MFM BROADCAST

Dear Colleague,

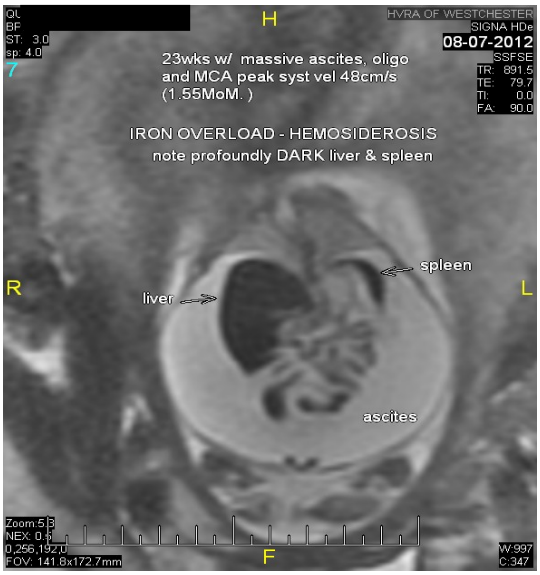
Welcome back to Fetal MR Interesting Case Presentation

I hope you enjoy the following case.

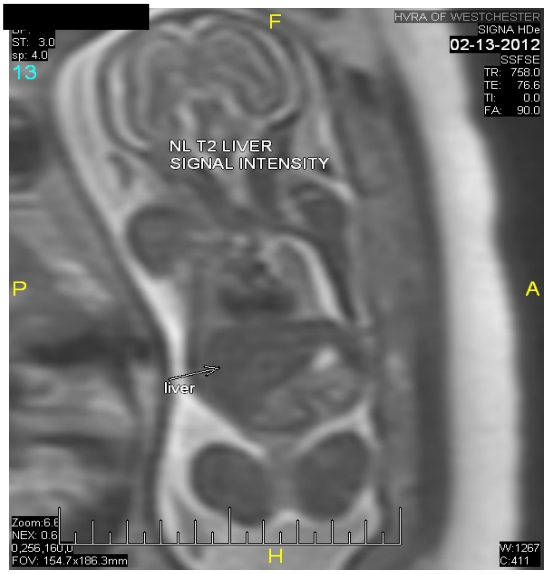
DIAGNOSIS: MR diagnosis of hepatic iron overload in a fetus with anhydramnios, massive ascites and no pre-MR clinical diagnosis.

HISTORY: 23-week gestation referred for fetal MR. Outside ultrasound studies demonstrated anhydramnios with massive ascites and nonnormalized kidneys. Prior to MR, normal peak systolic middle cerebral arterial velocities were demonstrated with normal maternal serum TORCH and parvovirus titers and no evidence for alloimmune hydrops and no causative structural malformations. Normal fetal echocardiogram was obtained at the time of MR.

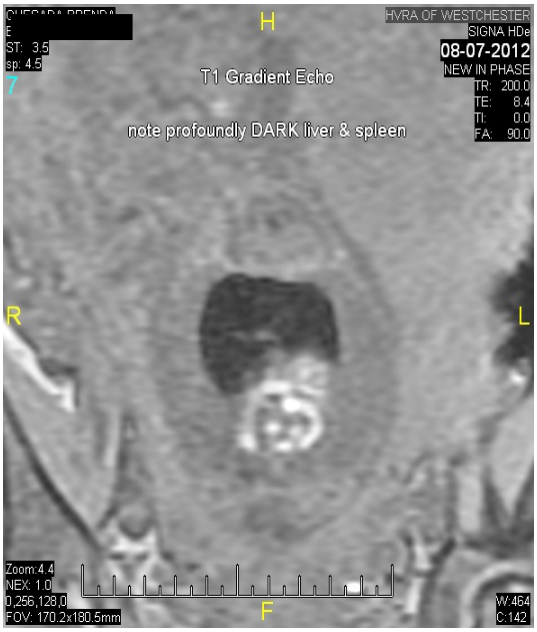
FETAL MR FINDINGS: Fetal MR demonstrated normal kidneys and markedly abnormal (dark) hyposignal intensity in an enlarged liver on both T1 and T2 pulse sequences.



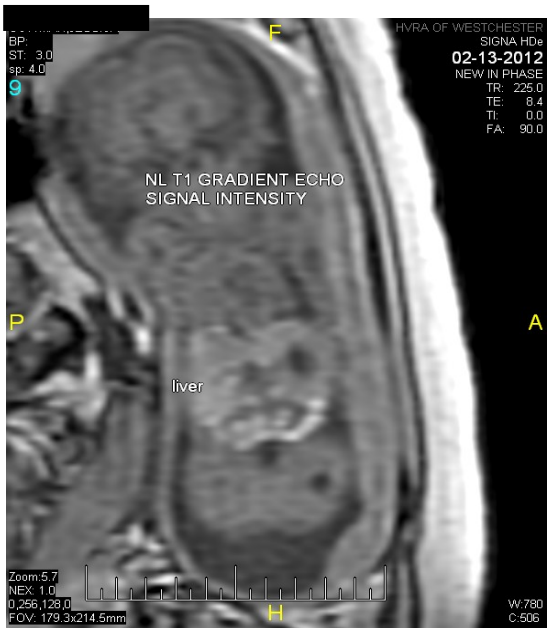
T2 iron overload - dark liver



Normal T2 liver



T1 iron overload – dark liver



Normal T1 liver

Ultrasound at the time of fetal MR demonstrated elevated middle cerebral artery peak systolic velocities at 48 cm/sec – greater than 1.55 MoM for gestational age – consistent with severe anemia. Between outside ultrasound studies and MR, no other structural malformations that might be causative for nonimmune hydrops were identified.

