

Vascular pathology in patients with Alveolar Echinococcosis: framework for assessment and clinical management



HEIDELBERG
UNIVERSITY
HOSPITAL

Paula Gieser¹, Uta Merle², Thomas Junghanss¹, Tim F Weber³, Marija Stojković¹

¹Infectious and Tropical Diseases, University Hospital Heidelberg, Heidelberg, Germany, ²Department of Internal Medicine IV, University Hospital Heidelberg, Heidelberg, Germany,

³Department of Diagnostic and Interventional Radiology, University Hospital Heidelberg, Heidelberg, Germany

Introduction

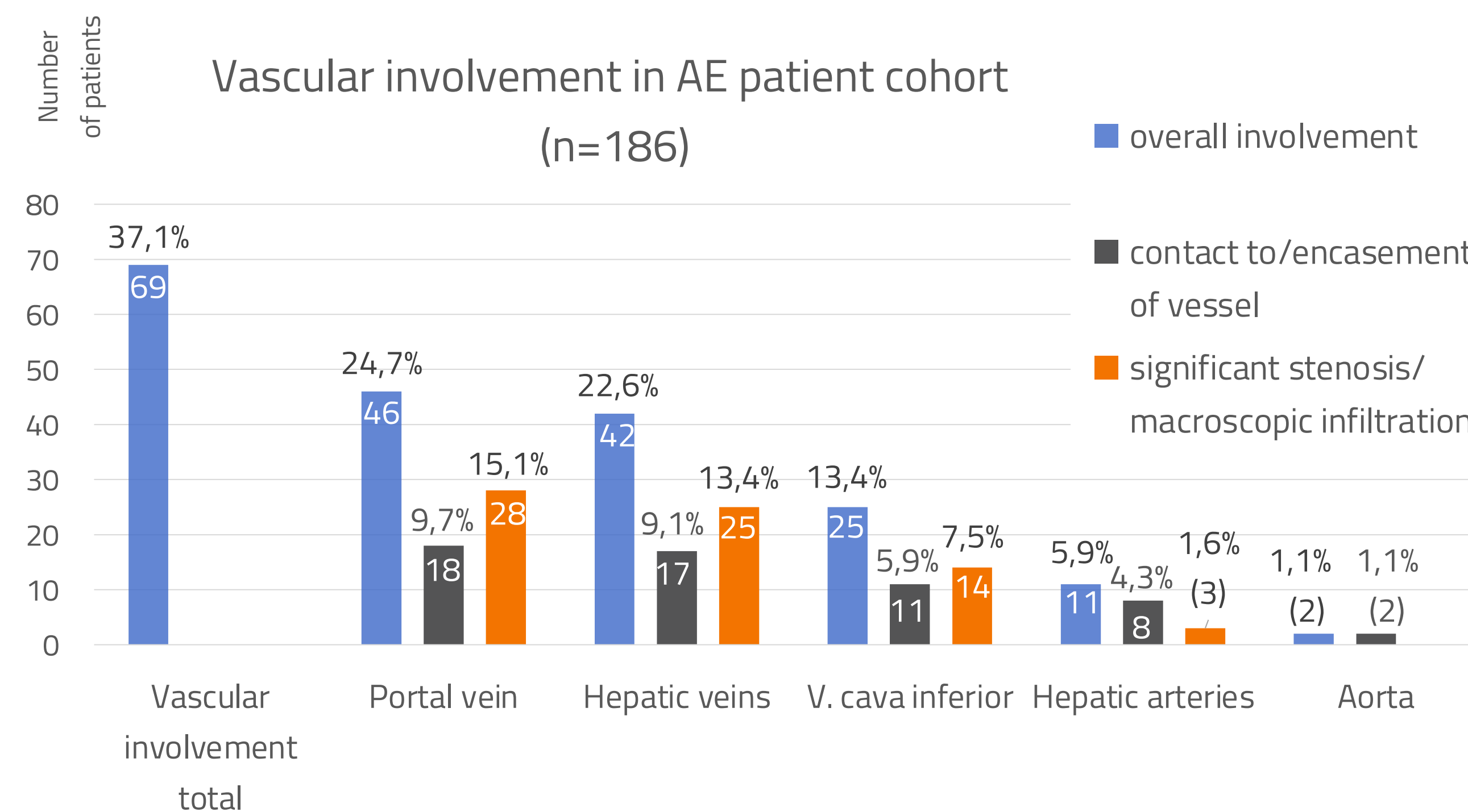
Alveolar echinococcosis (AE) is a rare parasitic liver disease, exhibiting all features of solid hepatic malignancies. Vascular impairment is a known complication in AE patients but often remains unnoticed until catastrophic events precipitate.

