

Radiographic features of Veno-occlusive disease: What signs should we be looking for in children?

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- Hepatic veno-occlusive disease (VOD), also known as sinusoidal obstruction syndrome is toxic injury of hepatic sinusoidal capillaries which obstructs small hepatic veins and can lead to liver failure.
- It is a complication of hematopoietic stem cell transplantation in those with hematological or solid organ malignancies.
- Mild VOD is treated conservatively and is self-limiting.
- Moderate and severe VOD can be life-threatening; an estimated 30-60% of affected children developing multi-organ dysfunction and liver failure[1], necessitating treatment with defibrotide (anti-fibrotic agent) and supportive therapy, with replacement of blood products and clotting factors. In cases of irreversible liver failure, a liver transplant is required and the mortality rate is high.

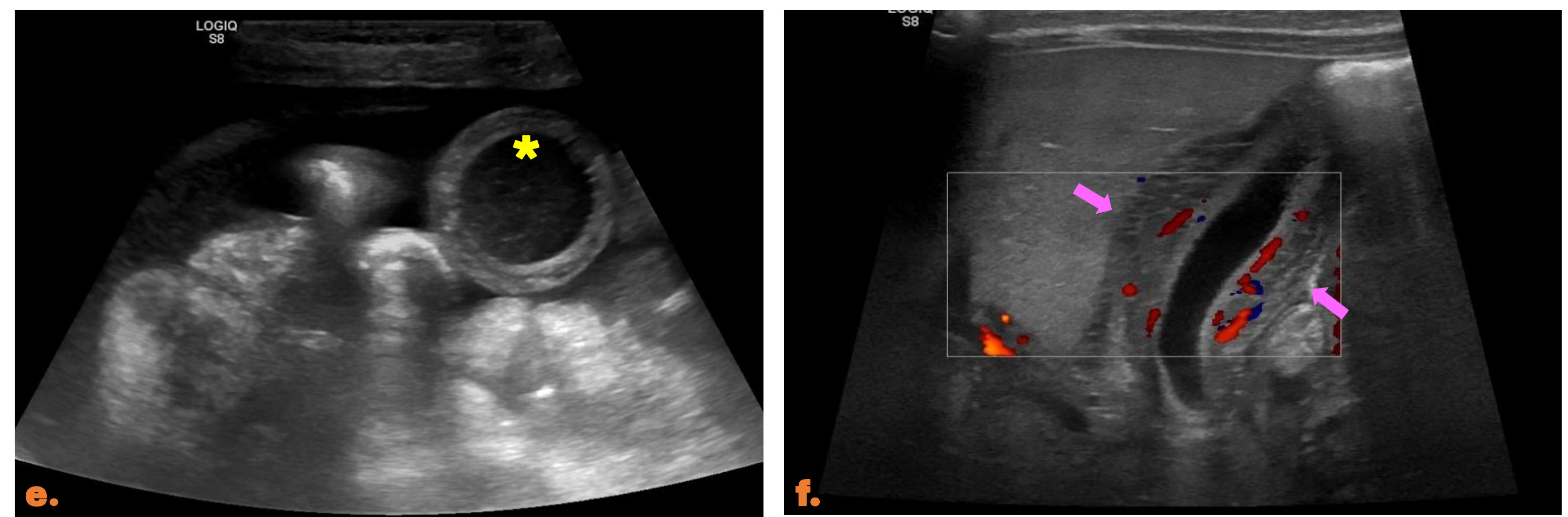
- Patients typically present with
 - Hepatomegaly, hepatic dysfunction or jaundice
 - weight gain secondary to fluid retention
 - ascites [2].
- Ultrasound is useful in **confirming clinical suspicion** of VOD, **assessing severity** and **monitoring treatment**
- This poster aims to demonstrate common ultrasound features seen in patients.

