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Physical development and cognitive performance in a monozygotic twins for biliary atresia: Report of a case and literature reviewing

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ABSTRACT

A case of monozygotic twins, one of whom suffered from biliary atresia and the other one was apparently healthy, is described herein. Children with biliary atresia appear to be vulnerable to developmental lags in the physical and cognitive performance, due to malnutrition and deteriorated liver function. In our case where the child with biliary atresia, who underwent a successful kasai operation, has achieved comparable growth development and cognitive performance in comparision with her sister, which suggest that the early diagnosis and timly kasai operation could allow children with biliary artesia to grow well in the short-term following time.

1. Case report

Biliary atresia (BA) is an idiopathic neonatal cholestatic disease characterized by the progressive inflammation and fibrosis of the biliary tree, involving the occlusion of the extra- and intrahepatic bile ducts. The incidence of BA in twins is fairly lower compared with the total prevelance of BA. Discordant presentations of biliary atresia in monozygotic twins were shown in our report.

The mother was 27 years old (gravida 1, para 2). Her past medical history included only hepatitis B. She didn't take any medicine during the pregnancy other than oral multivitamins. There was no history of surgery operation, trauma, blood transfusion, diabetes or hypertention during gestation. Written informed consent was obtained from both parents.

Following spontaneous labor on 22th, October 2014, at 37^{+3} weeks by dates, she underwent a cesarean section and delivered a couple of monozygotic twin sisters at the Central Maternity Hospital of Tianjin.

Twin 1: The first girl weighed 2350 g after birth, and the Apgar score was 10/10. She became icteric after birth and did not achieve free-jaundice so that was referred to the Children's Hospital of Tianjin at 46 days of life, during this period, she presented persistent jaundice, pale stools and dark urine.

Examinations after being admitted to hospital showed moderate jaundice with a total bilirubin of 8.61 mg/dL (normal, 0.3–1.1 mg/dL) and a direct bilirubin of 6.93 mg/dL (normal, 0.1–0.4 mg/dL). Transaminases were moderate elevated: aspartate aminotransferase (AST) 137U/L (normal, 8–40 U/L), alanine aminotransferase (ALT) 43U/L (normal, 0–40 U/L), and γ-Glutamy transpeptidase (γ-GT) 967U/L (normal, 7–50 U/L). An abdominal ultrasound showed an enlarged liver without a visible gallbladder or bile duct but a cystic structure. A percutaneous liver biopsy was not done for its difficulty to control.

Serology examination for toxoplasmosis, rubella, cytomegalovirus, herpes (TORCH) presented negative except EBNA-IgG positive, blood examination showed CMV-DNA$<10^3$ copies/mL, TOX-DNA$<10^3$ copies/mL, CMVpp65(+), the mother's milk test discovered that CMV-DNA 8×10^3 copies/mL, which suggested the baby maybe infected by EB virus. The monozygotic twin's chromosome G band karyotype analysis presented normal. No other abnormality was found.

Exploratory laparotomy at 49 days of life showed hepatomegaly, a small gallbladder and the thin, tortuous biliary structure during intraoperative cholangiogram. The intraoperative liver biopsy showed bile ducts proliferation, cholestasis, widened portal tracts, portal fibrosis and bile plugs, which were consistent with extrahepatic biliary artesia. So the kasai portoenterostomy was operated

