Raniopagus represents 2 to 6% of conjoined twins and is the rarest type of this disorder. It is generally believed that conjoined twins develop as a result of failure of complete separation (fission) of a single fertilized ovum and that this abnormality is established at the end of the 2nd week of gestation. Some authors, however, believe that the abnormality is a result of fusion of two separate embryos, with the junction occurring in the open cranial neuropore just before the end of the 4th week after fertilization. Conjoined twins are always genetically identical and share the same sex. Females are more commonly affected, with a male/female ratio of 1:4. No association with maternal age, race, parity, or heredity has ever been observed.

Documentation of conjoined twins has occurred throughout history. The term “Siamese twins” was first popularized following the birth of Eng and Chang Bunker in Siam (now Thailand) in 1811. Eng and Chang Bunker were not the first conjoined twins, but their world-wide fame helped coin the lay term Siamese twins.

Perhaps the oldest medical illustration of conjoined twins is a woodcut circa 1496 (Fig. 1), which shows twin girls joined at the forehead. The girls lived to the age of 10 years and died within hours of each other. At the time, their birth was interpreted as a sign of God’s displeasure. A little less dramatic, but just as interesting was the birth of the Brisbane twins on April 4, 2000. The Brisbane twins were born attached upside down and back to front, and this is believed to be the first published case of such a joining. Their birth and separation will be discussed in this paper.

Separation of craniopagus joined at the occiput

Case report

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Siamese or conjoined twins have intrigued both the physician and layperson for centuries. The craniopagus type (joined at the head) is exceedingly rare, with an incidence of one in 2.5 million births. Most clinicians never see a case of craniopagus, and those who do rarely see more than one. The authors present a case of the craniopagus type of conjoined twins born and recently separated in Brisbane, Australia. The prenatal diagnosis, subsequent investigations, separation, and outcome are presented.

Key Words • craniopagus • conjoined twins • surgical separation

Cranio-pagus represents 2 to 6% of conjoined twins and is the rarest type of this disorder. It is generally believed that conjoined twins develop as a result of failure of complete separation (fission) of a single fertilized ovum and that this abnormality is established at the end of the 2nd week of gestation. Some authors, however, believe that the abnormality is a result of fusion of two separate embryos, with the junction occurring in the open cranial neuropore just before the end of the 4th week after fertilization. Conjoined twins are always genetically identical and share the same sex. Females are more commonly affected, with a male/female ratio of 1:4. No association with maternal age, race, parity, or heredity has ever been observed.

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Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance.

Case Report

Cranio-pagus girls were born on April 4, 2000, to a 21-year-old gravida I, para 0 mother at the Royal Women’s Hospital in Brisbane, Australia. The craniopagus abnormality was diagnosed at 14 weeks of gestation by prenatal ultrasonography, followed by prenatal MR imaging. Both imaging methods revealed evidence of a left occipital–suboccipital joining based on and below the left transverse sinus. There was a suboccipital bone defect that allowed communication of dura mater and cerebrospinal fluid between the twins. Both brains appeared to be separate and normally developed. Twin 1 was otherwise normal; Twin 2 was noted to have a possible congenital heart defect.

Initial neurosurgical consultation occurred at 21 weeks of gestation. The parents were counseled on multiple occasions thereafter. The issues discussed included: 1) possible termination of the pregnancy; 2) increased birth risk to mother and babies; 3) the existence of a possible heart defect in Twin 2 and potential consequences; 4) the possibility and timing of separation; and 5) the chance of the babies surviving a separation operation.

With regard to the issue of termination, the treating doctors made no attempt to influence the parents’ decision. The facts were given to the parents and the possible risks of morbidity and mortality were discussed. The parents made their decision to proceed with the pregnancy based on their own social and ethical values.

At 33 weeks of gestation there was premature rupture of membranes and onset of labor. The twins were born by classic cesarean section after general anesthesia had been induced in the mother. The combined birth weight of the twins was 3.21 kg (Fig. 2). Twin 1 was assigned Apgar scores of 6 at 1 minute and 9 at 5 minutes after birth with
no significant medical issues; Twin 2 was assigned Apgar scores of 8 at 1 minute and 8 at 5 minutes. Twin 2 was found to have coarctation of the aorta, with a patent ductus arteriosum.

