Universal Fetal Cardiac Ultrasound-
At the Heart of Newborn Well-Being
HUDSON VALLEY RADIOLOGY ASSOCIATES

MR Case Study: CMV Fetopathy and Cardiovascular Doppler

Introduction: The case to be presented is that of CMV fetopathy and the unique contribution of MR and cardiovascular Doppler in identifying central nervous system injury and assessing risk for metabolic acidosis, stillbirth and postnatal neurodevelopmental challenges.

Case Study:
MR imaging was performed at 21 weeks, 3 days on a fetus whose outside studies demonstrated somatic growth restriction including a BPD three weeks smaller than expected. Outside ultrasound studies demonstrated a "dangling" choroid in an atrium that was less than 10 mm. Intracranial anatomy was otherwise unremarkable. Uterine and umbilical artery Dopplers were reported as abnormal. There were positive maternal CMV titers. At the time of the MR, amniotic fluid CMV studies were pending.

The MR protocol was primarily directed at intracranial assessment - was there evidence of direct parenchymal injury in addition to the generalized cranial and somatic growth restriction?

Image #1 is a coronal T2 image demonstrating no sylvian fissure indentation – abnormal for gestational age, which in conjunction with microcephaly probably represents a malformation of cortical development.

Image #1 shallow Sylvian fissure (SF) for gestational age.

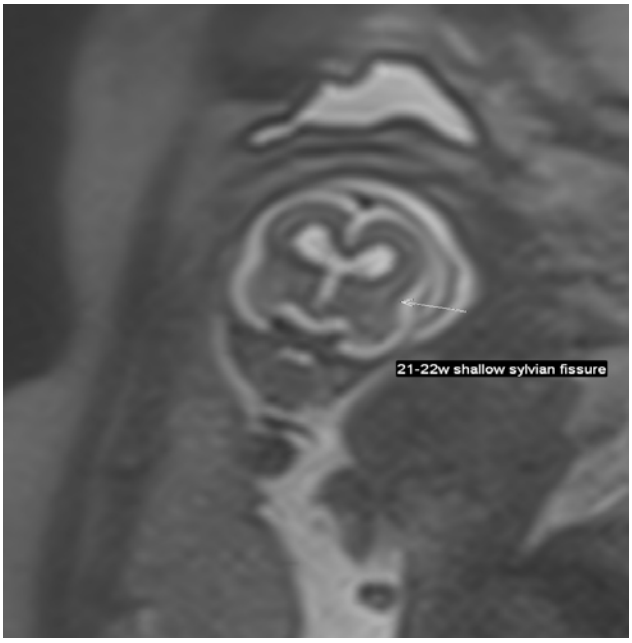


Image # 2 is a gestational aged matched no rmal demonstrating normal depth SF for comparison. (arrows)

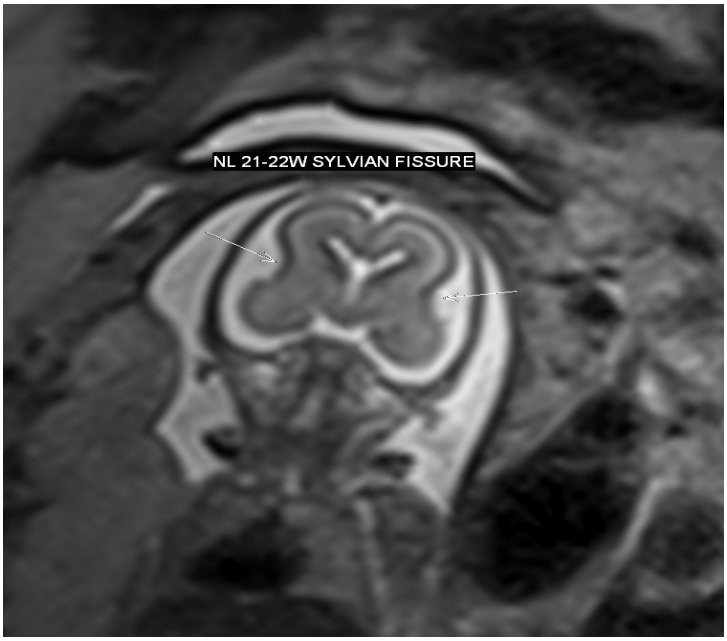


Image #3 is an axial T1 gradient echo sequence demonstrating absence of the normally seen trilaminar organization of brain parenchyma with abnormal increased basal ganglion signal intensity (brightness).

