DICEPHALY - A COMPLICATION OF TWIN PREGNANCY: A CASE REPORT

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ABSTRACT: A twin is one of the two offsprings produced in the same pregnancy.[1] Twins can either be monozygotic (identical) meaning that they develop from one zygote that splits and forms two embryos, or develop from two eggs, each fertilized by separate sperm cells.[2] Women who have family history of fraternal twins have a higher chance of producing fraternal twins themselves, as there is a genetically linked tendency to hyper-ovulate. There is no known genetic link for identical twinning.[3] Conjoined twinning is a rare phenomenon, occurrence being 1 in 50000 to 100000. However since 60% are stillborn or die shortly after, the true incidence is around 1 in 200000 live births.[4]
KEYWORDS: Monozygotic, Dizygotic, Hyper-ovulate, Twins.

INTRODUCTION: Conjoined twins form when one sperm fertilizes one egg, but at some point during the stage where the single egg splits, the process stops, and the twins develop attached to one another. Identical twins will only be conjoined if they separate after the 12th day of conception. No one knows why conjoined twins occur, although genetic and environmental factors have been explored. Conjoined twins are always of the same sex, same types (dicephalus & ischiopagus for example) may only have one set of external genitalia (or none at all).[5] They are most likely to occur in India or Africa than in China or the US (although the rates in Vietnam have been much higher in recent years, possibly due to Agent Orange exposure).
The conjoined twins when united laterally can lead to types like:

a) Parapagy - twins that share a conjoined pelvics, one symphysis pubis and one or two sacrum's united side by side. This type account for about 5%. It is possible for them to share a heart (if one has situs inversus of the heart), but rare. They may share the liver, kidneys and other organs.
b) Dithoracic parapagus - when the union is united to the abdomen and pelvis (does not involve thorax). This is a subset of parapagus twins. They will be of the dipus type. They will likely to have their own hearts, although some will have situs inversus and would possibly share a heart as well.
c) Dicephalic parapagus (dicephalus) - when the union includes the abdomen, pelvis and thorax. Dicephalus have one trunk with two heads. They are always of the dipus (two legs) type, and 3rd and 4th arms may present on their backs. This is a subset of parapagus twins. They may share a heart and other organs. The rarest type of pappagus twin is dicephalus dipus dibrachius.

- Dibrachius- two arms are present (about 10% of all dicephallus/dithoracic twins).
- Tribrachius-three arms are present (about 10% of all dicephalus/dithoracictwins).
- Tetrabrachius-four arms are present (about 80% of all dicephalus/dithoracic twins).

d) Diproscopic parapagus- a single trunk and single head with two faces. Various level of duplication of the face and brain can be seen. This is the most uncommon and rare type of conjoined twinning, also called as Monocephalic Diprosopos.[5]

MATERIALS AND METHODS: A still born female baby of 32 weeks gestation was donated to the department of anatomy, BMCRI, Bangalore on 25/5/2013 for preservation in museum. It was sent from a primary health care hospital, Singasandra, Kengeri, Bangalore. The only case history obtained was that it was still born on 25/5/2013 at 12:50pm. History of consanguineous marriage was obtained. The baby was dicephalic, dipus and dibrachius. Of the dicephalic heads, one was normal and other was anencephalic head with spina bifida and meningomyelocele. Eye balls of anencephalic head were protruding. Nasal orifice and oral orifices were normal. External ear of normal head on right side was normal while that of left side was smaller and compressed between two heads. Anencephalic head right side external ear was small in size being compressed between two heads that of left side was normal. A single umbilical cord was noted.

CASE PRESENTATION: A dead fetus with the feature of a rare clinical condition conjoined twin with dicephalus was noted. One head had a normally formed cranium and the other head was anencephalic. The accessory anencephalic head was located in the crook between the left shoulder and the neck of the normally formed head. There was a single thorax, single abdomen, two neurologically independent upper limbs, one complement of genitalia and an anus as well as two neurologically independent lower limbs.

