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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.

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

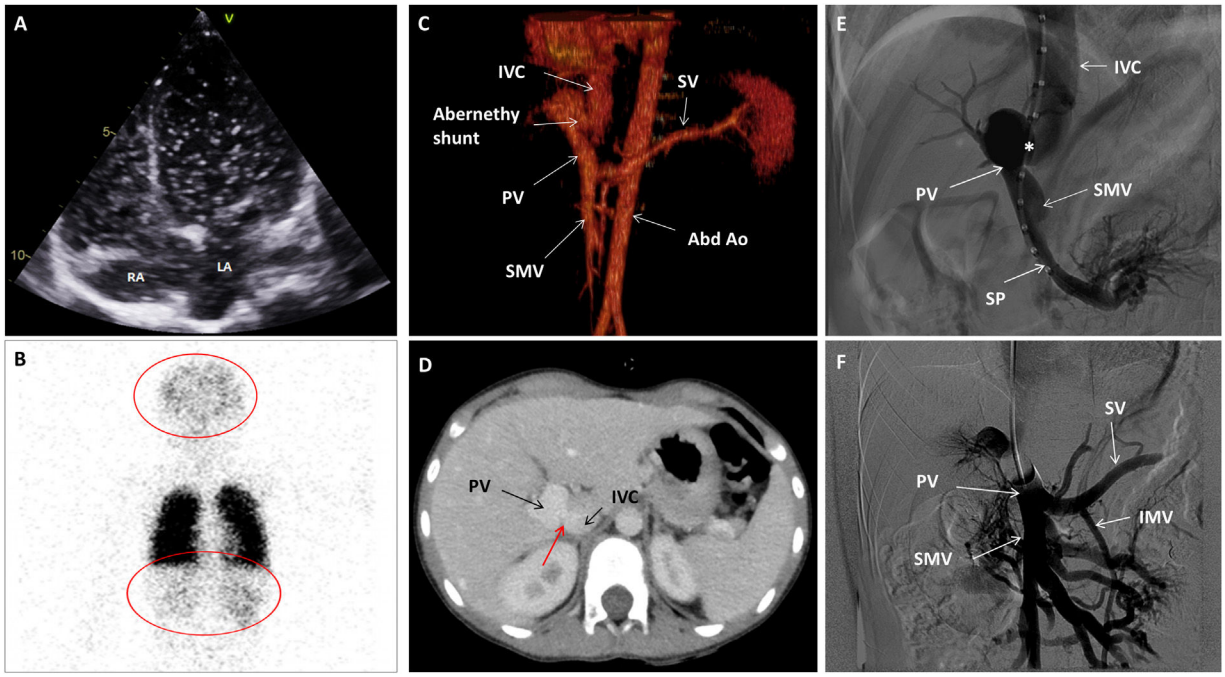

Fig. 1. (A) Echocardiography. Four chamber view depicting a shaked serum. After three cardiac cycles the left atrium was also opacified. (B) Nuclear scanning with Technetium 99m macro-aggregated albumin. A part from the lungs, brain and kidney also showed radioactivity due to tracing uptake (red circles). (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.

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 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 erytropoyetin 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.

Table 1
Main mechanisms of extrathoracic hypoxemia in pediatrics.

Extrathoracic hypoxemia mechanism	Disease example
Hypoventilation disorder	• 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-pulmonar syndrome	• Liver disease (cirrhosis, fibrosis,etc.) • Congenital extrahepatic portosystemic shunt (Abernethy)
Airway obstruction	• Congenital (tracheal atresia, laringomalacia, quist, mass) • Acquired (laryngitis, epiglottitis, abscess, strange body) • Both: paralyzed cord, tracheal stenosis
Blood disorder	• Anemia
Intoxications	• Cyanide • Carbon monoxide • Meta-hemoglobinemia
Reduced inspired pO_2	• Living at high altitude

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.

