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Fate of Non-Excisional Surgery in Choledochal Cyst in Children[1]

Woo-Ki Kim, Kwi-Won Park, Seong-Cheol Lee and Suk-Koo Lee

Department of Pediatric Surgery, Seoul National University Children's Hospital and College of Medicine, Seoul National University, Seoul 110-744, Korea

= Abstract =Drainage surgery between choledochal cyst and intestine was the standard mode of treatment until mid-1980 at the Department of Pediatric Surgery, Seoul National University, Seoul, Korea. A total of eight cases were treated with by-pass surgery between 1978-80 period. From mid-1980, excision of the cyst has been used as a standard mode of surgical treatment for the choledochal cyst. Twenty one excision were carried out in 1980-1986 period. Four of those patients who received excision of cyst had received bypass surgery as a primary treatment in the past. Repeated attack of cholangitis or jaundice were the main reasons for reoperation, i.e. the excision of cyst. From 30 choledochal cyts, 11 patient received non-exicisional surgery as a primary mode of treatment. Six required reoperations including five excisions. From eight initial drainage procedures, three patients required reoperation due to morbidity from internal drainage procedure of choledochal cyst. However, there are four patient [50%] who are free from the morbidity such as cholangitis, jaundice, recurrent pain or stricture formation. The fate of cysto-jejunostomy should be closely followed for a life time to clarify the possibility of development of cancer.

Key words: ***Choledochal cyst***

INTRODUCTION

The first article from Korea regarding choledochal cysts on international journal appeared in 1969 in Archives of Surgery (Lee *et al.* 1969).

They reviewed about 500 cases reported in the literature and added their own 9 cases. Like others, they concluded internal bypass surgery, cystojejonostomy in Roux-Y fashion is a safe, reasonable surgical operation. Most of the Korean surgeons followed their suggestion, since they were the pioneers of pediatric surgery in Korea. Even during one of authors' pediatric surgical training in the United States, which was between 1975-77, internal bypass surgery was performed without any doubt in mind all over the United States (Duckett *et al.* 1971; Kim 1981). Mortality and morbidity of by-pass surgery were low compared to that of excisional surgery reported initially (Tsardakas and Robnett 1956). However, those that used drainage procedures recognized that a significant number of patient develop recurrent cholangitis or stones from biliary stasis for a variety of reasons including inadequate cyst decompression and stricture at the anastomosis (Filler and Stringel 1980).

Though, excisional surgery of choledochal cyst began to appear on international journals in '70s (Ishida *et al.* 1970; Kasai *et al.* 1970), but their voices were not heard greatly outside of Japan.

Our first problem came from a 5 years old girl who had received cyste-dueodenostomy at age of 1 year 9 months for choledochal cyst. After surgery, she had developed many attacks of abdominal pain and occasional jaundice. At

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Table 1. Drainage vs. excision of choledochal cysts

Year	No. of total	Drainage	Excision	Others
1978	3	3		
1979	2	2		
1980	5	3	2	
1981	3		3	
1982	2		1	1*
1983	8		8	
1984	1		1	
1985	3		3	
1986	3		3	
Total	30	8	21	1*

(*: exploratory laparotomy)

reoperation, age of 5 year 6 months, her anastomotic opening was completely closed. Another bypass, cysto-jejunostomy Roux-en-Y fashion was made. Her jaundice disappeared immediately, but her abdominal pain did not go away. Because of this patient, we turned our attention to the excisional surgery of choledochal cyst.

The first excision was done in November of 1980 on 3 months old boy with choledochal cyst. Since then, excisional operation has been our choice of operation (Table 1).

This study was undertaken to elucidate the natural course of the patients who underwent bypass or drainage procedure between the cyst and intestine for the treatment of choledochal cyst.

MATERIALS AND METHODS

This study is based on the follow up study on the 11 patients (Table 2) who had non-excisional surgical procedure for their initial treatment from 30 patients with choledochal cyst during 1978-86 period. Two of these 11 patients received initial drainage procedure elsewhere. From these 30 patients, there were six reoperations. Five of them previously received drainage procedure, three from our hospital and two from elsewhere. Total 11 patients were selected for the non-excisional surgical group. Their postoperative courses were followed closely ad studied by U/S, UGI, LFT, Isotope and CT if necessary.

