Chulalongkorn Medical Journal

Volume 40
Issue 3 *March 1996*

Article 5

3-1-1996

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Chandrakamol, Bidhya; Vejchapipat, Paisam; Chittmittrapap, Soottipom; and Poovorawan, Yong (1996) "Biliary atresia: 10 - year experience at ChulalongkomUniversity Hospital," *Chulalongkorn Medical Journal*: Vol. 40: Iss. 3, Article 5.
Available at: https://digital.car.chula.ac.th/clmjournal/vol40/iss3/5

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นิพนธ์ต้นฉบับ

Biliary atresia: 10 - year experience at Chulalongkorn University Hospital

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Chandrakamol B, Vejchapipat P, Chittmittrapap S, Poovorawan Y. Biliary atresia: 10 - year experience at Chulalongkorn University Hospital. Chula Med J 1996 Mar;40(3): 193-202

Objective : *To review and evaluate biliary-atretic patients surgically treated at Chulalongkorn University Hospital*

Setting : *Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Chulalongkorn University Hospital, Bangkok, Thailand*

Design : *Retrospective descriptive study*

Subjects : *One hundred and eight jaundiced infants suspected with biliary atresia from January 1985 to December 1994*

Methods : *The data of all patients were reviewed in the areas of general informations, diagnosis,treatment,complications, and outcome of the operation*

Results : *There were 108 persistent cholestatic jaundice patients undergoing diagnostic laparotomy, 79 (73.15 %) of whom were diagnosed as biliary atresia intraperatively. Of these,49/79 (62.02 %) were operated on before 2 months old. The male:female ratio of the 79 was 1:1 (40:39). There were 4 cases*

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(5%) of type I biliary atresia and 75 cases (95 %) of type III biliary atresia. Seventy patients (88.60 %) were treated with Kasai hepatic portoenterostomy, 5(6.33 %) with hepatic portocholecystostomy and 4 cases (5.07 %) with hepatic choledochojejunostomy. Detalied data in (56 cases) indicated that ascending cholangitis was the most common post-op complication (46.43 %). Thirty-four patients [34 of 56 (60.7 %)] had bile drainage after Kasai operation and 20 of them had satisfactory results and free from jaundice. The other 22 cases classified as failures and 9 of them died during follow up. There were 4 patients underwent orthotopic liver transplantation after failed Kasai operation.

Key words : *Biliary atresia, Neonatal jaundice.*

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Received for publication. January 13,1996.

พิทยา จันทรกมล, ไพศาล เวชชพิพัฒน์, สุทธิพร จิตต์มิตรภาพ, ยง ภู่วรวรรณ. ท่อน้ำดีตีบตันแต่กำเนิด : ประสบการณ์ 10 ปี ในโรงพยาบาลจุฬาลงกรณ์. คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย. จุฬาลงกรณ์เวชสาร 2539 มีนาคม:40(3): 193-202

วัตถุประสงค์ : *เพื่อรวบรวมและประเมินผลการผ่าตัดรักษาผู้ป่วยทางเดินน้ำดีตีบตันแต่กำเนิด ของโรงพยาบาลจุฬาลงกรณ์ คณะแพทยศาตร์ จุฬาลงกรณ์มหาวิทยาลัย*

สถาบัน : *หน่วยกุมารเวชศาสตร์ ภาควิชาศัลยศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย โรงพยาบาลจุฬาลงกรณ์ สภากาชาดไทย*

รูปแบบการวิจัย : *การศึกษาย้อนหลังแบบพรรณนา*

การคัดเลือกผู้ป่วย: *ผู้ป่วยที่สงสัยว่าเป็นโรคทางเดินน้ำดีตีบตันแต่กำเนิด จำนวน 108 ราย ตั้งแต่เดือนมกราคม พ.ศ. 2528 ถึงเดือนธันวาคม พ.ศ. 2537 เป็นระยะเวลา 10 ปี*

วิธีการศึกษา : *รวบรวมศึกษาข้อมูลทั่วไป, การวินิจฉัย, การรักษา, ภาวะแทรกซ้อนและผลของการผ่าตัดรักษาผู้ป่วยท่อน้ำดีตีบตันแต่กำเนิด*

