

Management of Dual Diagnosis: Chilaiditi Syndrome and Biliary Atresia Complicated by Cirrhosis

Khadija Boualiten ^{a*}, T. Adioui ^a, I Mouslim ^a, S Berrag ^a,
F Nejjari ^a and M Tamzaourte ^a

^a *Department of Gastroenterology I Unit, Mohamed V-Souissi University, Mohamed V Military Teaching Hospital, Rabat, Morocco.*

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:

This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: <https://www.sdiarticle5.com/review-history/119968>

Received: 10/05/2024

Accepted: 11/07/2024

Published: 15/07/2024

Case Report

ABSTRACT

Chilaiditi sign is an incidental radiographic finding of a usually asymptomatic condition, where part of the intestine is situated between the liver and the diaphragm. However, the term "Chilaiditi syndrome" is used for symptomatic hepatodiaphragmatic interposition. We report the case of a 15-year-old patient presenting with chronic hepatic colic due to this syndrome, which was diagnosed by abdominal CT scan. Most patients with Chilaiditi syndrome can be managed conservatively. However, surgery is indicated for those who do not respond to conservative management or in cases of suspected serious complications such as ischemia or intestinal perforation. Biliary Atresia (BA) is a rare congenital malformation characterized by an inflammatory and destructive process that obstructs the intra- and extrahepatic bile ducts, leading to rapid progression to complete cholestasis and irreversible biliary cirrhosis, ultimately resulting in the child's death within the first few years of life. Our case is the rare in the literature indicating the association of Chilaiditi syndrome and biliary atresia.

*Corresponding author: Email: boualitenkhadija@gmail.com;

Cite as: Boualiten, Khadija, T. Adioui, I Mouslim, S Berrag, F Nejjari, and M Tamzaourte. 2024. "Management of Dual Diagnosis: Chilaiditi Syndrome and Biliary Atresia Complicated by Cirrhosis". *Asian Journal of Research and Reports in Gastroenterology* 7 (1):137-40. <https://journalajrrga.com/index.php/AJRRGA/article/view/142>.

Keywords: Chilaiditi syndrome; Biliary atresia; 15-year-old girl; treatment.

1. INTRODUCTION

“Chilaiditi syndrome is a rare condition defined by the presence of gastrointestinal symptoms associated with the radiological finding of segmental interposition of the intestine between the liver and diaphragm. Although rarely identified as a source of abdominal pain, Chilaiditi syndrome has clinical significance as it can lead to a number of serious complications including bowel obstruction, perforation and ischemia” [1-4]. “Most patients with this intestinal anomaly are asymptomatic throughout their lives; however, they can manifest with intermittent abdominal pain, distention, vomiting, anorexia, and constipation that on rare occasions require surgical intervention” [5-7].

2. CASE REPORT

A 15-year-old girl has a post medical history of neonatal jaundice revealing a type I biliary atresia complicated with liver cirrhosis. She underwent a Kasai procedure at the age of 1 month with smooth post-operative course and unremarkable follow-up. The patient presented with chronic hepatic colic without any other associated signs including no fever, jaundice, vomiting or constipation. Abdominal examination revealed slight tenderness in the right hypochondrium, and both biological and

Abdominal US results were unremarkable. Symptomatic treatment was initiated, but due to persistent symptoms, an abdominal CT scan was performed, revealing colonic interposition between the liver and the right hemidiaphragm (Fig. 1). The diagnosis of Chilaiditi syndrome was established. Following a multidisciplinary medical team discussion and considering the patient's complex medical history, analgesic treatment was recommended.

3. DISCUSSION

The Chilaiditi sign was initially identified by Antoine Béclère in 1899. However, in 1910, Demetrios Chilaiditi, a Greek radiologist first described three cases of right hemidiaphragmatic interposition of the colon [8]. In the general population, the prevalence of the Chilaiditi sign is estimated to be between 0.025% and 0.28%, but it could reach 1% in the elderly. The male-to-female sex ratio is 4:1[9].

