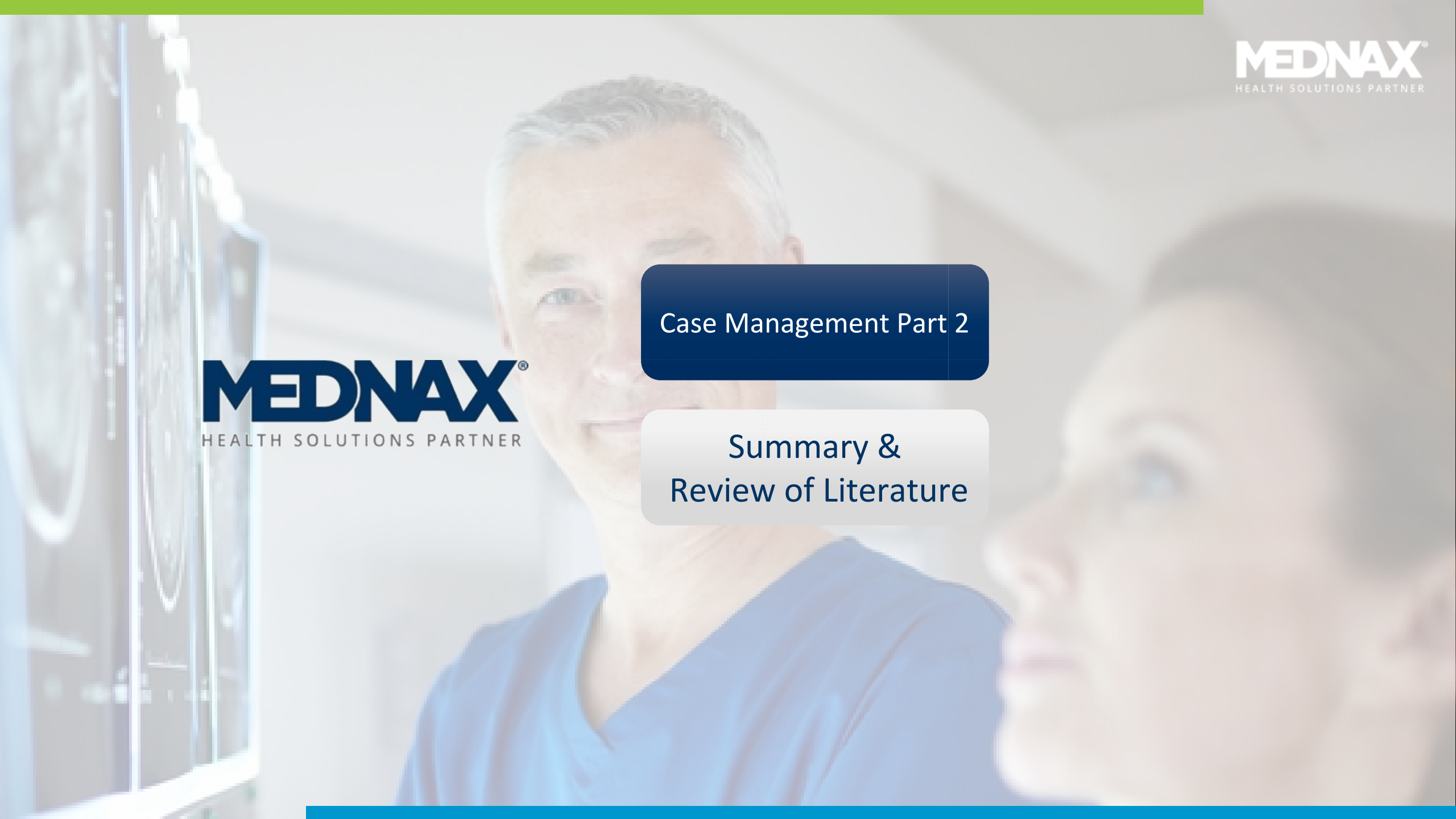


Case Management Part 2

Summary & Review of Literature

Part 2 – Case Management

The patient had a magnetic resonance imaging (MRI) at 25 weeks gestation, with findings consistent with a pial arteriovenous fistula. The arteriovenous malformation drained into the vein of Galen from the left basal vein of Rosenthal. Subsequent ultrasounds showed mild enlargement of the left lateral ventricle, likely indicating left cerebral atrophy/ ischemia, increase in fetal cardiomegaly, with tricuspid regurgitation. Fetal cardiac failure likely accounts for lack of umbilical artery end diastolic flow, and reversal of the ductus venosus A-wave.

