at the operation

The correlation between the age at the operation and prognosis is shown in (Fig. 8). with regards to the age at the operation, a considerable close correlation was found in our series but not so close correlation compared with Prefessor Kasai's data.

III) The degree of the liver fibrosis

We divided the patients into 3 groups accoding to our classification which we reported previously: the mild (F1), the moderate (F2) and the severe liver fibrosis (F3).

There is close correlation between degree of liver

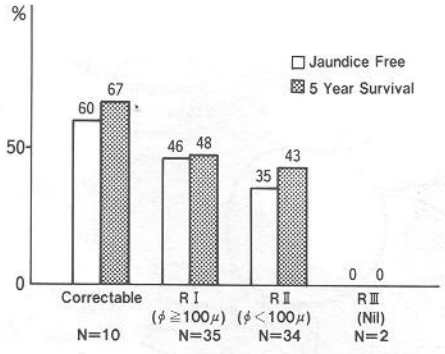
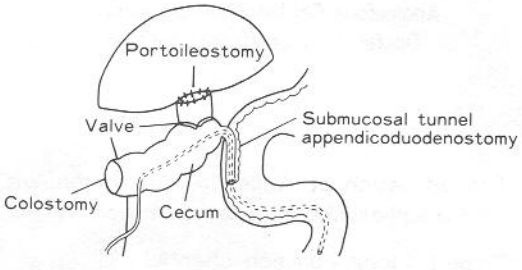


Fig.10. Correlation among the size of ductuli at porta hepatis, jaundice free and 5 years survival.



Operative procedure(I)

Hepatic portoduodenostomy with interposition of ileocecoappendix and colostomy for external biliary drainage

Fig. 11

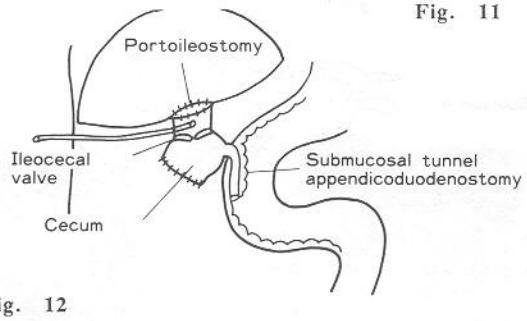


Fig. 12

Operative procedure(II)

Hepatic portoduodenostomy with interposition of ileocecoappendix

fibrosis jaundice free and five years survival (Fig. 9).

IV) The size of ductuli at the porta hepatis

We divided the patients into 4 groups according to the size of the ductuli at the porta hepatis as we reported previously: correctable, RI whose diameter of ductuli is over 100 micron, RII whose is under 100 micron and RIII whose has no ductuli (Fig. 10) shows the correlation between the size of ductuli and the jaundice free. We found the close correlation between these two factors and so did between the size of ductuli and five years survival.

Fig. 13. Operated cases with hepatic portoduodenostom with interposition of ileocecoappendix.

No.	Cases	Age at ope.days	Proce-dures	Post.op. periad	Cholan-gitis	Jaundice	Prognc
1.	S.Y.	55	I	17 m	+	--	gooc
2.	A.H.	60	"	14 m	-	--	ex.
3.	N.H.	100	II	12 m	-	--	ex.
4.	H.C.	65	"	3 m+	+	--	died
5.	S.I.	113	"	11 m	±	--	ex.

Operative procedures
I : Hepaticoduodenostomy with interposition of ileocecoappendix
II : same above without cecostomy

c. New modification of Kasai operation

Recently, we are doing new operative technique at Kasai operation. Firstly, I designed the hepatic portoduodenostomy with the interposition of the ileocecoappendix with cecostomy (procedure I; Fig. 11). Secondly, the cecum was closed primarily (procedure II; Fig. 12). This technique expects double antireflux mechanism of the ileocecal valve and the appendico-duodenostomy by the tunnel technique.

The first procedure was performed in two patients, and the second procedure in three patients (Fig. 13). All patients became jaundice free. The postoperative cholangitis, however, developed in three patients. One of the other two patients who had mild cholangitis shows moderate portal hypertension. Three patients including one patient who has mild cholangitis are doing verywell wit hout jaundice and obvious portal hypertention.

d. Analysis of the prognostic factors and the choice of the operative procedure

Based on our results, it enabiles to predict the prognosis at the initial operation by using frozen section. Patients with BA were divided into 3 groups according to the predictable prognosis and we considered the indication of Kasai operation retrospectively (Fig. 14). Group I includes all

Fig. 14. Operated cases with hepatic portoduodenostomy with interposition of ileocecoappendix.

