

Research Article



A Pair of Craniopagus Twins (O' Connell type III TV CPT)- a Puzzle of Nature Fascinating Neuroscientists and Embryologists

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ABSTRACT

Conjoined twins are monozygotic twins which develop from a single fertilized ovum. The zygote forms a blastocyst in which the inner cell mass splits into two identical masses which in turn develop into fetuses. They have a monochorionic monoamniotic placenta. The twins are of same sex and look alike. They have one placenta with two umbilical cords. The two partners can be completely separated by appropriate surgery if each of them receives separate vascular supply. They are classified depending on the site at which they are joined. Early diagnosis by ultrasound (US) and magnetic resonance imaging (MRI) is essential for management of conjoint twins. In addition, early counseling of parents and termination of pregnancy if indicated may be offered if serious congenital anomalies are associated.

Keywords: Conjoined Twins, Craniopagus, Ultrasonography

INTRODUCTION

Conjoined twins or double monsters are most uncommon (probably 6-10 per million births), highly fascinating anomaly of nature. The monozygotic twins develop from a single zygote. It occurs when there is incomplete embryonic division of the blastocyst occurs in the 2nd week of gestation resulting in varying degree of fusion between two fetus¹. The incidence of conjoint twins is 1 in 50000 to 1 in 100000 deliveries^{2,3}. The ratio between male and female is 3:1^{2,3}. The conjoint twins raise religious, moral, legal and ethical issues.^{4,5} Craniopagus twins are uncommon malformation found once in 2.5 million live births and represent only 2-6%⁶ of conjoint twins.

Approximately 40% of conjoined twins are stillborn and an additional one-third die within 24 hours, usually from congenital organ anomalies, leaving perhaps 25% to be considered for surgical separation^{7,8}. Probably 60%-40% die during pregnancy (Twin transfusion syndrome, parasitic twins, etc.) and most of the rest 35-40% do not survive due to complication at parturition and associated serious congenital anomalies⁹.

The incidence of different types of conjoint twins in the department of obstetrics and gynecology in SCB medical college diagnosed in 3rd trimester of pregnancy represented in table 1. Ref¹⁰

Case Report

A 24 year old primigravida, of poor socioeconomic status from Phiringia, Phulbani, Odisha, India was admitted to department of Obstetrics & Gynecology, DHH (District headquarters hospital), Phulbani. At 38th week of gestation the mother had undergone four ANC (antenatal

checkups) without any antenatal ultrasound. The mother gave birth to two babies by normal vaginal delivery, where the heads are joined together, at the vertex. The inter twin longitudinal angle is 90 degree Hence a TV type III CPT (CranoPagus Twins); (O' Connell classification).¹¹

The two babies cried immediately after birth and weight is 4 kg. There is no family history of twinning on either maternal or paternal sides and no history of exposure to X- ray, antenatally.

The meconium and urine were passed on day 1 and umbilical stump fell on day 4.

There was a single placenta and two separate umbilical cords. Both the babies had a normal cardio respiratory system and didn't need any support.

Table 1

Types	Incidence	Site of union
Thoracopagus	33%	Chest
Thoraco-omphalopagus	16%	Chest & abdomen
Dicephaius	11%	Lower body
Cephalothoracopagus	6%	Head and chest
Craniopagus	5%	Head
Pyopagus	4%	Sacrum
Ischiopagus	3%	Pelvis
Other combination	22%	-----



DISCUSSION

The exact etiology of conjoined twins is still a mystery. However there are two theories explaining the etiology. First is the “fusion theory” in which the embryonic tissue divides incompletely and remains fused incompletely at some point. Second is the “collision theory” in which the two separated embryonic masses fuse before tissues

differentiation. The former one is more acceptable.

At about 2nd week, in the normal twinning process, the inner cell mass splits into two separate and nearly equal masses, each producing a single individual¹². Conjoined twins are monozygous and therefore will be of same sex and usually with monochorionic and monoamniotic placenta⁹.

