СРПСКИ АРХИВ
ЗА ЦЕЛОКУПНО ЛЕКАРСТВО
SERBIAN ARCHIVES
OF MEDICINE

Address: 1 Kraljice Natalije Street, Belgrade 11000, Serbia
☏ +381 11 4092 776, Fax: +381 11 3348 653
E-mail: office@srpskiarhiv.rs, Web address: www.srpskiarhiv.rs

Paper Accepted*

ISSN Online 2406-0895

Case Report / Приказ болесника

Aleksandar Sretenović[1,2,♣], Đorđe Topličić[1], Rastko Sava Jeremić[1], Dragana Vujović[1,2]

Biliary atresia associated with intestinal malrotation-unusual intraoperative presentation

Билијарна атрезија повезана са малротацијом црева-необична интраоперативна презентација

[1]University Children's Hospital, Belgrade, Serbia;
[2]University of Belgrade, Faculty of Medicine, Belgrade, Serbia

Received: January 13, 2025
Revised: February 11, 2025
Accepted: February 12, 2025
Online First: February 14, 2025
DOI: https://doi.org/10.2298/SARH250113017S

*__Accepted papers__ are articles in press that have gone through due peer review process and have been accepted for publication by the Editorial Board of the *Serbian Archives of Medicine*. They have not yet been copy-edited and/or formatted in the publication house style, and the text may be changed before the final publication.

Although accepted papers do not yet have all the accompanying bibliographic details available, they can already be cited using the year of online publication and the DOI, as follows: the author's last name and initial of the first name, article title, journal title, online first publication month and year, and the DOI; e.g.: Petrović P, Jovanović J. The title of the article. Srp Arh Celok Lek. Online First, February 2017.

When the final article is assigned to volumes/issues of the journal, the Article in Press version will be removed and the final version will appear in the associated published volumes/issues of the journal. The date the article was made available online first will be carried over.

♣Correspondence to:
Aleksandar SRETENOVIĆ
University Children's Hospital, Tiršova 19, Belgrade, Serbia
E mail: alexmd64@gmail.com

Biliary atresia associated with intestinal malrotation-unusual intraoperative presentation

Билијарна атрезија повезана са малротацијом црева-необична интраоперативна презентација

SUMMARY

Introduction Biliary atresia (BA) is an idiopathic, progressive obliterative cholangiopathy of unknown etiology. The incidence of BA is 5–10 cases per 100,000 live births. Two clinical phenotypes of BA have been described: syndromic and non-syndromic form. From 1959. Kasai procedure is a standard therapeutic procedure. However, patients in whom there is an association of biliary atresia with other structural anomalies may have a worse outcome after the procedure. The goal was to present our patient with unusual form of BA associated with intestinal malrotation.

Case report We present a two-month-old female infant hospitalized because BA was suspected. On the echosonographic examination of the abdomen the gallbladder was not visible. Intraoperative diagnosis of BA was confirmed, and the Kasai procedure was performed. During the operation, intestinal malrotation with Ladd's bands was identified. In this case, after the complete Ladd procedure, we decided to trace the Roux coil through the mesoduodenum and then behind the duodenum towards the portal plate, where a portoenterostomy was then created in a standard way. During the "follow-up" the infant was free of complaints, the stools were normally discolored, and the values of liver function tests had a downward trend.

Conclusion Any doubt about the diagnosis of biliary atresia obliges us to determine the existence of all other possible anatomical abnormalities and associated anomalies, due to their potential importance in changing treatment plans and surgical approach, but also the impact on the outcome of treatment.

Keywords: biliary atresia; Kasai procedure; malrotation

САЖЕТАК

Увод Билијарна атрезија (БА) је идиопатска, прогресивна облитеративна холангиопатија непознате етиологије. Инциденција БА је 5–10 случајева на 100.000 живорођених. Описана су два клиничка фенотипа БА: синдромски и несиндромски облик. Од 1959. Касаи процедура је стандардна терапијска процедура. Међутим, пацијенти код којих постоји повезаност билијарне атрезије са другим структурним аномалијама могу имати лошији исход након операције. Циљ нашег рада је био да прикажемо случај БА повезан са малротацијом црева.