Group	Prog.	Neces.of Liver Trans.	Op. proce.	Proportion
I Correctable or R I in F1	Good	Low	Kasai or modified Kasai	31%
II R II in F1 or RI RII inF2	Fair	Moderate	Kasai No stoma	61%
III R III or R I or RII in F 3	Poor	High	No Kasai	8%

correctable or RI cases with F1, which were expected good prognosis by Kasai operation. In this group, we concentrate to do our best by Kasai operation or its modification with or without stoma. Group II includes the patients with RII and FI, RI or II and F2 whose prognosis may be fair and whose possibility of liver transplantation may be moderate. In this group, we do Kasai operation without stoma, which is thought to be hazardous for the liver transplantation. Group III includes the patients with RIII with RIII or F3 except correctable, which is impossible to obtain a good prognosis by Kasai operation. In this group Kasai operation should not be performed and prepared for the liver transplantation.

B) Congenital biliary dilatation (CBD)

A current topic on this lesion is that since Babbit's report [2], the high incidence of the association of the anomalous choledochopancreatic ductal junction (ACPDJ), that is, the long common channel formation is found approximately in 10% in patients with CBD and is closely correlated not only to the pathology of the CBD but also various biliopancreatic lesions.

ACPDJ, that is, the long common channel formation permits the pancreatic juice reflux into the bile duct which causes various biliary lesions, cholangitis, gallstone, perforation, and biliary tract carcinoma. Also bile reflux into the pancreatic duct causes various pancreatic lesion,

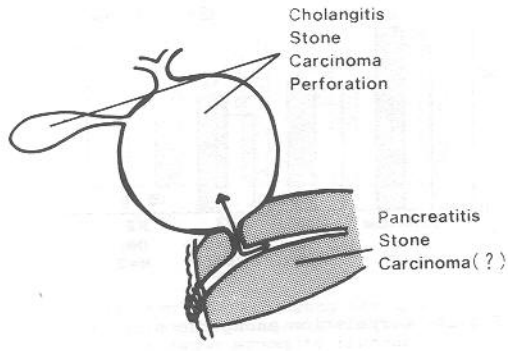


Fig. 15

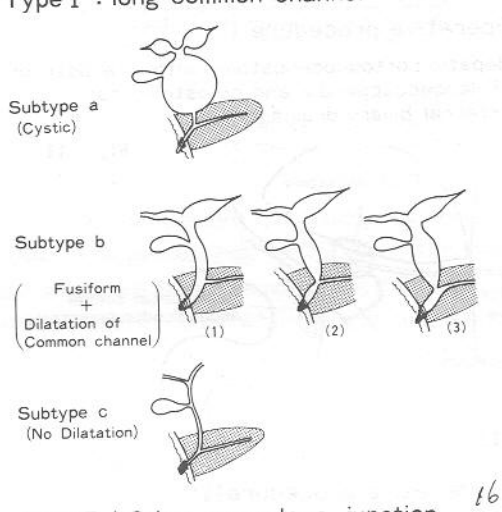


Fig. 16

panceratitis, stones, and may be carcinoma (Fig. 15).

Our classification of anomalous choledochopancreatic ductal junction is shown in (Fig. 16).

ACPDJ is divided into two main types:

Type (I)- the long common channel type
Type(II)- miscellaneous types of the anomalous junction

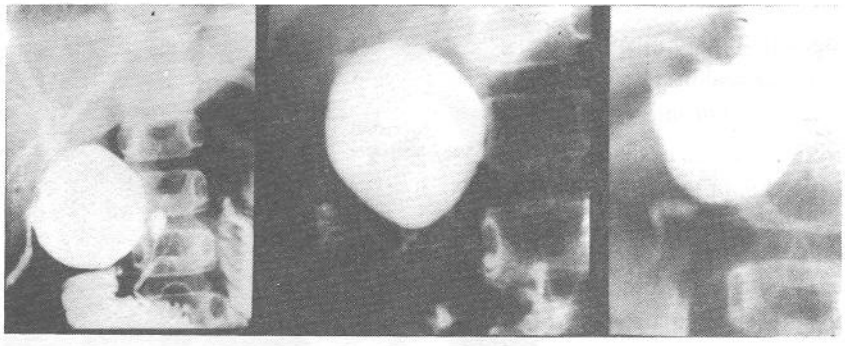


Fig. 17. Cholangiographies of Type Ia.