The Biliary Atresia (BA) is a rare congenital malformation characterized by an inflammatory, destructive process that obstructs the intra- and extrahepatic bile ducts, leading to rapid progression to complete cholestasis and irreversible biliary cirrhosis, ultimately resulting in the death of the child within the first few years of life [10,11].

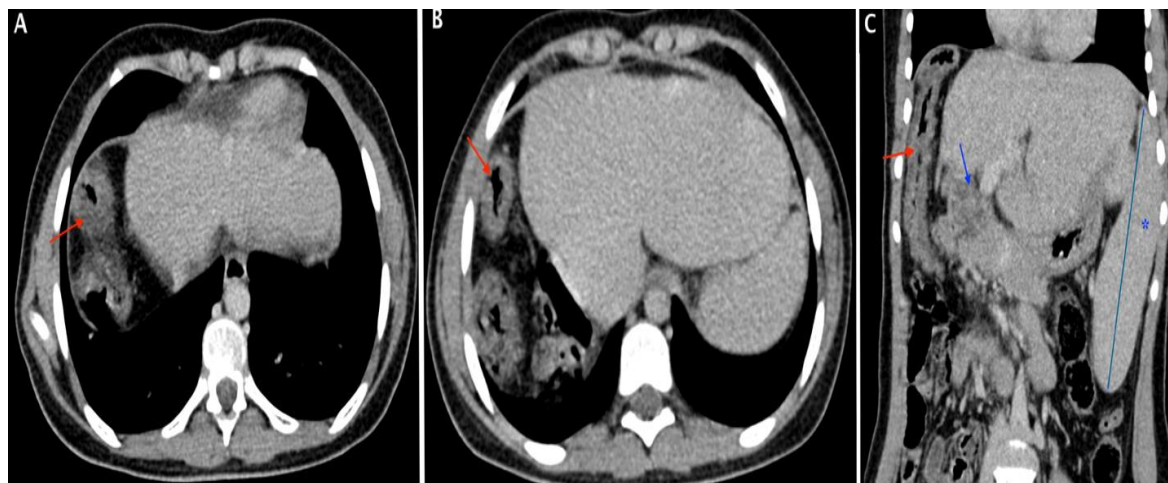


Fig. 1. (A and B) Axial section with (C) sagittal reconstruction of abdomen CR SCAN demonstrating the presence of interposed colonic loops between the right hemidiaphragm and liver (red arrows). Note the splenomegaly (blue asterisk) and stigmata of hepatopuertoenterostomy, also known as Kasai procedure (blue arrow)

The Chilaiditi sign is generally asymptomatic and is often discovered incidentally. However, if the patient is symptomatic, it is referred to as Chilaiditi syndrome, which may manifest as abdominal pain, constipation, nausea, vomiting, and occasionally dyspnea, respiratory distress and cardiac arrhythmias [9,12]. It can lead to complications such as obstruction, volvulus, or digestive perforation. Diagnosis is confirmed through a chest X-ray and abdominal CT scan. To diagnose Chilaiditi's sign from radiographic images, following criteria must be met in erect position: An elevation of the right hemidiaphragm compared to the liver, distension of the colon due to the presence of air, and the upper margin of the liver situated below the level of the left hemidiaphragm [9]. The Various contributing factors have been identified, including cirrhosis, absence or laxity of suspensory ligaments or the falciform ligament, aerophagia, increased abdominal pressure (due to conditions such as ascites, obesity, or pregnancy), elevation of the right hemidiaphragm (due to causes like paralysis or herniation), dolichocolon, chronic constipation, prolonged bed rest, intestinal malrotation, chronic bronchitis, and pulmonary emphysema. Other factors such as mental retardation or schizophrenia have also been identified [13,14].

In most cases, conservative medical treatment with analgesics or the use of a nasogastric is sufficient. However, severe complications may require surgical intervention. The primary treatment of biliary atresia is a hepatoportoenterostomy (Kasai procedure), which creates a bile-digestive shunt between the liver hilum and the jejunum. For our patient, after a multidisciplinary discussion, she received long-term conservative treatment due to her severe condition, which contraindicates surgical treatment, along with regular monitoring for her cirrhosis.

To our knowledge this is the first report in a paediatric patient of an association of biliary atresia associated with chilaiditi syndrome.

