Turkish Neurosurgery 2007, Vol: 17, No: 1, 27-32

Craniopagus, Laleh and Ladan Twins, Sagital Sinus

Zahid Husein KHAN[1]
Saeed HAMİDİ[2]
Seyed Mojtaba MİRİ[3]

[1] Azad University of Medical Sciences, Neurosurgery, Tehran, Islamic Republic of Iran,
[2] Tehran University of Medical Sciences, Neuroanesthesiology, Tehran, Islamic Republic of Iran
[3] Tehran University of Medical Sciences, Neurosurgery, Tehran, Islamic Republic of Iran

ABSTRACT

Adult craniopagus twins aged 29 years who succumbed to the surgical separation are being presented posthumously. Neuroimaging studies had revealed a total type 3 craniopagus with a common superior sagittal sinus beginning at its distal half and draining into a common Torcular Herophili. In July 2003, neurosurgeons at Singapore embarked on the mission to tackle the separation of the most complex and challenging adult craniopagus of the present millennium, unfortunately ending up with devastating results. The authors intend to expand upon their life style, shared intracranial structures, the potential risks of a one-stage procedure as carried out in this case and finally offer some suggestions in tackling such rare and complex cases in neurosurgery.

KEY WORDS: Craniopagus; Laleh and Ladan twins; Sagittal sinus; Unsuccessful separation.

Received: 31.01.2007
Accepted:13.02.2007

Correspondence Address:
Zahid Husein KHAN
Tehran University of Medical Sciences, Neuroanesthesiology, Tehran, Islamic Republic of Iran
E-mail : zhkhan51@yahoo.com

INTRODUCTION

Craniopagus twins is an exceedingly rare congenital anomaly occurring at a frequency of 4-6 per 10 million live births (24). Conjoined craniopagus twins might be due to development of two fetuses from the primary zygote but with incomplete cleavage at their craniums at the end of the 2nd week of gestation while some believe the abnormality is a result of the fusion of two separate embryos, with the junction occurring in the open cranial neuropore just before the end of the 4th week after fertilization (26). Conjoined twins are always genetically identical and share the same sex. Females are more commonly affected, with a male/female ratio of 1:4 (6). These twins can be joined at the vertex, at the side or at the forehead, the vertical type being the most common (23).

O'Connell's classification denotes three anatomical types for vertical craniopagus, based on relative facial orientation (type 1: face same direction; type 2: face opposite direction (140-180 degrees) and type 3: intermediate angle of rotation of the long axis of one head on that of the other) (21). Again, based on the continuity of the scalp down to the central nervous system, the craniopagi are classified into various types (type A: connected by the scalp and subcutaneous tissues and perhaps with bony fusion; type B: sharing dura matter, but not leptomeninges or brain; type C: sharing leptomeninges; and type D: a structurally continuous central nervous system) (30). A complex type III is a new subdivision forwarded by Khan et al (15) to elucidate their craniopagus twins sharing the interhemispheric fissure. Classification of craniopagi by the portions of crania that are connected (e.g. craniopagus parietalis, frontalis or occipitalis and temporoparietalis) (3) is descriptive, can be applied on first inspection, and is appealingly simple.

Surgical separation has often led to frustrating results, i.e., loss of one of the twins or both. Craniopagi reported so far in the literature are

being compiled (Table I), and their ultimate outcomes are being tabulated and catalogued. Since most of the surgical separations conducted so far have been carried out without the patients' consent or knowledge because of the young age, this particular case of adult craniopagus twins was all the more challenging and difficult, because the twins had lived together for almost 28 years, were fully grown up, totally aware of their surroundings and therefore would not surrender to surgical separation unless thoroughly briefed and fully appraised of the potential complications ingrained in such types of procedures.