ผลการศึกษา : *จากการผ่าตัดเปิดช่องท้อง ผู้ป่วยที่สงสัยว่าเป็นโรคท่อน้ำดีตีบตันแต่กำเนิด 108 ราย เพื่อยืนยันการวินิจฉัยด้วย intraoperative cholangiography พบว่าเป็นโรคท่อน้ำดีตีบตัน แต่กำเนิดจริง 79 ราย (73.15 %) ผู้ป่วย 49 ราย จากผู้ป่วยท่อน้ำดีตีบตันแต่กำเนิดทั้งสิ้น 79 ราย (62.02 %) ได้รับการผ่าตัดก่อนอายุ 2 เดือน เพศชายต่อเพศหญิงเท่ากับ 1:1 (40 ต่อ 39) ผู้ป่วย 75 ราย (88.60 %) ได้รับการผ่าตัดรักษาแบบ Kasai hepatic portoenterostomy, 5 ราย ได้รับการผ่าตัดแบบ hepatic portocholecystostomy (6.33 %) และ 4 ราย (5.07 %) ได้รับการผ่าตัดแบบ hepatic choledochojejunostomy จากข้อมูลที่รวบรวมได้ 56 ราย พบว่า ascending cholangitis เป็นโรคแทรกซ้อนหลังผ่าตัดที่พบบ่อยที่สุด (46.43 %), ผู้ป่วย 34 รายจาก 56 ราย (60.7 %) มีอาการเหลืองลดลงหลังการผ่าตัด และในจำนวน 34 รายนี้ 20 ราย (35.7%) อาการเหลืองหายไป ส่วนที่เหลือ 22 ราย อาการเหลืองไม่ลดลง หลังการผ่าตัด และผู้ป่วย 9 ราย เสียชีวิตขณะติดตามการรักษา*

The successful treatment of biliary atresia in infants has been developing for years. Prior to 1959 the 5-year survival rate of these patients was less than 5 %.[1] Since Kasai presented the hepatic portoenterostomy surgical technique, the survival rate has much improved, especially when the correction was performed within the first 2 months of life.[2] If Kasai operation cannot produce bile drainage, liver transplantation becomes another choice of treatment.

The objective of this study was to evaluate the treatment of biliary atresia at the Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Chulalongkorn University Hospital.

Patients and Methods

All infants with persistent cholestatic jaundice who underwent diagnostic laparotomy and intraoperative cholangiography between January 1985 and December 1994 were retrospectively reviewed. The relevant data was collected from the in-patient records, operating room records, out-patient files and pathological reports.

The retrospectively descriptive review included the number of patients, sex, final diagnosis, preoperative bilirubin level, age of the patients at the time of surgery, type of operations performed,complications, and the results of operation. In addition,the data indicated criteria for diagnosis. The anatomic type of biliary atresia and operative time were also reviewed.

Results

During the ten-year-period (1985-1994) there were 108 persistent cholestatic jaundice patients who underwent diagnostic laparotomy with intraoperative cholangiography at Chulalongkorn Hospital. The infants could not be definitely diagnosed either clinically or by means of ultrasonograph and hepatobiliary scan. Diagnostic laparotomy with intraoperative cholangiography was indicated to prove the biliary atresia. At laparotomy, biliary atresia was diagnosed in 79 cases (73.15 %), and biliary hypoplasia in 3 cases. The other 26 cases (24.07 %) were diagnosed as hepatitis or cholestasis of other cause. (Table 1). Sixty three (58.33 %) out of all 108 cases were operated on before the age of 2 months (Diagram 1).

Among the 79 biliary atresia patient,the male to female ratio was 40:39 i.e. 1:1 and 49 infants (62.02 %) were operated on before 2 months of age. Of the 79 biliary atresia patients, 4 cases (5.06 %) were type I biliary atresia and 75 cases (94.93 %) were type III - biliary atresia. There were no type II biliary atresia . Seventy infants (88.60 %) were treated by Kasai hepatic portoenterostomy. Five cases (6.33 %) had hepatic portocholecystostomy and 4 cases (5.07 %) underwent hepatic choledochojejunostomy.

Table 1. Final diagnosis of 108 jaundiced infants underwent diagnostic laparotomy

Final diagnosis	Male	Female	Total
Biliary atresia	40	39	79
Patent biliary tract*	18	8	26
Biliary hypoplasia	3	0	3

* diagnosed later of hepatitis or cholestasis

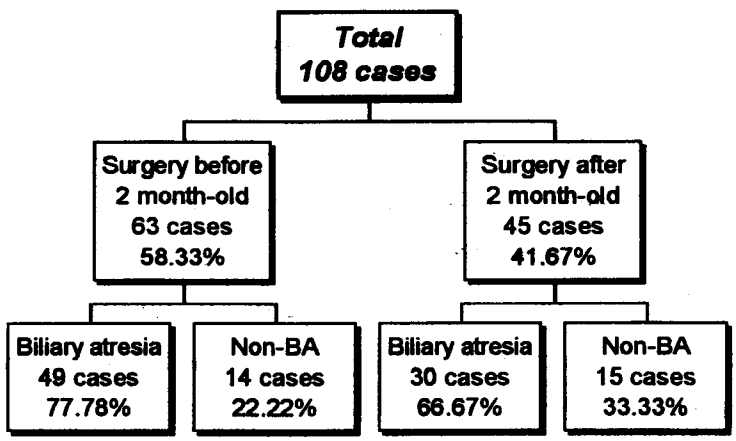

Diagram 1. The age at the time of the diagnosis.