Ultrasound variables and Criteria Measurements

Variable	Lassau's criteria	HokUS-10 scoring
Grey-scale ultrasound		
Hepatomegaly	Liver enlargement: increase in 2 of 3 measurements of >2cm in adults and >1cm in children relative to baseline	Hepatic left lobe vertical diameter ≥70mm (1 point); Hepatic right lobe vertical diameter ≥110mm
Splenomegaly	Spleen enlargement: increase > 1cm relative to baseline measurements in the greatest axis	Long axis increase
Gallbladder wall thickening	>6mm	≥6mm (1 point)
Portal vein diameter	≥8mm in children	≥12mm (1 point)
Ascites	Presence	Mild (1 point); moderate/severe (2points)
Paraumbilical vein	Visualisation	Diameter ≥2mm (2 points)
Colour Doppler ultrasound		
Absence/presence of flow	Flow recorded in paraumbilical vein	Appearance of paraumbilical vein blood flow signal (2points)
Flow direction	Reversed flow in the main portal vein	Congestion of hepatofugal flow in the portal vein (1 point)
Spectral doppler ultrasound		
Portal vein velocity/ density/ congestion	Flow demodulation (disappearance of velocity variations with breathing); decreased spectral density in portal vein; maximal flow in the main portal vein <10cm/s; portal vein congestion index ≥.1; monophasic flow in the hepatic veins	Velocity <10cm/s (1 point)
Hepatic artery resistive index	≥.75	≥.75 (1 point)

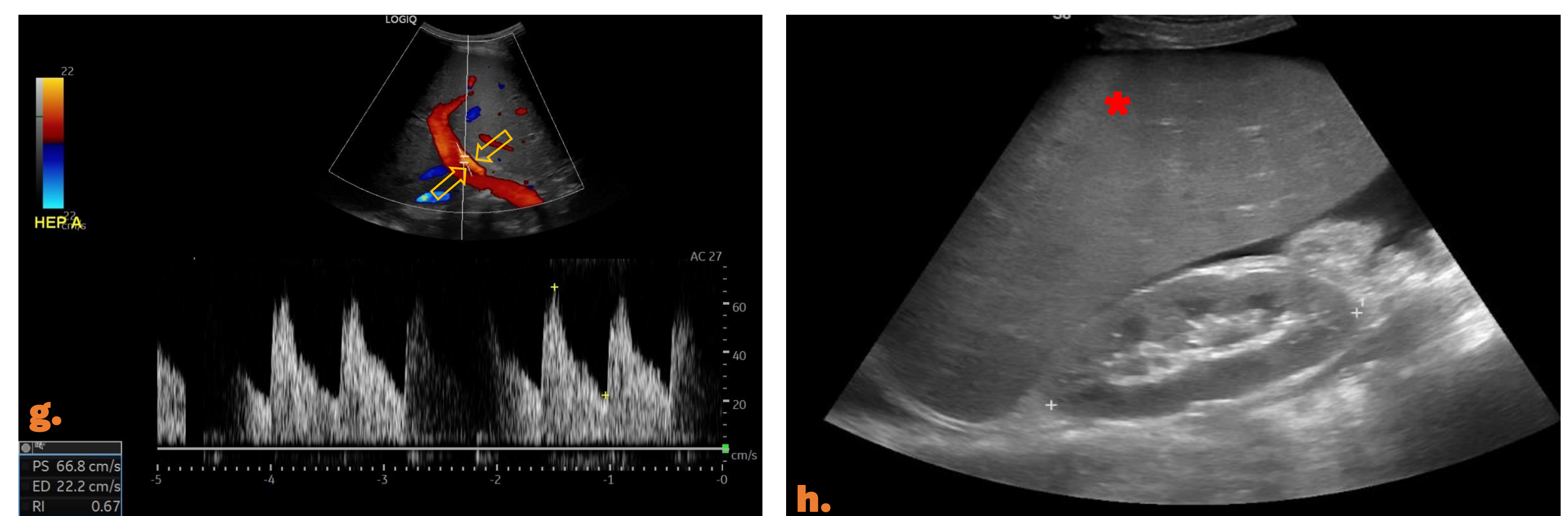
Table 1. Parameters which are significantly associated with a diagnosis of VOD [3].

