# Timing of operation in children with a prenatal diagnosis of choledochal cyst: A single-center prospective study

Xisi Guan

Department of Neonatal Surgery, Guangzhou Women and Children's Medical Center, Guangzhou, Guangdong, China 2022-10-23



# 1, Background

A choledochal cyst (CDC) is a congenital anomaly of the biliary system, which is more common in the Asian population [1]. If a CDC is not diagnosed and treated promptly, it often leads to a series of serious complications, including cholangitis, cyst rupture, cholestatic cirrhosis, and even cholangiocarcinoma. Infants with a postnatal diagnosis of CDC often present with symptoms, and to avoid the occurrence of serious complications, operative correction should be performed as soon as possible when their clinical conditions allow. However, in the current era with the improvement of prenatal screening technology, an increasing number of choledochal cysts are diagnosed prenatally in the fetus. In developed countries, as many as 15% of choledochal cysts are found before birth [2-5]. Some of these children receive intervention when they are asymptomatic at an early stage, while some have progressed to CDC-related symptoms before operative correction. The timing of operation for children with a prenatal diagnosis of CDC remains controversial. Redka et al. believed that infants with an antenatal diagnosis of CDC needed to be followed closely with ultrasonography and biochemical liver function tests and undergo elective reconstructive surgery at 3 months of age [6]. In contrast, Diao et al. performed a prospective cohort study on asymptomatic children with a prenatal diagnosis of CDC and divided the children into an early ( $\leq 1$ month) operative group and a late (> 1 month) operative group. They found that the extent of liver fibrosis was greater in the late operative group, and the postoperative recovery was statistically significantly delayed in patients with delayed surgery [7]. Therefore, the authors recommended that operative correction should be performed in the neonatal period. In addition, Japanese clinical guidelines for pancreaticobiliary malformations suggest that symptomatic neonates and infants should be operated on as soon as possible. Elective operations should be considered for asymptomatic cases at approximately 3–6 months of age along with postnatal monitoring of organ functions, primarily of the liver, to determine if laboratory symptoms develop to warrant earlier intervention [8]. Our previous study [9] showed that it is more advantageous to receive surgical treatment in the asymptomatic period for patients with prenatally diagnosed CDC. In addition, the age at operation (months) appears to be unrelated to intraoperative and postoperative complications, which is distinct from previous studies. More interestingly, we found that a specific cyst size (length > 5.2 cm and width > 4.1 cm)



Guangzhou Women and Children's Medical Center

suggested that clinical symptoms might appear and that the surgery should be performed as soon as clinically safe to proceed.

# 2. Objective

We tried to select the operation time according to the cyst size and evaluate the treatment effect.

# 3. Study design

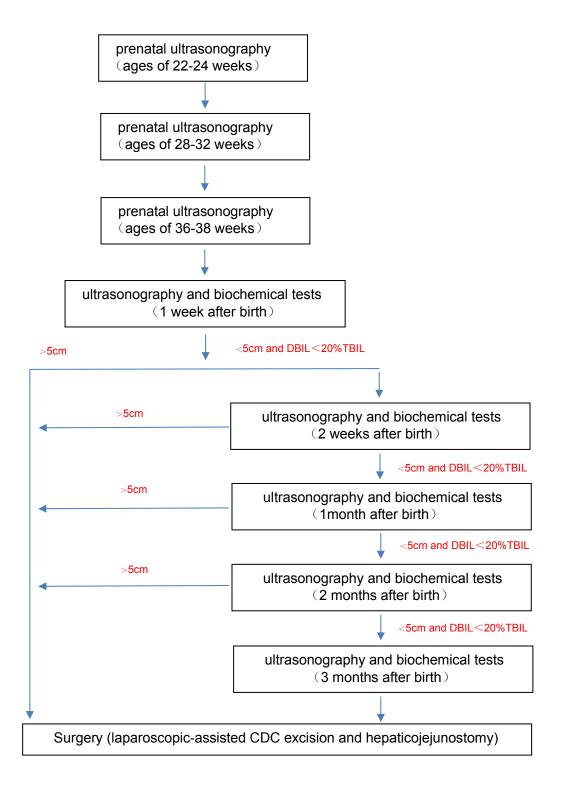
The follow-up process from antenatal to perinatal was established, and the maximum diameter of postnatal cyst was used as the standard for the selection of surgical opportunity. The curative effect was evaluated by comparing with previous treatment schemes.