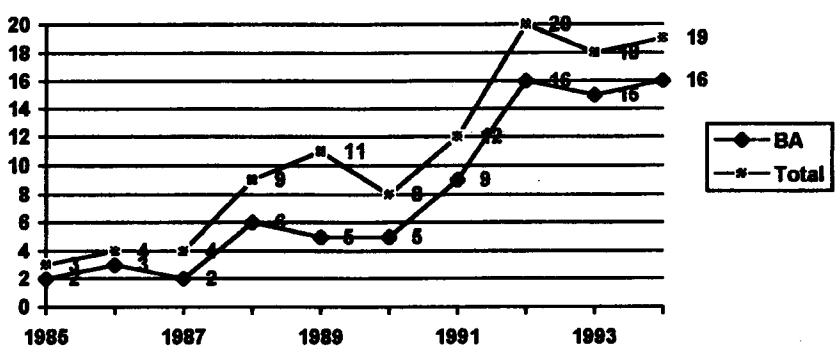

Diagram 2. The number of jaundiced and biliary-atresia-patients operated per year.

The number increased up to about 20 cases a year in the last 3 years. There were seven re-operations for revision of the Kasais operation. There were 4 liver transplantations in patients who had end-stage chronic liver failure.

Detailed data for 56 biliary atresia infants (70.88 %) was available for this study. Thirty-three of these cases (62.5 %) had jaundice before 1 month of age 13 cases (23.2 %) were affected at the age of 1.2 months and 8 cases (14.3 %) revealed jaundice detected after 2 months. Total average bilirubin level at admission was 14.19 ± 6.94 mg/dl (mean ± SD). The operative time was 190.80 ± 36.79 minutes. From the pathological reports, the histological liver examination showed that biliary cirrhosis was present in 47 patients (83.93 %) while there was only bile stasis without histological evidence of cirrhosis in 9 patients. Post-operative complications within 30 days after operation are shown in Table 2. Failure of bile drainage after surgery is separately discussed.

Follow-up data collected from the in and out-patient files were analysed to determine the results of the operation and the long term results. Four patients underwent liver transplantation and three have been living well. All patients were followed for more than three months. The average follow-up time was 15.63 ± 13.68 months (mean ± SD). The patients were classified into 4 groups according to the outcome after operation by the following criteria:

Group A: Clinical remission, jaundice free,total bilirulin < 2 mg %

Group B: Inadequate bile drainage with less jaundice total bilirubin level decreased from preoperative level but still > 2 mg %

Group C: Failure to create bile drainage, persistent jaundice, total bilirubin increased with early signs and symptoms of progressive chronic liver failure

Group D: Death from liver cause during follow-up

Table 2. 30 - day postoperative complications in the 79 cases underwent Kasai operation.

Complications	Number (cases)
Death	1
Wound dehiscence	1
Leakage of anastomosis	2
Gut obstruction	2
Cholangitis	27

In this series , thirty-four patients (34 of 56,60.7 %) had bile drainage after the Kasai operation. Twnenty (35.7 %) had satisfactory result and were free from jaundice, and 14 had partial bile drainage. The other 22 cases were classified as failure and 9 of them died during follow up. The result are shown in Diagram 3.

In the patients who had bile drainage (groups A and B) , 25 of 34 cases (67.9 %) underwent surgery before 2 months of age, whereas 15 of 22 patietns (67.3 %) who had no bile drainage after the procedure and had progressive liver damage (group C and D) underwent the operation after 2 months of age. There wer 4 cases who underwent orthotopic liver transplantation, after failure of Kasai operation. One of them died. The other three did have good result and have enjoyed normal life.

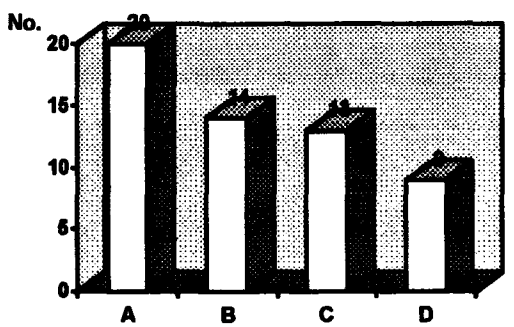

Diagram 3. The result of Kasai's operation classified by the bile drainage after surgery.

