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Abstract

Demographic characteristics and clinical data of all newly diagnosed BA patients in Shanghai were collected from January 1, 2015 to October 31, 2016. The total number of live births was 377 420 during the study period, and the incidence of biliary atresia in the Shanghai was 10.86 per 100 000 (95% CI: 7.8-17.74), with 62.9% and 45.7% cases retaining native liver survival at two and five years after Kasai procedure, respectively. Implementation of systematic screening measures for biliary atresia in China are 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 vary widely from approximately 1:5000 to 1:20,000 newborns among geographic regions [2]. Given the lack of a newborn disease tracking and reporting system, there is no accurate incidence data in Chinese mainland. Following the biliary atresia screening project carried out from 2015 to 2016 in Shanghai, this study is allowed to evaluate the incidence and long-term prognosis of BA in the Chinese population.

This study applied to the Shanghai Center for Women and Children's' Health for the total number of live births from January 1, 2015 to October 31, 2016. All newly diagnosed BA cases by intraoperative cholangiogram and pathological examination were registered at one of the four pediatric hospitals which are qualified for diagnosing and treating BA in Shanghai (Figure 1). This study was reviewed and approved by the Ethics Committee of Children's Hospital of Fudan University. The population cumulative incidence of BA was calculated by dividing newly diagnosed BA patients by the total number of live births. The 95% confidence interval (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 was diagnosed as BA, yielding an incidence of 10.86 per 100 000 (95% CI: 7.8-17.74, 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, 3 cases (7.3%) were premature, and one BA case with 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 one case having intestinal malrotation (Table

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Thirty-five BA cases underwent Kasai procedure at median age of 84 days (IQR: 49–89) days. Among which 11 cases (31.4%) received surgery before 60 days of age, and 8 infants (22.9%) were beyond 90 days of age. Twenty-four (68.6%) BA infants became jaundice-free at three months after the Kasai procedure. However, 11 BA cases eventually had to undergo liver transplantation due to persistent jaundice or cirrhosis. After two years and five years following up, 62.9% and 45.7% 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 more accurate estimate for such a rare disease, which is comparable to the reports in Japan and Taiwan [4,5]. The chance of underestimate in 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 part of the country because of the huge, diverse and geographic populations over the country.

Existing evidences suggest statistical benefit for Kasai before 60 days of age [1,6], but only 31% BA infants achieved it between 2015 and 2016. However, with the implementation of early detection measures, such as stool color card screening and serum or heel blood MMP-7 precise diagnostic testing, the 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 center.

References

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Associated malformations		
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Ages of Kasai Operation (median, day)		84 (49, 89)
	≤60	11 (31.4%)
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Outcomes		
	Jaundice-free 3 month after Kasai ^	24 (68.6%)
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Figure legends

Figure 1. Patient flow for screening study of biliary atresia

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The authors have no relevant financial or non-financial interests to disclose.

Author contributions

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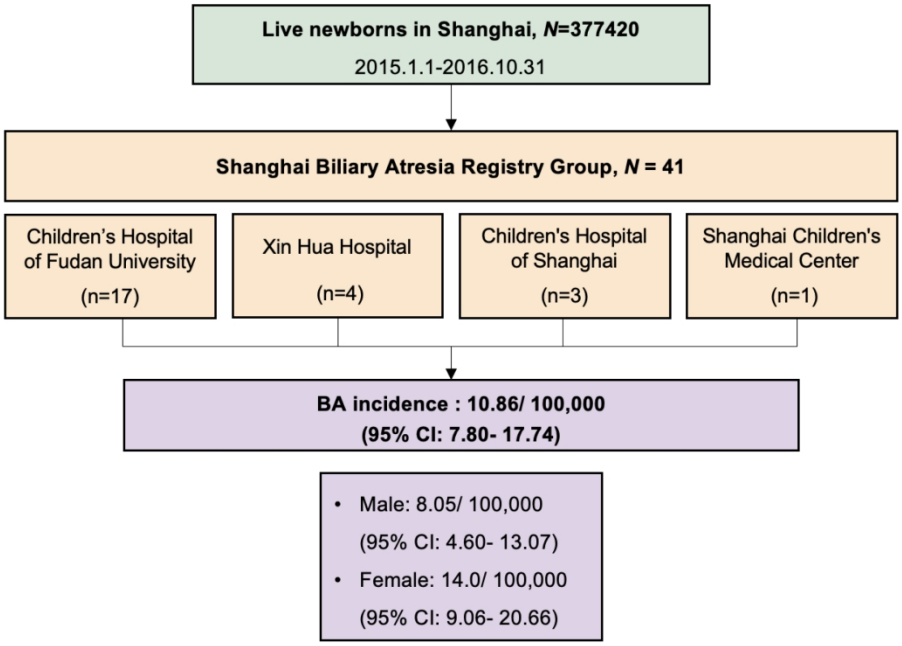
Not applicable.



