Screening for portal system anomalies: a 3-step protocol

Objective
To evaluate a 3-step screening protocol to detect fetal portal system anomalies.

Methods
A 5-year retrospective analysis of all the portal system anomalies were evaluated from stored images at our tertiary fetal medicine service using the following process: Step 1: confirm a normal gall bladder to umbilical vein relationship Step 2: confirm the presence of the DV Step 3: confirm normal direction of blood flow in the confluence of the main portal vein and portal sinus using colour Doppler. Only cases of isolated portal system anomalies and where satisfactory images of the 3 steps were available. All portal system anomalies were classified as described shown by Achiron et al. (Ultrasound Obstet Gynecol. 2016;47: 739-47).

Results
We were able to analyse 84 cases of portal system anomalies. There were 65 cases of Persistent Right Umbilical Vein (PRUV), 2 cases of Umbilical Vein Varix (UVV), 10 cases of Umbilical Vein Systemic Shunt (UVSS), 2 cases of Ductus Venosus systemic shunt (DVSS), 2 cases of Intrahepatic portosystemic shunts (IHPSS), 1 case of Extrahepatic portosystemic shunt (EHPSS) and 2 unclassified variants (UV). At least one of the 3 steps were abnormal in all except the two cases of IHPSS. PRUV UVSS UV DVSS IHPSS UV EHPSS n=65 n=10 n=2 n=2 n=2 n=1 Abnormal GB to 65 1 2 1 0 0 0 0 UV relationship DV absent 0 10 0 2 0 1 0 Abnormal flow in portal vein and sinus 65 3 0 0 0 1 1.

Conclusion
Although uncommon, fetal portal system anomalies are associated with complications like fetal growth restriction, cardiac overload leading to hydrops, hyper-ammonia in neonates and rarely frank liver failure. A 3-step screening protocol is feasible and appears effective at identifying the vast majority of fetal portal system venous anomalies.