At 2 days of age, Twin 2 suffered an acute and profound hypotensive episode associated with the heart defect. The hypotension resulted in hypoxic brain damage and acute tubular necrosis. She underwent open-heart surgery at 8 days of age. A patch repair of the coarctation and ligation of the patent ductus arteriosum was performed.

Twin 2’s acute tubular necrosis resulted in her having no urine output from Days 3 to 20. Her creatinine level peaked at 0.13 mmol/L (normal range 0.05–0.1 mmol/L). Renal investigations (including isotope scans) showed that Twin 1 was contributing 95% of renal function. Twin 1 experienced no problems performing the renal filtration for both infants.

Clinical assessment at 3 months of age confirmed that the twins were joined in the occipital–suboccipital region, based on and below the left transverse sinus and extending to the confluence of the sinuses in each baby. The conjoined area measured 25 cm in diameter. The twins were aligned upside down and back to front. Neurological examination of Twin 1 was unremarkable. Cerebral palsy had developed in Twin 2 as a result of the hypoxic episode that occurred soon after birth; she was hypertonic and hyperreflexic, with upper motor neuron signs and clonus. Computerized tomography scans, MR images, and MR venograms (Fig. 3) confirmed the occipital–suboccipital skull attachment. Twin 2 was noted to have significant cerebral atrophy on imaging. There was no evidence of shared brain tissue. There appeared to be two separate left transverse sinuses, but it was undetermined as to whether they shared a common midline wall. Both left transverse sinuses appeared to be dominant. Conventional angiography was not performed because it was believed that there would be an increased risk due to the age of the infants with little or no added information to be gained.

Magnetic resonance images of the brains obtained when

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Fig. 1. Woodcut showing the earliest documented case of craniopagus, circa 1496.

Fig. 2. Upper: Photograph of the Brisbane twins obtained soon after birth. The infants are joined upside down and back to front. This is believed to be the first documented case of this type of joining. Lower: Skull x-ray film obtained on Day 1 postpartum.

Fig. 3. Magnetic resonance venogram revealing separate transverse sinuses but leaving the question of whether the sinuses share a common midline wall.
the twins were 6 months old demonstrated herniation of a small portion of Twin 1’s cerebellum into Twin 2’s posterior cranial fossa; this posed a concern (Fig. 4). Also of concern was the skull asymmetry of the twins, as the degree of positional deformation was increasing over time.

In preparation for surgical separation, an acrylic model of the skull was made from three-dimensional CT reconstruction data (Fig. 5). The model showed the relationship of bone between the babies and alignment of the skulls; it also demonstrated the position of the venous sinuses. It provided valuable assistance in planning the separation. The model was used by the plastic surgery team to plan the surgical incisions and skin flaps to provide the best possible skin coverage. To do this, a tailor-made pressure garment (Second Skin Pty., Ltd., Brisbane, Australia) was fitted around the acrylic model. This was used to simulate the scalp skin in both patients. An attempt was made by the manufacturers to mimic the elastic properties of skin. The material was used to plan the dimensions of the scalp flaps and allow model surgery to be performed. This allowed greater confidence that the planned flaps reconstruct the defect appropriately.

Given that Twin 2 had significant medical problems and her long-term prognosis was guarded, it was believed to be unwise to subject both twins to the possible complications of tissue expansion, which in babies include infection and depressed skull fracture. The decision was made that the defect could be completely closed in Twin 1 by using a large occipital scalp flap. In Twin 2 the critical defect of exposed brain and dural grafts would be closed using an occipital scalp flap that had sufficient overlap to be anchored to the vascularized pericranium to gain a margin of good-quality vascularized scalp-flap healing beyond the critical defect. The remainder of the defect in Twin 2 would be covered with a split-skin graft.

