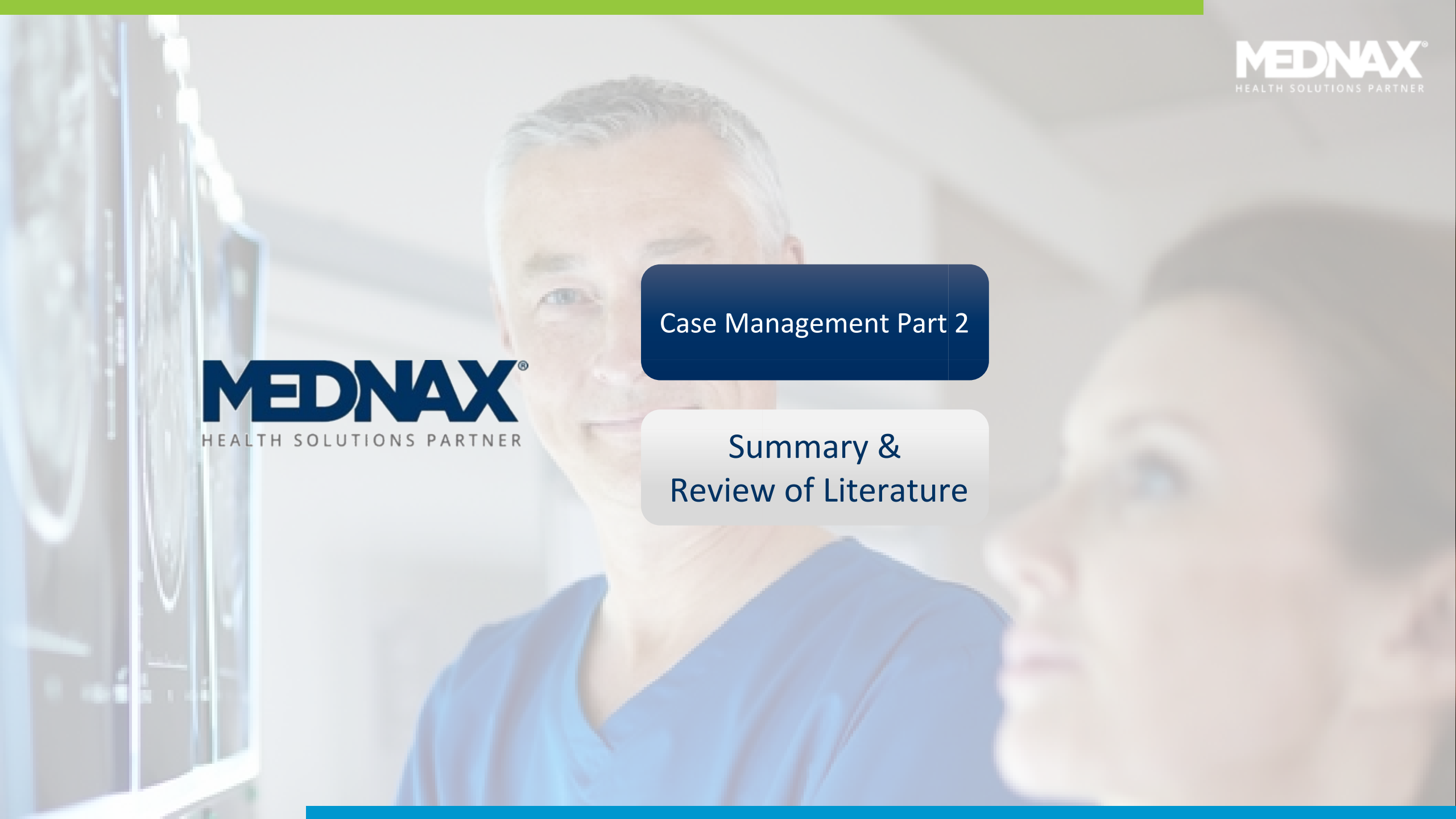

Case Management Part 2

Summary & Review of Literature

Part 2 – Case Management

Case Management: The patient was diagnosed with a di/di triplet pregnancy. Triplet A had its own distinct placenta and amniotic sac. Triplets B/C were a conjoined monochorionic/monoamniotic pair, consistent with conjoined cephalothoracapagus janiceps. There were found to be two distinct but conjoined thoracic cavities. The crania were conjoined. Four lower extremities were identified, although one was abnormal and shortened, with incomplete bone development. Two upper extremities were identified. Two hearts were identified as well.

