

# Lessons from Clinical Cases in (Rare) Vascular Liver Diseases

## Ensinaamentos a Tirar de Casos Clínicos em Doenças (Raras) Vasculares do Fígado

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**Palavras-chave:** Doenças do Fígado; Doenças Vasculares; Síndrome de Budd-Chiari.

Vascular liver diseases constitute one of the most complex and diagnostically challenging areas in hepatology. These disorders encompass a heterogeneous spectrum of conditions affecting hepatic vascular inflow, intrahepatic microcirculation, or venous outflow, often resulting in portal hypertension, hepatic dysfunction, and systemic complications. Although individually uncommon, their collective impact on clinical hepatology is significant and continues to expand as diagnostic tools and conceptual frameworks evolve.<sup>1</sup>

In recent years, advances in imaging, pathology, and clinical research have reshaped our understanding of hepatic vascular diseases. Updated international guidelines now emphasize the need to approach these diseases within a unified framework that integrates clinical presentation, vascular imaging, histopathology, genetics, and hemodynamic evaluation.<sup>1</sup> Within this spectrum, entities such as portal vein thrombosis (PVT), Budd–Chiari syndrome (BCS), porto-sinusoidal vascular disorder (PSVD), and congenital portosystemic shunts, among others, represent key clinical challenges requiring multidisciplinary expertise.

Among these conditions, PVT has gained particular attention due to its increasing recognition across different clinical settings, including cirrhotic and non-cirrhotic patients. The complexity of its pathophysiology, diagnostic evaluation, and therapeutic management continues to stimulate international collaborative research efforts aimed at refining classification systems and clinical trial endpoints.<sup>2</sup> Similarly, BCS remains a paradigmatic example of hepatic venous outflow obstruction requiring timely diagnosis and a structured, stepwise therapeutic approach.<sup>3</sup>

Case reports have historically played a central role in advancing knowledge in rare and complex diseases. In vascular liver disorders, where clinical presentations are frequently heterogeneous and pathophysiological mechanisms remain incompletely understood, individual clinical observations often provide critical insights that later shape broader clinical understanding. For this reason, case-based contributions remain particularly valuable in this field, complementing large cohort studies and guideline-driven clinical practice.

The present special issue of SPMI Case Reports, dedicated to vascular liver diseases, brings together a collection of illustrative clinical reports contributed by clinicians and researchers with recognized expertise in hepatology and vascular liver diseases. These cases highlight the remarkable diversity of clinical presentations and emphasize the importance of careful diagnostic reasoning and multidisciplinary management, serving as basis of an international course on this subject promoted by the COST Action EuroValdiNet 23146.

The clinical section of this issue includes reports spanning acute vascular emergencies, chronic vascular diseases, and complex systemic conditions with hepatic vascular involvement. Some cases illustrate unusual presentations or distinct diagnostic and therapeutic approaches, emphasizing the need for a comprehensive and multidisciplinary discussion involving clinicians, hepatologists, internists, pathologists, radiologists, interventional radiologists, hematologists, pediatricians, cardiologists, and pulmonologists.

Complementing these clinical reports, this special issue includes two mini-atlas sections dedicated respectively to the histopathological and radiological aspects of vascular liver diseases. Histological evaluation remains crucial for diagnosing entities such as PSVD, where subtle portal and sinusoidal abnormalities may represent the key diagnostic clue, as well as other conditions such as sinusoidal obstruction syndrome or ischemic cholangiopathy. Similarly, radiological imaging plays an essential role in identifying vascular abnormalities and guiding clinical decision-making. Doppler ultrasound, computed tomography, and magnetic resonance imaging allow increasingly detailed visualization of hepatic vascular architecture and hemodynamics, aiding in the diagnosis of PVT, BCS, and in distinguishing subtle differences between liver cirrhosis and PSVD, for example. The radiology mini-atlas included in this issue provides a visual overview of characteristic imaging patterns that clinicians

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may encounter in daily practice. Both mini-atlases may serve not only as quick reference tools but also as valuable didactic resources for educational settings, including lectures and teaching sessions.

Beyond its scientific contributions, this special issue also highlights the intersection between science and artistic interpretation. The cover artwork for this edition was created by Catarina V. Nery, a Belgian artist of Portuguese descent, whose work visually evokes the intricate branching architecture of the hepatic vascular network through the artist's perspective. The image symbolically reflects the complexity and elegance of hepatic vascular physiology, reminding us that disturbances within this delicate vascular system may profoundly alter liver function and systemic homeostasis.

Taken together, the contributions presented in this special issue illustrate the remarkable heterogeneity of vascular liver diseases and reinforce the importance of multidisciplinary collaboration in their diagnosis and management, as well as the central role of Internal Medicine in the integrated care of these patients. As research in hepatic vascular biology continues to evolve, further collaborative efforts will be essential to refine disease classification, identify therapeutic targets, and improve patient outcomes.

The scope of this special number of SPMI Case Reports, with this collection of clinical cases and illustrative atlases, is to provide readers with practical clinical insights while stimulating further interest and research in the fascinating and rapidly evolving field of vascular liver diseases. ■

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