One week before the operation, a pair of life-sized mannequins were joined in the precise upside-down back-to-front position as the conjoined twins to anticipate any potential problems that could occur during surgery. Numerous lines and tubes were attached to the mannequins and the surgical and anesthesiology teams practiced turning and transferring the mannequins while maintaining sterility and preventing the lines and tubes from kinking or entangling.

Neurological assessment of Twin 1 just prior to surgery yielded normal findings. Twin 2, however, was microcephalic and suffered from clonic–tonic seizures. She did not fix her gaze, smile, or follow, and was hypertonic with increased reflexes and clonus. Audiology revealed high-frequency hearing only.

At 7.5 months of age (corrected age 6 months taking into account premature birth), on October 24, 2000, the separation operation was performed. A team of 25 persons, including two neurosurgeons, two plastic surgeons, one pediatric surgeon, four anesthetists, and numerous operating room nurses and technicians was assembled. Anesthesia was induced in the twins and a Tenckhoff catheter was placed in Twin 2. Fascia lata grafts were harvested from both twins for later dural repair. The twins were initially placed with their left sides up (Fig. 6). The separation operation commenced and took approximately 7 hours. Occipital scalp flaps were elevated off the pericranium as designed...
by the plastic surgery team. Connecting occipital and sub-occipital bone on the left side was removed using a high-speed drill and upcutting punch and bone nibblers, and the conjoined dura was exposed. Using the sterilized acrylic model as a guide, the dura mater was incised and separated without damage to the transverse sinuses or torcular region in either twin. Some minor blood loss occurred with entry into osseous venous lakes. The twins were then turned over to allow bone and dural separation on the other side. In Twin 1 the herniated cerebellum was returned to the posterior cranial fossa without difficulty. Following separation, two teams of neurosurgeons, plastic surgeons, and anesthesiasts worked separately on each twin. The dural defects were repaired with fascia lata grafts. Bone chips from the conjoined site were laid over the bone defects. In Twin 1 the scalp defect was covered entirely with the designed occipital scalp flap. In Twin 2 the defect was repaired using a scalp flap to cover the bone defect and a split-skin graft to cover the rest of the defect. There was minimal intraoperative blood loss. The overall operation, including induction of anesthesia, lasted 12.5 hours.

The postoperative care team included intensive care specialists, pediatricians, and renal physicians. The separated twins made rapid progress and were extubated on Day 1. They remained in the intensive care unit for 2 days. The postoperative course of Twin 1 was uneventful except for feeding problems. The postoperative brain CT scan obtained in this twin was normal. Twin 2 encountered several minor postoperative problems. There was a small cerebrospinal fluid leak on postoperative Day 6 that required surgical revision of the wound. On postoperative Day 10, 8% of the split-skin graft was lost, exposing an area of pericranium. This was dressed with Intrasite gel and Duoderm. The exposed area healed within 6 weeks without further grafting.

In Twin 2 the postoperative CT scan revealed a small right subdural hemorrhage that was 5 mm thick. The collection caused no pressure effect because of its small size and the preexisting cerebral atrophy. A follow-up CT scan obtained several weeks later demonstrated that the subdural collection had resolved spontaneously without sequelae. Twin 2’s renal function improved postseparation and she did not require dialysis. The Tenckhoff catheter was removed on Day 10.

Twin 1 was discharged from the hospital 4 weeks postoperatively (Fig. 7 left). Twin 2 remained in the hospital because of her neurological problems. She also experienced recurrence of the aortic coarctation and underwent a second cardiac operation in February 2001. Her neurological condition remained static with features of moderate-to-severe cerebral palsy and seizures. She was eventually discharged home to her family in March 2001 (Fig. 7 right).

**Discussion**

Craniopagus is rare. Initial results of separation in the early 20th century were poor. In the 30 attempts to separate the craniopagus type of conjoined twins reported since 1928, only seven of the 60 infants emerged unscathed; 30 died, 17 were reported to be neurologically impaired, and the remainder were reported before the ultimate outcome could be determined.

As technology has improved so have outcomes. Most conjoined twins born today are detected during routine prenatal ultrasound examinations performed during the first or second trimester. The Brisbane team used a combination of ultrasound and fetal MR imaging to gain accurate information about the conjoined area and this information was used to counsel the parents during the prenatal period.

