# Introduction

Conjoined twins, one of the most fascinating human malformations, have been worshipped as gods and feared as monsters. Anesthesia for conjoined twins, either for separation surgery, or for Magnetic Resonance Imaging (MRI) or other evaluation procedures is an enormous challenge to the pediatric anesthesiologist. This is an extra challenging surgery because the anesthesiologists need to care for two patients at the same time instead of just one ***(Lalwani et al., 2011).***

The incidence of conjoined twins is very rare (1:200,000) live births. Inspite of their rare occurrence, several successful operative separations have been reported. Twinning is more common in Indian and African populations than in Caucasians ***(Lalwani et al., 2011).***

Surgery to separate conjoined twins may range from relatively simple to extremely complex depending on the point of attachment and the internal parts that are shared. Most cases of separation are extremely risky and life-threatening ***(Szmuk et al., 2006).***

Anesthesia for conjoined twins' separation surgery centers on many concerns like: Conjoined twins' physiology as crossed circulation, distribution of blood volume and organ sharing with their anesthetic implications. Long marathon

surgery with massive fluid shifts and loss of blood and blood components and their rapid replenishment accounts also for a major concern for the anesthesiologist. One of challenges also for anesthesia during conjoined twins operation is meticulous planning for organized management of long hours of anesthetic administration in two pediatric subjects simultaneously with multi surgical specialties involvement and their unique requirements ***(Chalam, 2009).***

Classification of conjoined twins is based on the site of union as thorax–40% (thoracopagus), upper abdomen–(xiphopagus), lower abdomen–33% (omphalopagus), sacrum-19% (pyopagus), pelvis-6% (ischiopagus) or skull-2% (craniopagus). Thoraco-omphalopagus is one of the most common types accounting for 74% cases ***(Thomas and Lopez 2004).***

# Aim of the Essay

The aim of this essay was to highlighten prenatal evaluation, classification and anesthetic management of conjoined twins.

# Classification and Prenatal Evaluation of Conjoined Twins

## Multiple births:

- **Definition:**

Multiple births refer to the delivery of twins and higher order multiples (e.g., triplets, quadruplets). Multiple births occur when multiple fetuses are carried during a pregnancy with the subsequent delivery of multiple neonates ***(Hibbs et al., 2010).***

- **Pathophysiology:**

The 2 types of twin pregnancies are dizygotic and monozygotic.Dizygotic twins develop when 2 ovum are fertilized. Dizygotic twins have separate amnions, chorions, and placentas (Fig.1). The placentas in dizygotic twins may fuse if the implantation sites are proximate. The fused placentas can be easily separated after birth ***(Qiu et al., 2008).***

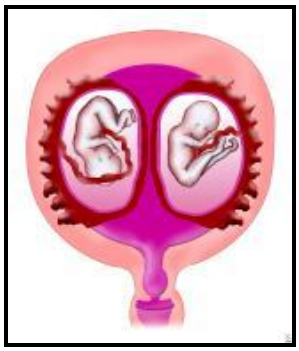

**Figure (1):** Diamniotic/dichorionic placentation ***(Qiu et al., 2008).***

Monozygotic twins develop when a single fertilized ovum splits after conception. An early splitting (i.e., within 2 day after fertilization) of monozygotic twins produces separate chorions and amnions. These dichorionic twins have different placentas that can be separate or fused. Approximately 30% of monozygotic twins have dichorionic/diamniotic placentas. Later splitting (i.e., 3-8 days after fertilization) results in monochorionic/diamniotic placentation (Fig. 2). Approximately 70% of monozygotic twins are monochorionic/diamniotic. If splitting occurs later (ie, 9-12 d after fertilization), monochorionic/monoamniotic placentation occurs (Fig.3).

