HERE are few important studies and after review of them simple questions needs to be answered. Protocol for management for patients posted for Kasai porto-enterostomy needs to be explored systematically.

BASIC QUESTIONS:

1. What ideal time for kasai?
2. What intraop must for pediatric surgeon dealing with kasai e.g length of roux n y limb ?
3. Role of steroids post kasai postop?
4. Role of prophylactic antibiotics?
5. Definition consensus on failure of kasai.
6. WHY DIFFERENCES IN data FROM EAST AND WEST.

POST /Intra KASAI, REVIEW OF RECENT STUDIES.

STUDY 1: doi:10.1016/j.cgh.2011.07.024
CLINICAL GASTROENTEROLOGY AND HEPATOLOGY 2011; 9:1086–1091

SAMPLE : January 1, 1977, and December 31, 1988, 106 patients,
Confirmed BA: by IOC,H/P.
Thirteen patients had a type I or II BA; 9 patients underwent a hepaticojejunostomy or a choledochojejunostomy. In 4 patients, a portoenterostomy was performed. Eighty-three patients were classified as type III BA and underwent the classic Kasai portoenterostomy.
Results:
1. The 20-year survival rate was not significantly associated with the age at surgical correction.
2. more than 25% of BA patients in The Netherlands survive at least 20 years with their native liver.
3. Interestingly, one fifth of the 20-year transplant free survivors had no signs of cirrhosis and a normal liver biochemically.
4. The 20-year transplant-free survival rates in the 1977 to 1982 cohort presented in this report were similar to those reported from France in the same period (20% vs 23%, respectively). In the 1983 to 1988 cohort the transplant-free survival rate increased to 32%. However, survival remained lower than that reported from Japan (58%).
5. Surgery could not show a beneficial effect of surgery before 45 days,
6. Survival rates tend to be higher in patients with type I or II BA (42% in types I/II; 25% in type III). These figures are comparable with those from a large French cohort, but inferior to numbers from Japan, which reported a 100% 20-year transplant- free survival rate in 7 patients.
7. Signs of severe cholestasis and fibrosis in the per-operative liver biopsy specimen are not necessarily indicative of a poor prognosis.
8. The presence of BASM does not exclude a favorable outcome.

Not used:
1. Postoperative corticosteroids
2. Prophylactic antibiotics.

STUDY 2: J Korean Surg Soc. 2011 October; 81(4): 271–275.

doi: 10.4174/jkss.2011.81.4.271

SAMPLE: 32 patients with KPE performed for biliary atresia. Between 1990 and 2000.
Results:

1. 76.2% 10-year survival rates with native liver
2. one-third of survivals over 10 years had portal hypertensive events
3. 26.3% have cholangitis in 19 survivors over 10 years.
4. One-third of long-term survivals remain alive without any problem.

STUDY 3:
Lykavieris P, Chardot C, Sokhn M, Gauthier F, Valayer J, Bernard O. Outcome in adulthood of biliary atresia: a study of 63 patients who survived for over 20 years with their native liver. Hepatology 2005;41:366-71

SAMPLE: between 1968 and 1983
RESULTS:

1. The 20-year survival was better in those who had undergone porto-cholecystostomy (35%) or cysto-jejunostomy (40%) than in those with porto-enterostomy (19%),
2. In those who were operated on before 90 days of age (28%) than later (12%) SURVIVAL BETTER.
3. Of 51 patients alive IN 2005 with native liver, 96% developed cirrhosis.
4. overall results of the Kasai procedure was poor, with only 47 of 271 (18%) children being able to avoid LTx in the long term

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STUDY 4: *Japanese Biliary Atresia Society*
Nio M, Ohi R, Miyano T, et al. Five- and 10-year survival rates after surgery for biliary atresia: a report from the Japanese biliary atresia registry. J Pediatr Surg 2003;38:997–1000. doi:10.1016/S0022-3468(03)00178-7
SAMPLE size: One hundred sixty-four patients (11.9%) had type I atresia of the common bile duct, 34 (2.5%) had type II atresia of the hepatic ducts, and 1,162 (84.1%) had type III atresia at The porta hepatis.
Results:
1.Impact of the age at operation on bile flow was not clear until 90 days of age, and after 90 days the bile
flow rate worsened.
2. over-all 5-year survival rate changed from 69.4% to 78.3%, the difference was not statistically significant.
3. 10-year follow-up results of the 108 patients initially registered in 1989, 72 (66.7%) and 57 (52.8%) survived with and without the aid of LTx, respectively
4. only 19 patients lost to follow-up among the 743 patients registered from 1989 to 1994.
5. The incidence of cholangitis in complex modifications such as the Roux-en with an intestinal valve and the Suruga II was not lower than that in the original Rouxen- Y procedure.
no antireflux procedure – intra op could successfully prevent cholangitis

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STUDY 5: JPGN 2010;51: 631–634

SAMPLE: Kasai operations were performed on a total of 103 patients(45 boys and 58 girls). definitive diagnosis was confirmed with operative cholangiogram. Patients would receive prednisolone at initial dose at 4 mg/kg/day on day 7 after the operation for 2 weeks. The dose was reduced to 2 mg/kg/day for another 2 weeks and finally 1 mg/kg/day for the last 2 weeks

Results:

1. The overall success rate of Kasai operation (as defined by the need for transplantation within 1 year) was 59.3%

2. the results of the Kasai operation are still variable across different centers.

