Craniopagus: second Brisbane case

Case report

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Craniopagus is a rare and intriguing condition with an incidence of one in 2.5 million births. The chance of a neurosurgeon seeing a case in a working lifetime is unlikely. The chances of two cases from the same community within 12 months are remote in the extreme. The authors present a second case of craniopagus born and separated in Brisbane, Australia, in 2001 and discuss the intricacies of surgical separation and the lessons learned.

KEY WORDS • craniopagus • conjoined twin • siamese twin • surgical separation

It became clear after the initial fetal imaging studies that the second set of Brisbane twins had far more extensive problems than the first set, who had been separated without difficulty in October 2000. The area that was joined was much larger, with a shared circular draining venous sinus. There was also apparent shared brain tissue but it was difficult to be certain of the extent based on the intrauterine imaging. Twin 2 had no kidneys or bladder, which was most probably incompatible with life, even if she were to survive a staged separation operation.

In January 2001 the parents attended a case conference with specialists from our institution’s administration and the Departments of Neurosurgery, Plastic Surgery, Neonatology, Medical Imaging, Nephrology, and Genetics for the purpose of discussing their conjoined twin pregnancy. At the case conference, all of the information contained in the radiology report was explained carefully to the parents and a considerable amount of time was spent ensuring that the parents were appropriately informed of the clinical situation.

Specifically, the parents were advised of the following complications if the pregnancy continued. 1) The pregnancy conferred an increased risk to the life of the mother because the twins would need to be delivered by cesarean section. 2) There was no guarantee that the pregnancy would go to full term. 3) There was a high risk of stillbirth. 4) One twin (Twin 2) had no kidneys or bladder and if she were born and subsequently separated from her sister and survived, she would require peritoneal dialysis and possibly later renal transplantation. The chances of Twin 2 surviving this scenario were assessed as minimal. At the end of the case conference the views of the various clinicians were summarized and repeated to the parents who then
informed the individuals attending the meeting that despite this information, termination of the pregnancy was not an option and that the pregnancy would be continued.

**Birth of the Conjoined Twins**

On May 3, 2001, the conjoined twins were successfully delivered by cesarean section at the Royal Women’s Hospital. They were born 3 weeks prematurely, although initially the condition of both babies was stable. Examination revealed conjoined calvaria with a circumferential base of 32 cm connected in the temporal, parietal, occipital, and posterior fossa regions of both babies. Postpartum CT and MR imaging demonstrated that both babies had normal but distorted cerebral and cerebellar hemispheres; however, it was still difficult to tell if there was any shared brain because the adjacent temporal lobes and posterior fossae were abutting and distorting each other. The CT venography (Fig. 4) and the subsequently fashioned acrylic model of the twins (Fig. 5) revealed significant sharing of the venous sinuses, with a large circular sinus around the perimeter of the area that was joined. Each baby had a single superior sagittal sinus draining into the shared circular sinus, with distorted right and left transverse sinuses as avenues for drainage. A complex and primitive deep venous system existed with eventual drainage into the shared circular sinus.

As with the first set of separated twins, conventional angiography was not used in this case due to an increased risk of the procedure in babies with little or no added information to be gained.

**Postnatal Condition of the Twins**

From May 10, 2001 (Day 8), Twin 2 began developing increasing hypertension, whereas Twin 1 maintained a normal blood pressure. The results of echocardiograms obtained 1 week apart showed doubling in the thickness of Twin 2’s left ventricle wall in response to the increase in blood pressure. Despite extensive investigation, there remained no identifiable cause of Twin 2’s hypertension. A diagnosis of malignant idiopathic hypertension causing left ventricle failure was made.

The usual course of action (if Twin 2 had been a singleton) would have been to treat the hypertension with medication. The risk in proceeding with conventional hypertension treatment, however, was that with shared circulation between the twins, medical intervention to lower Twin 2’s blood pressure would also cause a fall in Twin 1’s blood pressure. Twin 1’s blood pressure was not elevated, so lowering of her blood pressure could have compromised her brain or the one shared kidney. By not treating Twin 2’s elevated blood pressure to protect Twin 1, we were forced to leave Twin 2 at risk of the high blood pressure and progressive cardiac failure.