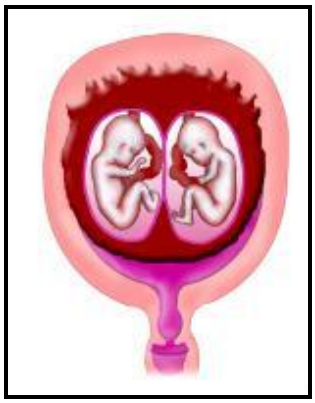

**Figure (2):** Diamniotic/monochorionic placentation ***(Qiu et al., 2008).***

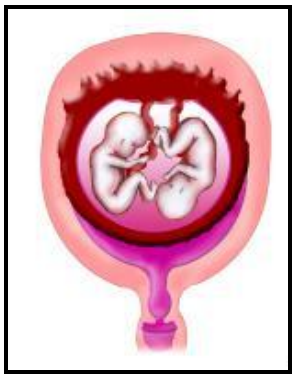

**Figure (3):** Monoamniotic/monochorionic placentation ***(Qiu et al., 2008).***

Monochorionic/monoamniotic twins are rare; only 1% of monozygotic twins have this form of placentation. Monochorionic/monoamniotic twins have a common placenta, with vascular communications between the 2 circulations. These twins can develop twin-to-twin transfusion syndrome (TTTS). If twinning occurs more than 12 days after fertilization, then the

monozygotic fertilized ovum only partially splits resulting in conjoined twins ***(Wright et al., 2007).***

Triplet pregnancies result from various fertilization, splitting, and development scenarios that involve ovum and sperm. For example, triplets can be monozygotic, dizygotic, or trizygotic. Trizygotic triplets occur when 3 sperms fertilize 3 ova. Dizygotic triplets develop from one set of monozygotic cotriplets and a third cotriplet derived from a different zygote. Finally, 2 consecutive zygotic splittings with a vanished fetus can also result in monozygotic triplets. Zygosity in quadruplets and higher order multiples also varies. Although the evaluation of the placenta or placentas after the birth is important in all multifetal pregnancies, the examination may not always help determine zygosity ***(Qiu et al., 2008)***.

## ➢ Epidemiology:

The birth rate of monozygotic twins is constant worldwide (approximately 4 per 1000 births). Birth rates of dizygotic twins vary by race. The highest birth rate of dizygotic twinning occurs in African nations, and the lowest birth rate of dizygotic twinning occurs in Asia ***(Wright et al., 2007)***.

## ➢ Mortality/Morbidity:

Multifetal pregnancies are high-risk pregnancies. Multifetal pregnancies are complicated by a higher incidence of hypertensive diseases, anemia, preterm labor, premature rupture

of membranes, hyperemesis gravidarum, placenta previa, polyhydramnios, and delivery complications (eg, Cesarean delivery, placental abruption, operative delivery, malpresentation, cord accidents, postpartum endometriosis) ***(Hibbs et al., 2010).***

Because of advancements in perinatal and neonatal care, the major issues that affect neonatal outcome of multiple fetal pregnancies include preterm delivery, low birth weight, and intrauterine growth retardation ***(Wright et al., 2007)***.

The mean gestational age at delivery is approximately 37 weeks for twins, 33 weeks for triplets and 28 weeks for quadruplets. Divergence from singleton growth curves occurs at approximately 32 weeks' gestation in twins, 29-30 weeks' gestation in triplets, and 27 weeks' gestation in quadruplets ***(Shinwell, 2005).***

Specific morbidities in multiple fetal pregnancies are controversial. Neonatal outcomes at specific gestational ages and birth weights are similar to singleton pregnancies. Neonates born to multiple fetal pregnancies may have a higher risk of acute respiratory morbidities, such as respiratory distress syndrome but do not have a higher incidence of chronic lung disease ***(Garite et al., 2004).***

Other major morbidities, including intraventricular hemorrhage, periventricular leukomalacia, retinopathy of

prematurity, necrotizing enterocolitis, patent ductus arteriosus, nosocomial infection, and length of hospital stay, demonstrate no statistical difference between singletons and multiples ***(Lee et al., 2006)***.

The risk of cerebral palsy in multiple fetus pregnancies parallels decreasing gestational age. A second association during the late preterm period (34-37 weeks' gestation) may correlate with the increasing maternal morbidities of multiple fetal pregnancies during this time frame (i.e., fetal growth restriction, hypertensive disorders, placental insufficiency) ***(Wright et al., 2008)***.

The neonatal mortality rate in multiple fetal pregnancies is similar to singleton rates and parallels decreasing gestational age. In monochorionic twins followed up from the first trimester until a mean age of 24 months, studies showed that twin-to-twin transfusion syndrome and assisted conception increased the risk of both death and neurodevelopmental impairment, whereas early onset discordant growth increased only the risk of death. Of the 136 pregnancies studied, 90% resulted in both twins surviving, 4% resulted in 1 survivor, and 6% resulted in no survivor. Overall, mortality was 8% and neurodevelopmental impairment occurred in 10% of infants ***(Chauhan et al., 2010).***

# Conjoined twins:

## ➢ Definition:

Conjoined twins are defined as monochorionic monoamniotic twins fused at any portion of their body as a result of an incomplete division of the embryonic disk, occurring after the 13th day post conception. The term 'conjoined' is actually a misnomer, since most authors consider the pathogenesis of the condition to result from failure of complete separation, rather than fusion of twins ***(Benirschke, 1998)***.