Post-mortem plain X-Ray findings showed a fully developed cranium with normal facial structure continuous with the main body. The second head was devoid of a cranium. Each cranium was connected via a separate spine. The accessory anencephalic spin terminated abruptly at the sacral area. Right clavicle was elongated entering the neck region of the accessory head.

AUTOPSY FINDINGS: The head with normal calvaria had a well formed brain whereas the anencephalic head had no forebrain. Two complement of neck organs and two cervical vertebral columns were demonstrable. The right sided trachea continued to a right sided pair of normal lung. The left trachea split to join left normal lung and collapsed hypoplastic lung tissue found between right and left lungs behind the heart. A single intestinal tract opened to the exterior as a well-formed anal canal. Marked hepatomegaly occupying upper half of abdomen was noted.

DISCUSSION: The relationship between conjoined twinning and anencephaly is not well understood. However, it has been observed that the incidence of congenital malformations is significantly increased in conjoined twinning probably due to the later incomplete fusion of the monozygotic embryo during embryogenesis or due to secondary union of two originally separate

monovular embryonic discs.[6] For this reason, it is claimed that the same aetiological factor could be responsible for both the conjoining process and congenital malformations.[7] Consequently there is failure of the neural tube at the cranial end during the fourth week of the development[8] causing the forebrain primordium to be abnormal and the development of the calvaria to be defective. This gives rise to anencephaly which is a fatal disorder. This disorder is also associated with a high risk of preterm delivery before 32 weeks due to the development of polyhydraminos,[9] possibly due to the fetuses lacking the neural control necessary for swallowing amniotic fluid.[10]

Conjoined twins occur in approximately 1 in 40,000 births, but only once in every 1,00,000 to 2,00,000 live births.[5] We therefore agree with Owolabi et al that termination of pregnancy should be advised in cases where dicephalic twins are detected early in utero, especially if there is discordance for anencephaly as in our case.[11] Screening the serum of pregnant women at 16 to18th week of gestation for alpha-fetoprotein can result in the detection of about 80% of fetuses with anencephaly and other neural tube defects.[12]

CONCLUSION: Many congenital defects of interest can now be detected before birth. A severe lesion as that found in our index case, which is incompatible with postnatal life, requires counseling. If detected on monitored antenatal care, it may indicate termination of pregnancy. Prenatally diagnosed dicephalus conjoined twins discordant for anencephaly have been reported but is rare.[13] It is also rare for such an anomaly to escape antepartum diagnosis with the current antenatal screening tests carried out in developed countries.

RECENT STUDIES:

- Establishment of anteroposterior axis is signated by cells at the cranial margin of the embryonic disc. This area, the anterior visceral endoderm (AVE), expresses genes essential for head formation, including the transcription factors OTX2, L1M1 and HESX1 and the secreted factors cerebrus and lefty which inhibit nodal activity in the cranial end of the embryo. These genes establish the cranial end of the embryo before gastrulation. [pg. 52, 12th edition T. W. SALDER LANGMAN'S Medical Embryology].
- If the gene Goosewid is over expressed in frog embryos, the result is a two-headed tadpole. Perhaps over expression of this gene explains the origin of this type of conjoined twins. [pg. 55. 12th edition T.W. SALDER LANGMAN'S Medical Embryology].

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Fig. 1

Fig. 2

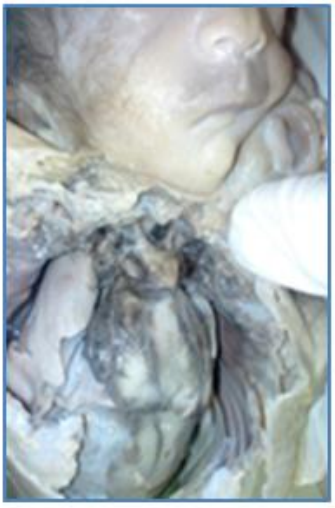

Fig. 3

Parameters	Normal head	Anencephalic head
Head circumference	27cms	14cms
Biparietal	10cms	6cms
Antero-posterior (nasion-inion)	15cms	5cms

Table – 1: Measurements

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