By Day 12, Twin 2’s cardiac function had deteriorated to a point at which she required ventilatory support due to heart failure and pulmonary edema. Her blood pressure ranged from 140 to 180 mm Hg systolic (normal 60 mm Hg systolic) with a mean arterial pressure of 100 mm Hg. Twin 1 also began to receive ventilator support to protect her from the effects of the anesthetic medications. Attempts to stabilize Twin 2’s condition were unsuccessful and she was unable to be weaned from the ventilator due to pulmonary edema secondary to heart failure. On Day 13, the opinion of the senior consultant staff in neonatology and nephrology was that cardiac failure could occur at any time for Twin 2 and cause progressive collapse over a period of days (to weeks), which if untreated, would cause her death.

A second case conference was organized between the parents and the medical team for the purpose of discussing the deterioration of the twins. The parents, after considering the options, requested surgical intervention, which was to be expected because they had elected to proceed with the pregnancy in the first instance, with a view to separating the twins if possible.

Because the operation would most likely lead to the premature death of Twin 2, albeit by hours to days, an urgent afterhours sitting of the Supreme Court of Queensland was held to determine the legality of surgery so that the parents and the surgical team would be pro-
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The Operation

On the morning of May 5, 2001 (Day 18), the twins, who were both receiving ventilator support, were taken from the intensive care unit to the operating room for surgical separation.

The babies were initially in a supine semisitting position. The surgical team had previously practiced the proposed operative positioning by using a pair of life-sized mannequins. Fascia lata grafts were harvested for later dural closure. Because tissue expansion was not an option, and Twin 2 was dying, a vascularized skin flap was raised to provide total coverage of the brain, sinuses, and soft tissue of Twin 1, allowing for a total reconstruction. If Twin 2 survived the operation, the plan was to close the defect with the previously harvested fascia lata. The skin on insertion. No attempt was made to repair the dural defect with the previously harvested fascia lata. The skin was closed with underlying heavy packing of Surgicel and split skin grafts.

At the bottom end of the wound, between the shoulders, was the largest abnormal venous sinus in Twin 2, which ran into the shared circular venous sinus. It was thought that this sinus should be ligated last to reduce the effects of venous hypertension; therefore, no attempt was made to ligate this sinus via the supine approach and it was hoped that it could be secured from the prone position.

At the completion of dissection in the supine semisitting position, the wound was packed and the babies were rolled into the prone position, once again with heads up as much as possible to decrease venous hypertension. Placing the babies prone proved to be a very difficult position in which to continue the surgery; it was hard to find a plane between bone and underlying venous sinuses/lakes. Progress was slow with moments of difficulty in controlling bleeding points. Eventually, all of the major sinuses in Twin 2 connecting to the shared circular sinus were ligated except the major draining sinus located at the bottom between the babies’ shoulders. Access to this area was very restricted due to the shoulders. Several attempts were made while the babies were prone to prepare Twin 2’s last connecting sinus for ligation but it was not possible to do it safely.

The decision was made to turn the babies back to the supine position to complete the separation. When preparing for the second turn, Twin 1 unexpectedly suffered cardiac arrest and required resuscitation for approximately 20 to 30 minutes. This appeared to be a hypovolemic arrest and the anaesthesiology team was able to restore cardiac output after cardiopulmonary resuscitation and urgent volume replacement. All sterile measures were abandoned at this stage during aggressive resuscitation. Twin 2’s cardiac performance continued to remain very weak, as it had through the course of the operation.

Following a period of stabilization, the babies were rolled back into the supine position. After a further 20 minutes of dissection, Twin 1 again arrested and required aggressive resuscitation and was again resuscitated after 30 minutes. During this time Twin 2 died, still joined to her sister by a large draining sinus and surrounding dura, soft tissue, and brain.

Twin 2’s remaining draining sinus was still causing major problems because it had not been fully exposed; however, if separation could not be performed imminent-ly, Twin 1 would also die of exsanguination. The decision was made to place a large clamp across the remaining area that was joined to complete the separation, while maintaining the patency of the shared venous sinus. During the first attempt to place the clamp a large volume of blood was lost; the clamp was the removed and the blood loss was controlled with digital pressure. Despite these difficulties Twin 1’s output remained adequate and the clamp was reapplied with success. Twin 2 was then separated from Twin 1 and the clamped area in Twin 1 was oversewn. Twin 2, now deceased, was cleaned and dressed by the operating room staff and presented to her parents to allow them the opportunity to grieve.