Table I: A historical overview of craniopagi separation

Year	Outcome	Report
1505	First division (one twin dead preop, the second dead 3 days postop.)	Munster et al.(20)
1928	Maiden attempt in 20th century. Both died on day of operation	Cameron et al.(4)
1949	First pair surviving separation(died 3 hrs postop.)	Barbosa et al.(2)
1953	First prolonged survival of one child	Grossman et al.(13)
1956	Successful (normal growth and development)	Baldwin et al.(1)
1957	First prolonged survival of both children	Voris et al.(28)
1964	One child lost during separation	O'Connell et al.(22)
1984	One twin discharged well. Second twin had severe developmental delay	Shively et al.(25)
1987	One severely developmentally delayed	Bucholz et al.(3)
1989	Both twins with severe neurological and developmental delay	Cameron et al.(5)
1991	Successful separation of both twins	Drammond et al.(9)
1991	Successful separation of both twins	Konovalov et al.(16)
1995	Successful separation of both twins, one died a month later	Rutka et al.(23)
1997	Unsuccessful separation	Maroof et al.(19)
1997	Prenatal diagnosis. Aborted	Costea et al.(8)
1997	Successful separation, one twin was delayed in development	Walker et al.(29)
1999	Both twins died during separation	Khan et al.(15)
2001	Minor disability in one twin and severe developmental delay in the other	Goh(10)
2003	Adult craniopagus. Both died during separation	Carson BS (7)
2004	Successful separation	Goodrich et al.(12)
2005	Craniopagus Parasiticus. Successful separation	Lotfy et al.(18)

The surgical separation was finally conducted in July 2003 at Singapore after a battery of sophisticated investigations and days of academic discussions, but despite the surgeons' and the public optimism the results were devastating, leading to the death of both twins on the operating table.

The authors deemed it necessary to add this exceedingly rare case of adult craniopagus to the realms of contemporary medical literature with a special emphasis on the different personalities of the twins, the complexity of this particular craniopagus and finally the imminent risks in a one-stage separation. Such rarely performed and complex surgeries should be submitted to medical journals for the purposes of archiving such events, advancing scientific knowledge, and helping future clinicians who may have to treat such cases (11).

CASE REPORT

Female craniopagus twins aged 29 who had finished their law course, and were normally built and looked healthy except for a massive attachment at the temporo-parieto-occipital region (Fig 1) are presented. Their attachment and the fact that they had managed to live to their third decade make them unique and probably never to be seen again.

While walking, their cranial attachment would pull the twins' neck and upper chest centripetally and the rest of their bodies flaring out centrifugally, a posture that would impart considerable pain and distortion to their nuchal muscles, the cervical vertebrae and the upper thoracic vertebral column. During childhood and adolescence, the two girls adapted themselves reasonably well to changing situations at home and in public life. They were of the opinion that under the most difficult circumstances, one should have a goal. They liked painting, flowers and cycling. During cycling, one would ride the bicycle whereas the other one would happily run on foot to keep pace with the rider. At last, fed up with this unnatural way of living coupled with innumerable psychological, social and privacy matters, they willingly and knowingly opted for separation choosing their own surgeons. The neurosurgical team at Singapore embarked on this mission to separate the most complex adult craniopagus of the present millennium but the results were devastating leaving an impression in the

minds of the medical community that there are some places and surgical horizons where the stakes are extremely high.

SKULL AND BRAIN ANATOMY OF THE TWINS

SKULL ANATOMY

Anatomically, the two skulls were attached anteriorly at the mastoid region, along with an attachment at the squamous part of the temporal bone. The auricles of the twins were explicitly separate but dysplastic changes could be seen in the middle and inner ears. The parietal bone at the place of junction of the twins was absent altogether, and a single cranial cavity was formed. However, the brains of the twins were demarcated. The posterior fossae of the twins were protected by a single occipital bone (Figure 1).

Figure 1: Anterior view of the twins, Face opposite direction.

NEUROVASCULAR ANATOMY

The anterior and posterior cerebral circulations of the twins did not have major anatomical variations that could be of clinical and surgical significance except for a terminal branch of the middle cerebral arteries anastomosed at the conjoined side. This anastomosis had a preferential flow from Laleh's to Ladan's MCA (17). The most complex communication was in the major cerebral superficial sinuses which made separation a distant reality and a utopian dream.

The superior sagittal sinuses (SSS), after some interconnection at the end of the middle third, eventually became connected to each other to form a conjoined SSS (CSSS). The straight sinuses, after traversing a short steep ascendant course, supracallosally opened in the CSSS 1.5 centimeter after fusion (17) and collectively formed torcular herophili (TH), the latter extending from CSSS at the straight sinuses and opening to both the transverse and tentorial sinus opening.

The Torcular Herophili (TH) which drains both the superficial and the deep venous sinuses was single in both twins, thus making a surgical decision of separation exceedingly difficult. Two of the transverse sinuses (TS) that drain the TH were visible centrifugally, but that at the conjoined side formed a common lake not depicted clearly in sinography. As the two cerebellar hemispheres were enclosed in a single occipital bone, a single occipital sinus existed for the two twins (Figure 2). Details of the vascular anatomy of Laleh and Ladan are provided by Lasjaunias et al (17).