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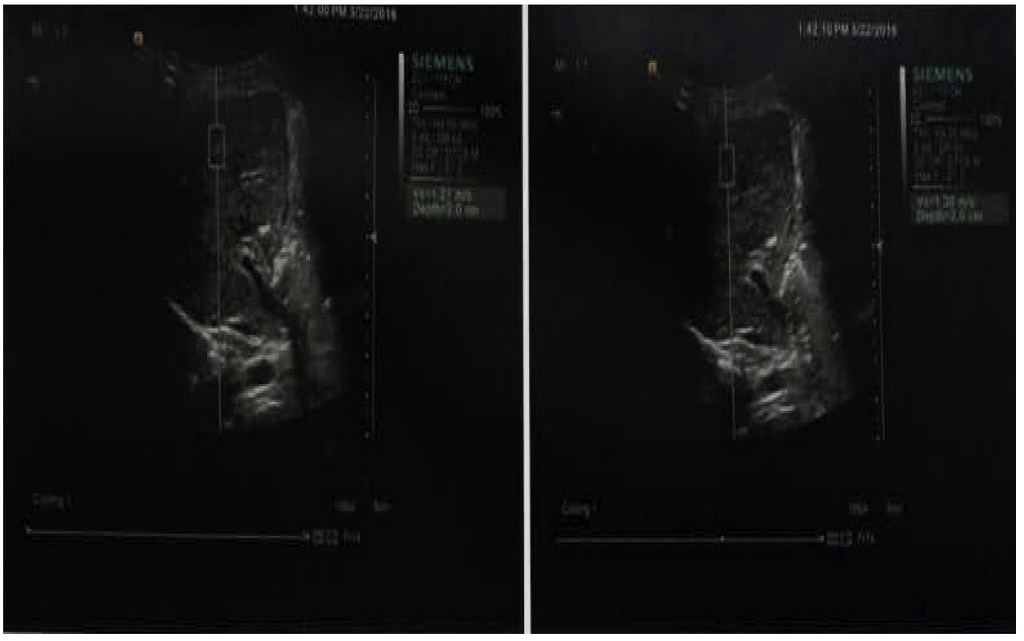
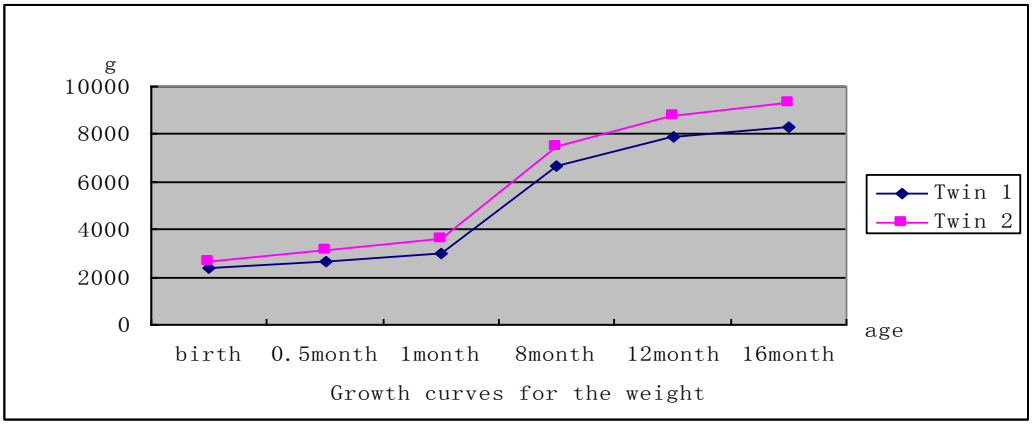


Fig. 1. Color Doppler ultrasound.



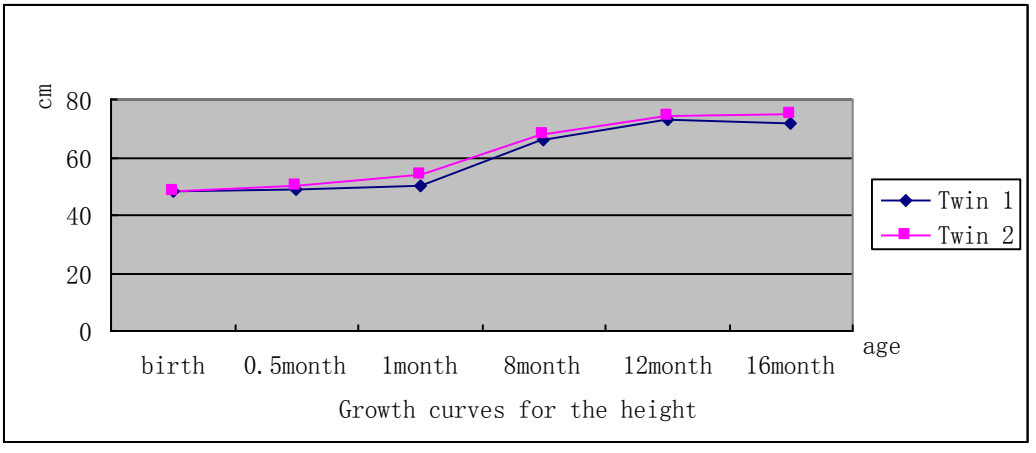


Fig. 2. Growth curves for the weight and height.