She underwent extensive counseling regarding her options, including termination of pregnancy, selective termination of the conjoined set, and expectant management. She opted for expectant management, with plan for palliative care after delivery.

Part 2 – Case Management

She continued on with routine care. The conjoined pair was noted to have slightly decreased "individual" biometry measurements. At 31 weeks' and 4 days' gestation, she was sent to labor and delivery due to contractions and severe polyhydramnios of the conjoined pair, with the deepest vertical pocket being 20cm. She was significantly symptomatic with abdominal pain and difficulty breathing. She was given a course of betamethasone and a decision was made to proceed with Amnioreduction, where 2.5 Liters of amniotic fluid was removed.

Three days later, she underwent premature preterm rupture of membranes (PPROM). Fetus A was breech with feet in the vagina. She was taken to the operating room (OR) for an emergent cesarean delivery. Fetus A delivered with Apgar's of 9 at 1 minute of age, and 9 at 5 minutes of age, weighing 1620 grams. Triplets B and C were conjoined with Apgar's of 1 and 1. Comfort care had been agreed to, and the conjoined pair died shortly after delivery. The patient and her family declined autopsy.

Image 6 – cephalothoracopagus janiceps

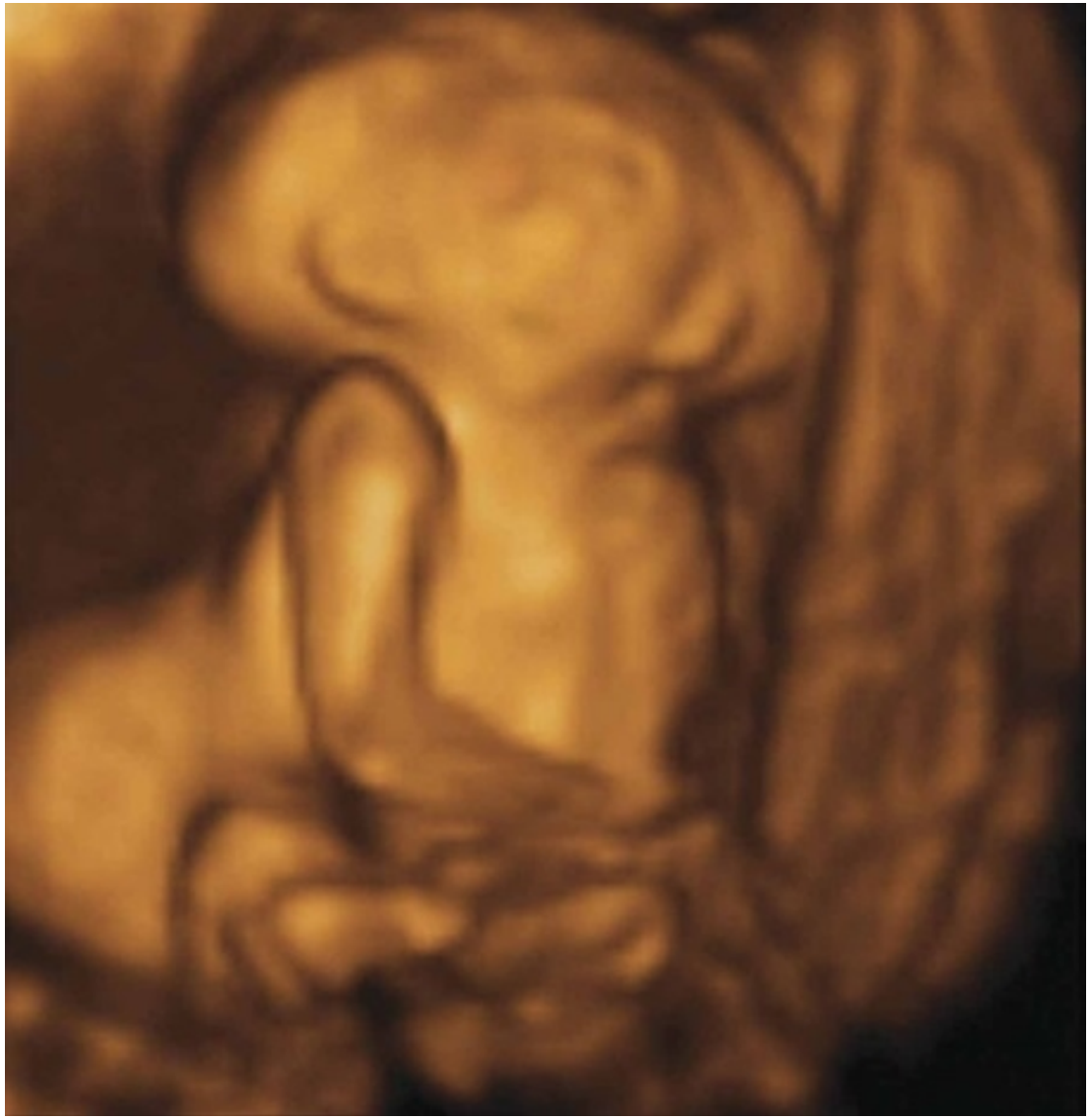

Image from Prenatal diagnosis of cephalothoracopagus janiceps: sonographic-pathologic correlation, by S. Luewan, K. Sukpan, Y. Yanase, and T. Tongsong. 2010. Journal of Ultrasound in Medicine

