

# Idiopathic Adult Ductopenia: Case report

Daniel Felipe Cerón-Caicedo,<sup>1</sup>  Jorge Steven Montealegre-Arturo,<sup>2</sup>  Viviana Mayor-Barrera.<sup>3\*</sup> 

## OPEN ACCESS

**Citation:**

Mayor-Barrera V, Montealegre-Arturo JS, Cerón-Caicedo DF. Idiopathic Adult Ductopenia: Case report. Revista. colomb. Gastroenterol. 2025;40(2):196-199.  
<https://doi.org/10.22516/25007440.1204>

<sup>1</sup> Internal Medicine Resident. Universidad Libre Seccional Cali. Cali, Colombia.

<sup>2</sup> General Practitioner. Home Care Physician at Sura. Cali, Colombia.

<sup>3</sup> Internist. Clínica Nueva Comfenalco. Cali, Colombia.

\*Correspondence: Viviana Mayor-Barrera.  
vivimayor66@gmail.com

Received: 29/04/2024  
Accepted: 10/02/2025



## Abstract

**Introduction:** Intrahepatic cholestatic ductomas can be associated with an inflammatory process that leads to the destruction of small or medium caliber biliary canaliculi with their subsequent disappearance. Adult idiopathic ductopenia is a diagnosis of exclusion characterized by the destruction of at least 50% of the portal ducts and requires the exclusion of autoimmune and mechanical causes, among others. **Case Report:** 23-year-old patient with no pathologic history who presented with progressive jaundice and incoercible pruritus. Cholangioresonance was performed, which ruled out mechanical obstruction or images suggestive of primary sclerosing cholangitis. Studies of infectious etiology were negative and serum levels of antibodies associated with destruction of biliary canaliculi were within normal ranges. Given the persistence of jaundice of unclear cause, it was decided to perform a liver biopsy which reported loss of ducts in the portal triads with focal destruction. **Discussion:** After analyzing the requested studies, the presentation and course of the disease, the diagnosis of adult idiopathic ductopenia was considered, and management with ursodeoxycholic acid was started, achieving gradual improvement and a follow-up appointment was made. **Conclusion:** The diagnosis of ductopenia was made after having negative serological studies for infectious/inflammatory causes, through a negative cholangioresonance and a liver biopsy as reported in the literature. Excluding secondary causes of ductopenia.

## Keywords

Bile canaliculi, cholestasis, intrahepatic, biliary tract diseases, adult, jaundice.

## INTRODUCTION

Idiopathic ductopenia (IAD) is a chronic condition classified within the spectrum of intrahepatic cholestatic diseases, characterized by the loss of septal and interlobular bile ductules in adulthood<sup>(1)</sup>. This disorder has the potential to progress to biliary cirrhosis and, in severe cases, may present as acute liver failure<sup>(2)</sup>. Since its first description in 1988 by Ludwig et al., the etiology remains unknown, though an inflammatory process leading to the destruction and eventual disappearance of bile ductules has been proposed. In recent years, few cases of idiopathic adult ductopenia have been reported in global literature, with even fewer in Latin

American medical literature<sup>(3)</sup>. According to reviewed literature, two clinical presentations have been described: a mild, asymptomatic form and a more severe form in which the only effective treatment is liver transplantation. This report presents the case of a young patient from Cali, Colombia, who presented with jaundice and was ultimately diagnosed with ductopenia based on histopathological findings during diagnostic and therapeutic evaluation.

## CASE REPORT

A previously healthy 23-year-old man, with no personal or family history of liver disease, no substance abuse, or

use of herbal medications, sought care at Clínica Nueva Comfenalco in Cali due to a one-week history of progressive jaundice and intractable pruritus. Blood tests revealed the following: total bilirubin (TB): 16.6 mg/dL (0–1 mg/dL), alkaline phosphatase (ALP): 349 U/L (40–129 U/L),  $\gamma$ -glutamyltransferase (GGT): 73 U/L (8–61 U/L), aspartate aminotransferase (AST): 67 U/L (0–38 U/L), alanine aminotransferase (ALT): 120 U/L (0–41 U/L). The patient was screened for viral infections, including human immunodeficiency virus (HIV) and hepatitis A, B, and C, all of which returned negative.