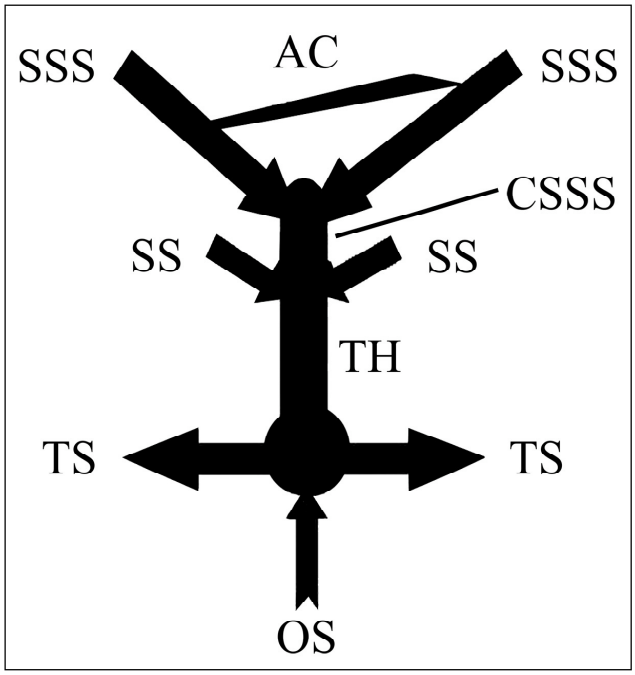

Figure 2: A schematic posterior view of the twins' draining venous sinuses. SSS: Superior Sagittal Sinus; CSSS: Conjoined Superior Sagittal Sinus; AC: Anastomosing Channels; SS: Straight Sinus; TH: Torcular Herophili; TS: Transverse Sinus; OS: Occipital Sinus

BRAIN PARENCHYMAL ANATOMY

Each twin had a well-formed brain right from cerebrum down to the cerebellum, anatomically separated from the adjoining brain (Figure 3) but with a single cranial cavity. The temporal lobe of the brain at the site of junction showed some degree of atrophy.

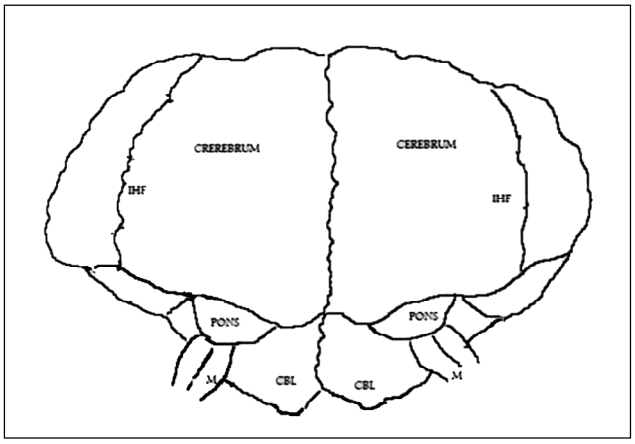

Figure 3: A schematic anterior view of the twin's brains. IHF: Inter Hemispheric Fissure; CBL: Cerebellum; M: Medulla oblongata.

SUGGESTED SURGICAL PROCEDURE FOR SUCH CRANIOPAGI

Leaving aside the issue of Laleh and Ladan, such types of craniopagi should preferably be separated in the first year of their lives. If they reach adulthood. a distant dream, separation becomes difficult or perhaps impossible because of two alive and neurologically intact separated twins. A surgical team forced to undertake the separation procedure for the adult type of craniopagus should carry it out as a staged procedure, and in the case of total craniopagus plan or consider to save one at the expense of the other rather than planning to save both and thereby losing both. This may be the only logical panacea to make such separation promising and possible.

The major problems in separating craniopagi stem from the anatomical interconnections, and the stage or type. If one can transform type III craniopagus into type I (minimal venous, arterial or parenchymal interconnections), the intraoperative complications during surgical separation would be modest and easily manageable. Taking this dictum into consideration, the vascular interconnections should be dealt with first and later in the second and third stages and separation of parenchymal, dural, skull and scalp interconnections carried out. Such a gradual and staged separation permits better adaptation to the new haemostatic conditions.