## ➢ Historical aspects:

The earliest example of conjoined twins is a 17 cm marble statuette portraying parapagus twins the double goddess dating from the sixth millennium B,C,. The statue of sisters of cattathoyuk is housed in the Anatolian Civilization Museum in Ankara, Turkey**.**Another early example is a stone carving of pyopagus twins dated to 80 B,C, discovered in Fiesole and housed in the San Marco *Museumin* ,Florence, Italy ***(Geroulanos et al., 1993).***

The earliest attempt at separation of conjoined twins took place in Kappadokia, Armenia when 1 of the male ischiopagus twins died at the age of 30 years, an attempt was made to save the surviving twin by separating him from his dead brother, but he died 3 days later ***(Geroulanos et al., 1993).***

The first successful separation of conjoined twins took place in 1689. The surgeon, Johannes Fatio, separated omphalopagus twins in Basel, Switzerland, by tracing the umbilical vessels to the navel where he tied them separately. He then tansfixed and tied the bridge between the two infants with a silken cord and cut the isthmus. The ligature fell off on the ninth postoperative day and both children survived ***(Lewis and Edward, 2003).***

The most celebrated pair of conjoined twins was Chang and Eng born on a river boat in Siam in 1811. They were joined at the xiphisternum by a short band that stretched so they were eventually able to stand side by side. They were taken by hunter to the United States,where they were exhibited by the showman Phineas Barnum. They married sisters, lived in separate houses in North Carolina spending 3 days in each house alternately, and had 22 children between them. They lived together for 63 years ***(Lewis and Edward, 2003).***

## ➢ Incidence:

Although the worldwide incidence of monozygotic twinning is the same in all ethnic groups, the incidence of conjoined twins appears to be higher in sub-Saharan Africa, ranging from 1:50 000 to 1:100 000 live births, or 1 in 400 monozygotic twin births. Thirty-one sets were born in Southern Africa between 1974 and 1982, of which 15 were stillborn, 7 were

considered inoperable and only 4 were successfully separated There was no information on 5 others ***(Rode et al., 2006).***

The natural history that follows a prenatal diagnosis of conjoined twins confirms that a large number of infants die either in utero (28%) or immediately after birth (54%); in fact, only 18% survive ***(MacKenzie et al., 2002)***.

## ➢ Embryology:

Embryologically, conjoined twins formation results either from failure of separation of the embryonic plate between 15 and 17days' gestation, or from secondary union of two separate embryonic discs at the dorsal neural tube or ventral yolk sac areas at 3 - 4 weeks' gestation. Extensive embryological studies appear to favor the latter theory, but this remains controversial ***(Rode et al., 2006).***

Four days after fertilization the trophoblast (chorion) differentiates. If the split occurs before this time, the monozygotic twins will implant as separate blastocysts each with their own chorion and amnion. Eight days after fertilization, the amnion differentiates. If the split occurs between the 4th and 8th days, then the twins will share the same chorion but have separate amnions. If a split occurs after the 8th day and before the 13th day, then twins will share the same chorion and amnion. This is a very rare condition and accounts for 1-2% of monozygotic twins. The embryonic disk starts to

differentiate on the 13th day. If the split occurs after day 13, then the twins will share body parts in addition to sharing their chorion and amnion ***(Sethi et al., 2004).***

The cause of this incomplete separation is unknown. Conjoined twins have been induced in experimental animals with such diverse agents as butyric acid, acetone, manganese deficiency and radiation. The cause of the striking female predominance in conjoined twins is still unknown ***(Kingston et al., 2001)***.

Figure (4) illustrates the germ layers in embryos of conjoined twins. The proximity of the segments determines how much shared tissue there will be. The further apart the segments, the greater the likelihood that the organs will develop fully in each fetus. If the segments are at their farthest point, there will only be a minimum of tissue and cartilage joining the twins, i.e. omphalopagus twins will develop ***(Kingston et al., 2001)***.

