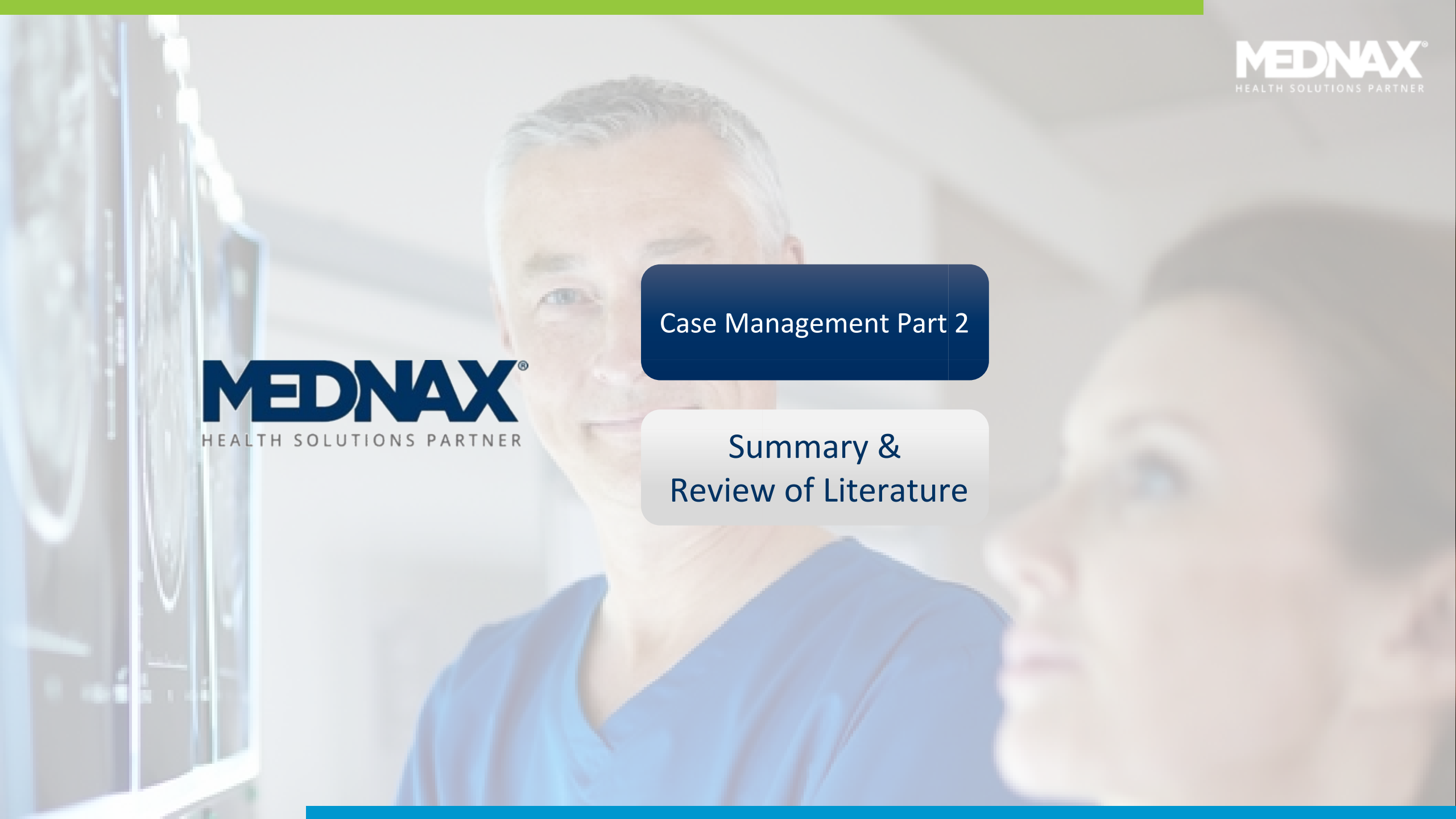


Case Management Part 2

Summary & Review of Literature

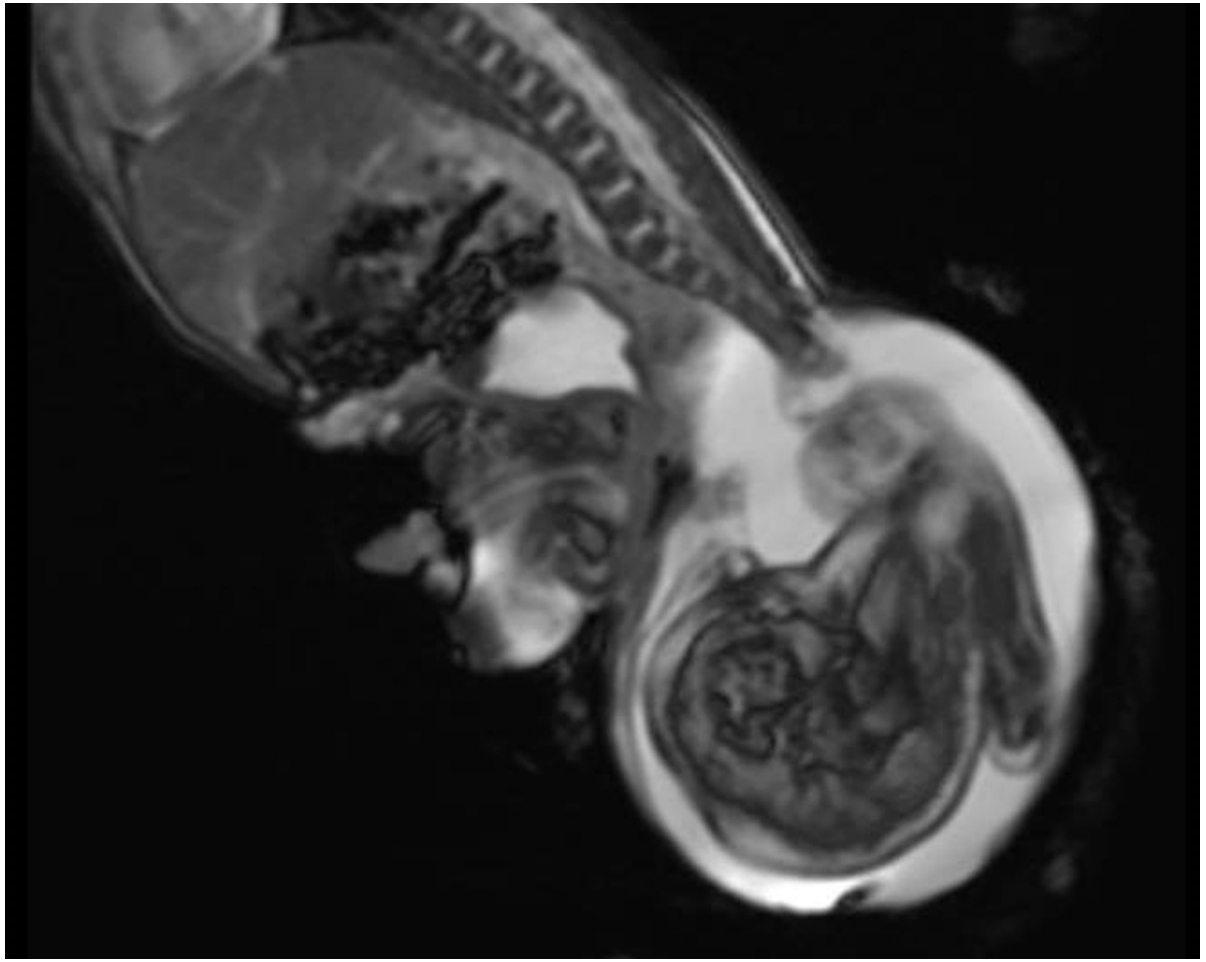
Part 2 – Case Management

The patient was seen in our ultrasound center every 3-4 weeks for follow-up evaluation of the fetal sacral mass, which was suspected to be a sacrococcygeal teratoma versus fetus in fetu. Amniocentesis results were 46XY. The mass was noted to have minimal vascularity and cystic and solid components, including areas with a long bone and foot structure. It was predominantly exophytic, but had an intrapelvic cystic component that extended to the level of the bladder. At 20 weeks the mass measured 4.1 x 4.5cm and had interval growth to 12.7x7.9cm by 36 weeks. Throughout our surveillance, the fetus had normal MCA dopplers, normal interval growth, and no signs of hydrops.

