

An urgent chest-abdomen computed tomography scan was performed, showing acute emphysematous cholecystitis (Fig. 1A) complicated with probable bilioperitoneum (Fig. 1B). An incidental finding was bilateral pleural effusion and collections in the mediastinum containing air, indicative of NM (Fig. 1C), and air in the retroperitoneum extending to the mediastinum (Fig. 1D). No esophageal ruptures were identified, which, together with the absence of subcutaneous emphysema, allowed us to reasonably rule out esophageal perforation.

Given the progression of the sepsis, support measures were intensified with high-dose amine supplementation, broad-spectrum antibiotic therapy, and orotracheal intubation. However, the patient developed multiorgan dysfunction. The combination of the fulminant nature of the symptoms, the rapid progression, hemodynamic instability, age and associated comorbidities led us to rule out aggressive surgical debridement of the infectious focus as being too risky, even though the symptoms were unlikely to improve with conservative management. The patient died a few hours later.

NM is a polymicrobial infection of the mediastinum caused by aerobic and anaerobic microorganisms^{4,5} which, despite combined treatment, still has high mortality rates. Early multidisciplinary management⁷ in specialized centers⁶ using broad-spectrum antibiotic therapy subsequently adjusted according to the results of sensitivity testing^{5,7} and aggressive surgical drainage of the primary focus and mediastinum are key to patient survival^{4,5}. A delay in diagnosis and surgery of more than 24 h has been associated with an unfavorable prognosis³.

The main risk factors and predictors of mortality include the presence of previous comorbidities, acute renal failure¹, and severe sepsis or septic shock^{2,5}. Patients with signs of sepsis and/or septic shock should be admitted to the intensive care unit²⁻⁴. The length of stay in the unit and severity at admission have a direct impact on mortality³.

Early diagnosis is therefore fundamental^{4,7}, though complex⁶, and a high level of suspicion is necessary² since the clinical picture is nonspecific, marked by the primary focus (such as trismus, dysphagia, or even abdominal pain)⁵, and by chest pain, dyspnea, and signs of sepsis⁴. Computed axial tomography remains the diagnostic technique of choice for establishing the origin of infection, assessing its extension, and planning surgery^{4,7}. The diagnosis can be defined by the presence of a remote focus with radiological findings indicative of mediastinitis³.

Spread of an abdominal infection to the mediastinum, though rare, has previously been described in the literature. Chang and Chen⁸ describe the first published case of mediastinitis marked by extensive necrosis and abscess formation in the mediastinum caused by a pancreatic pseudocyst. Chong et al.⁹ later reported a patient with acute pancreatitis complicated by the formation of a paraesophageal pancreatic pseudocyst associated with NM. Finally, Dajer-Fadel et al.¹⁰ described a patient with emphysematous pyelonephritis and mediastinal progression of infection with NM. They suggest that the route of dissemination is retroperitoneal spread of air and infection to the mediastinum. These cases illustrate the possibility of propagation to the mediastinum with the development of secondary ascending NM.

This is therefore the first case reported in the literature of ascending NM associated with emphysematous cholecystitis, and the fourth case of ascending NM from an abdominal focus. Given the rarity of this form of presentation, this complication should be considered

in patients with a history of abdominal infection and generalized decline. Combined treatment with antibiotic therapy and surgical debridement has been shown to reduce mortality. A high level of suspicion is necessary to allow early diagnosis and the rapid introduction of therapeutic measures.

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Abernethy Malformation: An Unusual Extrathoracic Cause of Chronic Hypoxemia in Pediatrics



Malformación de Abernethy: una causa extratorácica inusual de hipoxemia crónica en pediatría

Dear Editor,

Abernethy malformation is rare congenital extrahepatic portosystemic shunt that allows blood from the gut and spleen to reach the systemic venous circulation bypassing the liver filter. This situation leads in some cases to serious complications such as hepatopulmonary syndrome, portopulmonary hypertension, or hepatic encephalopathy. Specifically, hepatopulmonary syndrome may

present with chronic hypoxemia. Even though it is a rare condition, extrathoracic pathology should be seek after ruling out cardiac and primary pulmonary disease.