Image # 3

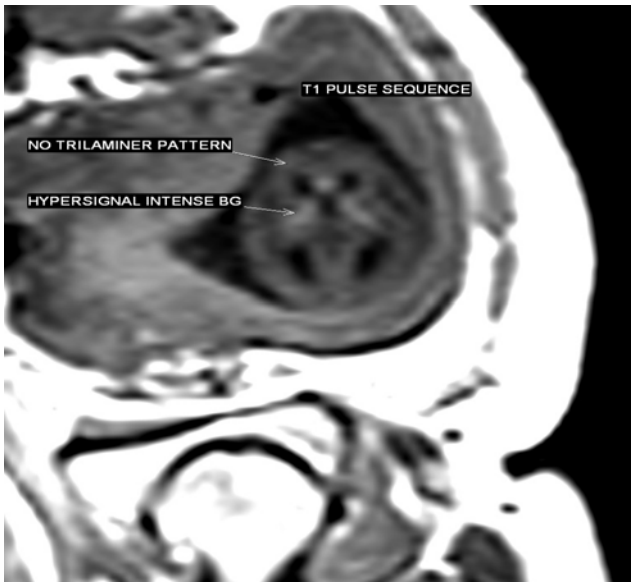


Image #4 is an age matched normal for comparison. Absence of trilaminar pattern of signal intensity probably reflects the disrupted patterns of neuronal migration from the ventricles to the surface of the brain accompany early CMV fetal infection. Abnormal T1 basal ganglion hypersignal intensity has been described in the acute phase of hypoxic ischemic injury in term neonates.

Image # 4

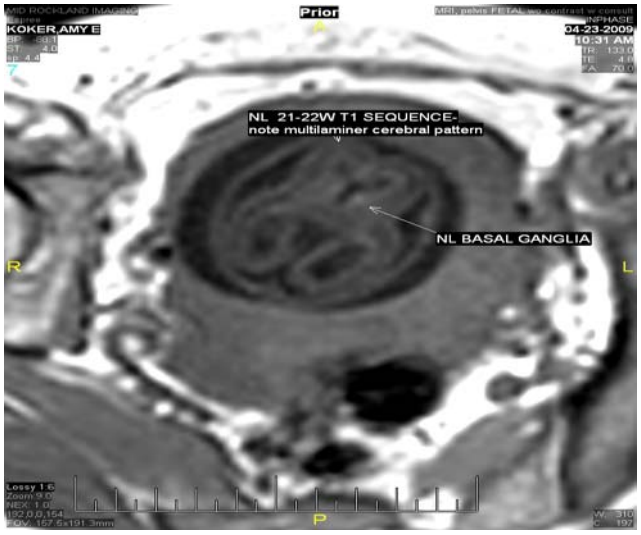
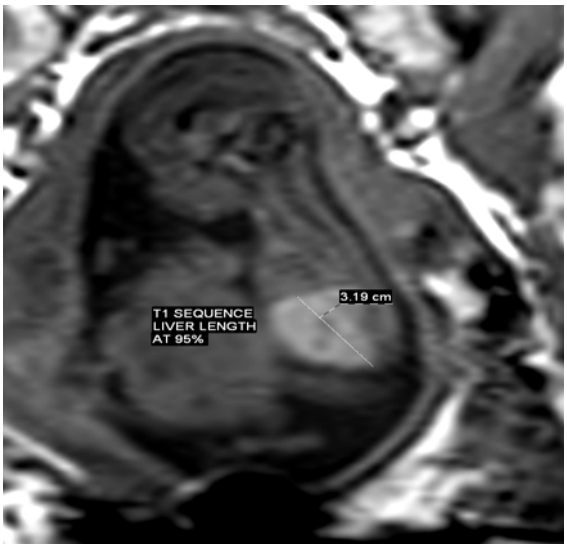


Image # 7



Sagittal T1 gradient echo **Image #7** demonstrates an enlarged liver measuring 3.2 cm, at the 95th percentile for gestational age. The T1 hypersignal intensity of liver and spleen allows for improved detection of hepatosplenomegaly in comparison to

Diffusion weighted imaging (DWI) was critical in this case to demonstrate direct cerebral injury. A significant weakness to standard T2 single shot fast spin echo sequences is that they are relatively insensitive to cerebral edema as demonstrated in this case by the absence of signal abnormalities on the T2 sequence (see Image #1 and Image #11).