Gallbladder



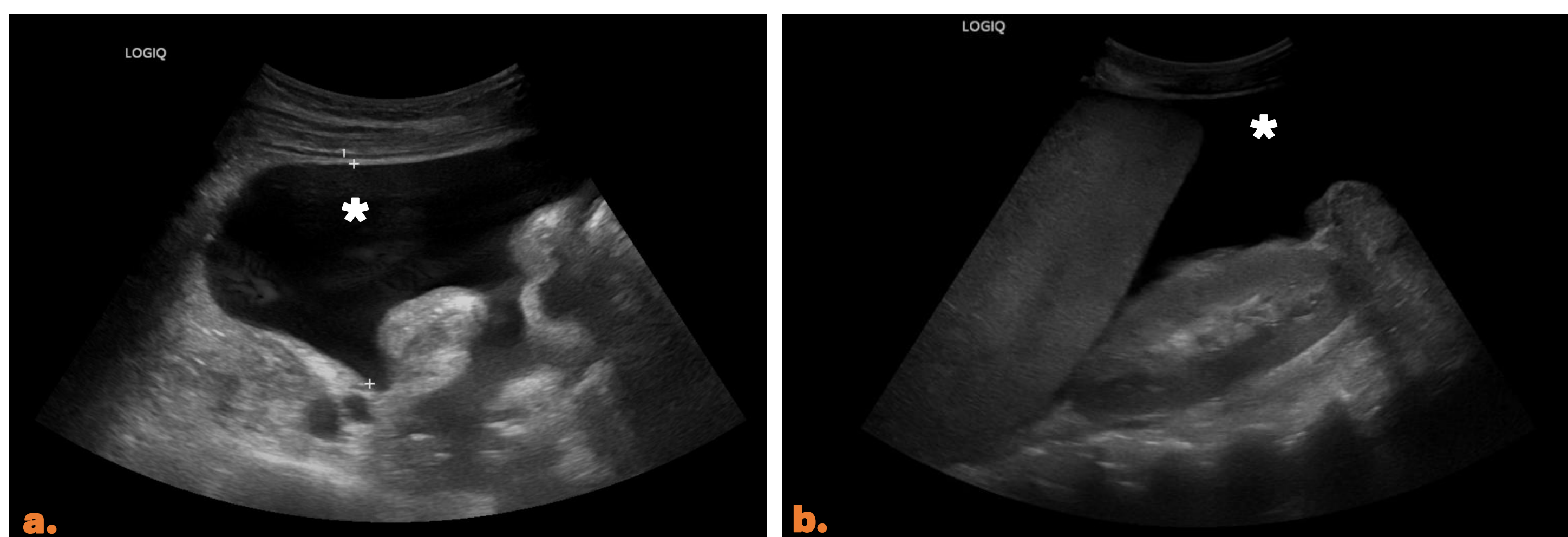
In patients developing or with VOD the gallbladder will become thickened and oedematous and should measure >6mm [3]. (e) transverse gallbladder with biliary sludge (*), thickened wall and surrounding ascites. (f) Oedematous gallbladder with increased wall vascularity (pink arrows).