A five year-old boy was admitted for an acute bronchitis and hypoxemia. On physical exam, perioral cyanosis and digital clubbing were discovered. The child had a past medical history of recurrent bronchitis and was followed in the pediatric neurology outpatient clinic under the suspicion of an autistic disorder. In the emergency department nebulized salbutamol was initiated and respiratory symptoms quickly subsided. Three days later, the child improved his clinical condition presenting no dyspnea and a clear cardiopulmonary auscultation but hypoxemia persisted. Orthodeoxia was observed with transcutaneous oxygen saturation decreasing from 88% to 81–82% when he changed from supine to a sitting position. Blood tests showed normal hematocrit (39%), normal liver function, and normal ammonia levels.

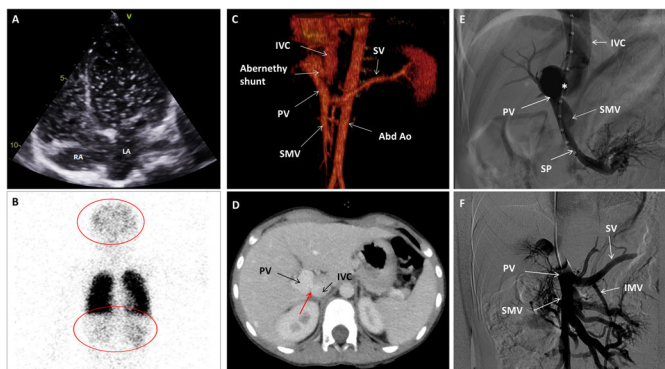


Fig. 1. (A) Echocardiography. Four chamber view depicting a shaken serum. After three cardiac cycles the left atrium was also opacified. (B) Nuclear scanning with Technetium 99m macro-aggregated albumin. A part from the lungs, brain and kidney also showed radioactivity due to tracing uptake (red circles). (C) CT Angiogram. 3D Volume rendering showing the Abernethy malformation. (D) CT Angiogram: Axial plane illustrating the Abernethy shunt (red arrow). (E) Fluoroscopy. Left oblique projection. The centimetered catheter reaches the portal vein from the inferior vena cava, through Abernethy shunt (asterisk). (F) Fluoroscopy. Antero-posterior projection. Balloon shunt occlusion test. Abbreviations: Abd Ao: abdominal aorta. IMS: inferior mesenteric vein. IVC: inferior vena cava. LA: left atrium. PV: portal vein. RA: right atrium. SMV: superior mesenteric vein. SP: splenic vein.

Some tests were performed to find the cause of hypoxemia. No evidence of pulmonary hypertension nor of intracardiac shunts were noticed on echocardiography with agitated serum, but quick pass of microbubbles to the left atrium was suggestive of an intrapulmonary shunt (Fig. 1A, Video 1). No lung disease and no evidence of macroscopic pulmonary arterio-venous fistulas were detected in the thorax angio Computed Tomography (CT). However, under the suspicion of an extra-cardiac right to left shunt, a lung nuclear scanning with Technetium 99m-labeled macroaggregated albumin was requested. Brain and kidney radiotracer uptake confirmed the presence of a right to left shunt of 36% (Fig. 1B). These findings were indicative of an intrapulmonary shunt suggestive of hepatopulmonary syndrome. An abdominal echography demonstrated a porto-caval latero-lateral shunt with a well-developed portal vein system. An abdominal angio-CT confirmed the diagnosis of Abernethy type-2 malformation (Fig. 1C and D).

