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# Incidence and outcome of biliary atresia in Shanghai, China from 2015 to 2016: a cohort study

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## ABSTRACT

Demographic characteristics and clinical data of all newly diagnosed biliary atresia patients in Shanghai were collected from 1 January 2015 to 31 October 2016. The total number of live births was 377 420 during the study period, and the incidence of biliary atresia in Shanghai was 10.86 per 100 000 (95% CI 7.8 to 17.74), with 62.9% and 45.7% cases retaining native liver survival at 2 and 5 years after Kasai procedure, respectively. Implementation of systematic screening measures for biliary atresia in China is needed.

Biliary atresia (BA) is one of the most serious neonatal cholestasis and the most common primary disease of liver transplantation in children.[1] The reported BA incidence varies widely from approximately 1:5000–1:20 000 newborns among geographical regions.[2] Given the lack of a newborn disease tracking and reporting system, there are no accurate incidence data in Chinese mainland. This study aims to evaluate the incidence and prognosis of BA in the Chinese population.

This study was applied to the Shanghai Center for Women and Children's Health to determine the total number of live births from 1 January 2015 to 31 October 2016. All newly diagnosed BA cases by intraoperative cholangiogram and pathological examination were registered at one of the four paediatric hospitals that are qualified to diagnose and treat BA in Shanghai (figure 1). The population cumulative incidence of BA was calculated by dividing newly diagnosed BA patients by the total

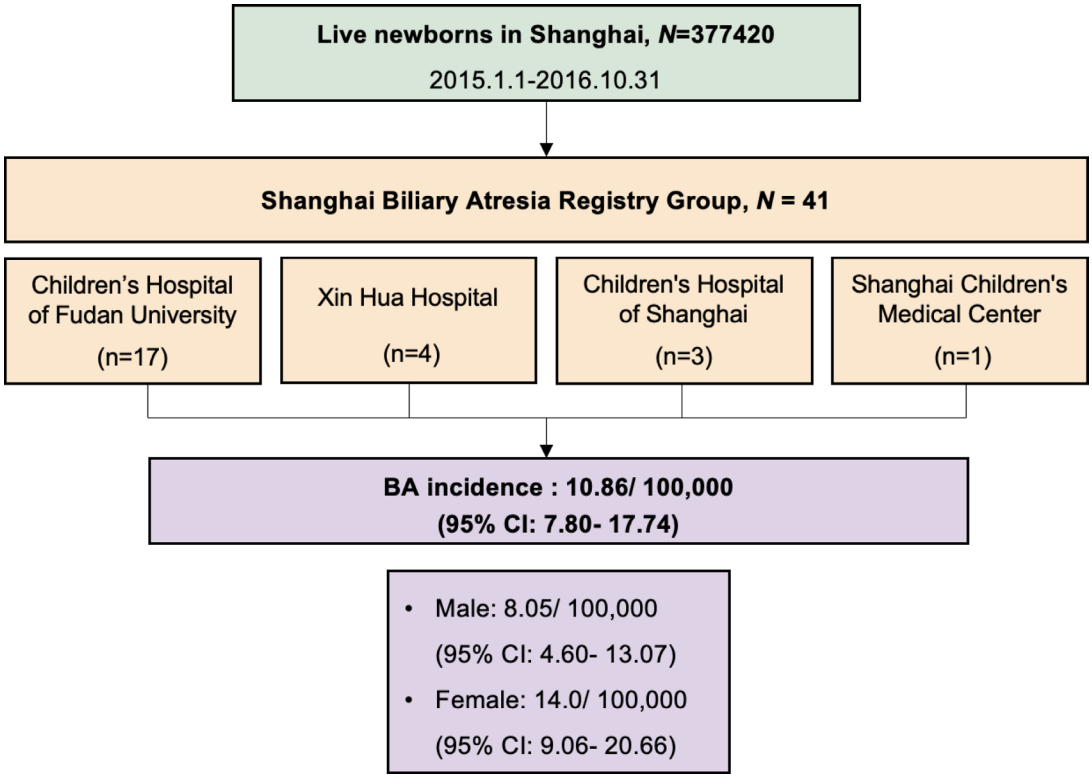


**Figure 1** Patient flow for incidence study of biliary atresia.



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Table 1 Characteristics and outcomes of BA infants during the study period