Приказ случаја У овом раду представљамо двомесечно одојче које је хоспитализовано због сумње на БА. На ехосонографском прегледу абдомена жучна кеса није била видљива. Интраоперативна дијагноза БА је потврђена и урађена је Касаи процедура. Током операције идентификована је малротација црева са Ладовим бридама. У овом случају, након комплетне Ладд процедуре, одлучили смо да усмеримо *Roux* вијугу кроз мезо дуоденума, а затим иза дуоденума ка портној плочи, где је потом на стандардни начин начињена портоентеростомија. Током редовног праћења одојче је било без тегоба, столица је нормално пребојена, а вредности тестова функције јетре су имале опадајући тренд**.**

Закључак Свака сумња у дијагнозу билијарне атрезије обавезује нас да утврдимо постојање свих других могућих анатомских абнормалности и придружених аномалија, због њиховог потенцијалног значаја у промени планова лечења и хируршког приступа, али и утицаја на коначни исход лечења.

Кључне речи: билијарна атрезија; Касаи процедура; малротација

INTRODUCTION

Biliary atresia (BA) is an idiopathic, progressive obliterative cholangiopathy of unknown etiology that affects the intra- and extra hepatic bile ducts, leading to cholestasis, progressive fibrosis and liver cirrhosis [1]. It is the most common cholestatic disorder of newborn age that

occurs with a variable incidence of 5–10 per 100.000 live born, with female to male ratio of 1.4:1 [2].

BA is most often an isolated defect but can be associated with other congenital anomalies in up to 16% of cases. There are two clinical phenotypes of BA, the syndromic or embryonic form, often associated with other congenital anomalies such as polysplenia, intestinal malrotation, portal vein abnormalities, situs inversus, absence of the inferior vena cava and congenital heart disease. The incidence of syndromic forms of biliary atresia is around 10-20% [3, 4]. The acquired or perinatal form of BA is more common and represents 80-90% of cases [4].

The association of biliary atresia with intestinal malrotation is rare and has a very variable incidence ranging from 0.3% to 12%. The appearance of the Kasai procedure in 1959 enabled the long-term survival of patients with BA [5]. In patients with associated congenital anomalies, the results of this procedure may be worse due to altered anatomical relationships [6, 7].

The goal of our paper was to present an unusual case of BA associated with intestinal malrotation.

CASE REPORT

A female full-term infant, born by spontaneous vaginal delivery with a birth weighing 3300g was hospitalized at the beginning of the third month of life (on the 67th day) due to jaundice and elevated values of liver function tests (LFT): direct bilirubin (DBil) 111.6 μmol/l, total bilirubin (TBil) 127 μmol/l, alanine aminotransferase (ALT) 173 U/L, aspartate aminotransferase (AST) 118 U/L. In the clinical findings at the beginning of hospitalization,

apart from icterus, the liver was palpated 2 cm below the right lower rib arch, while the rest of the physical findings were normal. Echosonographic examination of the abdomen after fasting revealed enlarged left lobe of the liver (cranio-caudal / diameter 45mm, caudate lobe antero-posterior /AP/ diameter 16mm), and the gallbladder was not visible. Considering the symptoms and signs of the disease, laboratory and ultrasound findings, BA was suspected and surgical treatment indicated. At the age of 71 days, the patient was taken for intraoperative cholangiogram and to proceed to a Kasai procedure if indicated. Hypoplastic gallbladder and the absence of extra hepatic bile ducts were found intraoperative which confirmed the diagnosis of BA and the need to perform the Kasai procedure (Figure 1). During the operation, intestinal malrotation was identified with the duodenojejunal transition in the right half of the abdomen, the cecum and the appendix in the left upper quadrant of the abdomen. After the Ladd procedure was performed, the problem of the direction of the Roux-en-Y loop, which in the original technique is oriented transmesocolic, arose. After the Ladd procedure was completed, we decided to trace the Roux loop through the mesoduodenum, which enabled the best anatomical position of the intestinal loop to create a porto-entero anastomosis (Figure 2). After the appendectomy, the coils of the colon were placed in the left half of the abdomen and a liver biopsy was taken.

The infant started enteral feeding on the third postoperative day, and on the fifth postoperative day had normal discolored stools. Full enteral intake was established seven days after surgery. Bilirubin values showed a decreasing trend after the surgery. The infant was discharged from the hospital seventeen days after surgery, on the 88th day of life. Bilirubin values at discharge were elevated (DBil 87.9 μmol/l, Tbil 104.2 μmol/l) as well as transaminase values (AST 221.0 U/L, ALT 389 U/L), stools were normally discolored, and body mass at discharge was 5020 grams. PH analysis showed congenital biliary atresia type 2.

