

[Symposium Lecture]

Screening of Biliary Atresia for Early Management

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Biliary atresia (BA) is a progressive cholangiopathy starting from fetal or neonatal period, which may lead to rapid liver cirrhosis and liver failure. BA is the most common cause of liver related death and transplantation in children. The incidences of BA in Taiwan (1.5-2.0/ 10,000 live birth) and French Polynesia (3.2 /10,000) were higher than other part of the world, such as Japan, Australia,, Europe or U.S.A.

Hepatic porto-enterostomy (Kasai operation) should be conducted as early as possible to improve the long term outcome of infants with BA. Prolonged neonatal jaundice can be caused by BA or non BA causes (such as unconjugated hyperbilirubinemia or intrahepatic cholestasis). Therefore BA neonates are easily overlooked, leading to delayed diagnosis and treatment. This phenomenon occurred often because breast milk jaundice is much more frequently found than BA. Screening for BA in infants is mandatory to enhance earlier diagnosis and intervention, which could save children's life.

There are several screening methods for BA, such as blood conjugated bilirubin, bile acids, urine sulfated bile acid, fecal conjugated bile acids, and stool color card. The concept of screening newborns for BA using a stool color card was initiated in Japan by Professor Akira Matsui in the early 1990s. With high incidence of BA, Taiwan established the world first universal stool color card screening program for BA since 2004. The rate of Kasai operation <60 days of age increased from 47% in the era before stool color card screening program to 60-74% after the program. The 5-year jaundice free native liver survival rate also increased from 27% before the program to 64% after the program. With the improvement of the rate of earlier Kasai operation and survival rate, still there are needs of further efforts to improve the rate of earlier Kasai operation and outcome of biliary atresia. We investigated the causes of delayed Kasai operation and found that continuous education of the caretakers and medical personnel are both very important.

In conclusion, screening to early detect BA is needed for early management and better outcome. Stool color card screening program is a simple, safe, low cost, non-invasive, applicable and effective screening method for BA.