Chilaiditi’s sign and syndrome: rare conditions but diagnostically important in pediatrics. Clinical cases

Signo y síndrome de Chilaiditi: condiciones infrecuentes pero con importancia diagnóstica en pediatría. Casos clínicos

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Abstract

Although infrequent, bowel interposition between diaphragm and liver, Chilaiditi’s sign or syndrome (without or with gastrointestinal symptoms), are a major clinical condition given the possibilities of differential diagnosis, such as pneumoperitoneum, diaphragmatic hernia and subphrenic abscess. 

Objective: To report the cases of two preschool patients with Chilaiditi’s sign and syndrome, as well as to highlight the importance of this clinical condition. Clinical cases: Case 1: A male preschooler evaluated by respiratory disease without abdominal symptoms. Thorax X-ray shows left retrocardiac infiltrates and air in right subdiaphragmatic region. Previous radiographies shows the same image. He was diagnosed with Chilaiditi sign associated with pneumonia, antibiotics were used before discharge. Case 2: A female preschooler, evaluated by abdominal distention and constipation. A previous thorax X-ray shows bowel interposition between diaphragm and liver. Barium enema confirmed the findings. Blood test were normal. A Chilaiditi’s syndrome was diagnosed. She received medical treatment with favorable evolution. Conclusion: These cases highlight the importance of this clinical condition that, despite being infrequent, constitutes a diagnostic challenge in the emergency services.

Keywords: Chilaiditi Syndrome, subdiaphragmatic air, subdiaphragmatic bowel, Pneumonia
Introduction

The radiological finding of the intestinal interposition between the right hemidiaphragm and the liver is known as Chilaiditi’s Sign in honor to the first report carried out of this condition in 1910 by the Greek radiologist Demetrius Chilaiditi. When, besides this imaging finding, the patient suffers from gastrointestinal symptoms such as abdominal pain, vomiting, constipation, abdominal distention, and loss of appetite the Chilaiditi’s Syndrome is constituted.

This radiological finding is extremely rare. It presents an incidence that ranges from 0.025 to 0.28% in all radiological examinations of the thorax and abdomen, with a preference for persons over the age of 65 and with a higher frequency in men than women, with men: women ratio of 4:1. Despite this well-described epidemiological behavior, when reviewing the literature, extraordinary case reports of neonatal affection and many others in the pediatric population of different ages can be found. Consequently, it is necessary that pediatricians have subject knowledge.

Three clinical presentations have been described for this condition: 1) transverse colon or small intestine interposition in the right anterior subphrenic space; 2) colon or stomach interposition in the right extraperitoneal space; 3) transverse colon interposition in the right posterior subphrenic space. The presence of air will be evident in the subdiaphragmatic space with these forms, this finding has great clinical importance when is accompanied by pain since it doesn’t always imply a surgical emergency. Therefore, the clinical suspicion and awareness of this pathological condition could prevent unnecessary surgical interventions.

The aim of this article is to present the cases of two preschool patients with Chilaiditi’s Sign and Syndrome and highlight the importance of this clinical condition in relation to the likelihood of confusion and differential diagnosis.

Clinical cases

Case 1

3-year-and-11-months-old male patient, with a history of asthma in outpatient management with montelukast, was taken to the emergency services due to clinical picture of 2 day of evolution, characterized by rhinorrhea and productive, emetizing cough associated with fever up to 102.2 °F (39°C) that did not decrease with antipyretic. He did not have an epidemiological background of contagious or previous migratory movements.

The physical examination showed a symmetrical thorax with fine bibasilar rales. The abdominal examination and the rest of the physical examination were within the normal ranges. The laboratory tests evidenced leukocytosis (17,090 c/mm³) with left shift (PMN: 13,940 c/mm³, monocytes 1,300 c/mm³, lymphocytes 1,850 c/mm³), and C-reactive protein 117.62 mg/L (cut-off point 12 mg/L).

Chest X-ray showed left retrocardiac infiltrates with aerial bronchogram and the presence of air in right subdiaphragmatic region (Figure 1).

The mother was asked again about abdominal symptoms, but she denied them. The patient was diagnosed with acute pneumopathy antibiotic treatment was prescribed based on amoxicillin at a dose of 90 mg/Kg per day over 8 days.

The patient was clinically stable when reassessing two weeks later. For this medical evaluation, the mother provided radiographs from the previous year where the same presence of air in the right subdiaphragmatic region was evident. Hence, after concluding that the patient presented Chilaiditi’s sign, he was discharged.