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Tabaco y coronavirus: una oportunidad para dejar de fumar

COVID-19 and Smoking: An Opportunity to Quit

Estimado Director:

El año 2020 se presentaba en nuestro país con unas altas expectativas en la deshabituación tabáquica, debido a la esperada financiación de 2 de los 3 fármacos de primera línea para dejar de fumar (vareniclina y bupropion) por el régimen de la Seguridad Social. Pero la llegada en marzo de 2020 de la pandemia por enfermedad por SARS-CoV-2 (COVID-19), con el confinamiento domiciliario y la ansiedad y el estrés provocados, supuso un cambio de escenario. Pese a que *a priori* podríamos pensar que en época de COVID-19 no sería el momento ideal para dejar de fumar, en nuestra experiencia existen datos que muestran lo contrario.

Analizamos la abstinencia durante el confinamiento de los pacientes valorados en nuestra unidad de tabaquismo que habían comenzado un intento de dejar de fumar desde el 1 de enero de 2020 hasta la declaración del estado de alarma el 13 de marzo. Se trataba de 100 pacientes, el 46% varones, con una edad media de 59 ± 9 años, un 35% padecía hipertensión arterial (HTA), un 8% diabetes y un 26% dislipemia. Como antecedentes respiratorios, un 42% enfermedad pulmonar obstructiva crónica (EPOC) y un 20% apnea obstructiva del sueño (AOS). A todos se les prescribió tratamiento con vareniclina. Respecto al tabaquismo, puntuaron 8 ± 2 puntos en la escala analógica visual de motivación, 6 ± 2 puntos en el test de Fagerström y cooximetría de 16 ± 12 ppm. Edad media de inicio de tabaquismo de 17 ± 5 años, consumo medio de 18 ± 8 cigarrillos al día y acumulado de 43 ± 18 a/p. El seguimiento en la unidad consiste en unas 6-7 visitas hasta cumplir un año de abstinencia, siendo la primera a las 2-4 semanas del día D. Con la llegada del confinamiento y la suspensión de las visitas presenciales, se realizaron las visitas previstas de seguimiento de manera telefónica por el neumólogo responsable de la unidad, resolviendo dudas y prescribiendo medicación de forma electrónica. Se preguntó acerca de la abstinencia al mes, 3 y 6 meses, y durante el periodo de confinamiento (del 14 de marzo al 21 de junio de 2020). Un 56% de los pacientes se declararon abstinentes durante el confinamiento. Si analizamos la abstinencia por meses, obtuvimos un 67% de abstinencia el primer mes, un 52% a los 3 meses y un 47% a los 6 meses. De los que no dejaron de fumar, un 12% manifestó haber reducido a más de la mitad el número de cigarrillos consumido.

Estudios previos han mostrado un porcentaje de éxito de abandono del tabaquismo de un 25-35% los primeros 6 meses y hasta el primer año, mediante la combinación de tratamiento farmacológico, apoyo psicológico y supervisión especializada[1,2]. Pensamos que varios factores pueden contribuir al éxito obtenido en nuestro caso, a pesar del confinamiento y la situación generada.

En primer lugar, el tabaquismo ha mostrado ser un factor de riesgo para la evolución grave de la COVID-19, multiplicando por 2 la posibilidad de evolución grave de la enfermedad[3–7]. Una mayor preocupación por la salud y una mayor percepción del riesgo de enfermar se asocia con una mayor motivación para dejar el tabaco y un mayor número de intentos exitosos[8]. Podríamos comparar esta vulnerabilidad con la hospitalización, situación ya descrita previamente como una oportunidad para dejar de fumar[9]. Todo esto pese a que el distanciamiento social, el confinamiento y sus consecuencias sociales y económicas como el desempleo, aumenten la prevalencia de estrés y ansiedad, que son factores que pueden contribuir al aumento del consumo de tabaquismo y a las recaídas[10]. Una encuesta realizada en EE. UU. durante la pandemia en fumadores de puros mostró que, pese a que un 40% declaró haber fumado incluso más cantidad los primeros días, el 76% tenía una mayor percepción del riesgo de complicaciones de la COVID-19. Un 70% mostró predisposición a dejarlo en los siguientes 6 meses y hasta un 46% declaró haber solicitado ayuda para ello[11]. Otro estu-