CLINICAL PROFILE AND OUTCOME OF PATIENTS DIAGNOSED WITH BILIARY ATRESIA: A 20 YEAR EXPERIENCE IN PHILIPPINE CHILDREN'S MEDICAL CENTER

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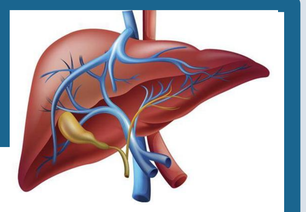

BACKGROUND

Biliary atresia is a progressive cholangiopathy that leads to death without timely intervention. It commonly presents as *jaundice beyond 2 weeks* of life with findings of cholestasis commonly ignored and misdiagnosed. Determining the clinical profile and outcome can help identify areas to improve management in the local setting.

OBJECTIVES

To determine the clinical profile and outcome of patients diagnosed with biliary atresia in Philippine Children's Medical Center in the past 20 years.

METHODOLOGY

A *descriptive cross sectional study* was done on children 0 to 18 yrs old *diagnosed* with Biliary atresia *through liver biopsy and/or biliary exploration with intraoperative cholangiogram*. Clinical profile, diagnostic, and therapeutic data were gathered.

RESULTS

Review of 330 charts identified **122 cases** in the **past 20 years**.

- *Male* patients presenting with *jaundice within the first 3 weeks of life and acholic stools* were predominant
- Majority had normal nutritional status (n=66, 81.5%) with hepatomegaly (n=115, 99.1%) and umbilical hernia (n=20, 17.4%)
- *Consultation and diagnosis were delayed* at a median of 6.9 weeks and 17.2 weeks respectively
- Baseline laboratories showed direct hyperbilirubinemia of 8.4 mg/dl (±2.94) & elevations of ALT, AST, alkaline phosphatase and GGTP
- On *ultrasonography,* none had triangular cord sign but had *non-visualized or abnormal gallbladder findings (45.9%)*
- *Liver biopsy* showed *majority with microscopic cirrhosis* at the time of diagnosis (75.5%)
- Eighty-one patients (68.6%) underwent Kasai portoenterostomy (KPE) with only 6 patients documented to have liver transplantation. Comparison showed *increased admissions for ascending cholangitis in those who underwent KPE* versus *those who did not that had more admissions for gastrointestinal bleeding and pneumonia.*

CONCLUSION & RECOMMENDATIONS

The study showed early onset of jaundice and symptoms but significant delay in diagnosis and management. Emphasis to pediatricians in recognizing cholestasis for early referral and evaluation should be done.

✓ A registry to document data and prospective studies on patients with biliary atresia can help improve outcomes by changing management strategies and improving treatment.

KEYWORDS jaundice, cholestasis, biliary atresia, Kasai portoenterostomy