Case 2

2-years-and-10-months-old female patient, previously healthy, was referred to Pediatric Gastroenterology due to a clinical picture that started a month before, consisting of rhinorrhea, cough, and respiratory distress. The chest X-ray showed an image of the right colon between the hemidiaphragm and the liver (Figure 2), but not another significant cardiopulmonary finding. At the time of the medical consultation, the mother mentioned the improvements related to the respiratory symptomatology. However, she did manifest that the patient presented abdominal distension associated with hard stool consistency, five times a day.

The physical examination highlighted a preschoo-ler in regular general conditions with global abdominal enlargement, tympanic, soft to the palpation, no palpable masses or visceromegaly, in the rest of the physical examination there were no significant findings. In the light of this radiological finding, laboratory test and complementary images were requested. The patient was treated with magnesium hydroxide and nonpharmacological measures (increased water intake and daily fiber) to improve constipation.

The patient was reassessed with requested tests. The abdominal ultrasound reported liver and bile ducts without lesions, spleen and kidneys without anomalies, and abundant gas was found in the epigastrium-mesogastrium by the left lobe, pancreas, and great vessels. The barium enema confirmed colonic interposition between the diaphragm and the liver (Figure 3). The tests of thyroid function, transaminase, and baseline glycemia were within normal range.
Based on the clinical picture and the laboratory tests, it was concluded that the patient presented Chilaiditi’s syndrome. With the correct management, the patient presented an improvement in her bowel habits and abdominal distention. Therefore, the treatment was maintained, and the patient was monitored by Gastroenterology.

**Discussion**

While is extremely rare, Chilaiditi’s sign is an important imaging finding since the differential diagnosis possibilities such as pneumoperitoneum, diaphragmatic hernia, intestinal pneumatisis, subphrenic abscess, and hydatid cyst. Nevertheless, it can be differentia-
From the free intraperitoneal air by identifying colonic haustra and by observing how the image is not affected by the patient’s position changes. The vast majority of cases, like ours, correspond to incidental findings when doing a radiography for other reasons. In particular, our cases were identified in pediatric patients, despite being a condition that most often affect older adults.

This report supports the idea mentioned by Iturralde et al., who indicate that only one radiography is sufficient to diagnostic Chilaiditi’s sign or syndrome. In case 2, given the gastrointestinal symptoms manifested by the patient (abdominal distention and constipation), the barium enema examination was requested in order to discard other pathologies, considering existing association of Chilaiditi’s syndrome with other congenital malformations as intestinal level and the importance of discarding a possible differential diagnosis.

Despite not having full clarity of its etiology, many factors have been implicated in this entity’s development. The most common cause is the flaccidity and elongation of the intestinal suspensory and hepatic ligaments. Furthermore, the predisposing factors include hepatic, intestinal, diaphragmatic, and miscellaneous factors, which are listed in Table 1. It is an entity of rare presentation and affects equally men and women, and its diagnosis increases with the age and the amount of abdominal fat. Congenital and anatomical alterations have been mainly seen in children.

Most patients with Chilaiditi’s sign are asymptomatic, as in the case 1. But occasionally, as in the case 2, they can present nausea, vomiting, abdominal pain, constipation, intermittent intestinal obstruction, chronic abdominal pain, and even acute abdomen cases have been reported. In case of presenting any of these symptoms, the term Chilaiditi’s syndrome is used.

In general, patients with Chilaiditi’s sign do not require managing. On the other side, patients with Chilaiditi’s syndrome normally improve with conservative symptomatic management, noticing that the symptoms have improved with bed rest, hydration, intestinal decompression, laxatives, and at times enemas. Only some patients will need surgery, those cases with persistent pain or development of complications, within which has been described: intestinal obstruction, perforation, ischemia, and volvulus. In the both presented cases, it was decided an expectant management with good subsequent evolution.

### Conclusion

The importance of this rare radiological finding lies in a proper diagnostic orientation due to the high complexity pathologies that can be mistaken for Chilaiditi’s sign and syndrome. An inadequate interpretation of the imagining finding can lead to misdiagnosis, which means excessive managing with no benefits to the patients. In this line, the exhaustive evaluation of the images and a correct contextualization with the manifestations are the roadmap to discard differential diagnosis of great severity.

### Ethical Responsibilities

**Human Beings and animals protection:** Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

**Data confidentiality:** The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

**Rights to privacy and informed consent:** The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

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### Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

### Table 1. Factors that have been associated to the Chilaiditi’s Sign

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<td>Elevation of right hemi-diaphragm</td>
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<td>Hepatic Factors</td>
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<td>Relaxation/elongation of suspensory ligaments</td>
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<td>Intestinal factors</td>
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<td>Relaxation/elongation of suspensory ligaments</td>
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<td></td>
<td>Constipation</td>
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<td>Colonic elongation/hyper-motility</td>
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<td>Others</td>
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References