Infiltration of splanchnic vessels is common but not systematically evaluated. Our aim is to describe vascular pathology in alveolar echinococcosis and to raise attention for vascular complication. We propose a framework for structured evaluation as the basis for treatment decisions and monitoring.

Materials & Methods

- Retrospective case series
- National and international clinical reference center for echinococcosis at University Hospital Heidelberg.
- Cohort: patients with confirmed alveolar echinococcosis and at least one follow-up with primary presentation between April 2000 and October 2021
- Extraction of clinical data and subdivision into 3 groups (A: no vascular involvement, B: contact to or encasement of vessels, C: significant stenosis or macroscopic infiltration of vessels)
- Description of vascular involvement by mapping course of disease (clinically and radiologically) and interventions
- Analysis of vascular baseline and follow-up examinations in relation to vascular complications
- Development of multidisciplinary framework to identify, monitor and treat vascular complication based on long center-based experience

Results

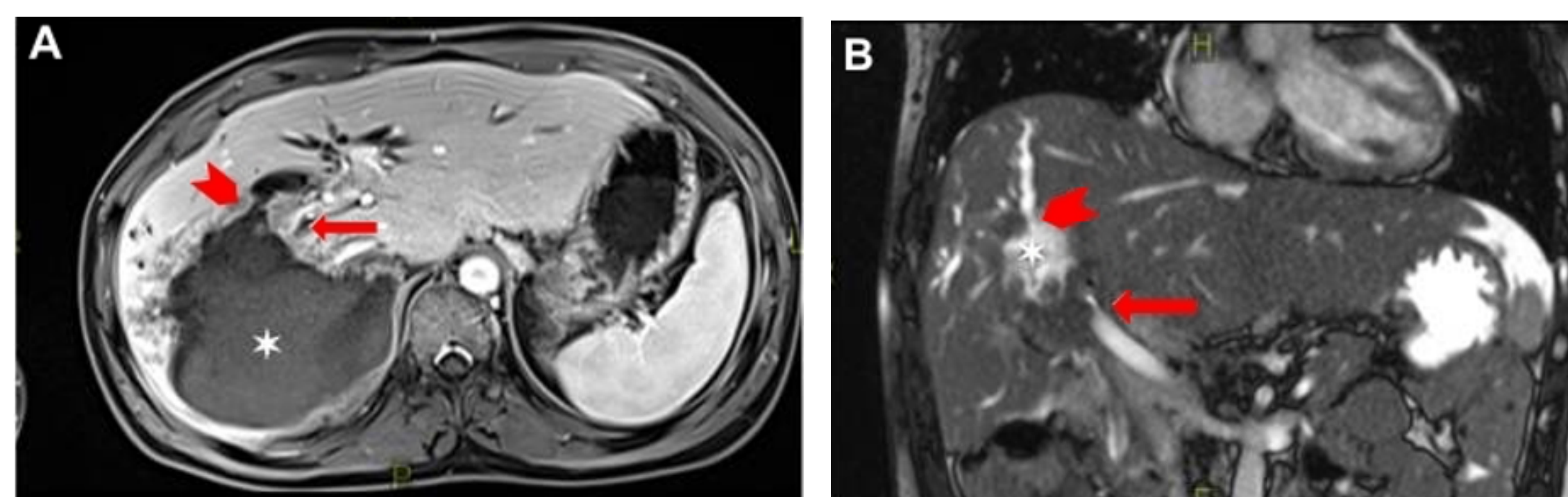


Exemplary cases & Radiological presentation

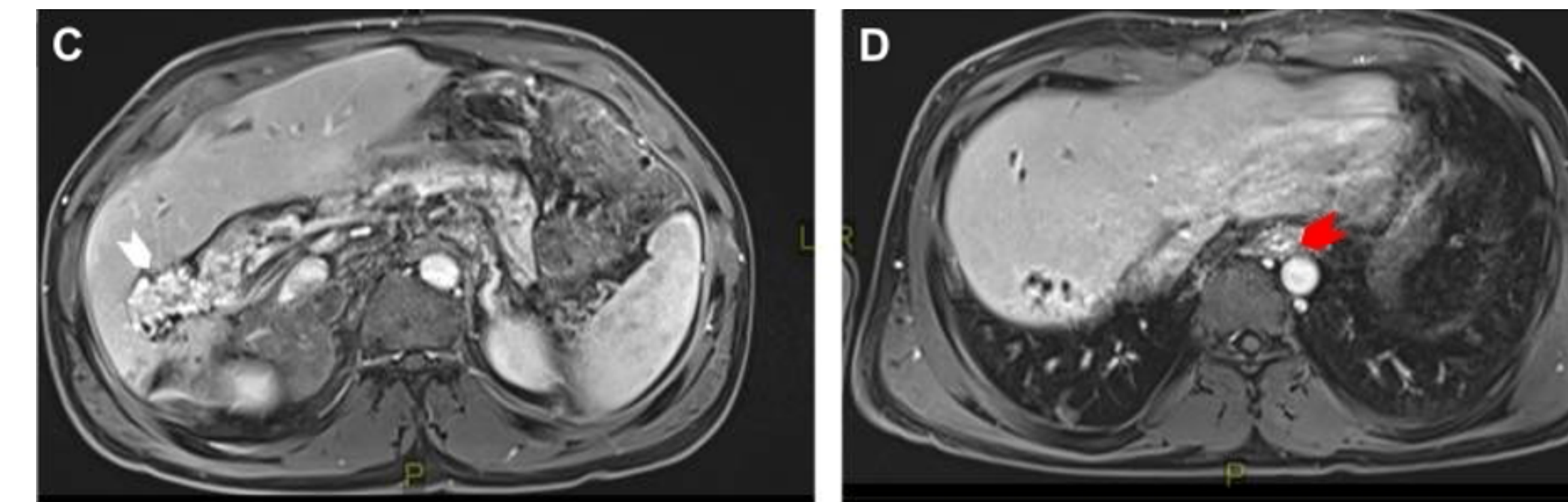
I. Significant portal vein stenosis

29-y-o male patient with alveolar echinococcosis P3 N1 M1

⚠️ Complications	⊕ Clinical management
• Cholestasis and cholangitis	→ ERCP and stenting of biliary duct → antibiotic prophylaxis → Albendazole 2x400mg
• Portal vein occlusion with oesophageal varices III° and acute variceal bleeding	→ Beta-blocker (NSBB) Carvedilol → Endoscopic band ligation of varices → Follow-up intervals: 3, then 6 months → Anticoagulation Apixaban 2x2,5mg



Baseline-MRI: ★ hypovascular, necrotic Alveolar echinococcosis lesion in right hemiliver, → communication and upstream dilation of bile ducts, ← subtotal portal vein occlusion.