At the follow-up examination three weeks after the operation, the bilirubin values remained unchanged, while the LFT values were decreasing, and the stools were usually discolored. Control abdominal ultrasound was performed one month after the operation showing no signs of intrahepatic bile ducts dilatation. Two months after the operation, the stools were usually discolored, with significant decrease of bilirubin and LFT values (Direct bilirubin: 65.8 umol/L Total bilirubin: 83.7 umol/L, AST: 104 U/L ALT: 152 U/L).

The authors declare that the article was written according to ethical standards of the Serbian Archives of Medicine as well as ethical standards of institutions for each author involved. Informed consent was obtained from all the subjects involved in the study.

DISCUSSION

Biliary atresia is a chronic progressive cholangiopathy, which occurs in a syndromic form with a frequency of 10 to 25% [1]. The association with intestinal malrotation is about 12% [2]. Although BA is known to coexist with different anatomical variations, studies have varied results on the prognosis of isolated BA compared with BA that is associated with congenital anomalies [3, 4, 5].

The outcome after surgery also depends on the conditions maintained in syndromic forms of BA [5]. However, due to limited number of cases the data on prognosis after the surgery is not yet adequately established [6].

Abdominal heterotaxy can have several surgical implications, primarily related to the orientation of the Roux-en-Y loop due to altered normal anatomical relationships [7, 8]. In the case of intestinal malrotation or non-rotation, the colon is located in the left side of the abdomen, while the duodenojejunal transition is located on the right side of the spinal column,

in the right side of the abdomen which can altered the normal orientation and positioning of the Roux-en-Y loop. Although it is unknown how these anatomical relationships would affect an infant's prognosis for biliary atresia, recognizing this abnormal anatomy is crucial both for the first exploration of hepato-portoenterostomy and for liver transplantation.

It can be expected that the presence of extra-hepatic anomalies and altered anatomy will negatively affect the outcome of surgical treatment. Differences in survival are primarily attributed to associated cardiac anomalies, however, studies showed that in experienced centers there is no significant difference in the outcome of surgical treatment or in long-term biliary drainage [9]. The long-term follow-up of patients after portoenterostomy is necessary because the establishment of bile drainage alone does not mean a cure for such patients, because the Kasai procedure does not affect progressive fibrosis as a basic characteristic of the disease [7].

It is very important that these anatomical relations become recognized on time because even though it is unknown how they affect prognosis of BA, it interferes with surgical approach and selection of technique to be applied [10, 11].

The timely diagnosis of biliary atresia and associated conditions provides more effective management and good outcome [12].

If there are some associated conditions confirmed, they are not always diagnosed antenatal, but if they were, the diagnosis is always challenging and management usual involves surgical correction in the immediate postnatal period, or later on if possible [13].

For the next decade there are some expectations for the identification of new therapeutics for infants with cholestasis and potential use of precision medicine to optimize therapy for each infant [14, 15].

Even though that the liver transplantation is a gold standard for treatment of patients with BA, and the vast majority of patients reach adulthood, waitlist mortality negatively affects the overall prognosis and in that case it is not related to associated anomalies [16].

Although syndrome forms of BA are rare, the diagnosis of BA should always indicate further evaluation for another anatomical abnormalities because of their importance and potential influence on treatment plans and the surgical approach.

ACKNOWLEDGMENTS

We would like to thank the patient.

Funding: This research received no external funding

Conflict of interest: None declared.