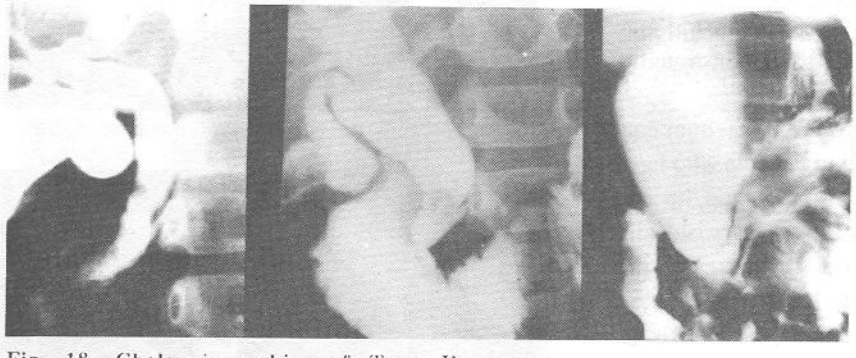
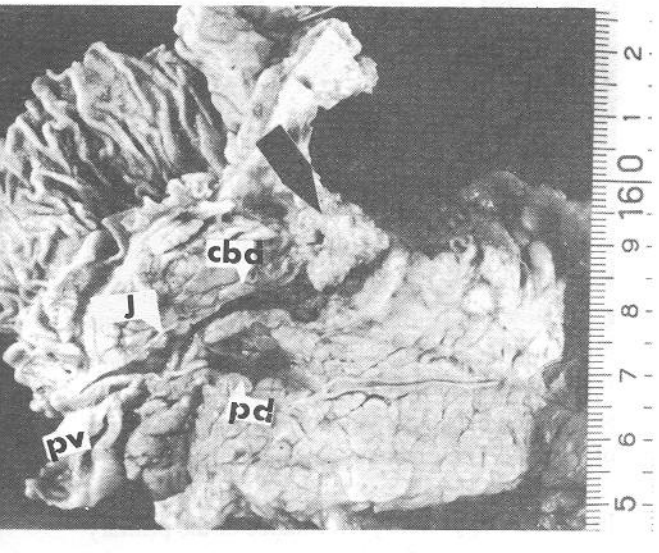


Fig. 18. Cholangiographies of Type Ib.



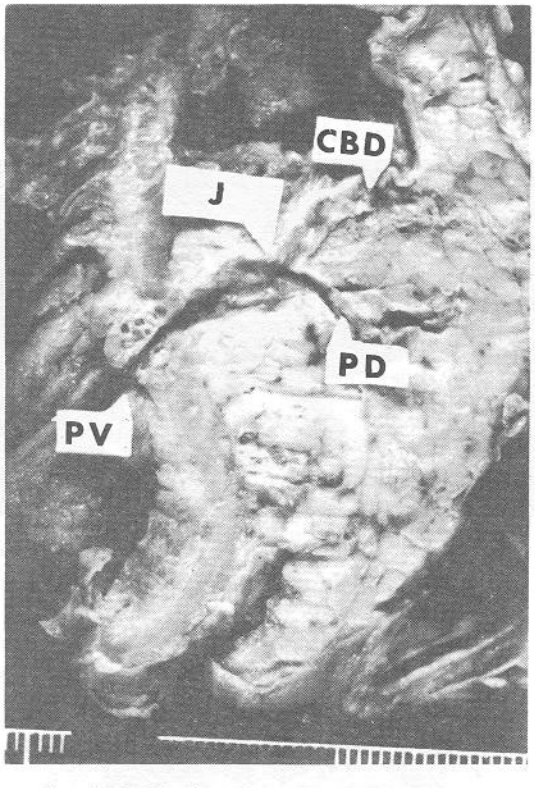


Fig. 19 - Fig. 20. CBD: common bile duct, PD: pancreatic duct, J: junction, PV: papille of vater

Type (I) was divided into three. Type (I-a) shows cystic dilatation of the common bile duct, Type (I-b) shows fusiform dilatation and Type (I-c) shows no dilatation (Fig. 17,18). Type (I-a) and Type (I-b) are so-called choledochal cyst. Based

on this classification, the congenital biliary dilatation is considered to be one of anomalous choledochopancreatic ductal junction syndrome or long common channel syndrome.