Image 7

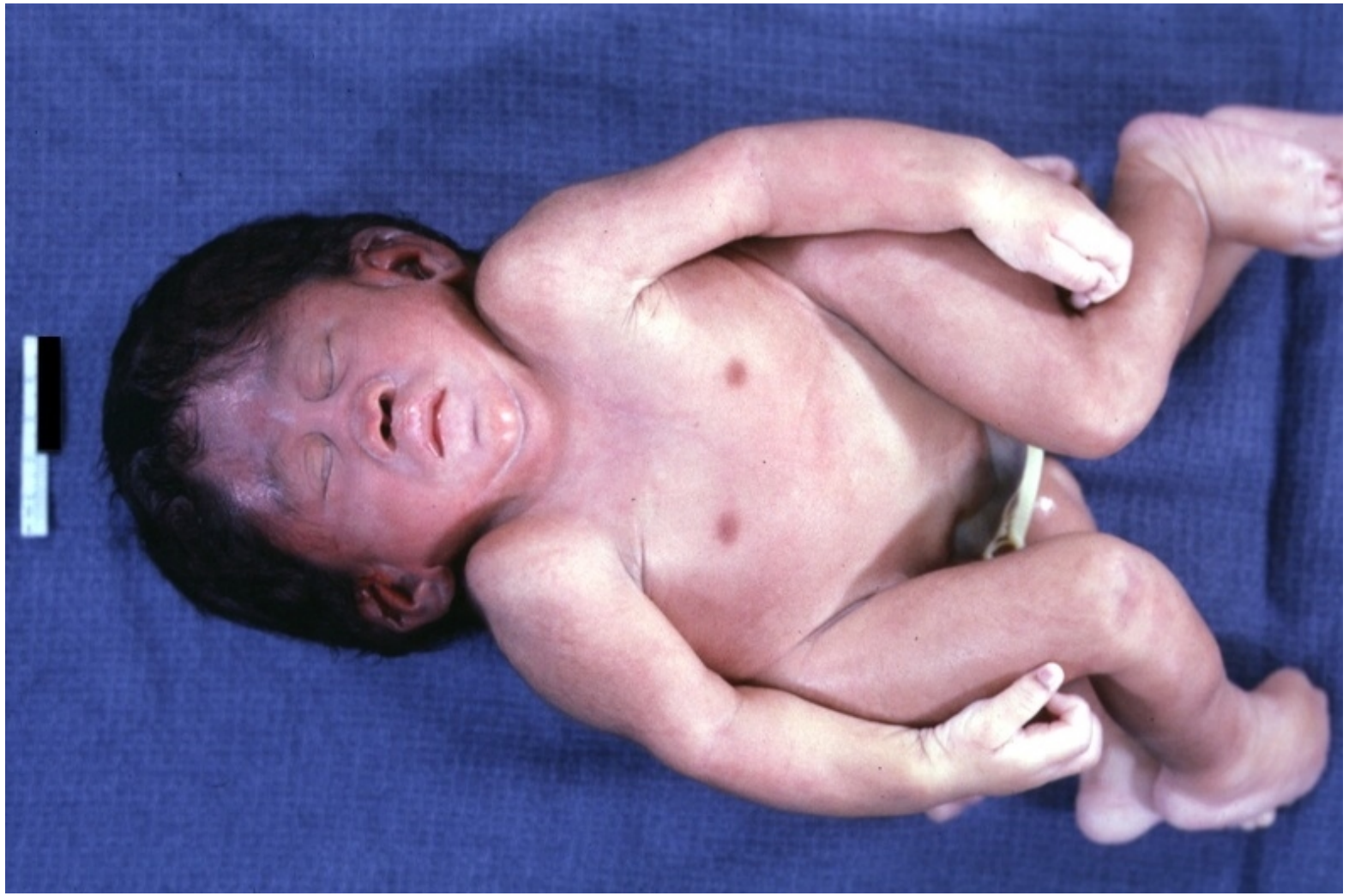

Image GROSS: BODY: Conjoined Twins Cephalothoracopagus Janiceps: Gross natural color anterior view, by P. Anderson, 2013, http://peir.path.uab.edu/library/picture.php?/7458


MEDNAX
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Summary &
Review of Literature

Summary & Review of Literature

Conjoined twins are the rarest subtype of monochorionic twins. They occur in 1 in 50,000 to 1 in 100,000 births. (Mathew, R., et al., 2017) They are believed to occur when incomplete division of a monozygotic embryo occurs after 13 days. There appears to be a female predominance in conjoined twin sex, higher prevalence in Latin American continents, and an association with assisted reproductive technology (Chen et al., 2011; Mutchinick et al., 2011).

Conjoined twins are classified by region of fusion, including head (cephalopagus), thorax (thoracopagus), umbilicus (omphalopagus), lower abdomen and pelvis (ischiopagus), cranium (craiopagus), vertebra (rachipagus), and sacrum (pyopagus). When multiple regions are fused, such as including both head and thorax, the nomenclature can be modified as indicated, e.g. “cephalothoracopagus” which includes fusion of both the head and thorax. (Baken et al., 2013) This particular case is one of cephalothoracopagus janiceps, which refers to a fused head, thorax, with the single fused head having two faces looking in opposite directions.

Summary & Review of Literature

Diagnosis can at times be challenging in early pregnancy, but conjoined twins have been diagnosed as early as 7 weeks' gestation in the literature. (Kongling, Shi, & Yao, 2014; Lam et al., 1998) Findings suggestive of conjoined twins include two fetuses which cannot be visualized separately in a single sac, bifid appearance of the fetal pole, heads and body at the same plane, and failure of the fetuses to change position to each other relatively over time. (Taner et al., 2009)