Thereafter the surgery in Twin 1 continued for 3 to 4 hours. Coagulopathy caused ongoing oozing of blood from the conjoined site. Diathermy was unsuccessful in stopping the ooze and additional management was performed with thrombin soaked packs of Surgicel and avetine. By 16 hours after initiation of surgery the ooze had slowed sufficiently to allow us to close the wound. A piece of parietal bone had been harvested from Twin 2’s skull after her death to be used to cover the exposed osseous defect in Twin 1 but it was thought best to leave it out until a later date for fear of causing further bleeding on insertion. No attempt was made to repair the dural defect with the previously harvested fascia lata. The skin was closed with underlying heavy packing of Surgicel and

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avetine. A drain was inserted to divert any CSF while the wound was healing.

Postoperative Course

Twin 1 was transferred to the pediatric intensive care unit in critical condition. Her postoperative recovery was hindered by some seizure activity, but this was well controlled with phenobarbitone. On Day 5 postoperatively, Twin 1 was returned to the operating room and the extra packs and small collection of extradural blood were removed and the wound was resutured. Again, a drain was inserted to divert any CSF while the wound was healing.

During the next 3 to 4 months a large pseudomeningocele collection developed under the conjoined area skin flap; however, CT scanning revealed no evidence of hydrocephalus and the venous sinuses were patent. Initially, the collection was treated with weekly percutaneous CSF taps; it continued to increase in size and eventually the taps became daily. A lumbar drain was then inserted for 5 days but complications arose due to an episode of meningitis. Eventually, the decision was made to place a dural patch and at the same time return the bone plate (which had been taken from Twin 2 at surgery, and sterilized and stored at $-80^\circ$C). Local pericranium was used for the dural patch. The bone plate was inserted using LactoSorb plates and sheets to cover any residual bone defects; a drain was inserted. In the following weeks, there was further collection of CSF under the skin but it eventually subsided.

Currently, Twin 1 is 2.5 years of age. She is developmentally a little behind, which is to be expected following two cardiac arrests, separation of an area of conjoined brain, and an extended period of time in hospital. She has a normal vocabulary for her age and is able to construct appropriate short sentences (five to six words). She commenced walking unaided at the age of 18 months, albeit with a slight limp due to mild left-sided weakness. Her treating pediatrician believes she will attend a normal school.

A series of CT scans obtained of the brain 12 months after separation demonstrated good brain volume and no evidence of hydrocephalus (Fig. 6 left); the lower cuts showed gliosis of the left temporal region (Fig. 6 right).

Discussion

Craniopagus is a rare condition. Various authors have collected the published cases from the literature at different times, although the number of cases identified varies according to when the reports were published and whether the search was confined to the English-language journals only and the extent of the retrospective searches.

In a historical review, in 1987 Winston reported the following: 79 cases of craniopagus occurring from 1495 to 1987; for the cases in which survival information is documented, only 11% of patients were alive by 10 years of age (without surgery); 30 reported cases of surgical experience in separating craniopagi in the 20th century were identified between 1928 and 1987; approximately 50% of patients died during or immediately after the operation; 26 of 60 patients appeared to be long-term survivors of separation; and limited information is available on the health of the survivors.

In 1974 Todorov and coworkers reported on 14 cases of craniopagus between 1928 and 1972, including a 24-year-old patient surviving craniopagus. Hoyle reviewed the literature on surgery for conjoined twins and found 167 reported cases. Of these, 27 pairs (16%) were craniopagus and there was approximately a 50% survival rate.

Since the publication of the aforementioned papers, several additional cases have been described, including

![Fig. 4. A CT venogram reconstruction demonstrating a large shared circular sinus around the perimeter of the joined area.](image)
our current report. In October 2000, twins joined at the occiput underwent separation at our institution. In April 2001, Nepalese craniopagi were successfully separated in Singapore. In 2002, twins were successfully separated in the US (unpublished data). Because there is limited information available about the health of long-term survivors, it is important that these cases be documented and followed over the next 10 to 20 years.

No one hospital or center in the world has significant experience with the surgical separation of craniopagi. Importantly, because of the differences in the way that twins are joined and what is shared, each case is unique. It follows that each surgical separation involves mostly a unique set of circumstances.

A review of the literature revealed a case report of the Baragwanath twins, who were separated in 1987; this case involved a similarly joined area to that in our case, which helped our team with the planning of the operation. More case reports are needed to promote understanding of this condition and to aid with decisions as to whether surgical intervention is appropriate.

