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Dicephalus dipus dibrachius An unusual case of conjoint twins

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Abstract : A conjoint twin is a rare presentation occurring in 140000 births and 1100000 to 200000 of live births. Its occurs due to late twinning events after 13th day of fertilization. Nearly 60 of conjoint twins are still born and 35 die within 24 hrs of life. Incidence is more among girls. Here we report an extremely rare form of conjoint twinning namely dicephalus dipus dibrachius parapagus twins which had two heads with anencephaly, two arms, two legs and single thorax and abdomen with duplication of spine. This case is reported due to its rarity.

Keyword : Conjoint twins, monochorionic, Dicephalus, dipus

INTRODUCTION:

Conjoined twins result from late twinning events when the body axes have been molecularly specified and are beginning to separate.(1)The study of conjoined twins is important because they may be diagnosed prenatally and may be surgically separable. The usual types of conjoined twins are thoracopagus, xiphipagus, pygopagus, craniopagus and ischiopagus. The case presented here is a rare form of conjoint twins with two heads, two arms and two legs with spinal duplication.

CASE REPORT:

A 23 year old primi gravida with non-consanguinity, presented with labour pains and draining per vaginum for 7 hours. She was not booked and registered, had no antenatal visits, iron and folic acid tablets, or had any antenatal USG examinations throughout her pregnancy. No prior miscarriages. There was no history of diabetes, hypertension, radiation exposure and maternal intake of drugs during pregnancy. Period of infertility for 3 years and conceived spontaneously without any assisted reproductive techniques. We received stillborn preterm female conjoint twins weighing 500gms delivered by normal vaginal route with **two heads, two arms, two legs, single thorax, abdomen and pelvis** (figure 1).

Figure 1. Dicephalus Conjoint twins showing two heads, Two arms and two legs With single thorax and abdomen

Both heads showed anencephaly (figure 2). There was **duplication of spine** with fusion at the lower lumbar region. **Single umbilical cord with single umbilical artery** was present. Placenta was monochorionic weighing 600 grams.

Figure 2. Anencephaly with duplication of spine

Figure 2. Anencephaly with duplication of spine

Figure 3. monochorionic placenta with Single umbilical cord and single Umbilical artery.

DISCUSSION:

Conjoined twins are particularly important because they may be diagnosed prenatally and can be surgically separable.(2) The incidence of conjoint twins is 1:50000 births of which duplicata incompleta is 1:100000 to 200000 of live births more common in females.(1,2)Conjoint twins may be caused by factors being influenced by genetic and environmental conditions . Assisted conception techniques such as IVF and ICSI may be a factor. (5) Currently the role of **HOX** and **PAX** genes in early embryogenesis are being evaluated.

EMBRYOLOGY:

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.(2) 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. (3,4) Conjoint twins are classified based on their ventral, lateral and dorsal point of union. Dicephalus is a lateral union-terata catadidyma and is further classified according to their extremities such as di, tri, tetrabrachius etc. (6)

ANTENATAL DIAGNOSIS:

Ultrasonographic identification of any of the following classical signs may suggest the diagnosis: Lack of separating membrane,

inability to separate fetal bodies, 3 or more vessels in cord, or alternatively 2-vessels cord (7), both fetal heads in the same plane, unusual backward flexion of the cervical spine, no change in the relative position after maternal movement and manual manipulations and inability to separate fetal bodies after careful observation.(8)Most frequent anomalies associated with conjoined twinning are duplication of visceral organs, omphalocele, facial clefts, meningomyelocele and imperforate anus.

(5) **Cardiac defects** such as single atrium, single ventricle, hypoplastic left ventricle,TAPVC, pulmonary and tricuspid atresia are frequently reported findings.(9) Conjoined twins show extensive sharing of the common viscera. The placenta of conjoined twins is always monochorionic. (10) A single umbilical artery is a common finding. Because division is so late and incomplete, only a few organ systems are duplicated. This makes surgical division impossible. Many variations of conjoined twins are possible.

DIFFERENTIAL DIAGNOSIS:

Multiple gestation, Teratoma, Cystic hygroma, Neoplasm, Parasitic twin.

CONCLUSION:

This case was reported due its extremely rare presentation with fewer than 70 cases being reported in the literature. (8) A severe defect such as found in our case, which is incompatible with postnatal life, requires counselling. If detected early enough during a properly monitored antenatal care, it may indicate termination of pregnancy. Moreoverthis mother who hasn't registered her pregnancy denotes lacunae in our antenatal coverage and needs concern.

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