REFERENCES

1. Mathur P, Gupta R, Soni V, Ahmed R, Goyal RB. Biliary atresia associated with polysplenia syndrome, dextrocardia, situs inversus totalis and malrotation of intestines. J Neonatal Surg. 2014;3(1):9. [PMID: 26023480]
2. Gupta L, Bhatnagar V. A study of associated congenital anomalies with biliary atresia. J Indian Assoc Pediatr Surg. 2016 Jan–Mar;21(1):10–3. [DOI: 10.4103/0971-9261.158095] [PMID: 26862288]
3. Guttman OR, Roberts EA, Schreiber RA, Barker CC, Ng VL; Canadian Pediatric Hepatology Research Group. Biliary atresia with associated structural malformations in Canadian infants. Liver Int. 2011;31(10):1485–93. [DOI: 10.1111/j.1478-3231.2011.02578.x] [PMID: 21819536]
4. Mirza B, Iqbal S, Sheikh A. Biliary atresia associated with polysplenia syndrome, situs inversus abdominus, and reverse rotation of intestine. APSP J Case Rep. 2012;3(2):14. [PMID: 22953308]
5. Davenport M, Savage M, Mowat AP, Howard ER. Biliary atresia splenic malformation syndrome: an etiologic and prognostic subgroup. Surgery. 1993;113(6):662–8. [PMID: 8506525]
6. Allarakia J, Felemban T, Khayyat W, Alawi A, Mirza A, Alkhazal B, et al. Biliary atresia with an unusual abdominal orientation: A case report. Int J Surg Case Rep. 2019;55:152–5. [DOI: 10.1016/j.ijscr.2019.01.028] [PMID: 30738370]
7. Elebute OA, Timo MT, Alakaloko FM, Seyi-Olajide JO, Bode CO, Ademuyiwa AO. Biliary Atresia Splenic Malformation: A Case Report from A Tertiary Centre in West Africa and the Lessons Learnt. J West Afr Coll Surg. 2023 Jan–Mar;13(1):106–10. [DOI: 10.4103/jwas.jwas_195_22] [PMID: 36923807]
8. Chen SH, Chang KC, Wu JF, Chen HL, Lin WH. Case Report: Kasai Operation in Biliary Atresia After Extensive Bowel Resection. Front Surg. 2022;8:802859. [DOI: 10.3389/fsurg.2021.802859. [PMID: 35141270]
9. Vazquez J, López Gutierrez JC, Gámez M, López-Santamaría M, Murcia J, Larrauri J, et al. Biliary atresia and the polysplenia syndrome: its impact on final outcome. J Pediatr Surg. 1995;30(3):485–7. [DOI: 10.1016/0022-3468(95)90062-4] [PMID: 7760248]
10. Sabra Abdelazeem T, Abdelmohsen Magdy S, Abdelazeem B. Biliary atresia with rare associations: a case report. Annals of Pediatric surgery. 2023;19–37. [DOI: 10.1186/s43159-023-00269-5]
11. Choong Yew Hoe E, Mohd Shah MS, Mohamad IS, Leow VM. Biliary Atresia Associated With Jejunal Atresia: A Case Report. Cureus. 2024;16(7):e65463. [DOI: 10.7759/cureus.65463] [PMID: 39184670]
12. Solanki S, Mahajan JK, Singhai P, Mitra S. Colonic Atresia Associated with Biliary Atresia. J Indian Assoc Pediatr Surg. 2023 Jul–Aug;28(4):339–41. [DOI: 10.4103/jiaps.jiaps_184_22] [PMID: 37635893]
13. Mohamed F, Basavaraju U. Biliary Anomalies in VACTERL Syndrome: A Case Report. Cureus. 2024;16(8):e66700. [DOI: 10.7759/cureus.66700] [PMID: 39262547]
14. Feldman AG, Sokol RJ. Neonatal Cholestasis: Updates on Diagnostics, Therapeutics, and Prevention. Neoreviews. 2021;22(12):e819-e836. [DOI: 10.1542/neo.22-12-e819] [PMID: 34850148]
15. Chung PHY, Zheng S, Tam PKH. Biliary atresia: East versus west. Semin Pediatr Surg. 2020;29(4):150950. [DOI: 10.1016/j.sempedsurg.2020.150950] [PMID: 32861448]
16. Esmati H, van Rosmalen M, van Rheenen PF, de Boer MT, van den Berg AP, van der Doef HPJ, et al. Waitlist mortality of young patients with biliary atresia: Impact of allocation policy and living donor liver transplantation. Liver Transpl. 2023;29(2):157–63. [DOI: 10.1002/lt.26529] [PMID: 35702029]