Imaging differential diagnosis for markedly decreased liver signal intensity on MR most commonly reflects iron overload.

Given evidence of hepatic iron overload and elevated middle cerebral arterial systolic velocities in the severe anemic range, the differential diagnosis statistically favors severe hemolytic anemia over the less likely possibility of congential hemochromatosis. The fetus was treated empirically with an intraperitoneal transfusion but expired shortly thereafter.

After fetal death, the PCR studies of fetal ascites were returned as positive for parvovirus.

Fetal autopsy confirmed markedly increased iron in the liver with paucity of extrahepatic iron within the adrenal medulla. No iron was found in the heart, thymus, kidney or brain mitigating against congenital hemochromatosis. The autopsy report made no mention of hepatitis or cirrhosis.

DISCUSSION: Fetal hepatic iron overload can be seen in association with the following disease processes:

- Hemolytic anemia
- Neonatal/congenital hemochromatosis – its most common cause in a fetus is gestational alloimmune liver disease (GALD)
- Infection - parvovirus and TORCH
- Intestinal disease - esophageal atresia, jejunal atresia, anal-rectal malformation, VACTERL.

TEACHING POINTS: Fetal MR contributed to the management of this case in the following ways:

- In a patient with anhydramnios and poor ultrasound viewing conditions, MR correctly identified normal kidneys.
- Fetal iron overload and elevated MCA velocities in the severe anemia range are nonspecific observations which can be seen in both an infectious-related hemolytic anemia as well as congenital hemochromatosis.
- In the absence of maternal or fetal evidence of infection (in the current case, fetal evidence was not established until after fetal death), **fetal congenital hemochromatosis** represented the leading alternative differential. This is an important differential to consider given the options of maternal treatment with intravenous immunoglobulins, its severe morbidity and mortality and its very high recurrence rate in future pregnancies.
    - Gestational alloimmune liver disease (GALD) is the most common cause for fetal/neonatal hemochromatosis. It is frequently accompanied by ascites, anemia, and thrombocytopenia. This presentation can therefore mimic other more common causes of immune and nonimmune fetal ascites/hydrops.
    - The prognosis in severe fetal hemochromatosis is generally very poor. The average life expectancy of the average severely effected newborn is days to a few weeks. Congenital hemochromatosis is a prominent cause of severe fetal liver injury and should be suspected in case of late intrauterine fetal demise in the absence of other definable cause. It should be suspected in any sick newborn with evidence of liver disease as it is the cause of most cases of newborn liver failure and/or cirrhosis. (Whitington).

Fetal MR demonstration of hepatic iron overload could be helpful in several clinical scenarios –

- Unexplained fetal ascites and/or hydrops – the presence of liver iron overload serves as a "red flag" for pathologies poorly assessed by ultrasound but can be diagnosed on fetal MR such as – liver disease/cirrhosis, esophageal atresia, anal atresia. As with many cases of ascites/hydrops, the coexisting pathologies may be merely associations and not necessarily directly causative to the ascites/hydrops and/or liver iron overload.

- The MR identification of fetal hepatic iron overload might also be helpful in evaluating those fetuses at risk for biliary atresia – persistent nonvisualization of gallbladder or right upper abdominal quadrant cystic mass. Biliary atresia can cause fetal hepatitis/cirrhosis which will not be visible on ultrasound. MR identification of altered liver signal intensity and/or iron overload would be supportive evidence of significant biliary disease with hepatic involvement.

REFERENCES:

Iron Overload in Gestational Alloimmune Liver Disease: Still More Questions Than Answers. Journal of Prenatal Diagnosis, 2012, 32, 810-812. Vanden _ Eijnden.

Fetal Liver Iron Overload: The Role of MR Imaging. European Radiology, 2011, 21: 295-300. Cassart.

Neonatal Hemochromatosis: A Congenital Alloimmune Hepatitis. Seminars in Liver Disease, Vol. 27, #3,007, 245-250. Whitington.

Fetal and Infantile Hemochromatosis. Hepatology. Vol 43, #4, 2006, 654-660. Whitington.

My personal score card in the MR and Ultrasound evaluation for placenta increta/percreta.
January 2008 through October 2012. 95 patients studied.

True positive:	27
True negative:	57
False positive:	7
False negative:	4
Sensitivity:	87%
Specificity:	89%
Positive Predictive Value:	79%
Negative Predictive Value:	93%
False Positive Rate:	11%
False Negative Rate:	13%

Daniel J. Cohen, M.D.
HUDSON VALLEY RADIOLOGY ASSOCIATES
danjcohen@optonline.net
914-391-0109

Same Day/Next Day Scheduling for Obstetrical MR
and Fetal Cardiac Ultrasound
Scheduling Department: (845) 634-9729
*18 Squadron Boulevard, New City, NY 10956
*115 Main Street, Tuckahoe, NY 10707