The most serious biliary complication is the carcinoma (Fig. 19) shows the carcinoma of the common bile duct associated with very long common channel formation in 77 year-old male.

(Fig. 20) shows an another patient with carcinoma of the common bile duct in 62 years old male. Very long common channel is demonstrated.

Now the high incidence of the biliary tract carcinoma is widely recognized in patients who have ACPDJ [3]. With regard to pancreatic lesion, (Fig. 21) shows the CT of a two years old girl with a huge choledocal cyst. In this case, the pancreas was felt to be quite hard, so we took a biopsy which shows a chronic pancreatitis as shown in (Fig. 22).

(Fig. 23) is ERCP of 4 years old girl with cystic dilatation. You can see quite dilated P.D. which suggests chronic pancreatitis. So I want to stress that you should pay an attention to this association when you have pancreatitis in children.

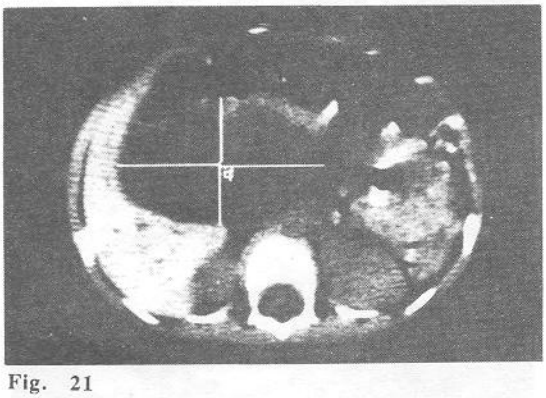


Fig. 21

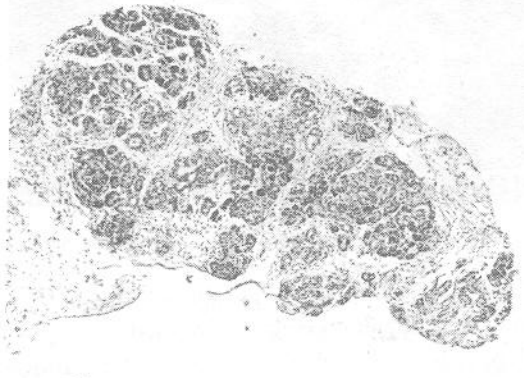


Fig. 22

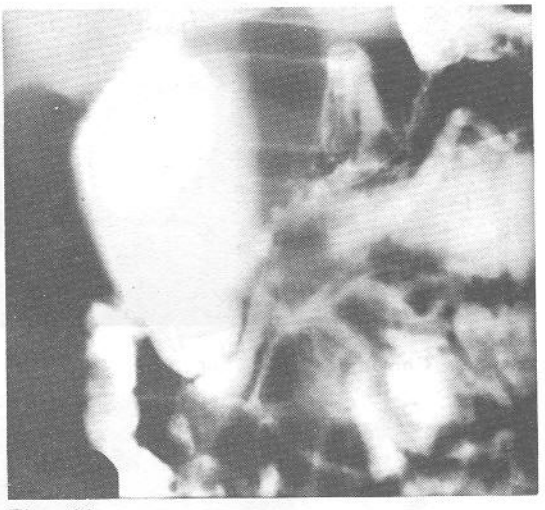


Fig. 23

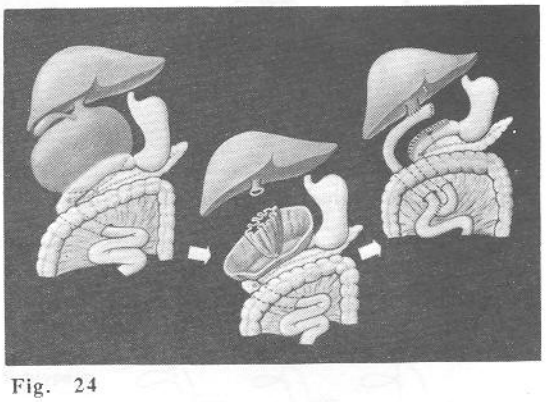
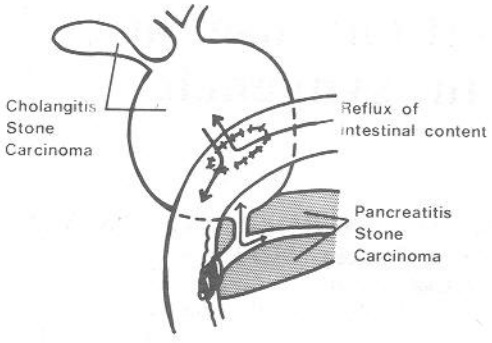


Fig. 24

With regard to the treatment of CBD, the basic principle has already been established. Firstly, total excision of dilated common bile duct should be carried out primarily. Secondly, with regard to reconstruction of the biliary tract, hepaticojejunostomy (R-Y) is most commoly used in Japan with very satisfactory result, but some people are still doing hepaticoduodenostomy after excision of the cyst. Their results are quite satisfactory.