4. CONCLUSION

Chilaiditi syndrome is a benign condition characterized by the interposition of the colon between the liver and the diaphragm, which can sometimes lead to various complications. While Chilaiditi sign is seen on imaging, it's crucial to exclude other serious conditions that may necessitate surgical intervention. Typically,

treatment for Chilaiditi syndrome involves conservative management, but if symptoms persist, more invasive treatment options may be required.

Interestingly, Chilaiditi syndrome is also a rare association or manifestation of liver cirrhosis. our case is the first in the literature where Chilaiditi syndrome is associated with biliary atresia complicated by cirrhosis.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that generative AI technologies such as Large Language Models, etc have been used during writing or editing of manuscripts. This explanation will include the name, version, model, and source of the generative AI technology and as well as all input prompts provided to the generative AI technology.

CONSENT

As per international standards, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Michele F, Raffaele C, Desirée SS, Jonel N, Vladislavs S, Karaorman M. Management of Chilaiditi syndrome: our experience and literature review. *Journal of Surgery and Research*. 2021;4(2):270-7.
2. Weng WH, Liu DR, Feng CC, et al. Colonic interposition between the liver and left diaphragm - management of Chilaiditi syndrome: A case report and literature review. *Oncol Lett*. 2014;7: 1657-1660.
3. Cawich SO, Spence R, Mohammed F, et al. The liver and Chilaiditi's syndrome: significance of hepatic surface grooves. *SAGE Open Med Case Rep*. 2017;5: 1-4.
4. Zvezdin B, Savic N, Hromis S, et al. Chilaiditi's sign and syndrome: theoretical

- facts and a case report. *Vojnosanit Pregl.* 2016;73:277-279.
5. Gaith Khair MD. Chilaiditi Syndrome Complicated by a Closed-Loop Small Bowel Obstruction. *Gastroenterology & Hepatology.* 2012;8(4):275.
 6. Al-Salem AH, Al-Salem AH. Congenital Intestinal Atresia and Stenosis. *Atlas of Pediatric Surgery: Principles and Treatment.* 2020;445-66.
 7. Beyer D, Mödder U, Benz-Bohm G, Horwitz AE. Special Features of Acute Abdominal Disorders in Children.
 8. Chilaiditi D. Zur frage der hepatoptose und ptose im allgemeinen im anschlussan drei falle von temporarer, partieller leberverlagerung. *Fortschr Geb Rontgenstr.* 1911;16:173-208.
 9. Moaven O and Hodin RA. Chilaiditi syndrome: A rare entity with important differential diagnoses. *Gastroenterol. Hepatol.* 2012;8(4):276.
 10. Safwan M, Ramachandran P, Vij M, Shanmugam N and Rela M. Impact of ductal plate malformation on survival with native liver in children with biliary atresia. *Pediatr Surg Int.* 2015;31:837-843.
 11. Sinha CK and Davenport M. Biliary atresia. *J Indian Assoc Pediatr Surg.* 2008;13(2):49-56.
 12. Yin AX, Park GH, Garnett GM and Balfour JF. Chilaiditi syndrome precipitated by colonoscopy: A case report and review of the literature. *Hawii J Med Publ Health.* 2012;71(6):158.
 13. Farkas R, Moalem J and Hammond J. Chilaiditi's sign in a blunt trauma patient: A case report and review of the literature. *J Trauma Acute Care Surg.* 2008;65(6):1540-1542.
 14. Altomare DF, Rinaldi M, Petrolino M, Sallustio PL and Guglielmi A, et al. Chilaiditi's syndrome. Successful surgical correction by colopexy. *Tech Coloproctol.* 2001;5:173-175.

Disclaimer/Publisher's Note: The statements, opinions and data contained in all publications are solely those of the individual author(s) and contributor(s) and not of the publisher and/or the editor(s). This publisher and/or the editor(s) disclaim responsibility for any injury to people or property resulting from any ideas, methods, instructions or products referred to in the content.

© Copyright (2024): Author(s). The licensee is the journal publisher. This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

The peer review history for this paper can be accessed here:

<https://www.sdiarticle5.com/review-history/119968>