DISCUSSION

The incredible task of separation of craniopagus twins has been attempted since the last century and the results have sometimes been satisfactory but also discouraging. Of the 48 patients reviewed by Bucholz and colleagues, 24 died in the postoperative period, and 2 were left with severe neurological dysfunction. They considered two determinants of operative survival as the area of junction in craniopagus and the venous connections (3). The present case, by these standards and yardsticks, had all the paraphernalia such as a common sagittal sinus, a massive attachment at the temporo-parieto-occipital region giving rise to osseous anomalies and skulls with the configuration of a partially common cranium to earmark it as a case with the worst prognosis (11, 14). The courage of the neurosurgeons embarking on a mission to tackle the separation of the most complex adult craniopagus of the present millennium at Singapore is admirable, although the results were devastating. What makes surgical separation difficult in such cases is a plethora of major problems such as complex cerebral or cerebellar connections, arterial connections, and above all the most dreaded problem of a common venous drainage system (30). The skull defects and skin scarcity also pose major problems, but they are amenable to modern reconstructive surgical techniques (9).

Employing a single-stage separation that used cardiopulmonary bypass and hypothermic circulatory arrest to separate an occipitally-joined craniopagus resulted in handicapped children with one suffering from a neurological dysfunction, and the other severely retarded (5). Both infants survived the marathon surgery in this attempt, but an alarming ethical issue arises as to whether are we morally permitted to conduct such operations at the expense of ending up with two handicapped infants with neurological dysfunction and severe mental retardation.

When one confronts a total type of craniopagus with cerebral and arteriovenous connections, the issues that stand foremost are neurovascular protection and a justifiable separation (30). The surgical indication for separation in such critical unions may either be gross difficulties incompatible with normal living because of anomalous configuration or else the unavailability of informed consent by the twins or their parents (15). In the case of Laleh and Ladan, there was no gross difficulty or a life-threatening situation that could be construed as an impediment to leading a normal life. Although

there existed no imminent indication for separation, the twins were fed up from living together, and had given their informed consent to undergo separation at the hands of the neurosurgical team whom they trusted and who had an international fame in conducting such a case successfully in the past. Separation should be attempted if there is a possibility of success (22), but success cannot be guaranteed. In Laleh and Ladan's case, the neurosurgeons were highly optimistic prior to surgery, but the grafted channels got thrombosed leading to edema, profuse bleeding and parenchymal engorgement triggering a vicious cascade of events that could not be controlled despite all the options and skills at hand.

Craniopagii Types C and D are difficult cases and are usually the ones who either die during surgical separation, or else suffer neurological injuries. In such cases, it appears appealing and scientific to first divide the shared venous system by gradual occlusion of common venous channels by means of endovascular or epidural balloon occlusion.

As the cerebral and vascular channels are tackled earlier, this procedure would proceed safely and without much of a problem if unexpected events did not occur. In the next session, the brains and their arterial connections are dissected and ligated, and kept apart by a silastic membrane preventing re-adhesions till the last stage (3) while the dura and skin are repaired in an anatomical fashion.

The final surgical session initiated weeks to months later consists of total dural and bony separation. In simple cases, the aforementioned three stages can be performed in a single session, whereas complex connections should preferably be reduced to less complex ones stage by stage, making the separation more tolerable, achievable, and conceivable for the twins (6).

Other intraoperative complications may also occur during such separations. Air embolism is always a risk as large veins (including venous sinuses) may be opened and the blood loss is significant.

Even if the scalp, the sinuses and the bony problems are solved by availing the best expertise and employing state of the art management, the parenchymal problem cannot be solved and would pose itself as the neural conundrum for those dealing with such cases. The strategy of separation however should remain the final option for such type of craniopagii in the case of an imminent danger of death or an evolving life-threatening illness endangering one of the twins.

In conclusion, we would say that adult craniopagi cases who have managed to live all their way to their third decade of life, and are on their way to graduate as licensed lawyers as was the case of Laleh and Ladan, should preferably have been left to lead their lives which although would apparently not have been fully acceptable, and of course devoid of privacy and self satiety, would nevertheless have been reasonably tolerable to appreciate the divinely and heavenly pleasures associated with it (27).

