Clinical profile and outcome of biliary atresia – a single centre appraisal from a tertiary care teaching hospital under public sector in Kerala

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Abstract

Background:

Biliary atresia is a rare *birth defect*, with prevalence of *1 in 8,000 – 18,000* live births and accounts for most of the pediatric liver transplantation (LT) globally. Timely work-up of neonatal cholestasis (NC) is of key importance to performing Kasai portoenterostomy (KPE) *before* 60 days of life, should biliary atresia be the diagnosis. For those who do not have successful drainage after operation, LT remains the only option. Biliary atresia meets the *disease-specific criteria* for *newborn screening*. Universal screening, using *stool colour cards* may improve age at diagnosis and treatment, leading to *improved outcomes*. The recently published *integrated neonatal cholestasis (NC) card* from an Indian study proposes to recognize NC at an early stage irrespective of aetiology. It is a major step towards *public health benefit* both at the *community* as well as *physicians' levels* to enable *early detection and timely referral and management*. There is no published data from Kerala on biliary atresia and its outcome.

Objectives:

To study the *clinical profile* and *outcome* of biliary atresia from a tertiary care teaching hospital under public sector in Kerala.

Methodology:

The prospectively collated clinical registry of children (<12 years) diagnosed with chronic liver disease (CLD) {based on clinical, biochemical [liver function test (LFT) and ultrasound abdomen +/- liver biopsy +/- transient elastography +/- genetic testing +/- upper gastrointestinal (GI) endoscopy} and the registry of children (<12 years) who underwent *liver transplantation* maintained in the Pediatric Gastroenterology (PGE) services, Dept. of Pediatrics, SAT Hospital, Govt. Medical College, Thiruvananthapuram from 12th February ,2014 to 30th November,2021 were retrieved and the data of children diagnosed with *biliary atresia* were extracted and analyzed. The protocol included a clinical examination, examination of the stool colour, and liver function tests to confirm the presence of conjugated hyperbilirubinemia.A toxoplasmosis, rubella virus, cytomegalovirus, herpes virus (TORCH) serology was done in most cases and hepatobiliary iminodiacetic acid scan

(HIDA) scan was carried out after priming with ursodeoxy cholic acid/phenobarbitone.Fasting ultrasound abdomen was routinely done to look for a distended gall bladder, the anatomy of the biliary tree, the liver echotexture, and exclude the presence of choledochal cyst. Patients in whom biliary atresia could not be excluded pre-operatively underwent percutaneous liver biopsy. The clinical presentation of children who underwent KPE and not underwent KPE and their survival outcome with *native liver* and *post liver transplantation* during the study period was analyzed.

Results:
During this study period *266* children were diagnosed to have CLD and out of this, 66 children were diagnosed to have biliary atresia. This constituted around *25%* of CLD diagnosed in children and the most common aetiology for CLD in less than 6 years of age among the study population. In our study, there were 30 males and 36 female (M: F- 1:1.2) which is in concordance with available descriptive epidemiology. *Jaundice* and *pale coloured stools* were present in all cases and the mean age at onset of jaundice was 8 days. Mean age at presentation was 1.4 months +/-0.26 months. 69% children had severe malnutrition. Out of this 66, 54 (82%) *children underwent KPE* and *the mean age at KPE was 69 days*. The delay in surgery from presentation may be attributed to delay in recognition of NC, referral, expeditious workup and delay in intervention.12 children were not offered KPE due to delayed presentation and refusal of parental consent.17 underwent liver transplantation (LT) [12 children – post KPE and 05 children – primary] which constituted the single most common indication (n= 32/53%) for LT facilitated through our PGE services.18 children were lost to follow-up after diagnosis including 16 cases post KPE.Out of the remaining 48 children who could be followed up during the study period, 32 children are alive [survival with native liver and post LT 50% each]. 33% mortality (n=16) was noted among the children who could be followed up during the study period [post KPE (n= 15) and post KPE LT (n= 01).The eldest survivor with native liver after Kasai's surgery was 12 years old upon follow up.

Conclusions:
This study highlights the *burden of biliary atresia* as the *single most common etiology* of CLD in children less than 6 years of age and the *most common indication of liver transplantation* in children from our cohort. As early diagnosis is the prime factor for survival of children with BA as revealed from this single centre study, to ensure early referral, there is an urgent need to *sensitize* pediatricians, obstetricians, other primary-care physicians and ASHA on the need for early evaluation for the cause of NC. For this it is suggested to incorporate the recently proposed *integrated neonatal cholestasis (NC) card* in the *well-baby cards* issued by Government for *newborn screening of NC.*

Key words: *Neonatal cholestasis; biliary atresia ; screening*

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