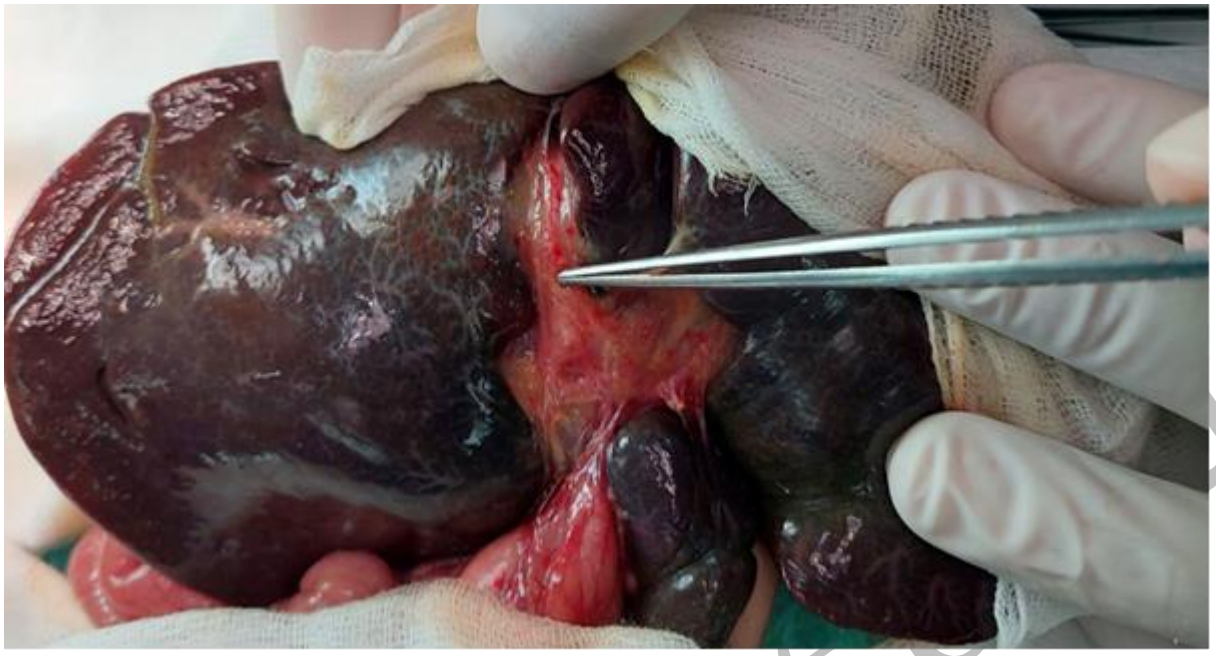

Figure 1. Macroscopic intraoperative finding of fibrotic liver parenchyma and absence of extrahepatic bile ducts

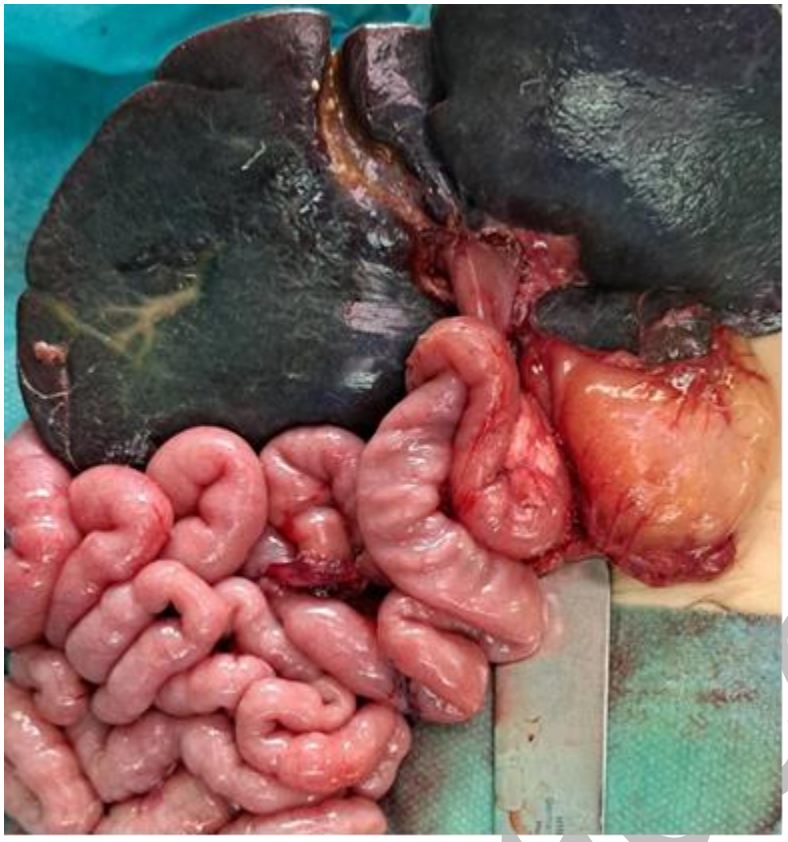

Figure 2. Porto-entero anastomosis – roux loop traced through the meso-duodenum