Perinatal and neonatal outcome of conjoined twins is difficult to assess, because a large proportion of those diagnosed are terminated upon diagnosis. One series by Chen, et al (2013) demonstrated that out of 75 cases detected in the first trimester, 80% terminated, 10.7% had fetal death, and only 8% carried to term. Out of the six sets of twins (12 infants), only four individual twins survived. The largest case series on 383 multinational conjoined series demonstrated a live birth rate of 45.6%; this figure, however, includes countries where termination of pregnancy is not practiced or available, and does not denote neonatal survival rate (Mutchinick et al., 2011)

Summary & Review of Literature

In terms of postnatal survival, the rate depends highly on the anatomic levels, the level of shared organs involved, and the ability of surgical expertise. A series in Brazil on 21 conjoined pairs over 20 years demonstrated that out of those 21, 9 (43%) could not be separated due to complexity of organs (liver and heart). Out of the remaining separated twins, the surgical survival rate was 66.7% (Tannuri, Batatinha, Velhote, & Tannuri, 2013).

If the diagnosis of conjoined twins is made, thorough counseling should be made with the patient regarding the extremely high fetal risk, poor prognosis, and neonatal risk. Termination of pregnancy should be offered, with earlier gestational ages being preferred. Multidisciplinary team should be involved including perinatology, obstetrics, anesthesia, neonatology, and pediatric surgery. If possible, a clear plan after delivery should be delineated, such as palliative care only, or aggressive surgical separation and intensive care. Delivery should be via cesarean. (O'Brien, Nugent, & Khalil, 2015)