The provision of treatment to craniopagus twins generally remains a significant medical challenge. Surgery is usually recommended to be performed sometime in the first 3 to 12 months of life but the timing may be dictated by other circumstances such as when one twin becomes seriously ill or there are other considerations favoring separation.

As part of the surgical planning, there must be a detailed operative strategy and its execution must ideally be performed by a multidisciplinary team after wide (international) consultation. This was the practice followed by the staff at the Royal Children’s Hospital, Brisbane, in 2000 with respect to the twins joined at the occiput and again with the second set of twins joined side by side in the present case. Virtually the same team participated in both operations.

As part of the operative strategy there must be a plan for each tissue that will be encountered, including the difficulties with skin coverage at the end of the procedure. Additionally, there must be a plan to address blood loss and resuscitation and, as such, the role of the anesthesiology team is critical. Postoperative concerns include CSF leak, hydrocephalus, meningitis, and control of seizure activity.

In 1987 Bucholz, et al., reported on the separation of temporoparietal craniopagus and noted that significant hemorrhage can occur late during the operation when the largest sinuses are left to be divided. The situation is often made worse by the anatomy resulting in less than optimal exposure. This was our experience as well and is a point of note.

Winston, et al., reported that rapid and large-volume blood loss had been a problem emphasized in several papers. They stated that should one child die before the division is complete, the surviving child’s circulation no longer encounters significant resistance from the other twin’s circulation, resulting in rapid exsanguination.

The acrylic models of the bone and underlying venous sinuses were the most useful in understanding the anatomy of the joined area before and during both separation operations. Two models were used prior to surgery in both sets of twins. One model was complete with the full skull and face of each twin attached. In the other model the faces were removed at each end to enable the surgical team to look inside and more readily view the complex venous anatomy. The models were made by stereolithographic biomodeling, a technology that allows three-dimensional CT and MR imaging data to be used to manufacture solid plastic replicas of anatomical structures. In particular, the relationship of the shared venous structures and the bone was clearly demonstrated.

Bucholz and colleagues have cautioned about postop-
erative CSF leak, which can cause significant morbidity. We were able to control CSF flow by diverting wound drainage until the wound had healed. Thereafter, any persistent fluid buildup may require repeated lumbar puncture or ventriculoperitoneal/lumboperitoneal shunt placement.

Conclusions

The crucial points to derive following the separation of two sets of craniopagi are the following. 1) Craniopagus twins present a rare and unique medical challenge. 2) In the past the statistics on patient survival following separation have been evenly balanced, although advances in medical investigation, surgical techniques, anesthesia, and intensive care have precipitated small improvements in recent years. 3) There are ethical and potential legal problems if operative separation causes the death of one or both individuals who are relatively intact in the conjoined state. 4) The decision to separate conjoined twins has to be evaluated on an individual basis. 5) Prior to surgery it is essential to seek consultation. It is important to include ethical and legal consultations. 6) It is essential to assemble a multidisciplinary team. 7) Craniopagi have an unusually high incidence of associated congenital abnormalities, especially those involving the heart and kidneys, which can cause sudden death of one or both twins. 8) All investigations should be performed early and repeated later if needed so as to be prepared for any sudden deterioration of one or both of the twins resulting in the need for urgent separation. Also, the anatomy of the joined area is often complex, so it can take a considerable amount of time to comprehend. 9) Surgeon fatigue is a major problem. Craniopagus operations can last from 6 hours to more than 50 hours. These issues need to be addressed prior to commencing surgery and consideration to appointing two separate surgical teams working in shifts should be given to any operation lasting for more than 18 to 24 hours. It is important to remember that the most difficult part of the operation may be toward the end when the surgeon is fatigued and therefore more vulnerable to making mistakes or poor decisions. 10) The expectations of the parents/relatives and the general public should be kept low with regard to outcome, because catastrophic events can occur and are easier to accept if the family is forewarned. 11) Finally, to date, attempts to separate craniopagus twins have produced mixed results. So what of the future? Imaging, anesthetic, and surgical advances will probably lead to a small improvement in outcomes with time. Perhaps the major breakthrough with regard to improving outcomes will be with interventional radiology, allowing gradual closure of shared sinuses over a period of time prior to any definitive staged operation.

References


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Fig. 6. Postoperative CT scans obtained at 12 months of age. *Left:* There was good brain volume and no evidence of hydrocephalus. *Right:* Postoperative CT scan at 12 months of age demonstrating gliosis of the left temporal region.