Liver



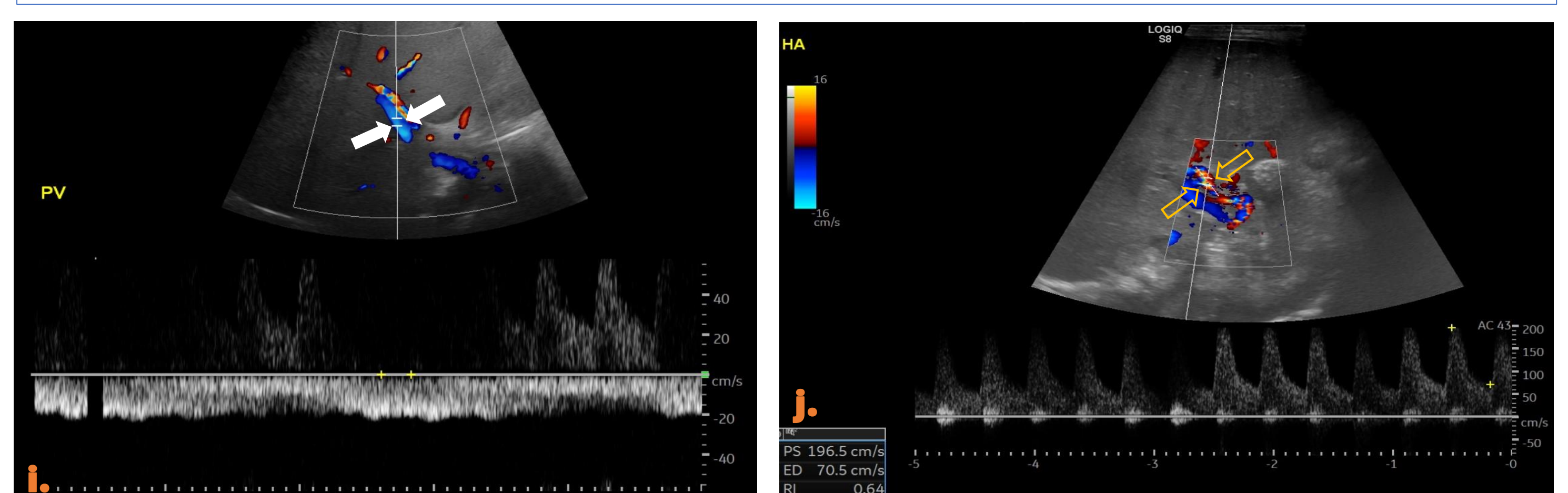
Sinusoidal endothelial cells in the liver may become obstructed post HSCT or chemotherapy which can lead to VOD. (g) Hepatopetal directional flow in the portal vein with increased hepatic artery resistive index (orange arrows). (h) Hepatomegaly (*) is a common sign for VOD, this is best confirmed on imaging.

Ascites

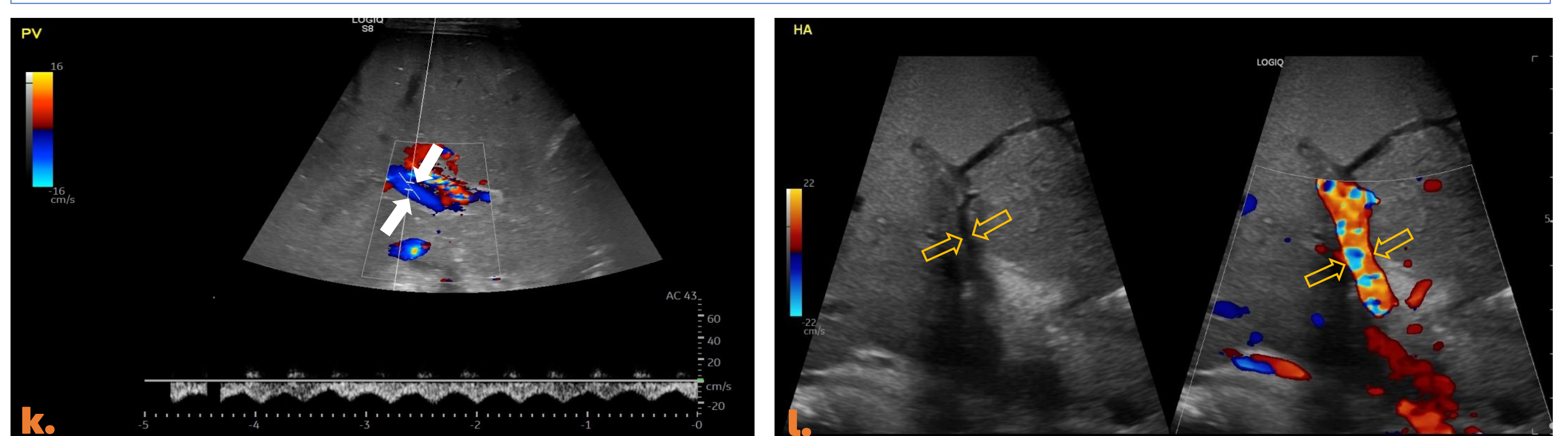


(a,b) Ascites (*) which is best confirmed on imaging. The presence of ascites alone does not confirm VOD, the diagnostic criteria requires ≥2 of the following:

- Unexplained consumptive and transfusion-refractory thrombocytopenia
- Otherwise unexplained weight gain on 3 consecutive days despite diuretics
- Hepatomegaly above baseline value
- Ascites above baseline value
- Rising bilirubin from a baseline value on 3 consecutive days or bilirubin ≥2mg/dL within 72 hours [3]