One of the main difficulties the Brisbane surgical team faced was that there was very little current information in the literature regarding surgical separation in cases of craniopagus. Of all cases of craniopagus that have been published, we could find no other case with similar anatomy, that is, twins joined at the left transverse sinus region, upside down and back to front.
Separation of craniopagus joined at the occiput

With regard to the timing of the operation, there are published opinions, but no firm evidence concerning when surgical separation should be performed in cases of craniopagus. We believed that delaying surgery until the infants were approximately 6 months of age was preferable to enable the babies to grow in size and increase their blood volumes.

Over the 3-month period leading up to the operation, the surgical team consulted with experts locally, nationally, and internationally in an effort to gain knowledge and information regarding the best way to attempt the separation. One useful piece of advice we were given was to watch closely for other congenital problems. This proved to be the case with Twin 2, who was born with a coarctation of the aorta. She suffered a hypotensive episode soon after birth, resulting in hypoxia to the brain and kidneys, and subsequent development of cerebral palsy and transient renal failure due to acute tubular necrosis. Had Twin 2 died as a result of this insult, an emergency separation would have been required. The potential risk for Twin 1 would have markedly increased in such a situation because of her small blood volume and immature immunity, as well as a lack of appropriate investigations and a lack of consultation and planning.

Following consultation, the general consensus on the operative plan was to try to find a cleavage plane between the sinuses of the twins and preserve both sinuses. If there was no cleavage plane or excessive bleeding was encountered, then it was believed that Twin 2’s sinus should be closed down past the confluence. Because Twin 2 already had cerebral palsy, it was believed that she would be affected to a lesser extent than her sister should a sinus need to be closed down. The parents of the twins were made aware of and consented to the surgical plan. Further consent was obtained from the Brisbane Royal Children’s Hospital ethics committee.

With regard to plastic surgical issues, it was believed unwise to subject the healthy twin to a prolonged period of tissue expansion and possible morbidity, given the poorer long-term prognosis of Twin 2. This was the basis of our decision to attempt a one-stage procedure to reconstruct both twins’ defects. The defect was measured, both preoperatively and on the acrylic model. The design process, as previously outlined, allowed a one-stage operation to reconstruct the defects in both twins appropriately with the prospect of tissue expansion and removal of the residual split-skin graft on the vertex of Twin 2 at a later date, if that was considered appropriate once the child had grown further. Twin 1 has a durable, well vascularized, and healed scalp flap, which remains cosmetically very acceptable, given that all scars are concealed within the hairline. There was some minor skin graft loss (5–8%) from Twin 2, which was not adjacent to the critical defect. This was managed conservatively and provided no ongoing morbidity. Twin 2 has a well-healed, durable, vascularized scalp flap over the critical defect and a stable split-skin graft over the remaining vertex.

With regard to the risks of the procedure, the team’s approach was to explain to the parents that, because of the rarity of the condition, it was difficult to provide accurate percentages of morbidity and mortality, but that the available published evidence was consistent with a 33% chance that both babies might die as a result of surgery, a 33% chance that one would die and one would survive, and a 33% chance that both would survive. We can expect that survival rates will continue to improve as investigations, anesthetic and surgical skills, and perioperative intensive-care skills improve over time.

Fortunately Twin 2’s prenatal renal failure proved not to be permanent and her renal function progressively improved after surgical separation, avoiding the need for dialysis even on a temporary basis. The ethical issues surrounding the possible management of chronic renal failure postseparation had been discussed with the parents before surgery. It is now 23 months since the birth of the Brisbane twins and 16 months since their separation. Twin 1 is meeting her normal milestones in every respect, whereas Twin 2 has moderate-to-severe cerebral palsy, which predated the surgical separation.

Conclusions

In summary, craniopagus is a rare and interesting multidisciplinary challenge. It is likely that the risks of morbidity and mortality associated with surgical separation will continue to decline with improved imaging and anesthetic technology. The cornerstone to success, however, is likely to remain teamwork, consultation, and preoperative planning.

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