The case was presented to an interventional radiology team of the reference hospital. A catheterization procedure documented a 10 mm side-to-side shunt between the portal vein and the inferior vena cava (IVC) (Fig. 1E). The basal portal vein and IVC pressures were 9 and 3 mmHg, respectively, and a balloon shunt occlusion test evidenced a moderate increase in portal pressure up to 23 mmHg (Fig. 1F). The anatomic (broad and short) shunt morphology was not suitable for any endovascular device. Therefore, the patient was presented to the pediatric hepato-digestive surgery team and underwent a surgical shunt closure through laparoscopy using a hem-o-lok® clip system. Thereafter the patient followed a favorable clinical course. Liver function and liver blood flow were monitored daily through blood test and echography, and remained normal. Before patient discharge, hypoxemia persisted with a basal transcutaneous hemoglobin saturation of 90% and, hence, a home oxygen therapy with nasal cannula (3 L/min) was provided. During follow-up the patient experienced a progressive improvement of his hypoxemia and six months after shunt closure oxygen therapy was withdrawn.

Abernethy malformation consists in a rare congenital extrahepatic portosystemic shunt which allows blood from the gut and spleen to reach the systemic venous circulation bypassing the liver.¹⁻⁶ It was first described in 1793 by the London surgeon John Abernethy who observed for the first time a congenital absence of the portal vein with a mesenteric-caval shunt.^{3,5,7,8} The estimated incidence of the Abernethy malformation is reported to be 1/30,000 live births in countries where screening for galactosemia is routinely performed.¹ This shunt may be classified in two main types: type 1, in which the portal blood is completely diverted into the IVC through a side-to-end anastomosis and where there is an absence or a vague remnant of intrahepatic portal vein system; and type 2, in which the

Table 1
Main mechanisms of extrathoracic hypoxemia in pediatrics.

Extrathoracic hypoxemia mechanism	Disease example
Hypoventilation disorder	<ul style="list-style-type: none"> Respiratory depression due to dysfunction of the respiratory center (meningitis, head trauma, congenital hypoventilation, sedative drugs) Neuromuscular disease (muscular atrophy or dystrophy, Guillain Barre sind, etc.)
Hepato-pulmonary syndrome	<ul style="list-style-type: none"> Liver disease (cirrhosis, fibrosis, etc.) Congenital extrahepatic portosystemic shunt (Abernethy)
Airway obstruction	<ul style="list-style-type: none"> Congenital (tracheal atresia, laringomalacia, quist, mass) Acquired (laryngitis, epiglottitis, abscess, strange body) Both: paralyzed cord, tracheal stenosis
Blood disorder	<ul style="list-style-type: none"> Anemia
Intoxications	<ul style="list-style-type: none"> Cyanide Carbon monoxide Meta-hemoglobinemia
Reduced inspired pO ₂	<ul style="list-style-type: none"> Living at high altitude

portal system is hypoplastic but patent and communicated side-to-side with systemic veins, usually the IVC.^{1-6,8-11}

Pathophysiologically, Abernethy malformation could be considered as an infrequent cause of hepatopulmonary syndrome which is characterized by a deficient arterial oxygenation due to pulmonary capillary dilatation in the context of liver disease.^{4,11-14} Blood coming from the gut via superior mesenteric vein and spleen via splenic vein is deviated partially or totally to the IVC. This bypass could imply an imbalance between vasodilator and vasoconstrictor constituents that may lead to vasodilation of pulmonary capillaries, allowing a direct mixed venous blood shunt to the pulmonary veins without being oxygenated thereby causing subsequent hypoxemia.^{1,4,12,14}

Abernethy malformation in children may present a broad clinical spectrum. While some patients can be asymptomatic, others may exhibit marked cyanosis, severe hypoxemia, pulmonary arterial hypertension, digital clubbing, vascular anomalies like spider nevi, hepatic encephalopathy, or liver tumors.^{1,2,4,6,8-10,14} In the case described, digital clubbing, a clinical sign suggestive of chronic hypoxemia, was not consistent with a repeatedly observed normal hematocrit (39%) rather than an increased one as expected in view of the hypoxemia. It may be that it was too soon to observe a compensatory polyglobulia, or perhaps hypoxemia should have been more severe to stimulate erythropoietin production. Orthodeoxia was also observed, a clinical finding that may accompany the hepatopulmonary syndrome.^{4,12}

A mild autism was recognized but liver dysfunction was not detected and a normal liver parenchyma was visualized in the abdominal echography and in the CT. Thus, it is unclear whether or not the autistic features could correspond to an incipient form of hepatic encephalopathy.