| | BA (n=41) |
|---|---|
| **Sex** | |
| Male | 16 |
| Female | 25 |
| **Gestational age** | |
| Term | 38 |
| Late preterm | 3 |
| Prenatal-detected abnormality | |
| Cystic | 1 |
| Gallbladder atrophy or absence | 0 |
| BA Type | |
| Cystic | 2 |
| Isolated | 39 |
| Associated malformations | |
| Intestinal malrotation | 1 |
| Age at first visit (median, day) | 56 (38, 68) |
| Received Kasai operation | 35 (85.4%) |
| Age at Kasai operation (median, day) | 84 (49, 89) |
| ≤60 | 11 (31.4%) |
| 60–90 | 16 (45.7%) |
| ≥91 | 8 (22.9%) |
| Outcomes | |
| Jaundice-free 3 month after Kasai* | 24 (68.6%) |
| Native liver survival 2 years after Kasai | 22 (62.9%) |
| Native liver survival 5 years after Kasai | 16 (45.7%) |

*Serum total bilirubin <34 µmol/L (2.0 mg/dL).
BA, biliary atresia.

number of live births. The 95% CI of BA incidence was calculated using the Poisson distribution.

During the nearly 2-year study period, the total number of live births was 377 420 in Shanghai, and 41 cases were diagnosed as BA, yielding an incidence of 10.86 per 100 000 (95% CI 7.8 to 17.74, figure 1). The ratio of male to female BA cases was 1:1.56 (16/25), showing non-statistical gender difference in the BA incidence (8.05 per 100 000 vs 14.0 per 100 000, p=0.08). Among the BA infants, three cases (7.3%) were premature, and one BA case with an extrahepatic biliary cyst was detected by prenatal ultrasound. Two cases were diagnosed as cystic BA, and the remaining 39 cases were all isolated BA, with 1 case having intestinal malrotation (table 1).

The first outpatient visit age of 41 BA patients was 56 (IQR: 38–68) days, and the 35 BA cases underwent the Kasai procedure at the median age of 84 (IQR: 49–89) days. Among these, 11 cases (31.4%) received surgery before 60 days of age, and 8 infants (22.9%) were beyond 90 days of age. 24 (68.6%) BA infants became jaundice-free at 3 months after the Kasai procedure. However, 11 BA cases eventually had to undergo liver transplantation due to persistent jaundice or cirrhosis. After 2 years and 5 years following up, 62.9% and 45.7% of BA patients retained native liver survival, respectively (table 1).

The incidence of BA in Chinese mainland was reported earlier as 1.3 per 10 000 live births in 29 799 live births in Beijing.[3] We reported an incidence of 1.086 per 10 000 live births based on a population of more than 300 000 live births in Shanghai, making it a more accurate estimate for such a rare disease, which is comparable to the reports in Japan and Taiwan.[4 5] The chance of underestimating the incidence is small since the possibility is extremely low for local children to seek medical treatment elsewhere. In addition, parents have a relatively strong awareness of seeking medical diagnosis and treatment, which greatly reduces the probability of giving up treatment or dying before diagnosis in Shanghai. However, this incidence may not represent that in other parts of the country because of the huge, diverse and geographical populations of the country.

Existing evidence suggests statistical benefit for Kasai before 60 days of age,[1 6] but only 11 BA infants (31%) achieved it between 2015 and 2016 due to the lack of awareness and recognition of BA by primary medical staff and parents. This directly led to 6 out of 41 BA patients did not undergo the Kasai procedure due to older age at the time of diagnosis. However, with the implementation of early detection measures, such as stool colour card screening and serum or heel blood MMP-7 precise diagnostic testing, the early diagnosis proportion had increased to nearly 60%, and the mean age of Kasai procedure had decreased to an average of 56 days in 2022 in our single centre.

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