- (1) Inclusion Criteria:
- · Infants with a prenatal and postnatal diagnosis with CDC
- · Prenatal and postnatal hepatobiliary ultrasound data were complete
- · Age of visit  $\leq$  3 months

Exclusion criteria:

- · Unable to tolerate surgery after birth
  - (2) Flow Chart for prenatal diagnosis of choledochal cyst





# Prenatal ultrasonography [10]:

- (1) visibility of the right hepatic artery (RHA)
- (2) The maximum lumen length and maximum width of the GB and cyst from inner wall to inner ,as well as the wall thickness of the GB

## 4. Follow-up scheme

Follow-up scheme during the post-operative period included 1 week post-operative day, 1 month, 3-month, 6-month, then every year visits thereafter.

Follow-up Patients' follow-up after surgery mainly consisted of ultrasonography and biochemical tests (alanine aminotransferase (ALT), aspartate aminotransferase (AST),  $\gamma$ -glutamyl transpeptidase (GGT), and direct bilirubin (DBIL))

# 5. Outcomes and Efficacy Evaluation

- (1) For the case—control studies, each patient treated with the new protocol was matched with two control patients treated with old protocol. Cases and controls were matched by all of the following: (1) gestational age at birth (±2 weeks), (2) birth weight (±0.5 kg), (3) gender, (4) Age of visit < 3 months, (5) All patients received laparoscopic-assisted CDC excision and hepaticojejunostomy.
- (2) Old protocol <sup>[8]</sup>: symptomatic neonates and infants should be operated on as soon as possible. Elective operations should be considered for asymptomatic cases at approximately 3–6 months of age along with postnatal monitoring of organ functions, primarily of the liver, to determine if laboratory symptoms develop to warrant earlier intervention
- (3) Comparison of baseline material of patients in the two groups

  The median estimated gestational age (EGA), cyst diameter at the initial time of detection of the cyst, gestational age, cesarean rates, mean birth weight, sex ratio, age at operation in months, weight in kg at operation, type of CDC.
  - (4) Outcomes and Efficacy Evaluation
- · Comparison of preoperative complications

Guangzhou Women and Children's Medical Center

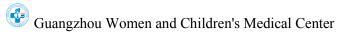
preoperative symptom rate, cyst rupture, preoperative laboratory results performed within 1 week before the operation (ALT,AST, GGT, DBIL, TBIL, DBIL/TBIL)

- Comparison of intraoperative conditions between the 2 groups.
   operative time in minutes, Body weight-corrected blood loss in ml/kg
- Comparison of preoperative complications
   bile leakage, anastomotic stricture, bowel obstruction, pancreatic

fistula, incision dehiscence

### 6. References

- [1] Lipsett P A, Pitt H A. Surgical treatment of choledochal cysts[J]. Journal of Hepato-Biliary-Pancreatic Surgery, 2003, 10(5):352-359.
- [2] Lugo-Vicente HL. Prenatally diagnosed choledochal cysts: Observation or early surgery?[J]. Journal of Pediatric Surgery, 1995, 30(9):1288.
- [3] Okada T, Sasaki F, Ueki S, et al. Postnatal management for prenatally diagnosed choledochal cysts[J]. Journal of Pediatric Surgery, 2004, 39(7):1055-1058.
- [4] Tanaka H, Sasaki H, Wada M, et al. Postnatal management of prenatally diagnosed biliary cystic malformation.[J]. Journal of Pediatric Surgery, 2015, 50(4):507-510.
- [5] Bancroft J D, Bucuvalas J C, Ryckman F C, et al. Antenatal diagnosis of choledochal cyst.[J]. Journal of Pediatric Gastroenterology & Nutrition, 1994, 18(2):142-5.
- [6] Redkar R, Davenport M, Howard E R. Antenatal diagnosis of congenital anomalies of the biliary tract[J]. Journal of Pediatric Surgery, 1998, 33(5):700.
- [7] Diao M, Li L, Cheng W. Timing of surgery for prenatally diagnosed asymptomatic choledochal cysts: a prospective randomized study[J]. Journal of Pediatric Surgery, 2012, 47(3):506-512.
- [8] Kamisawa T, Ando H, Suyama M, et al. Japanese clinical practice guidelines for pancreaticobiliary maljunction[J]. J Gastroenterol, 2012,47(7):731-759.
- [9] Xisi Guan #, Junting Li , Zhe Wang , Jixiao Zeng , Wei Zhong , Jiakang Yu. Timing of operation in children with a prenatal diagnosis of choledochal cyst: A single-center retrospective study. J Hepatobiliary Pancreat Sci.2022 Apr 18. doi: 10.1002/jhbp.1155



 $[10] \ Chen\ L\ ,\ He\ F\ ,\ Zeng\ K\ ,\ et\ al.\ Differentiation\ of\ cystic\ biliary\ atresia\ and$   $choledochal\ cysts\ using\ prenatal\ ultrasonography. [J].\ ULTRASONOGRAPHY,\ 2021.$