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ANESTHETIC MANAGEMENT OF CRANIOPAGUS TWINS

Essay

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in Anesthesiology

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LIST OF ABBREVIATIONS

Abbr.	Title
BIS.....	Bispectral Index
CT.....	Computerized tomography
DISIDA.....	Diisopropyl iminodiacetic acid
EEG	Electroencephalogram
EHBS.....	Extrahepatic biliary system
ETTs	Endotracheal tubes
GIT	Gastrointestinal tract
GOR	Gastro-esophageal reflux
MRI	Magnetic Resonance Imaging
NIRS.....	Near infrared spectroscopy
TCD	Transcranial Doppler
TEG	Thromboelastogram
TTTS	Twin-to-twin transfusion syndrome
TV	Total Vertical
US.....	Ultrasonography
VSD	Ventricular septal defect



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Mohammed Hamed Abd El Aziz

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

وَقُلْ رَبِّ
زِدْنِي عِلْمًا

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الآية (١١٤)

INTRODUCTION

Conjoined twins, one of the most fascinating human malformations. 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. In spite 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*).

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*).

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*).

AIM OF THE WORK

The aim of the work is to discuss and show the recent updates in anesthetic management of craniopagus twins.

Chapter (1)

CLASSIFICATION AND PRENATAL EVALUATION OF CONJOINED TWINS

Multiple births:

- **Definition:**

*M*ultiple 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*).

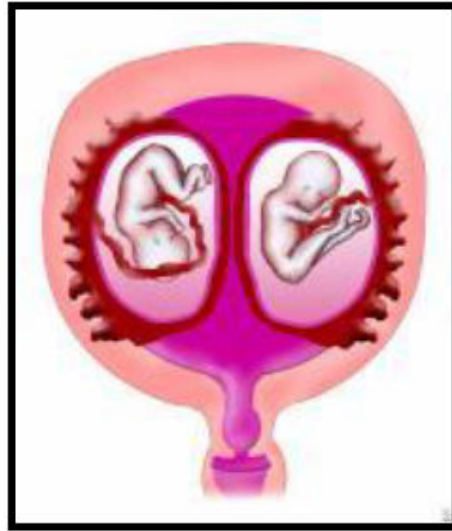


Figure (1): Diamniotic/dichorionic placentation (*Qiu et al., 2008*).

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).

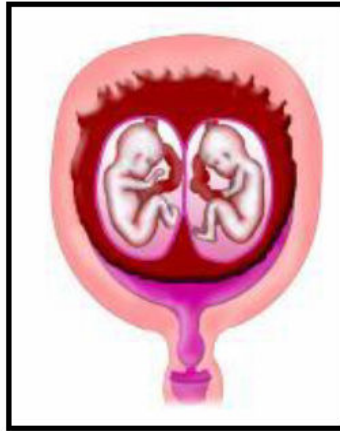


Figure (2): Diamniotic/monochorionic placentation (*Qiu et al., 2008*).

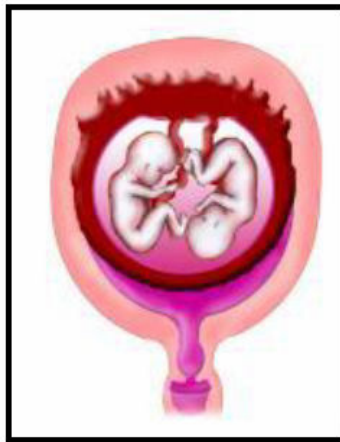


Figure (3): Monoamniotic/monochorionic placentation (*Qiu et al., 2008*).

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 cotriple derived from a different zygote. Finally, 2 consecutive zygotic splittings with a vanished fetus can also result in monozygotic triplets. Zygoty 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 zygoty (*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*).

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., 2007*).

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