Craniopagus

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In craniopagus parasiticus, a parasitic twin head with an undeveloped body is attached to the head of a developed twin.

Fewer than a dozen cases of this type of conjoined twin have been documented in literature, occurring in about 2 to 3 of 5,000,000 births [1)]

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Development

The exact development of craniopagus parasiticus is not well known. However, it is known that the underdeveloped twin is a parasitic twin. Parasitic twins are known to occur in utero when monozygotic twins start to develop as an embryo, but the embryo fails to completely split. When this happens, one

embryo will dominate development, while the other's development is severely altered. The key difference between a parasitic twin and conjoined twins is that in parasitic twins, one twin, the parasite, stops development during gestation, whereas the other twin, the autosite, develops completely

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In normal monozygotic twin development, one egg is fertilized by a single sperm. The egg will then completely split into two, normally at the two-cell stage. If the egg splits in the early blastocyst stage, two inner cell masses will be present, eventually leading to the twins sharing the same chorion and placenta, but with separate amnions. However, the egg can split into two, but still have one blastocyst. This will lead to one inner cell mass and one blastocyst. Then, as the twins develop, they will share the same placenta, chorion, and amnion.

Nursing care

A multidisciplinary group of professionals in a tertiary paediatric care setting in Italy developed a comprehensive protocol for the nursing care of craniopagus twins in hospital, informed by the findings of a literature review and the expertise of its members. The protocol was applied to the management of a pair of craniopagus twins who successfully underwent separation surgery at the authors' hospital. The protocol describes the crucial role of nurses in the care of craniopagus twins and their families before, during and after separation surgery [2)]

Case reports

A paper of Caceres et al. reviewed the plausible etiological mechanisms, clinical features, preoperative analysis, and documented modern-day craniopagus parasiticus surgical separation attempts as well as an historical review of the few cases documented in the literature.

They describe the successful separation of a 28-week preterm newborn from its parasite sibling twin bearing lethal congenital anomalies associated to Cantrell's pentad and sirenomelia. Description of the case, plausible explanations on the mechanisms of conjointment along with the associated congenital abnormalities of the deceased twin are examined along with an historical revision of craniopagus parasiticus and their separation attempts with special attention to the previously undocumented attempt of the Dominican CP separation surgery by Lazareff et al. RESULTS: The use of the deceased twin cranial vault tissues (skin, bone, and duramater) as an autologous implant due to the identical genetical profile served to remodel and close the skull of the surviving twin with good esthetic results and no tissue rejection. To our knowledge, this is the youngest preterm set of craniopagus parasiticus separated in an emergency fashion with good functional and esthetic outcome.

Craniopagus parasiticus is an infrequent subvariant of this rare form of twin conjointment which may require urgent separation due to the associated malformations of the parasitic twin; therefore, the fact that both siblings are genetically identical may prove as an advantage to use duramater, bone, and soft tissues from the parasitic twin as ideal grafts for covering the resultant defect after the separation has been performed [3)].

A case of craniopagus parasiticus, diagnosed at 16 weeks of gestation by ultrasound screening. To the best of authors knowledge, the case that we present is the first CP case that was diagnosed at such an early gestational age. The formed fetus was found to harbour complex cardiac anomalies. In view of poor prognosis of survival after delivery and upon permission from the couple, the pregnancy was terminated at 17 weeks of gestation [4)].

A 38-year-old multigravida (gravida V para IV) woman of Amhara ethnicity was referred from a rural health center to our hospital due to prolonged second stage of labor at 42+1 weeks. On her arrival at our hospital, an obstetrician decided to do a caesarean section because she was unable to deliver vaginally. A live baby girl weighing 4200 g was delivered. The placenta was single and normal. Her Appearance, Pulse, Grimace, Activity, and Respiration scores were 7 and 9 at 1 and 5 minutes, respectively. She appeared to be grossly normal except for the parasitic co-twin attached to her cranium. After a week of extensive counselling and investigation, a successful separation operation was done. Postoperation, she comfortably suckled on the breast and had no neurological deficit. Two weeks after separation she was discharged in a good healthy condition with an arrangement for postnatal follow up.

The causes of craniopagus parasiticus are still unknown due to a rarity of cases and a limited number of studies on it. There have been only nine to ten cases of craniopagus parasiticus, of which only three survived past birth and were documented in the literature. Genetic scientists and researchers continue to investigate this case because they might find explanations for the birth defect, and provide answers to improve the prognosis and the life chances of twins with craniopagus [5)]

An apocryphal case of craniopagus parasiticus: the legend of Edward Mordake [6)].

Kansal R, Kale C, Goel A. Craniopagus parasiticus: A rare case. J Clin Neurosci. 2010 Oct;17(10):1351-2. doi: 10.1016/j.jocn.2010.01.053. Epub 2010 Jul 23. PMID: 20655232 [7)].

Lotfy et al. operated on a patient with craniopagus parasiticus at Benha Pediatric Hospital in Egypt, 45 km north of Cairo. The child was 10 months old when the surgery was performed. By minimizing the time of surgery and adequate control of intraoperative bleeding, a successful surgical separation was achieved. Computed tomography, magnetic resonance imaging, magnetic resonance angiography, and computed tomographic angiography provided the information necessary to perform surgery.

The child underwent operation at the age of 10 months; the duration of surgery was 9 hours. Bleeding was the most serious problem, with the child receiving four liters of blood. The main arterial supply to the parasite was via the middle cerebral artery and was ligated in the Sylvian fissure. Bleeding, however, was mostly venous and was mainly controlled by diathermy and thrombin soaked packs of Surgicel, as well as clipping. After separation of the parasitic head, the dura was repaired using artificial dural grafts. Free bone flaps from the parasite were used to cover the osseous defect in the autosite. Skin flaps from the parasite were also used to cover the cranium.

This is the second case of craniopagus parasiticus in which separation was attempted. The first patient, operated on in the Dominican Republic, died 7 hours after surgery. In the present case, the child is still alive and without neurological deficit. [8].

A separation of one set of craniopagus parasiticus conjoined twins was attempted, but abandoned owing to major vascular and brain sharing [9]

A case of craniopagus parasiticus is described in which the parasitic twin is more fully developed anatomically than in any of the previous reports. Somatic and placental vascular anastomoses between the twins and hypoplasia of the umbilical cord of the parasite were also observed. These findings support the hypothesis that craniopagus parasiticus results from compromise of the blood supply to one of a pair of craniopagus conjoined twins [10].

Wang DM, Zhang PL. [A case report of craniopagus parasiticus (clinical features and the histological study of the accessory brain)]. Zhonghua Zheng Xing Shao Shang Wai Ke Za Zhi. 1985 Mar;1(1):31-3. Chinese. PMID: 3939788.

An extremely rare case of parasitic head protruding from the right side of the face is presented. It differs from all the other cases previously reported in the literature. The patient had a parasitic head with an abnormal brain, two eyebrows, two underdeveloped eyes, nose, mouth, 12 teeth, a tongue and plenty of hair. The parasitic head was excised. The nature of the embryological abnormalities is discussed and the literature of polygnathism reviewed [11].

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