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ORIGINAL ARTICLE

Incidence of biliary atresia associated congenital malformations: A retrospective multicenter study in China

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Summary *Background:* Some patients with biliary atresia (BA) have associated anomalies. Our study aimed to investigate the incidence of BA-associated malformations in mainland China, and compare the results with those reported in the Western literature.
Methods: Clinical data were collected retrospectively from five medical centers in mainland China. BA patients were diagnosed and confirmed by laparotomy with intraoperative cholangiography and liver biopsy. Cases were divided into isolated type BA and BA with associated anomalies, including polysplenia, situs inversus, intestinal malrotation, and cardiovascular anomalies.
Results: A total of 851 BA patients were recruited from Tianjin, Beijing, Wuhan, Guangzhou, and Shenzhen. Patients were grouped as follows: Type I, 13 cases (1.5%); Type II, five cases (0.6%); Type III, 833 cases (97.9%). Forty-two (4.94%) patients had 54 associated congenital abnormalities. The intra-abdominal anomalies included polysplenia ($n = 4$, 1 fusion between liver and spleen), situs inversus ($n = 2$), and intestinal malrotation ($n = 3$). The cardiovascular anomalies included atrial septal defect and ventricular septal defect ($n = 29$), patent foramen ovale ($n = 1$), patent ductus arteriosus ($n = 4$), and other cardiac malformations ($n = 3$, including coronary sinus dilation, left superior vena cava, Tetralogy of Fallot).

Conflict of interests: The authors declare that they have no conflicts of interests.

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Conclusion: Our data showed that spleen anomaly is not as common as reported in the Western literature. The difference may suggests different genetic and environmental risk factors for BA.

1. Introduction

Biliary atresia (BA) is one of the most common causes of obstructive jaundice in infancy, characterized by progressive biliary fibrosis, the cause of which is unclear.[1] Most authors believe that the clinical phenotype of BA is associated with genetic heterogeneity. Chromosomal changes have been discovered in some BA patients, and there are reports suggesting the role of genetic predisposition in its pathogenesis of BA.[2] Since Helwig[3] first reported a BA associated with polysplenia in 1929, other associated congenital malformations with BA were also added including situs inversus, intestinal malrotation, cardiopulmonary dysplasia, and other anomalies. The incidence of BA-associated congenital malformations has been reported to range from 3% to 20%.[4–6] Davenport et al[6] divided BA patients into two categories—acquired/nonsyndromic BA (90%) and embryonic/syndromic BA (10%)—and pointed out that BA patients with spleen malformations may have onset of the pathological process during the embryological phase of organ development.[6] In this study, our aim was to investigate the incidence of BA with associated malformations in the Chinese population and compare our results with those in the Western literature.

2. Patients and methods

The clinical data of BA patients were collected retrospectively from five medical centers in China between October 2009 and September 2014, including Tianjin Children's Hospital (Tianjin), Beijing Children's Hospital (Beijing), Wuhan Tongji Hospital (Wuhan), First Affiliated Hospital of Sun Yat-sen University (Guangzhou), and Shenzhen Children's Hospital (Shenzen). BA patients were diagnosed and confirmed by laparotomy with intraoperative cholangiography and liver biopsy. The preoperative examinations including electrocardiograms, chest X-ray, and abdominal ultrasound were performed routinely after admission; patients with abnormal results would be further examined by echocardiography or computed tomography (CT), and magnetic resonance imaging (MRI) to determine if there is any abnormality. Congenital malformations were assessed with abdominal ultrasound, echocardiography, CT, MRI, etc. All five medical centers used the same form of questionnaire for data collection (e.g., sex, age at surgery, surgical approach, and malformation type). Cases without complete information were excluded. The BA patients were divided into isolated type BA and BA with associated malformations. The latter group includes patients with one or more of the following anomalies: spleen anomalies, laterality anomalies, cardiac anomalies, intra-abdominal vascular anomalies, pancreatic anomalies, and malrotation, etc. This study was approved by the local ethics committee of each center.

Descriptive data were summarized. The age at the time of Kasai operation between isolated BA and BA with associated malformations was compared using t test, and a p value < 0.05 was considered statistically significant.

3. Results

In this study, a total of 851 patients with BA were recruited. The majority of patients had isolated BA (754, 89%). There were 42 BA patients with associated malformations. According to the classification criteria of BA by the Japanese Association of Pediatric Surgery, the patients in our group can also be classified based on the most proximal level of obstruction as follows. Type I refers to atresia of the distal bile duct with a patent common hepatic duct, gall bladder, and cystic duct, with or without cyst (13/851; 1.5%). In Type II, atresia of the common hepatic duct is at different levels; in some cases, the common bile duct, cystic duct, and gall bladder are patent but the common hepatic duct is atretic (5/851; 0.6%). Type III, the most common type, occurs in almost 90% of cases; here, the entire extrahepatic biliary system including the common hepatic duct, gallbladder and the common bile duct are atretic (833/851; 97.9%).[7]

There were 446 boys and 405 girls in our study. Table 1 illustrates the different BA types and the male/female ratio in the five medical centers. The male/female ratio is 1.09/1. The sex distribution of patients with malformations is shown in Table 2. The patients with congenital associated malformations were well distributed in Types I, II, and III. Patients with associated malformations had an extraordinarily high percentage of Type III BA, with malformations more frequently being presented in boys.

Forty-two (4.94%) patients with 54 associated congenital abnormalities were reported. The intra-abdominal anomalies included polysplenia ($n = 4$; 1 case is abnormal fusion between liver and spleen), situs inversus ($n = 2$), and intestinal malrotation ($n = 3$). Cardiovascular anomalies included atrial septal defect (ASD) and ventricular septal defect (VSD; $n = 29$), patent foramen ovale ($n = 1$), patent ductus arteriosus ($n = 4$), and other cardiac malformations ($n = 3$; including coronary sinus dilation, left superior vena cava, Tetralogy of Fallot). The list also included intestinal atresia and anorectal malformation, as shown in Table 3.

Among eight patients with two or more malformations, one boy had splenic and cardiac abnormality; one boy had intestinal atresia and patent foramen ovale; one boy had intestinal atresia, patent foramen ovale, patent ductus arteriosus, and duodenal diaphragm; one boy had ASD and pulmonary stenosis; one boy had ASD, patent ductus arteriosus, coronary sinus dilation, and left superior vena cava; and three girls had ASD and patent ductus arteriosus.