Part 2 – Case Management

The patient met with a neonatologist who discussed in length the severity of the symptoms, potential complications and outcomes. These included cardiac failure, hydrops and fetal or neonatal death. They chose to continue the pregnancy. The patient opted for cell free DNA after genetic counseling which came back negative. She also had consultations with pediatric cardiology and neurology.

At 29 weeks gestation, the patient was admitted due to increased cardiomegaly and poor fetal Doppler flow. The plan was to get the fetus to at least 32 weeks gestation, at which time surgical intervention would be an option. The fetal monitoring showed an increase in cardiac failure, fetal hydrops and worsening Doppler's. At 32 weeks 3 days the fetus was delivered via cesarean section.

Apgar scores at 1, 5, and 10 minutes were 3, 6, 6 respectively. An echocardiogram was performed which showed moderate right ventricular dysfunction and elevated pulmonary pressures. The baby's condition became progressively worse and passed away one day after birth.

Image 5 – Magnetic Resonance Imaging Scan

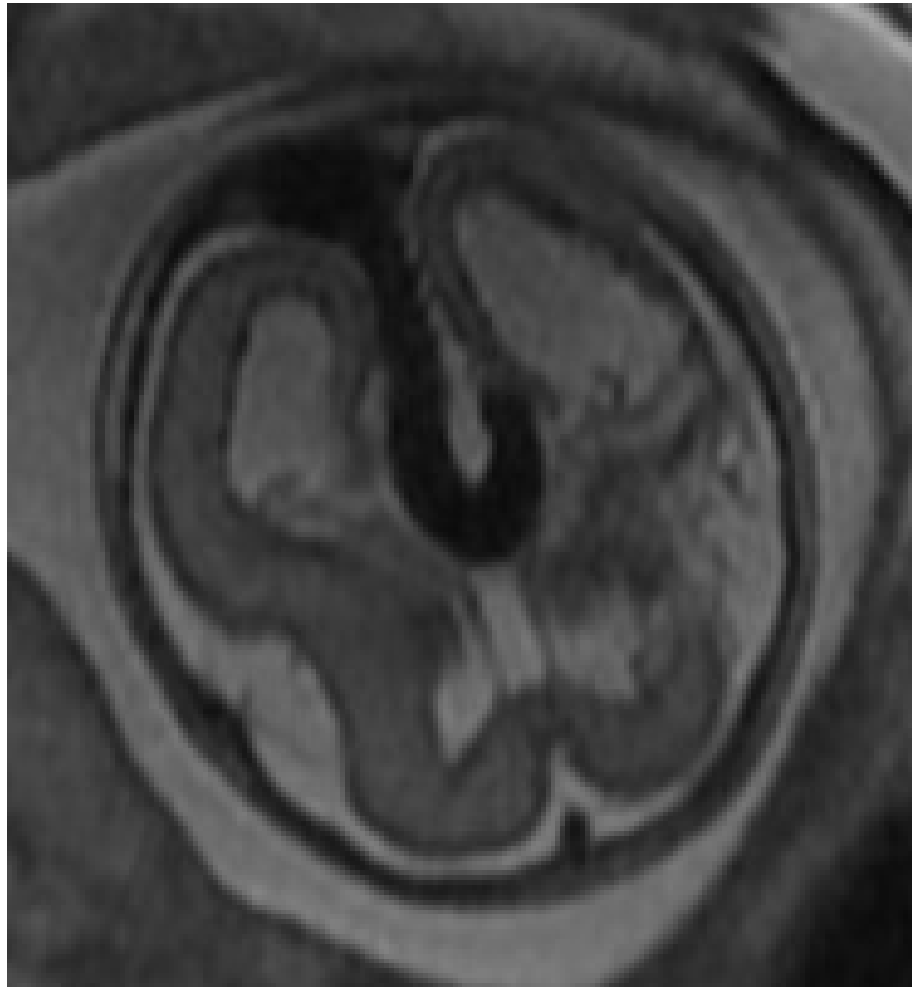
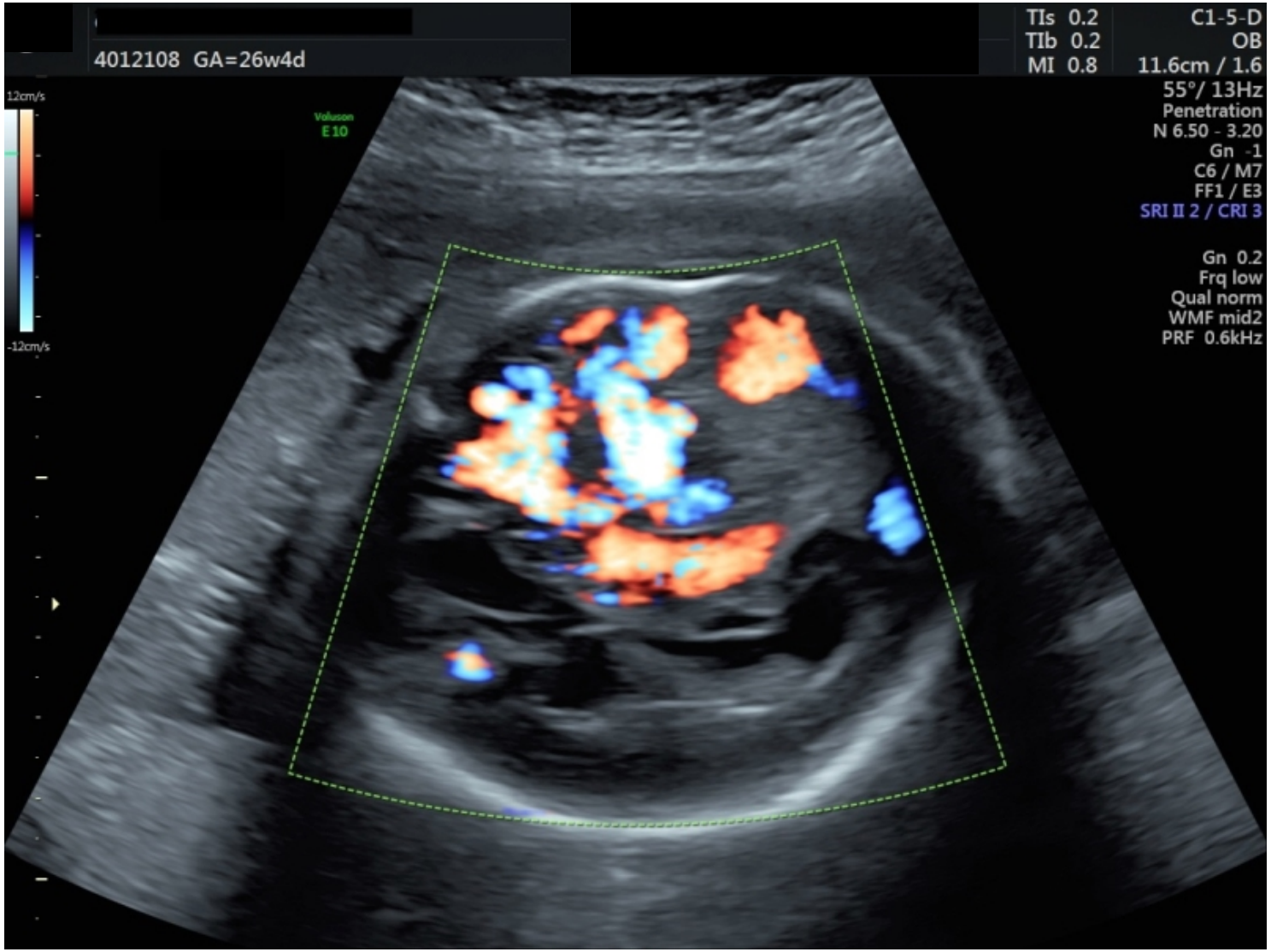