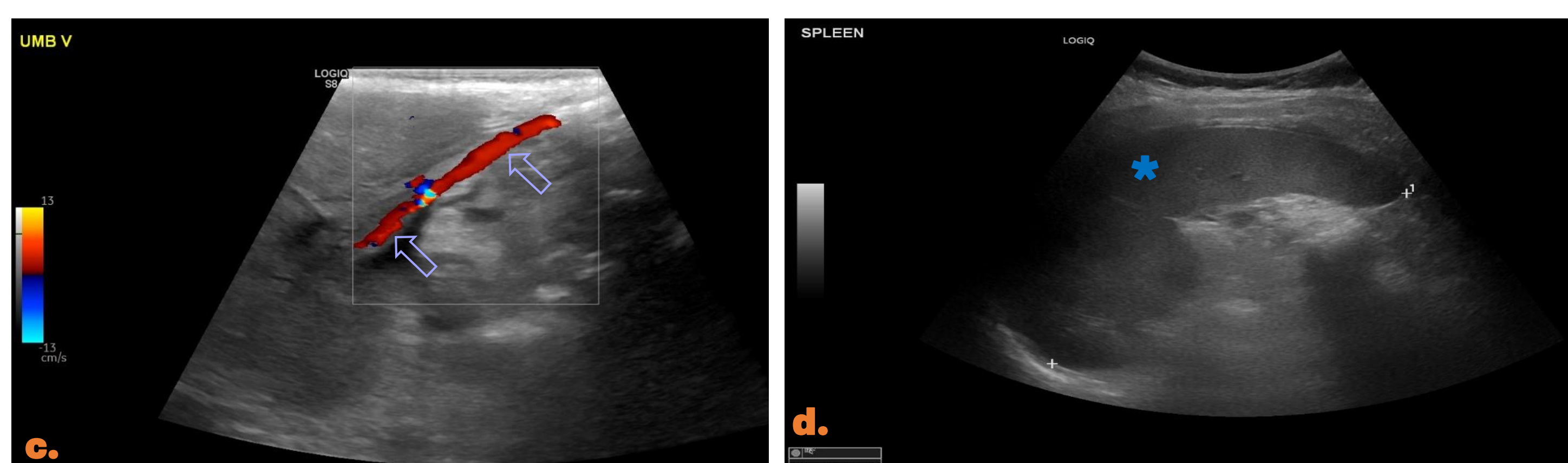


(i) Hepatofugal (reversed) portal venous flow (white arrows) with aliasing hepatic artery and (j) increased resistive waveform in the hepatic artery (orange arrows).



(k) pulsatile hepatofugal flow in the portal vein (white arrows). (l) mild prominence of the hepatic artery with aliasing flow (orange arrows).

Other abdominal appearances



(c) Recanalisation of the umbilical vein (purple arrows) at the falciform ligament. (d) Splenomegaly (*)

Conclusion

VOD is a serious condition that can be associated with substantial morbidity and mortality. As ultrasound is generally well tolerated by paediatric patients, the use of imaging can help predict the risk of VOD and assist in confirming a diagnosis and monitor progression before and after treatment. Early detection of VOD and prompt treatment are critical for optimizing patient management.

References:
1. Corbacioglu S, Carreras E, Ansari M, et al. Diagnosis and severity criteria for sinusoidal obstruction syndrome/veno-occlusive disease in pediatric patients: a new classification from the European society for blood and marrow transplantation. Bone Marrow Transplant. 2018;53:138–145. doi: 10.1038/bmt.2017.161.
2. Bohte A, Dierselhuys M, Van Noesel M, Lequin M. Imaging features of hepatic sinusoidal obstruction syndrome or veno-occlusive disease in children. Paediatric Radiology. 2022; 52(1): 122-133. doi: 10.1007/s00247-021-05174-w
3. Chan, S.S. et al. (2020) 'Imaging in hepatic veno-occlusive disease/sinusoidal obstruction syndrome', Biology of Blood and Marrow Transplantation, 26(10), pp. 1770–1779. doi:10.1016/j.bbmt.2020.06.016.