Part 2 – Case Management

The patient underwent an uncomplicated low transverse primary cesarean section at 39 weeks. A postnatal MRI was performed which confirmed the presence of a 12cm sacrococcygeal teratoma with a dominant solid mass and cystic surrounding component which extended to the presacral space with a 2cm intrapelvic portion. On day of life 2 the neonate underwent uncomplicated resection of a 400g mass via posterior approach with no internal pelvic component identified. On pathologic examination, the specimen had a well-formed right foot with 5 normal toes, calf, knee joint, and femur. Additionally, there was a round "head"-like structure covered by several skin tags suggestive of proboscis, black mature hair, and 1cm mass of CNS tissue with ganglion and glial cells. Mature intestines and colon with normal organization of ganglion cells were present and covered by a rudimentary diaphragm. There were also rudimentary cystic structures consistent with the collecting system of a kidney, and normal bilateral adrenal glands. Finally, there were gonads with testicles that appeared completely histologically normal. There was no immature or malignant tissue. Due to the strikingly mature development of several organ structures, including normal testicles, the pathologist confirmed the diagnosis of parasitic twin (fetus in fetu).

Image 5 – Neonatal MRI

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Summary &
Review of Literature

Summary & Review of Literature

Fetus in fetu (FIF) is a rare anomaly in which a fetus incorporates well-differentiated tissues of its parasitic monozygotic twin. It is estimated to occur in 1:500,000 live births (Iyer et al., 2003). FIF occurs most commonly in the retroperitoneum of the upper abdomen, although it has been reported to occur in the scrotum, skull, mediastinum, mouth, and adrenal gland (Aoki et al., 2004; Brand et al., 2004). FIF may be confused with meconium pseudocyst or a teratoma when identified during antenatal imaging, and most diagnoses of FIF cannot be made until pathologic evaluation of the specimen. Additionally, there is considerable controversy regarding the definition of FIF, and how it can be distinguished from a highly differentiated teratoma. Willis' theory, developed in 1935, states that FIF "is a mass containing a vertebral axis often associated with other organs or limbs around this axis." (Willis 1935) More recently, Gonzalez-Crussi have expanded the definition of FIF to include any structure in which the fetal form is in a very high development of organogenesis or associated with the presence of a vertebral axis (Gonzalez-Crussi, 1982). In the current case, no vertebral structures were identified, but based on the presence of a fully mature organ (testes) and long bones, the pathologist favored the diagnosis of FIF.

Summary & Review of Literature

In this case, distinguishing between FIF and well-differentiated sacrococcygeal teratoma (SCT) could not be accomplished antenatally. It may therefore be challenging to counsel parents because the two conditions are associated with very disparate antenatal and neonatal prognoses. The natural history of fetus in fetu is remarkably good, usually having no impact on the prenatal course of the pregnancy with the exception of bowel compression, hydronephrosis, and polyhydramnios from mass effect. In contrast, the mortality rate is as high as 52% among cases where SCT is diagnosed prenatally (Bond et al., 1990). Hydrops in SCT is usually, but not always fatal. Therefore, when the diagnosis is unclear, the fetus should undergo intense SCT surveillance with serial ultrasound monitoring for early evidence of hydrops, rapid tumor growth, anemia, and placentomegaly, all of which are associated with fetal morbidity and mortality. Mode of delivery is determined by the size of the mass; vaginal delivery may be possible with some small masses. However, when the sacral mass is over 5-10cm, as in our case, cesarean delivery is recommended to avoid trauma-induced hemorrhage or dystocia (Chervenak et al., 1985). Delivery should take place in a tertiary care facility with a NICU and pediatric surgical, and radiologic expertise immediately available.

Summary & Review of Literature

After birth the neonate should undergo imaging of the mass with MRI or CT scan to further characterize the lesion. Surgical resection of the mass is typically performed once the neonate has been stabilized. It is critical to distinguish FIF from SCT on pathologic evaluation as the latter has risk of malignancy developing in 7 to 30% of patients (Hedrick et al., 2004). There has only been a single case of malignant recurrence in fetus in fetu.