There were no statistical differences between excisional group and non-excisional group on the matter of clinical settings, such as age at the onset of symptoms, age at the operations, intervals between onset and surgery, and the presenting symptoms and signs. Most of the patient were getting medical treatment at the earlier age. Table 3. shows the age at operation in all choledochal cysts.

All choledochal cysts we experienced belonged to type I, i.e. fusiform dilatation of common bile duct except one who had a cystic dilatation at left hepatic duct.

RESULT

1. Fate of 8 patients who received drainage procedure in 1978-80 period as an initial surgical treatment

These patients were followed for 8-10 years

Table 2. Summary of 11 non-excisional group of choledochal cyst

I.D. No.	Age	Sex	Initial surgery	2nd Surgery
1201933 (1)	10	F	cysto-jejunostomy	not required
1203814 (2)	10	F	cysto-jejunostomy	not required
1217054 (3)	14	M	cysto-jejunostomy	lost in F/U
1220575 (4)	2	F	cysto-jejunostomy	not required
1247018 (5)	6	F	cysto-jejunostomy	excision at 13
1373587 (6)	5M	F	cysto-jejunostomy	excision at 2
1376644 (7)	1	F	cysto-duodenostomy	cysto-jejunosotmy, 5
1409416 (8)	2M	M	cysto-jejunostomy	not required
1697632 (9)	3	M	cysto-duodenostomy	excision at 5
1707020* (10)	10M	F	closure of perforation	excision at 3
1868941* (11)	3	F	cysto-duodenostomy	excision at 5

*: initial surgery was done elsewhere

Table 3. Age at the operation (No. of choledochal cyst)

Age at the operation	No. of choledochal cyst
less than 1 year	8
1-5 years	13
over 6 years	9
total	30

from their initial internal drainage procedure between cyst and intestine. Two patients eventually received excision of the cyst after 2 and 7 years later. They suffered from frequent attack of cholangitis and abdominal pain. Liver biopsy showed liver damage in both patients. One patients who received excision 7 years later showed a picture of early biliary cirrhosis. Closure of anastomosis was observed in one patient who had excision 2 years after the initial cysto-jejunosotmy.

One patient as mentioned earlier still refused to receive any further surgical procedure although she is symptomatic from mild pancreatitis evidenced by the elevated serum amylase. Her cysto-jejunostomy is patent. But she has a huge cyst evidenced by the ultrasound.

Four patients are asymptomatic, and having normal life. These four patients are followed by ultrasound, liver function test. Their growth and nutritional curve are within normal limit. The family members are well aware the risk of drainage procedure and are willing to receive further surgical procedure if it is necessary. In two, no residual cyst can be demonstrated by either ultrasound or CT scan. Their ages at operation were 2 months and 2 years respectively. The other two still have residual cyst demostrable on CT scan although they are completely asymptomtic.

One patient, who will be 24 years old in 1988 was lost to follow-up. She was perfectly healthy until age of 18 (1982). Table 4. represents the summary of these eight patients.

2. Reoperation on 11 non-excisional group/or in total 30 patient.

There were six reoperations on these 11 patients who did not receive excision of the cyst as an initial surgical procedure. There were no reoperations among patient treated with excision. Four patients (two thirds) required excision of choledochal cyst after initial internal drainage procedure. All four were symptomatic. In three of these, there were strictures at the anstomosis. Their liver function usually recovered very slowly after the establishment of proper drainage. The longer the interval between initial drainage and excision, the worse the liver pathology was. (Table 5) A three years old infant who had bile peritonitis at age of 10 months due to the spontaneous perforation of choldeochal cyst also required an excision. A patient with a revised cysto-jejunostomy definitely requires another operation to be free from her present symptoms.

Table 4. Fate of 8 drainage procedure for choledochal cyst

1 conversion from obstructed cysto-duodenostomy to cysto-jejunostomy: symptomatic, refusing surgery
2 excision of cysto-jejunostomy, 2 & 7 years later with liver damage (portal fibrosis & cirrhosis)
4 normal growth/asymptomatic, 8-10 years postop.
1 lost during follow-up/well until age of 18