Group A: Clinical remission, jaundice free, total bilirubin < 2 mg %

Group B: Inadequate bile drainage with less jaundice,total bilirubinlevel decreased from preoperative bilirubin but still > 2 mg %

Group C: Failure to create bile drainage, persistent jaundice,total bilirubin increased with early developed signs and symptoms of progressive chronic liver failure.

Group D: Death during follow-up according to the liver causes.

Discussion

Prior to the success of Kasai et al on portoenterostomy procedure in the treatment of biliary atresia, the outlook for patients with this disorder was almost always fatal. The average survival for patients with untreated biliary atresia was 18 months with survival rates of 4 % at 4 years.[3] After hepatic portoen- terostomy was

introduced, the outcome for biliary aresia patients brightened. Two-thirds of the patients improved. Unfortunately, not all patients can be sucessfully treated by hepatic portoenteros- tomy. If this technique is unsuccessful increating bile drainage, survival beyond 12 months is unlikely. Recently, liver transplantation has become a commonly accepted therapy for children with end-stage liver disease. Over a decade of clinical experience with liver transplantation is now available to assess progress. Otte et al[4] reported a 1-year survival rate of 85 % after full-size graft liver transplantation. Beath et al[5] noted that the five-year survival rate for patients with liver transplantation for biliary atresia was 65 %. However,there were a number of patients who died while waiting for the transplantation due to the lack of donors. Considering the shortage of donors in our country, the relationship between the timing of the oparation and results, the Kasai operation should remain the primary treatment for biliary atresia and at least 50 % of cases should be successful or at least improved.

Poshyachinda and Poovorawan[6] revealed that the DISIDA (diisopropyliminodiacetic acid) hepatobiliary scan was very helpful. After introduction of the DISIDA scan, the percentage of biliary atresia cases in the jaundiced patients who underwent diagnostic laparotomy increased from 50-60 % in the 1980's to 70-80 % in the 1990 's

The two major complications of the Kasai operation are cholangitis and esophageal varices from portal hypertension. Chiba et al[7] found that 28/50 (56 %) patients had cholangitis after the Kasai operation. Howard[8],in a review of the literature, noted that esophageal varices in long-term survivors ranged from 31 % to 65 %. In this series there were 48 % cases of cholangitis and 14 % cases of esophageal varices after the Kasai operation occuring in both short and longer period of follow-up.

The timing of surgical intervention for biliary atresia remains a critical factor in the care of infants with this disease. Mieli et al[2] reported that if the Kasai operation was performed before 2-months of age the bile flow was greater than after 2-months of age. The inverse relationship between age at the time of operation and the result of treatment was widely accepted. During the period of this study , 25 of the 36 children operated on at less than 2 months of age (69.44 %) had good, result compared with the failure group of 7 of the 22 children (31.8 %) who underwent surgery before 2 months old. Grosfeld et al [9] reported the long-term result of hepatic portoenterostomy that 35 % patients had jaundice-free 5-year survival, 27 % had 3-year survival with jaundice,and 38 % with less than 3-year survival and progressive liver deterioration.

Ohi et al[10] reported on a large series of long-term survivors that 48 out of 251 had more than 10-year survival with normal quality of life and 73 % of the 48 were operated on before 2 - months age. The result of this study indicated that 35.7 % had good results and were jaundice free, 25 % got partial bile drainage, and 39.2 % failure rate which is close to the results of pediatric surgical

centers in Japan and the western countries.

The important factor in improving the result of the Kasai operation is early diagnosis and prompt management with proper surgical technique.[11] and the age at the time of operation affects the result of treatment. The Kasai operation is without doubt the first-choice treatment for biliary atresia before the age of 2 months. If the Kasai operation cannot create the bile flow or the patients deteriorate due to the progressive liver damage, liver transplantation, is an alternative for those patients with end-stage biliary atresia.

Conclusion

Biliary atresia is one of the most interesting disease in the field of pediatric surgery of which the treatment is surgical intervention . Early diagnosis is the key of successful treatment. By hepato-biliary scintigraphy, and ultrasonogram it can not make the accurate diagnosis. The diagnosis is made by, the diagnostic laparotomy Kasai operation is the treatment of choice, liver transplantation is another alternative for those who have pregressive liver damage after Kasai operation. Among 34 patients who underwent Kasai operation did have bile flow (60.7%), 20 cases (35.7%) with satisfactory result.

Acknowledgement

The authors wish to thank Assistant Prof. Voravit Sriwatanawongsa M.D. American Board of Surgery who performed the successful liver transplantation to those infants who failed to Kasai's operation.

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