Initial suspicion was primary sclerosing cholangitis, prompting further autoimmune testing. Serological identification samples were taken for autoimmune markers, including anti-smooth muscle antibodies, anti-mitochondrial antibodies, antinuclear antibodies, asthma antibodies, antinuclear antibodies (ANA), anti-mitochondrial antibodies (AMA), anti-microsomal antibodies, liver and kidney (LKM1), all of which yielded negative results. Immunoglobulin levels were measured as follows: immunoglobulin A (IgA): 193 mg/dL (63–484 mg/dL), immunoglobulin M (IgM): 85 mg/dL (22–240 mg/dL), immunoglobulin G (IgG): 978 mg/dL (540–1822 mg/dL). Further subclass quantification revealed: IgG1: 1.31 g/L, IgG2: 0.47 g/L, IgG3: 0.1 g/L e IgG4: 0.24 g/L. Protein electrophoresis showed an alpha-1 and alpha-2 peak with a decrease in gamma A.

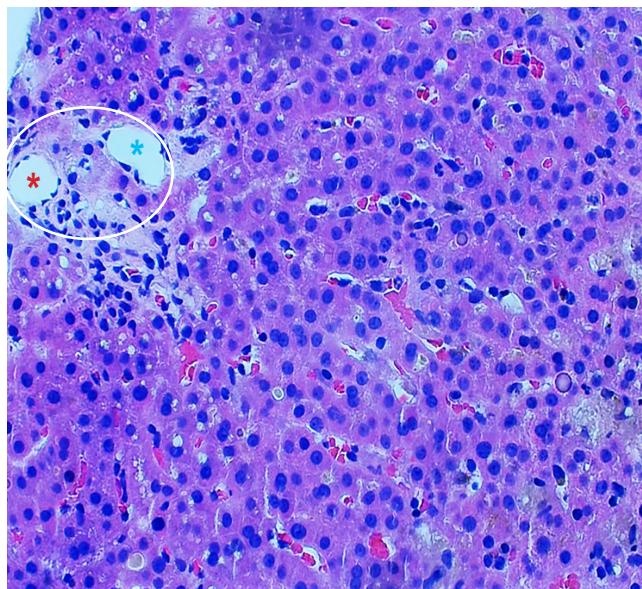
Leptospira infection was ruled out (negative IgM), and blood smear testing excluded hemoparasite-related diseases. Endoscopic studies showed no significant findings except for antral hyperemic gastritis. An initial abdominal ultrasound detected hepatomegaly without other abnormalities. Subsequent magnetic resonance cholangiography ruled out choledocholithiasis or biliary duct dilation. Finally, a percutaneous liver biopsy revealed marked cholestasis and loss of bile ducts in portal triads. Immunohistochemical staining confirmed focal duct destruction in portal triads (**Figures 1, 2, and 3**).

The patient was diagnosed with idiopathic adulthood ductopenia. Initial management with corticosteroids was initiated, achieving slow progressive reduction of bilirubin levels. For symptomatic management, cholestyramine 4 g was administered orally every 12 hours without achieving symptom control. Subsequently, ursodeoxycholic acid 300 mg was added orally every 12 hours, resulting in notable improvement.

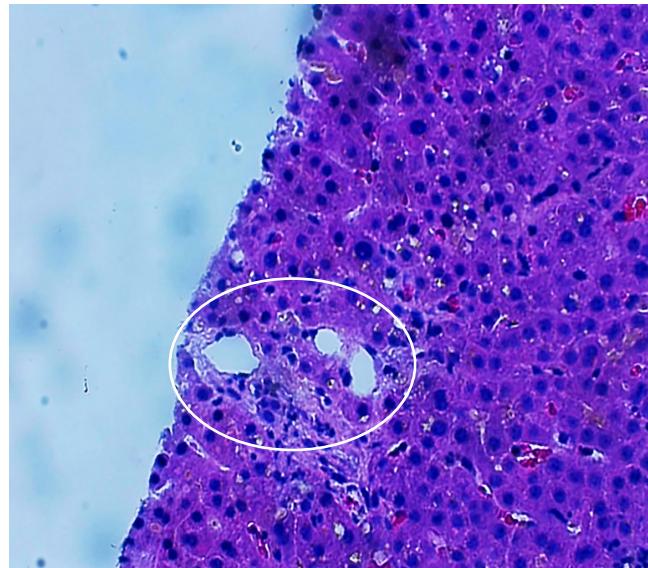
The patient was discharged with minimal residual symptoms, with plans for continued hepatology evaluation. However, due to clinical improvement, the patient voluntarily declined further testing.