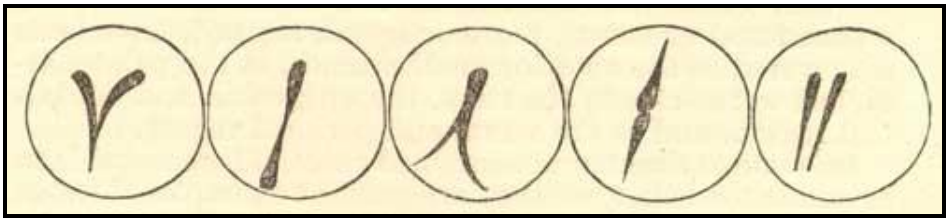

**Figure (4):** Germ layers in embryos of conjoined twins ***(Kingston et al., 2001)***.

## ➢ Classification:

Conjoined twins are classified according to the area of the bodies where the fusion takes place and the involvement of internal organs. The symmetrical and equal forms, in which the twins have equal or nearly equal duplication of structures, are called duplicata completa. When there is an unequal duplication of structures they are called duplicata incompleta, and this category includes the most severe types of conjoined twins in which just few organ systems are duplicated (fig.6). The most frequent varieties of conjoined twins are thoracopagus (40%), omphalopagus (33%), pygopagus (18%), ischiopagus (6%) and craniopagus (2%) (fig.5) ***(Dalmia, 1997).***

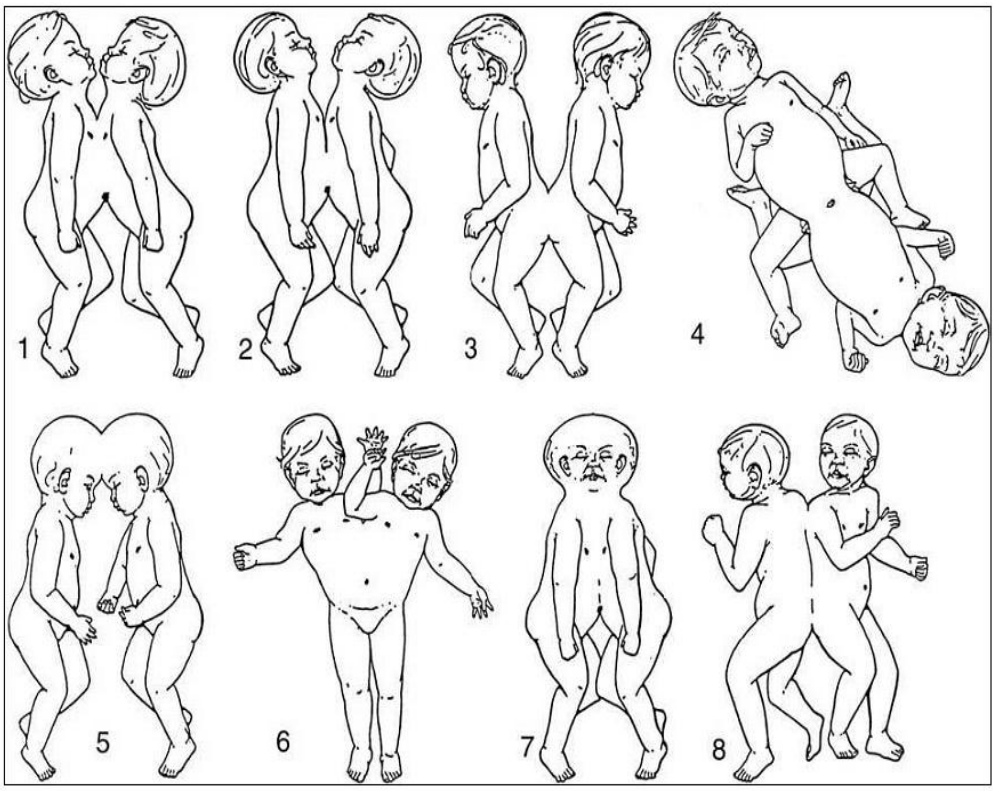


**Figure (5):** Schematic of the different types of conjoined twins: 1-thoracopagus, 2-omphalopagus, 3-pygopagus, 4- ischiopagus, 5-craniopagus, 6- parapagus, 7-cephalopagus, 8 _ rachipagus ***(Kingston et al., 2001).***