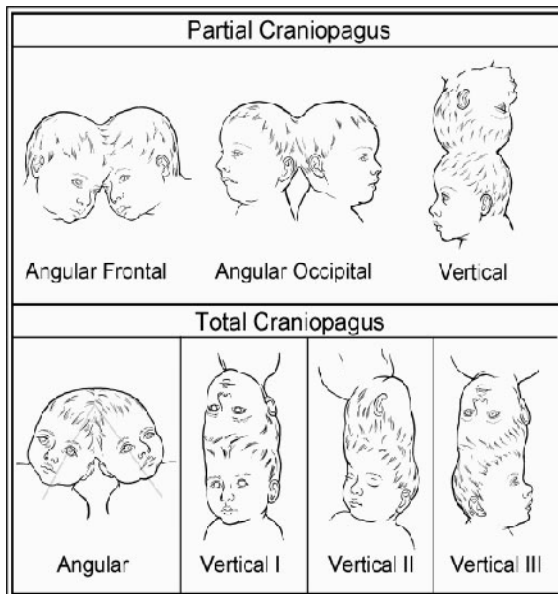


Fig 2, ref¹¹

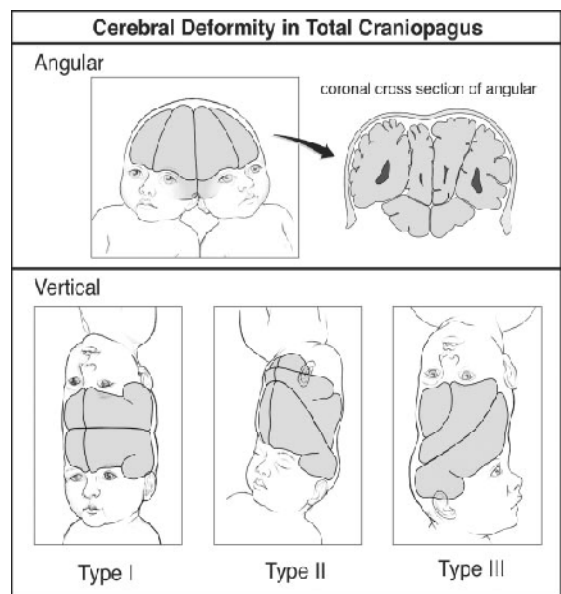


Fig 3, ref¹¹

In CPT the face and foramen magnum are not primarily involved, the skulls are usually joined in roughly homologous regions but asymmetries are also common¹³.

Types found may be classified depending on whether a significantly shared dural venous sinus system- SDVS is present- (total versus. partial): TA, TV, PA & PV (Total Angular, Total Vertical, Partial Angular, Partial Vertical)¹¹.

In Partial CPT, the unions are usually frontal^{14,15} and less commonly occipital^{16,17,19} or vertical biparietal¹⁸. The junctional diameter is often smaller in the partial forms

and occasionally an incomplete layer of bone may be present between the twins^{19,14}.

Partial craniopagus is thus essentially an extracranial abnormality and total vertical craniopagus a gross intracranial one. The partial unions may be frontal, parietal, or occipital; Total craniopagus is, from its nature, likely to be biparietal and, since it is a very wide union, it is thought to be better described as vertical.

Type I- The rotation of one head on the other is minimal and the faces are directed to the same side with inter-twin axial rotation < 40 degrees²⁰⁻²⁴. Type II- The rotation is

maximal (140-180degrees) and the faces are directed to opposite sides²⁵⁻²⁷. Type III-The rotation is (40 to <140) degrees; intermediate between the other two types.^{28,29}

Some CPT twins existed as total angular with more acute inter-twin longitudinal angulation and SDVS accompanied by complex interconnecting venous channels (or CVS), and markedly distorted cerebral hemispheres³⁰⁻³². Partial CPT children usually undergo successful separation at an earlier age than total CPT, and the separation more often results in survival of both children who may lead normal lives. Anomalies of other organ systems may jeopardize life.^{19,32}

The suitable time of separation of conjoined twins are related to the condition of those twins and to the type of twins³³. The optimal age of separation is 3-12 months.

CONCLUSION

The early diagnosis of conjoined twins is very rare, until the date of parturition. It is mostly found in rural settings where the modern diagnostic procedures are out of reach. Ultrasonography is the screening method for diagnosis of these malformations. The prognosis of conjoined twins depends on the degree of union and number of shared organs. CPT twins have significant brain compression which is an obstacle for the normal structural growth and development of the brain. CPT happens to be the most challenging work for the neurosurgeon, neuroanesthetics etc for successful separation of the babies. It is also important for the embryologist and anatomists for the developmental anomaly of the fetus.

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