3. The paradigm of early surgery may not apply to all patients experiencing isolated BA.

4. There was no difference histologically between patients who had surgery before 60 days and those between 61 and 80 days, with little or no progression to cirrhosis until at least beyond 100 days of life. This would suggest that although there were higher levels of bilirubin in patients who had the Kasai operation for 60 days of life, the overall effects on the liver were not as significant as once thought. Furthermore, because the age of onset of isolated BA is variable, and the success of the surgery determined mainly by the degree of damage of biliary ductules and the degree of fibrosis, it would not be logical to use the age at operation alone to predict the outcome.

5. optimal age for performing the Kasai operation should be between 61 and 80 days: well developed surgical portal path.

STUDY 6: Chirurg. 2009 Jul;80(7):628-33.

SAMPLE: 85 BA patients from 1993 to 2003 was aimed to evaluate prognostic factors using the log rank test

RESULTS:

1. 20% survive in the long term with their native livers.
2. Age at Kasai operation (P=0.46), degree of liver fibrosis (P=0.95), and all laboratory test results before Kasai failed to correlate with outcome
3. Normal levels of bilirubin 3, 6, and 12 months after Kasai and of aspartate aminotransferase with gammaGT after 6 months are associated with survival with native liver.
4. lack of predictive factors must prevent primary liver transplantation in BA

STUDY 7 : J Pediatr Gastroenterol Nutr. 2009 Apr;48(4):443-50.

Long-term outcome of children with biliary atresia who were not transplanted after the Kasai operation: >20-year experience at a children's hospital.

Sample:Group of 80 patients who had undergone theKasai operation between 1970 and 1986 .

RESULTS:

1. 5-, 10-, and 20-year survival rates of patients with their native livers were 63%, 54%, and 44%, respectively.
2. By age 20, nearly half of the adult survivors had already developed liver cirrhosis and its sequelae
3. Five female patients gave birth to a total of 9 children, and 1 male patient fathered a child

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STUDY 8 : British paediatric surveillance unit study of biliary atresia: outcome at 13 years.
J Pediatr Gastroenterol Nutr. 2009 Jan;48(1):78-81.

SAMPLE: 93 cases of biliary atresia in the United Kingdom and Ireland diagnosed between March 1993 and February 1995 were followed up prospectively.

RESULTS:

1. When the portoenterostomy was successful, 40 of 50 patients (80%) are alive without liver transplantation.
2. The 13-year actuarial survival without liver transplantation is 43.8% overall and is better in children treated at centres that treat more than 5 cases yearly
3. 45% underwent liver transplantation at a median age of 1 year (range 0.5-9), with 90% survival.

STUDY 9 : Long-term prognosis of patients with biliary atresia: a 25 year summary
J Pediatr Gastroenterol Nutr. 2006 Feb;42(2):190-5.

SAMPLE: 185 children were diagnosed with BA, 22 underwent exploratory laparotomy withoutKasai operation, and 163 underwent Kasai operation, of which 141 cases had long-term follow-up

RESULTS:

1. Five and 10 year survival rates with native liver were 35% and 31%, respectively
2. Liver transplantation was performed in 19 patients (all but 2 with a living-related donor), and 15 (79%) survived

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STUDY 10: A long-term experience with biliary atresia. Reassessment of prognostic factors. Ann Surg. 1991 Nov;214(5):590-8.

RESULTS: 1.Survival was not influenced by earlier age at operation, size of ductal remnants, or the use of an external biliary vent.

2. Good bile flow was predictive of a favorable outcome.

STUDY 11: Perioperative factors affecting the outcome following repair of biliary atresia Pediatrics. 1989 May;83(5):723-6.

RESULTS:

1. When performed earlier than 61 days after birth, surgery resulted in adequate bile flow in 64.7%
2. CHolangitis developed in three of the five (60%) with a Roux-en-Y limb length less than 40 cm and in two of the 19 (10.5%) with limb lengths greater than 40 cm (P less than .02).
3. Subsequent long-term antibiotic prophylaxis favors the best bile flow and reduces the occurrence of cholangitis, resulting in the best outcome

Long term data FROM ABOVE STUDIES:

	Period	Number of patients with surgical correction	Transplant-free survival at 20 y	OLT before the age of 20 y	Overall survival at 20 y
France (Lykavieris et al[6])	1968–1983	271	23%	32 (12%)	—
Japan (Nio et al,[19] 2003)	1970–1986	80	44%	5 (6%)	49%
Sendai, Japan (Nio et al,[27] 1996)	1953–1993	289	22 (8%)	—	—
The Netherlands (present study)	1977–1982	49	10 (20%)	8 (16%)	17 (35%)
	1983–1988	55	18 (32%)	16 (29%)	28 (51%)
	1977–1988	104	28 (27%)	24 (23%)	45 (43%)