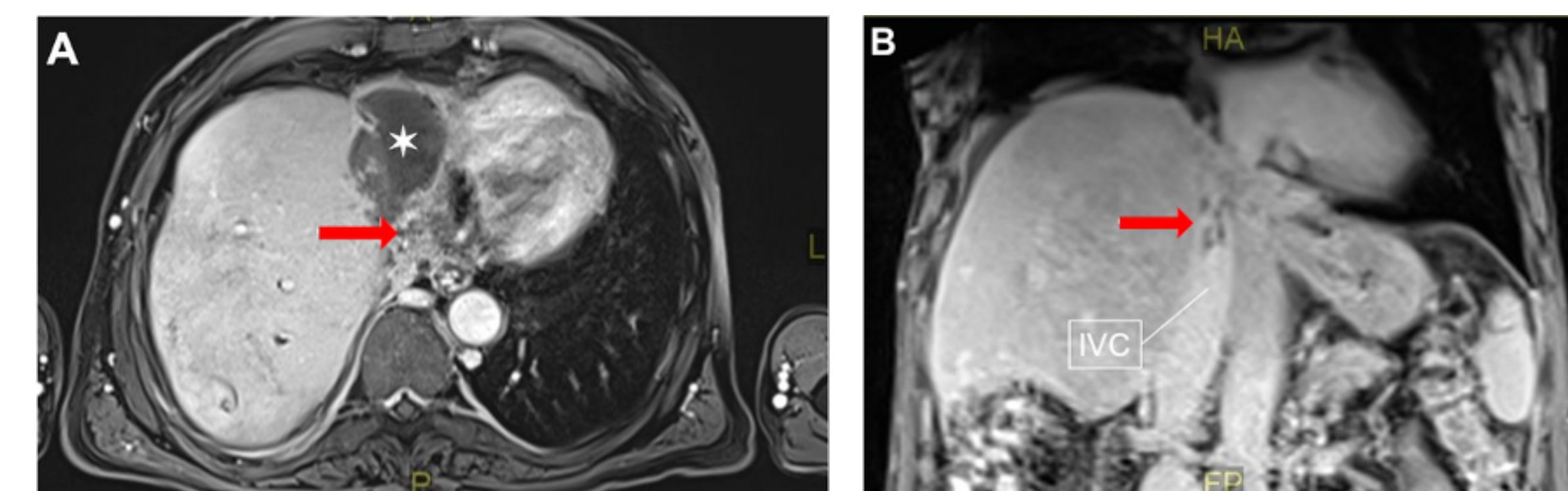


Follow-up MRI shows cavernous transformation (→) and oesophageal varices (→).

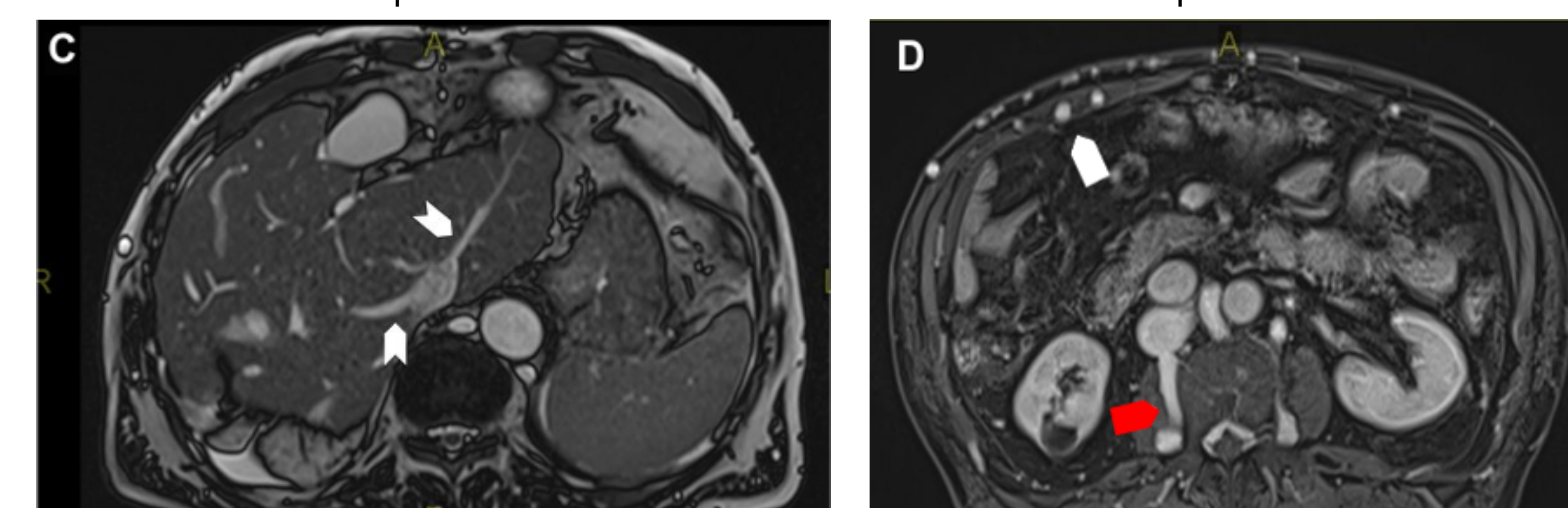
II. Significant inferior vena cava occlusion