## DISCUSSION

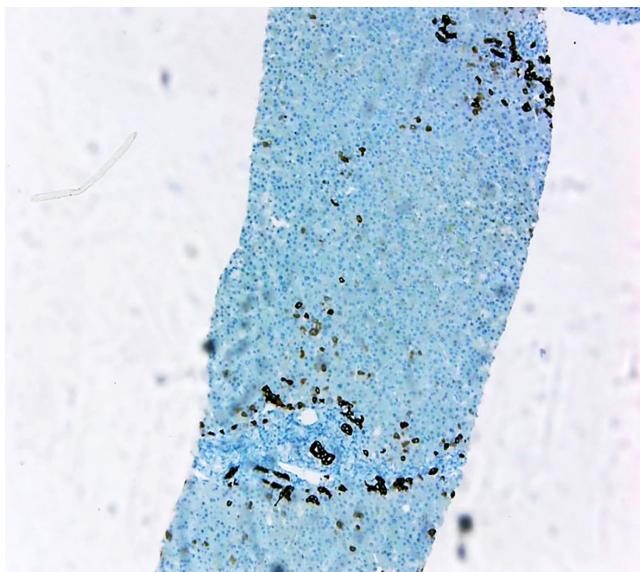
Primary biliary cholangitis (PBC) and primary sclerosing cholangitis (PSC) represent the most common causes of



**Figure 1.** The histological section shows hepatic parenchyma with a portal space (circle), where portal vessels (artery and vein) are identified but no bile duct is observed (\*red: artery, \*blue: vein). Image property of the authors.



**Figure 2.** The section shows hepatic parenchyma with a portal space, where a normal portal triad was identified containing portal vessels (artery, vein) and bile duct. Image property of the authors.



**Figure 3.** Immunohistochemical study with CK7 confirming the presence of ducts and adjacent hepatocytes with biliary metaplasia, demonstrating a cholangitic injury pattern. Image property of the authors.

chronic hepatic cholestasis in adults<sup>(4)</sup>. However, a small patient subset presents with chronic cholestasis lacking antimitochondrial antibodies (suggestive of PBC) or cholangiographic abnormalities (suggestive of PSC). These cases are frequently classified as AMA-negative PBC, small-duct PSC, or idiopathic adulthood ductopenia (IAD)<sup>(5)</sup>.

IAD describes chronic hepatic cholestasis of unclear etiology, typically beginning in adulthood and associated with intrahepatic bile duct loss. Multiple pathogenic mechanisms for intrahepatic biliary destruction have been implicated, including biliary atresia development, immunological causes from aberrant MHC class II antigen expression, infections, ischemia, or chemical injury<sup>(1-3,6)</sup>. Several medications (chlorpromazine, prochlorperazine, organic arsenicals, tolbutamide) have also been associated with bile duct destruction. These were ruled out in our patient, as their identification would exclude IAD diagnosis.

Current diagnostic criteria require: biochemical cholestasis, normal endoscopic studies excluding inflammatory bowel disease, absent antimitochondrial antibodies, no granulomatous cholangitis, absent histiocytosis X, no suppurative neutrophilic cholangitis, no lymphoma or neoplasia on liver biopsy, and histopathological confirmation of ductopenia. The latter requires loss of  $\geq 50\%$  of septal/interlobular bile ducts in a sample containing  $\geq 20$  portal

tracts, without granulomatous cholangitis or florid ductal injury<sup>(1,2,6-9)</sup> (**Figures 1 and 3**).

Clinical manifestations vary. Some patients remain asymptomatic, though the primary symptom is jaundice with typically intense pruritus. Biochemical analysis reveals a predominantly cholestatic pattern with direct hyperbilirubinemia and elevated alkaline phosphatase/ $\gamma$ -glutamyltransferase<sup>(1,10)</sup>.

Literature describes two IAD presentations: Type 1 (often asymptomatic, may show chronic cholestasis symptoms, with less duct destruction on biopsy) and Type 2 (more severe, potentially progressing to biliary cirrhosis with extensive duct destruction histologically, where liver transplantation remains the only proven effective treatment)<sup>(2,8,10,11)</sup>.