Table 5. Summary of 6 reoperation on choledochal cysts

1 conversion of cysto-duodenostomy (1 yr. 8 mos.) to cysto-jejunostomy at age of 5 yr 6 mos.
1 excision of cyst at 3 yr 6 mos after closure of perforation of cyst at 10 mos. of age.
4 excision of cyst after previous drainage procedure,
  a. all 4 were symptomatic at time of reoperation,
  b. interval: 1.5 yr., 2 yr., 7 yr., 10 yr.
  c. anastomosis site: only one is patent at reoperation
  d. cirrhotic liver: 2/4
  e. reason for excision: 2 cholangitis, 2 pain, 2 jaundice

DISCUSSION

This study proves what we already knew. Morbidity (recurrent pain, jaundice, stricture formation or obvious cholangitis) and re-operation rate after the drainage procedures are reported as high as 58 per cent and 38 per cent among 93 patients with choledochocysto-duodenostomies, a 34% morbidity and 13% reoperation rate among 53 with cysto-jejunostomies by Flanigan (1975). In our study, three patients from eight cystojujunostomies required reoperation (37.5%).

The major controversy regarding the treatment of choledochal cysts remains at the choice between internal drainage and excision as the procedure of choice. The chief advantage of the internal drainage procedures is the technical ease with which the operation may be performed with the low operative mortality (Spitz 1978). It is no wonder why these drainage procedures were so popular. However, it is also well known that these drainage procedures are against basic surgical principle of achieving a mucosa-to-mucosa anastomosis (Grove 1957), and of removing a dilated chamber with no musculature. In some cases, thick walled cyst do not collapse after decompression. Also it is impossible to place the anastomosis at a site on the cyst which will ensure dependent drainage when the child is either in the upright or supine position (Filler and Stringel 1980). Failure to provide mucosa-to-mucosa anastomosis, and destruction of lining cells of the cyst probably account for the stricture or complete closure of anastomosis seen our case 6,9 and 11. Also the initial cystoduodenostomy of case 7 was found to have complete obstruction of anstomosis.

Another disadvantage of drainage procedure is a possibility of persistent reflux of pancreatic juice into the bile duct. This pancreatic juice is blamed for the further destruction of mucosa and cyst wall, and for the development of cholangitis. Higher serum amylase level observed in case 5 and 7 probably do represent the sequellae of persistent pancreatic reflux. Pancreatitis might be responsible for the pain attack (Okada 1983). Pancreatic reflux into bile duct secondary to the anomalous arrangement of the pancreaticobiliary ductal system (APBD) is regarded as one of the main pathogenesis for the choledochal cyst (Todani *et al.* 1984; Komi *et al.* 1988).

Complete excision probably eliminates the possibility of bile duct carcinoma which has been reported in patients with choledochal cyst. Estimated incidence of 3 per cent was reported, which is about 20 times greater than expected (Kagewa *et al.* 1978). However, there are no concrete evidence that secondary excision of the cyst which was effectively drained and functioning will prevent biliary cancer (Spitz 1978). For the cases 1,2,4 and 8 only a warning from the surgeon were given at this time.

Improvements in surgical skills, and in pre/postoperative management have eliminated the many hazard of cyst excision (Lilly 1979). To the experienced hand, excision of cyst are safe and effective way to treat this disease.

Finally, one question should be answered: does the asymptomatic, internally drained patient require excision to prevent cancer?

In conclusion, three from eight patients with internally drained choledochal cyst eventually required an excision of cyst. Six out of 11 non-excisional procedures required secondary surgery. Further close observation is needed to evaluate the drainage procedure, especially the cancer risk.

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= 국문초록 =

소아 담관낭종에서 비절제수술의 예후

서울대학교 의과대학 소아병원 소아외과

김우기 · 박귀원 · 이성철 · 이석구

1980년 중반까지 서울대학교병원 소아외과에서는 담도낭종을 절제술이 아닌 방법, 즉 낭종과 소장을 연결하는 방식으로 치료하였다. 모두 8명의 환자가 이 방식으로 치료하였는데, 3명의 환자에서 재수술이 필요하여 낭종절제술로서 치료했다.

이후에는 모두 낭종절제술로 치료했으며, 도합 21명의 환자가 절제술을 받았다 (1986년 말까지). 본연구의 목적은 절제술을 못받은 8명의 환자를 포함하여 모두 11명의 환자가 겪는 경과를 밝히기 위함이다.

예측한바대로 절제술을 못받은 환자는 상당수(37.5%)에서 재수술이 필요했으나 50%는 정상적인 생활을 하고 있음이 밝혀졌고, 좀더 장기간의 추적조사가 필요함을 알 수 있다.