Image #9 is an axial DWI sequence at the level lateral ventricle demonstrating global cerebral hypersignal intensity not present on the axial T2 image for comparison. (Image # 11)

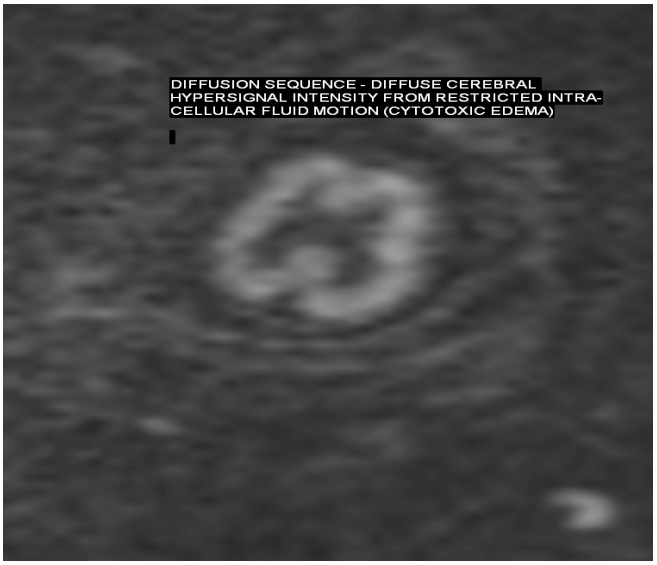
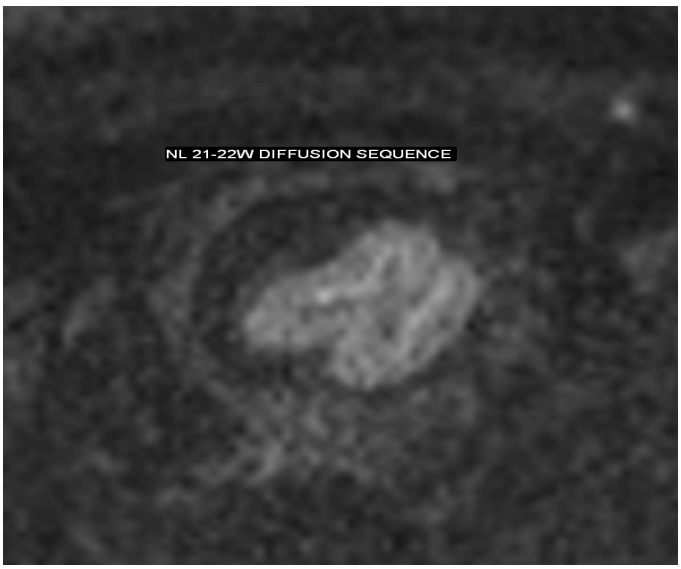


Image #10 is age matched normal DWI for comparison.



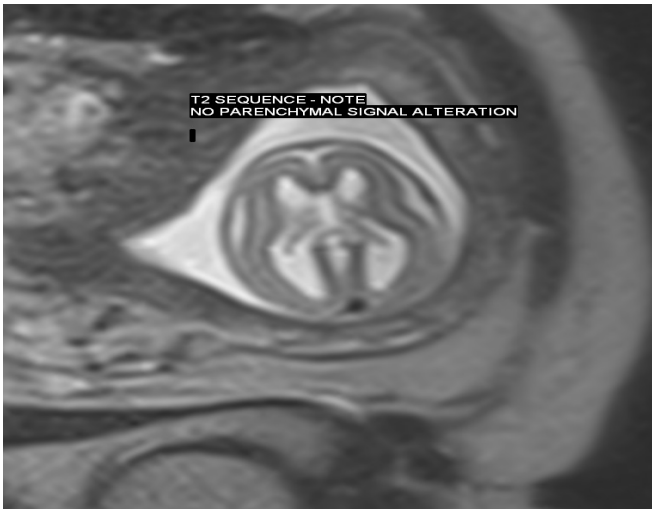
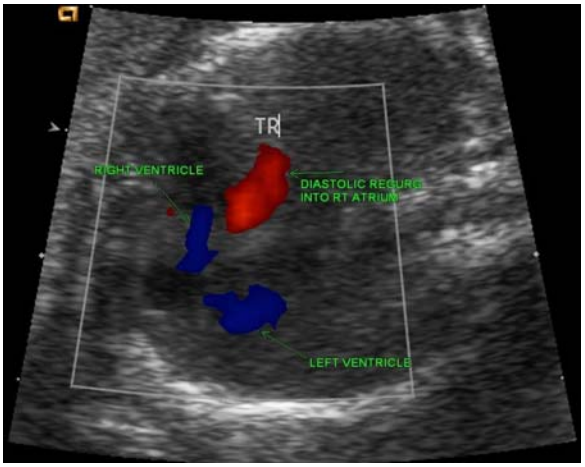
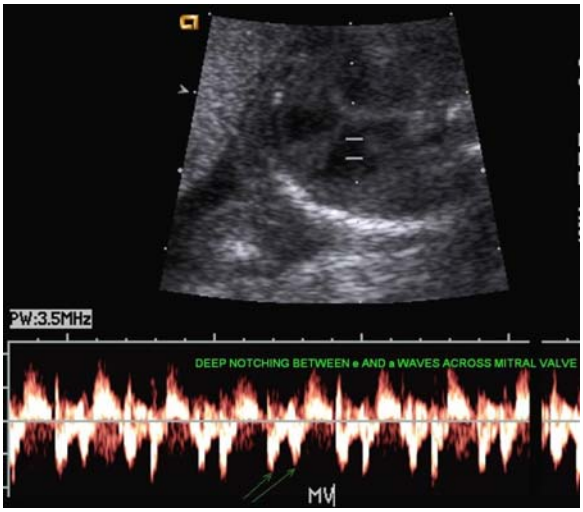


Image # 11 is normal T2 of current case.