For mild cases, ursodeoxycholic acid has demonstrated symptomatic benefit in multiple reports, though without documented disease progression impact. Clinical outcomes depend on ductopenia severity. Reviewed literature agrees that IAD typically progresses to end-stage liver disease when  $>50\%$  duct destruction exists, while lesser involvement usually follows a benign course<sup>(2,6,10)</sup>. Our patient showed  $<50\%$  duct abnormality on biopsy with progressive jaundice improvement. Determining whether his course will remain benign remains unclear due to potential recent diagnosis and loss to follow-up.

## CONCLUSION

Idiopathic adulthood ductopenia is a rare cholestatic liver disease diagnosed by excluding differential diagnoses while demonstrating biochemical cholestasis and biopsy-proven bile duct loss. While recognizing this entity is important, first excluding more common pathologies remains vital for survival. When biochemical-histopathological diagnosis is confirmed, prompt management of both cause and symptoms (when possible) impacts patient survival and quality of life.

## Acknowledgments

The research team.

## Conflicts of Interest

None reported.

## Funding Source

None.

## REFERENCES

---

1. Ludwig J. Idiopathic adulthood ductopenia: an update. *Mayo Clin Proc.* 1998;73(3):285-91.  
<https://doi.org/10.4065/73.3.285>
2. Khanlou H, Sass D, Rothstein K, Manzarbeitia C, Reich D, Jacobson L, et al. Idiopathic adulthood ductopenia: case report and review of the literature. *Arch Intern Med.* 2000;160(7):1033-6.  
<https://doi.org/10.1001/ARCHINTE.160.7.1033>
3. Timoniuk CI, Acosta M, Vizcaino AA, Panzardi MY. Ductopenia idiopática del adulto. Servicio de Cirugía Del Hospital "Dr. José R. Vidal". *Revista de Posgrado de la Vía Cátedra de Medicina.* 2008;180:7-11.
4. Kaplan MM. Primary biliary cirrhosis. *N Engl J Med.* 1996;335(21):1570-80.  
<https://doi.org/10.1056/NEJM199611213352107>
5. Kim WR, Ludwig J, Lindor KD. Variant forms of cholestatic diseases involving small bile ducts in adults. *Am J Gastroenterol.* 2000;95(5):1130-8.  
<https://doi.org/10.1111/j.1572-0241.2000.01999.x>
6. Burak KW, Pearson DC, Swain MG, Kelly J, Urbanski SJ, Bridges RJ. Familial idiopathic adulthood ductopenia: a report of five cases in three generations. *J Hepatol.* 2000;32(1):159-63.  
[https://doi.org/10.1016/S0168-8278\(00\)80203-2](https://doi.org/10.1016/S0168-8278(00)80203-2)
7. Douglass KM, Willner IR, Glenn DJ, Jones RM. Idiopathic Adulthood Ductopenia Causing Cirrhosis. *ACG Case Rep J.* 2020;7(3):e00349.  
<https://doi.org/10.14309/CRJ.00000000000000349>
8. Park BC, Park SM, Choi EY, Chae HB, Yoon SJ, Sung R, et al. A case of idiopathic adulthood ductopenia. *Korean J Intern Med.* 2009;24(3):270-3.  
<https://doi.org/10.3904/KJIM.2009.24.3.270>
9. Peniche-Moquel P, Pérez-Hernández JL, Cordova J. Chronic cholestasis in a 20-year-old young man: a case of idiopathic adulthood ductopenia. *Gastroenterol Hepatol.* 2013;36(10):628-30.  
<https://doi.org/10.1016/J.GASTROHEP.2013.07.003>
10. Rios R, Herrero JI, Quiroga J, Sangro B, Sola I, Pardo F, et al. Idiopathic adulthood ductopenia: long-term follow-up after liver transplantation. *Dig Dis Sci.* 2001;46(7):1420-3.  
<https://doi.org/10.1023/A:1010679502324>
11. Ludwig J, Wiesner RH, LaRusso NF. Idiopathic adulthood ductopenia. A cause of chronic cholestatic liver disease and biliary cirrhosis. *J Hepatol.* 1988;7(2):193-9.  
[https://doi.org/10.1016/S0168-8278\(88\)80482-3](https://doi.org/10.1016/S0168-8278(88)80482-3)