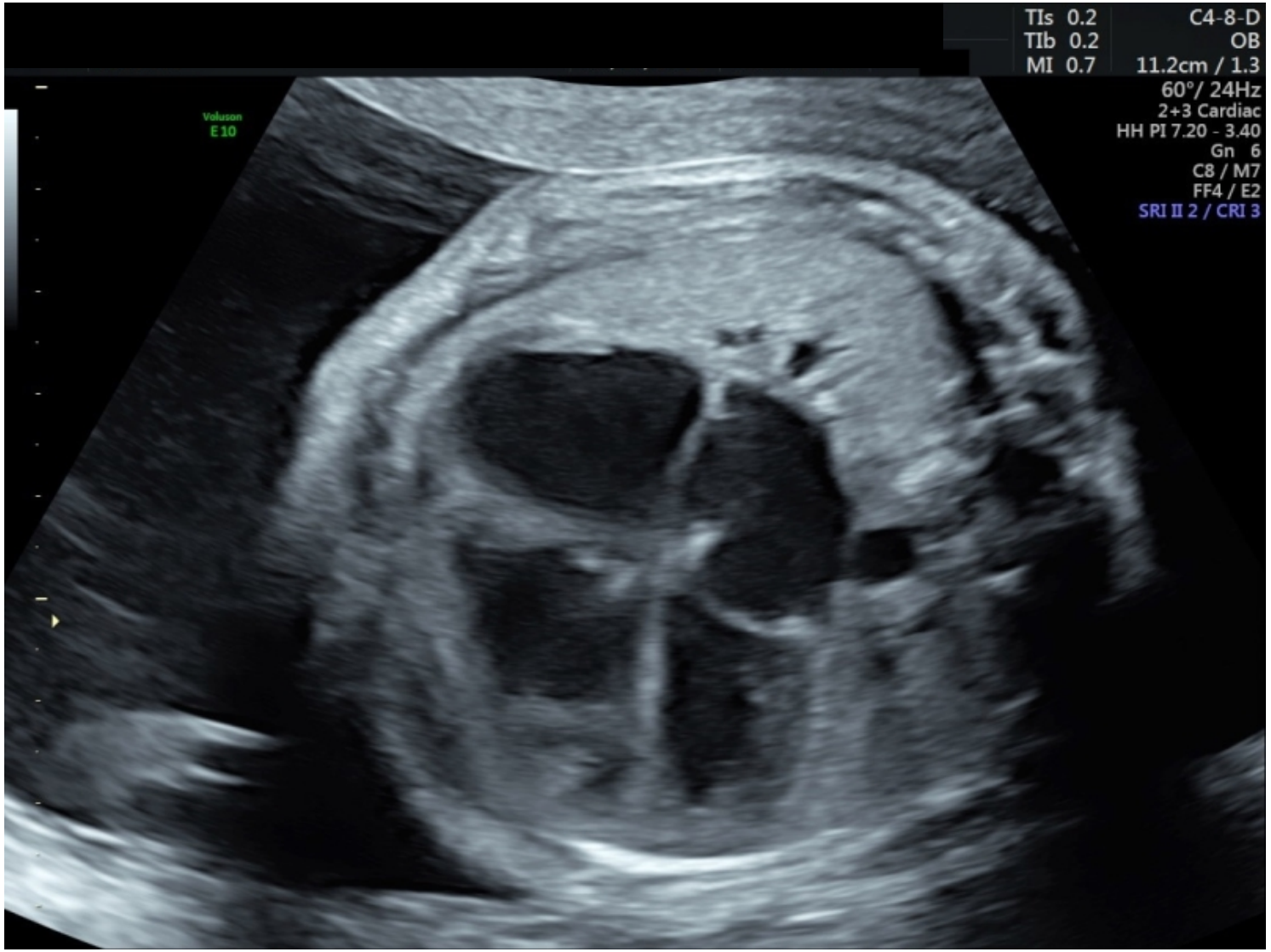
Image 6 – Enlarged AVF



Ultrasound images courtesy of:

Desiree Becker, RDMS, RDCS, RVT, Nichole Hamilton, RDMS and Joy Dobrinen, RDMS, RDCS. Seattle Ultrasound. Seattle, WA (USA)

Image 7 – Cardiomegaly



Ultrasound images courtesy of:

Desiree Becker, RDMS, RDCS, RVT, Nichole Hamilton, RDMS and Joy Dobrinen, RDMS, RDCS. Seattle Ultrasound. Seattle, WA (USA)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Summary &
Review of Literature

Summary & Review of Literature

Intracranial pial arteriovenous fistula (AVF) is a rare cerebrovascular malformation. According to a series reported by Halbach et al., (1989), pial AVF accounts for 1.6% of all intracranial vascular malformations. Intracranial pial AVFs have single or multiple arterial connections to a single venous channel (Lee, Son & Kim, 2008). Pial AVFs can cause severe morbidity and mortality, particularly in neonates. Congenital arteriovenous fistulas tend to have larger connections and increased flow. They are frequently associated with increased blood flow through the midline venous system, causing aneurismal dilatation of the vein of Galen (Köroğlu et al., 2006).

Pial AVFs correspond to an abnormal communication between arteries and veins. Most of the arteries feeding the pial AVF open into a single ectatic draining vein. (Garel, Azarian, Lasjaunias & Luton, 2005). Dilation of the vein of Galen results from the drainage of the fistula into the venous system.

Arterial malformations have two vascular patterns: arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs). Arteriovenous malformations (AVMs), microfistulas, are multiple arterial feeders joined via a nidus to draining veins. AVFs, macrofistulas, are direct shunts between large arterial and venous channels (Moradian, Nokhostin-Davari, Merajie, & Pouraliakbar, 2013). It is important to differentiate between a vein of Galen aneurysmal malformation (VGAM) and a vein of Galen aneurysmal dilation (VGAD) resulting from an arteriovenous fistula. In a VGAM, an arteriovenous malformation directly involves the vein of Galen, where in the VGAD, the vein becomes dilated due to a parenchymal AV fistula with the blood draining into the vein of Galen.

Summary & Review of Literature

There are distinct features of a pial AVF by ultrasound that differentiate it from a VGAM. The prognosis and treatment of these two types of cerebral arteriovenous malformations may vary depending on severity of symptoms and the type of malformation itself. It is vital to distinguish between them.

The sonographic features of a pial AVF are unexplained cardiomegaly with increased velocities and without cardiac malformation. Arteriovenous fistulas (AVFs) (and VGAM) may be responsible for high-output congestive cardiac failure (Garel et al., 2005). The most distinguishable difference between the two will be in the color and pulse Doppler imaging. In the case of a VGAM, Doppler will show mixed arterial and venous flow within the VGAM and venous-type flow in the draining vessels. In the case of a pial AVF, however, there will be a venous-type flow both in the dilated vein of Galen and in the vessels draining into this vein (Garel et al., 2005).

Summary & Review of Literature

Other sonographic features include the reversal of flow in the aorta, dilatation of the superior vena cava and cardiomegaly. The common causes of reversal of flow in the aortic arch are lesions in which a communication exists between the ascending aorta, the proximal aortic arch or its branches, and a lower pressure chamber or channel such as systemic veins, that results in “stealing” of blood from the distal aortic arch and descending aorta (Moradian et al., 2013). On fetal echocardiography, there is typically dilation of the superior vena cava, right ventricle and pulmonary artery. As the right heart dilates, the tricuspid valve annulus can stretch, resulting in tricuspid valve regurgitation. Flow across the aortic isthmus may reverse in diastole due to the cerebral steal. Combined cardiac output is increased, which results in high output congestive cardiac failure (Davey et al., 2012).

Management of pial AVF includes consultation and a care team involving maternal fetal medicine, pediatric cardiology, pediatric neonatology and pediatric neurology. A fetal magnetic resonance imaging (MRI) is helpful to confirm the diagnosis. Continual close fetal surveillance is necessary as heart failure and hydrops is likely.