In case of impposible performance of the primary excision because of various reasons, for example, huge cyst, perforation and very poor general condition, only external biliary drainage should be carried out. Internal drainage operation should not be done.



Postoperative Complications in patient with Choledochocystoduodenostomy

Fig. 25

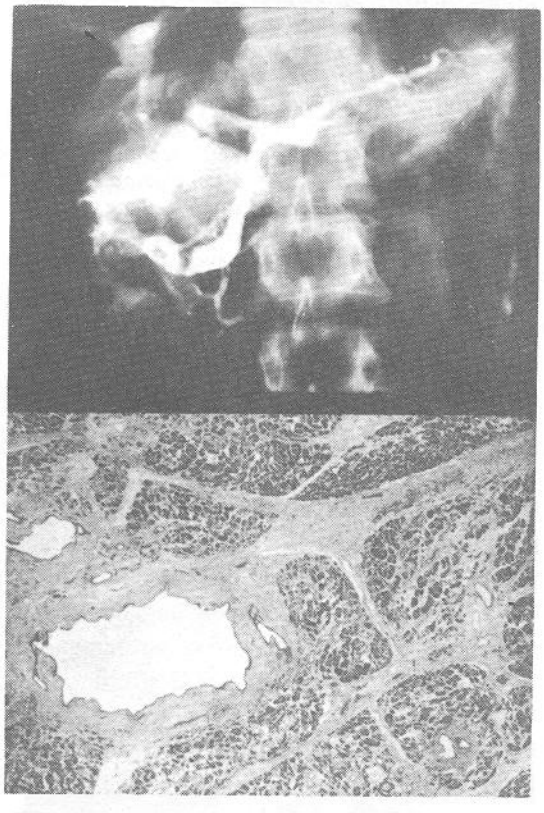


Fig. 26

If it is difficult to perform the total excision, especially the distal part of huge cyst, mucolysis of distal common bile duct should be carried out instead of total excision in order to avoid carcinoma (Fig. 24) shows the technique of mucolysis to the distal part of the cyst.

Choledochocystoduodenostomy was frequently performed in the past, but various serious complications occurred frequently, for example, cholangitis, stone, carcinoma in the bile duct and also pancreatitis, stone in the pancreas (Fig. 25).

(Fig. 26) is an ERCP at the histology of the pancreas at the bottom of 26 years old female whose choledochocystoduodenostomy has been performed at 6 year of age. The pancreaticoduodenectomy has been done for her chronic pancreatitis recently.

Therefore in old patinets whose internal biliary drainage operation had been already performed in the past, revision should be carried out if the patient presents any clinical symyptoms. If there is no symptom, very careful follow-up is necessary.

References

1. Kasai M: Current status and problems in the treatment of congenital biliary atresia J Jpn Surg Soc (Jpn), 88:1401, 1987.
2. Babbit D.P.: Conginital choleodochal cyst: New etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol 12:231, 1969.
3. Suda K, and Miyano T: An abnomal pancreatico-choleodochal-ductal junction in cases of biliary tract carcinoma. Cancer, 52:2086, 1983.