Case Report
Successful separation of craniopagus conjoined twins


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ABSTRACT
Craniopagus conjoined twins represent a rare phenomenon of congenital malformation/dysmorphism. The clinical pathology of this complex entity is reviewed and placed in perspective. Confusion surrounds the severity of craniopagus conjoined twins especially in relation to the difficulty of separation and subsequent outcome. Successful separation of craniopagus twins remains a rarity, however modern neurosurgical techniques have created opportunities for successful separation and brought hope for a normal survival of these children who in the past were often left as historical footnotes or put on display as oddities of nature. We report on a craniopagus conjoined twins from Sudan who had been successfully separated.

Key Words:
Craniopagus; Conjoined; Twins; Sudan.

INTRODUCTION
Conjoined twinning is one of the most challenging human malformations. Treating conjoined twins can be a daunting challenge for the surgeon. Moreover, these cases often raise religious, moral, ethical and legal issues [1,2].

Craniopagus twins (CPT) i.e. twins joined at the head are an uncommon malformation found once in 2.5 million live births and represent only 2–6% of conjoined twins [3]. 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 [4,5]. Nevertheless, worldwide a handful of CPT separation attempts occur yearly. In the last half-century, with the many advances in medicine including brain imaging, neuroanaesthesia and neurosurgical techniques, a successful outcome is possible
following separation of total CPT. In CPT, the face
and foramen magnum are not primarily involved, the
skulls are usually joined in roughly homologous re-
gions but asymmetries are common, and both vertical
and non-vertical or angular forms are found [6,7].
Stone and Goodrich [8] have recently sub classified
craniopagus into four varieties, depending on whether
a significantly shared dural venous sinus system (total
versus. partial) is present, and whether the inter-twin
longitudinal angulation is below 140 degrees.
We report on craniopagus female twins from Sudan who
had been successfully separated. To our knowledge this is
the first case report of craniopagus twins from Sudan.

CASE REPORT
The diagnosis of CPT was made during routine ante-
natal ultrasound scan in the first trimester. The mother
was a 26-year-old primigravida. The delivery was by
an elective Caesarian section at 36 weeks gestation
on the 22nd of September 2010. The two babies cried
immediately after birth with Apgar scores of nine at
one and five minutes for both. Birth weight was 6.5
kg. The twins were transferred to the special care
baby unit for further assessment. Clinical examina-
tion revealed that both babies were well, sharing one
cranium vertically with completely separate bodies
on each side (Figure 1). There were no dysmorphic
features and systemic examination was normal for
both. Initially the babies were kept on intravenous
10% Dextrose and later started on breast-feeding.
Investigations including complete blood counts, liver
and renal function tests, cranial and abdominal ul-
trasound were normal. Echocardiography revealed
normal anatomy for both hearts. A 3D cranial com-
puted tomography (CT) with reconstruction showed
craniopagus conjoined twins (vertical type 3) sharing
a single bony cranium with defective parietal bones.
Magnetic resonance imaging (MRI) showed cere-
brospinal fluid (CSF) cleft communicating between
the ventricular systems through the posterior parietal
region. Magnetic resonance venography (Figure 2)
revealed evidence of venous sharing through the su-
perior sagittal sinus at its posterior occipital extent.
Magnetic resonance angiography (MRA) showed no
obvious arterial sharing. Both babies passed urine and
meconium normally. The twins were discharged home
and arrangements were made for surgery in UK.
The parents contacted the charity organization, Facing The
World (http://facingtheworld.net/), through the BBC which

Figure 1 - Craniopagus conjoined twins at birth.

Figure 2 - Magnetic resonance venography (MRV) showing
evidence of venous sharing through the superior sagittal
sinus at its posterior occipital extent. (SSS=superior sagittal
sinus; TS= transverse sinus; SS= sagittal sinus).
made all the necessary arrangements to transfer the twins to
the United Kingdom accompanied by their parents.
The surgeons at Great Ormond Street Hospital in
London agreed to do this extremely hazardous sur-
gery. The surgery took place in four stages, two op-
erations in May, one in July and the final one was in
August 2011. Tissue expanders were inserted in July
2011. The post operative course was uneventful and
both girls were able to go to the general ward within a
few days with no residual neurological deficit. Figure
3 shows the twins after separation at the age of 11
months interacting and playing.

Figure 3 - The craniopagus conjoined Sudanese twins
(Rital and Ritaj) at the age of 11 months after successful
separation.

DISCUSSION
A number of reviews have detailed the complex an-
tomical and surgical problems in separating total
forms of craniopagus conjoined twins, notably shared
dural venous sinuses and the subsequent negative im-
lications towards survival and quality of life [9,10].
Experience with large numbers of conjoined twins is
limited to a few centers in the world (13 sets described
by O’Neill et al [11] from the Children’s Hospital of
Philadelphia, 14 sets reported by Mackenzie et al [12]
from the same hospital, 17 sets reported by Spitz and
Kiely [13] from Great Ormond Street Hospital, Lon-
don, and 46 sets reported by Rode et al [14] from Red
Cross Children’s Hospital, Cape Town, South Africa.
Recently, surgical experience with conjoined twins
has been reported in other parts of the world, such as
China, Saudi Arabia, and New Zealand [15-18].
There are few examples of successful and unsuccess-
ful separation of craniopagus twins, notably in adult
life. In Singapore in 2003, skull and brain separation
of 29-year-old craniopagus Iranian twins was unsuc-
cessful, and the sisters died of exsanguinations on the
operating table [17].
Craniopagus conjoined twins from Egypt were sep-
parated at the Children’s Medical Center in Dallas,
Texas. They were born in Egypt on June 2, 2001,
and were separated on October 12, 2003, during a
34-hour operation [19]. The operation involved skin
grafting and separation of the venous sinuses. Later,
they underwent skull reconstructive surgery, received
rehabilitation therapy, and, finally, left for Egypt on
November 19, 2005. Another successfully staged sep-
aration of craniopagus twins took place from 2003-
2004 at Montefiore Medical Center in New York [20].
Conjoined twins is one of the most intriguing and pro-
vocative anomaly that has challenged mankind over
the millennia. During the past half century craniopa-
gus twins have challenged the capability of neurosur-
geons. With modern neuroimaging and neurosurgical
techniques it is now possible to separate such twins
successfully.

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REFERENCES