Albeit diagnosis of Abernethy malformation in children remains challenging, once suspected clinically its presence can be easily confirmed by non-invasive imaging techniques such as abdominal echography, CT or Magnetic Resonance Imaging.^{1,3,5,9} Although this malformation can be a cause of extrathoracic hypoxemia, additional causes are summarized in Table 1.¹⁵

According to others and to our experience, optimal management of this congenital shunt requires a multidisciplinary team.⁸ Medical treatment, a prophylactic or therapeutic shunt closure, and a liver transplant are the main healing options.^{1-3,5,7-11} In our patient, and given the unfavorable anatomy for interventionism, a surgical closure through laparoscopy was elected. In general, it appears reasonable to perform an early rather than a late shunt closure in order to reduce the development of complications.^{1,7,9} Shunt closure restores intrahepatic circulation in most patients allowing a clinical improvement of hypoxemia, hepatic encephalopathy, and in some instances of the pulmonary hypertension.^{1,8,9} Our patient needed six months to recover a normal transcutaneous hemoglobin saturation and one year to experience a notable improvement in his behavior.

The case described contributes to increase the awareness of extrathoracic hypoxemia causes such as Abernethy malformation when facing a chronic hypoxemia in children and once cardiac and primary pulmonary causes have been excluded. A prompt diagnosis and proper management may prevent the development of serious complications.

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Conflict of interest

The authors have no conflicts of interest to disclose.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.arbres.2021.02.005](https://doi.org/10.1016/j.arbres.2021.02.005).

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COVID-19 and smoking: An opportunity to quit[☆]



Tabaco y coronavirus: una oportunidad para dejar de fumar

To the Editor:

The start of 2020 heralded high expectations for smoking cessation in Spain, due to the planned funding of 2 of the 3 first-line drugs for quitting smoking (varenicline and bupropion) by the Social Security regime. However, the arrival in March 2020 of the SARS-CoV-2 pandemic (Covid-19), with home confinement and the ensuing anxiety and stress, changed the landscape. Although logically we might think that the Covid-19 crisis would not be the ideal time to quit smoking, data are emerging that suggest the opposite.

We analyzed abstinence during confinement in patients followed in our smoking unit who had started an attempt to quit smoking between January 1, 2020, and the declaration of the state of alarm on March 13. There were 100 patients, 46% men, with a mean

age of 59 ± 9 years; 35% had high blood pressure, 8% had diabetes, and 26% had dyslipidemia. Respiratory history included chronic obstructive pulmonary disease (COPD) in 42% and obstructive sleep apnea (OSA) in 20%. All were prescribed treatment with varenicline. Smoking habits were as follows: 8 ± 2 points on the visual analogue motivational scale, 6 ± 2 points on the Fagerström test, and 16 ± 12 ppm on co-oximetry. Patients started smoking, on average, at 17 ± 5 years, and average consumption was 18 ± 8 cigarettes a day with a cumulative index of 43 ± 18 pack-years. Follow-up in our unit comprises about 6–7 visits until 1 year of abstinence is completed, the first visit taking place 2–4 weeks after quit date. With the arrival of confinement and the suspension of face-to-face visits, planned follow-up visits were made by telephone by the treating pulmonologist who resolved questions and prescribed medication electronically. Patients were asked about abstinence at 1, 3, and 6 months, including during the confinement period (March 14 to June 21, 2020). Over half (56%) of patients confirmed abstinence during confinement. If we analyze abstinence by months, we see a rate of 67% abstinence at 1 month, 52% at 3 months, and 47% at 6 months. Of those who did not stop smoking, 12% reported reducing the number of cigarettes consumed by more than half.

Previous studies have shown a 25%–35% success rate in smoking cessation in the first 6 months and up to the first year through the combination of pharmacological treatment, psychological support,

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