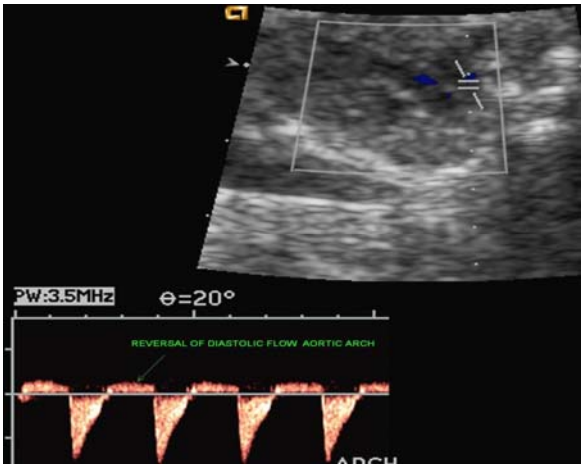
Building upon the outside ultrasound Doppler demonstrations of abnormal maternal and umbilical artery waveform patterns, I performed fetal cardiovascular Doppler to assess risk for metabolic acidosis, stillbirth, perinatal mortality and/or poor neurologic outcome.



Cardiac Doppler demonstrated massive tricuspid regurgitation **(Image #12)** Doppler regurgitant velocities exceeded 150 cm/sec.



Markedly widened and deeply notched E-A waves across the mitral valve **(Image 13**). These findings are associated with biventricular decreased diastolic compliance, cardiac dysfunction due to hypoxic cardiomyopathy (Mari. Journal Ultrasound Medicine. (26:1469-1477. 2007).



Color Doppler of the aortic arch demonstrates marked reversal of diastolic flow**, Image #14**. This observation is associated with poor newborn neurologic outcome (Acta Pediatrica. Vol 82, Issue 12. 919-924. Eronen. Jan 2008). Reversal of flow in the ductus venosus during atrial contraction (not shown) corroborates biventricular decreased cardiac diastolic compliance.

KEY IMAGING POINTS:

- The ability to identify direct brain parenchymal injury is dependent upon the specific pulse sequences employed at the discretion of the radiologist responsible for the case. As demonstrated in this case, the T1 type gradient echo and diffusion weighted sequences were necessary to demonstrate direct brain parenchymal injury as well as disruption of the trilaminar developmental organization of brain parenchyma. These technically more challenging sequences are site specific and time consuming due to their marked susceptibility to fetal motion.

- Contribution of intracardiac, aortic and ductus venosus Doppler to identify ischemic cardiac dysfunction and optimize risk assessment for stillbirth and/or future neurologic compromise.

- Diffusion weighted sequences are essential in evaluating any cause of fetal CNS ischemic injury such as monochorionic co-twin death, trauma, maternal hypotension, abruption, fetal-maternal hemorrhage.

PATHOLOGY RESULTS: Amniotic fluid titers were markedly positive for CMV. CMV inclusions were present in all organs including the lungs, liver, kidney and brain. Within the placenta, foci of acute villitis with CMV inclusions were seen.

Pathology of the umbilical cord demonstrated a 1 cm umbilical cord stricture just proximal to the umbilicus where it narrowed to a minimal diameter of 0.1 cm. Histology demonstrated fibrosis. There were no signs of a more generalized funisitis.

There was a spontaneous intrauterine demise one week after MR. Between demise and dilatation and evacuation, there was extensive autolysis precluding gross morphologic autopsy of fetal brain.

It is interesting to question whether or not growth restriction and abnormal Dopplers were due to a chronic primary umbilical cord stricture with CMV placental villitis and fetal infection as comorbid conditions versus the latter being a primary event with an umbilical stricture – presumably due to torsion. In either case, the MR findings of brain atrophy and absent sylvian fissure suggest a late first and early second trimester diffuse insult whose etiology could be either ischemic (umbilical cord) or infectious in origin. CMV fetopathy in the late first and early second trimester is known to disrupt neuronal migration resulting in lissencephaly and microcephaly.