REFERENCES

1. Baldwin M, Dekaban A. The surgical separation of Siamese twins conjoined by the heads (cephalopagus frontalis) followed by normal development. J Neurol Neurosurg Psychiatry 1958; 21:195-202
2. Barbosa A. Tentative cirurgica em um caso de craniopagus. Rev Bras Ciru 1949; 18:1047-50
3. Bucholz RD, Yoon KW, Shively RE.Temporoparietal craniopagus. J Neurosurgery 1987;66: 72- 9
4. Cameron HC. A craniopagus. Lancet 1928;1:284-85
5. Cameron DE, Reitz BA, Carson BS. Separation of craniopagus twins using cardiopulmonary bypass and hypothermic circulatory arrest. J Thorac Cardivasc Surg 1989;98: 961-7
6. Carnpbel S, Theile R, Stuart G, Cheng E, Sinnott S, Pritchard G, Isles A. Separation of craniopagus joined at the occiput. Case report. J Neurosurgery 2002;97: 983- 7
7. Carson BS. Comments; regarding the deaths of twins Ladan and Laleh Bijani. 2003; Available at: www.hopkinschildrens.org/pages/news/presdetails.cfm?newsid=144
8. Costa DM, Vladesco R, Vasiliu C, Leonte N, Virteg P. Craniopagus twins. Acta Obstetrica et Gynecologica Scandinavica 1997; 76:53
9. Drummond G, Mackay SD, Lischitz R. Separation of the Baragwanath craniopagus twins. Br J plast Surg 1991; 44: 49-52
10. Goh KYC. Separation surgery for total vertical Craniopagus twins. Child's Nervous System, 2004; 20(8-9): 567-75
11. Goh KYC. Surgeon's reply. Singapore Med J 2005; 46(7): 2
12. Goodrich JT, Staffenberg DA. Craniopagus twins: clinical and surgical management. Child's Nervous System, 2004; 20(8-9)
13. Grossman HJ, Sugar O, Greeley PW, Sadove MS. Surgical separation in craniopagus. JAMA 1953;153: 201-207
14. Khan ZH. Separated but oceans apart and fathoms deep: The conjoined twin's legacy (letter). Singapore Med J 2005; 46(7): 1
15. Khan ZH, Tabatabai SA, Saberi H. Anesthetic and surgical experience in a case of total vertical craniopagus. Surg Neurol 1999; 52: 62-7
16. Konovalov AN, Vaichis CHM. The successful separation of a craniopagus. Zh Vopr Neirokhir Im NN Burdenko 1991; 2: 3-10
17. Lasjaunias P, Kwok R, Goh P. A developmental theory of the superior sagittal sinus(es) in craniopagus twins. Child's Nervous System 2004; 20(8-9): 526-37

18. Lotfy M, Sakr SA, Ayoub BM. Successful separation of craniopagus parasiticus. Neurosurgery 2006; 59(5): E1150
19. Maroof M, AL-Rabeeah A, Bonsu A, McDevitt E, AL-Faryan A. Anesthesia for craniopagus and air embolism. Anesth Analg 1997; 84: S442
20. Münster S. La Cosmographiae Universalle. Basle, H Pierre, 1995; p 706
21. O'Connell JEA. Craniopagus twins: Surgical anatomy and embryology and their implications. J Neurology Neurosurg Psychiat 1976; 39: 1-22
22. O'Connell JEA. Investigation and treatment of craniopagus twins. 1. Introduction. Br Med J 1964; 1: 1333
23. Rutka JT, Souweidane M, Brugge KT, Armstrong D, Zuker R, Clark H, Creighton R, McLeod E, Khoury A, Hoffman HJ. Separation of craniopagus twins in the era of modern neuroimaging, interventional neuroradiology, and frameless stereotaxy. Child's Nervous System 2004; 20(8-9): 587-92
24. Sathekge MM, Venkannagari RR, Clauss PP. Scintigraphic evaluation of craniopagus twins. Br J Radiol 1998; 71: 1096-9
25. Shively RE, Bermant MA, Bucholz RD. Separation of craniopagus twins utilizing tissue expanders. Plastic Reconstr Surg 1985; 3: 763-80
26. Spencer R. Theoretical and analytical embryology of conjoined twins: Part I: embryogenesis. Clin Anat 2000; 13:36-53
27. Todorov AB, Cohen KL, Spilotro V. Craniopas twins. J Neurol Neurosurg Psychiatry 1974; 37: 1291-8
28. Voris HC, Slaughter WB, Christian JR, Cayla ER. Successful Separation of craniopagus twins. J Neurosurgery 1957; 14: 548-60
29. Walker M, Browd SR. Craniopagus twins: embryology, classification, surgical anatomy, and separation. Child's Nervous System 2004; 20(8-9): 554-66
30. Winston KR (1987) Craniopagi: Anatomical characteristics and classification. Neurosurgery 21: 769-81