69-y-o male patient with alveolar echinococcosis P4 N1 M1

⚠️ Complications	⊕ Clinical management
• Alveolar echinococcosis mistaken for malignancy • Albendazole toxicity with myelosuppression	→ Drainage of necrotic cavity with histopathological diagnosis AE → Albendazole at reduced dose 2x200mg in therapeutic range → Evaluation for Budd-Chiari-syndrome → Diuretics → Moderate cardiovascular practice → Anticoagulation Rivaroxaban 2x2,5mg
• Occlusion of hepatic vena cava segment with ascites, peripheral oedema and reduced fitness	



Baseline-MRI: ★ large AE lesion in left hemiliver. → Complete obstruction of vena cava at the level of the hepatic vein orifices with evidence of macroscopic vascular invasion.



Alternative venous drainage over accessory hepatic veins (→) into subhepatic vena cava segment. Venous collaterals via *Azygous system* (→) and veins of abdominal wall (→).

Conclusion & Recommendations

Radiology

- First approach: COMPLETE EVALUATION of VASCULAR INVOLVEMENT at PRIMARY PRESENTATION using STRUCTURED REPORTING TEMPLATE*.
- Monitor patients by evaluating hepatic vasculature and vena cava inferior by colour-coded duplex ultrasonography at 3-6 months intervals.



Vascular pathology identified? → Complete formal evaluation and start treatment, prophylaxis or monitoring.

Gastro-enterology

- V. cava or portal vein obstruction? → Evaluate for oesophageal varices and treat according to grade.
- V. cava or hepatic vein obstruction? → Evaluate for Budd-Chiari-syndrome and discuss interventional approach.

Infectious Diseases

- Benzimidazoles should be started as soon as vascular and/or biliary complications are treated and liver function is stable (ideally normal ALAT).
- Patients with vascular and/or biliary complications may not tolerate full benzimidazole dose.
- Start benzimidazole treatment with low dose and under close monitoring for drug toxicity. Increase dose stepwise.

Multidisciplinary workup

- Discuss indication for anticoagulation weighing benefit against risk of bleeding.
- Evaluate surgical treatment options to prevent long-term complications of vascular pathology.
- Consider LTx listing if liver function deteriorates.

References:
Gieser P, Merle U, Junghanss T, Weber TF, Stojković M. **Vascular pathology in patients with alveolar echinococcosis: framework for assessment and clinical management - a retrospective case series.** BMJ Open Gastroenterol. 2023 Aug;10(1):e001181. doi: 10.1136/bmjgast-2023-001181.
*Radiological Reporting template:
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10423801/bin/bmjgast-2023-001181supp001.pdf>