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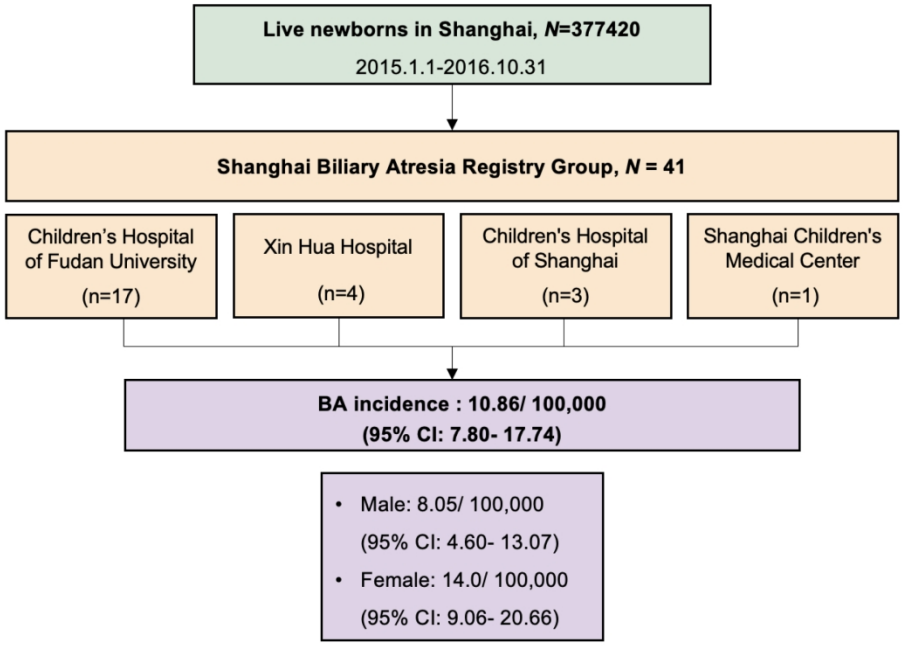

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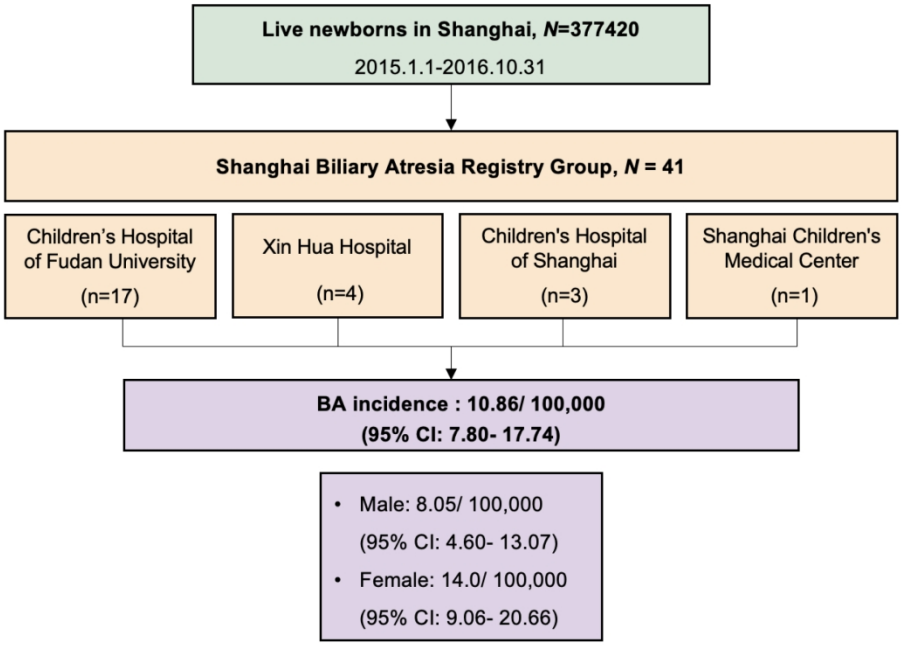

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