# Craniopagus Twins

# 11

## Surgical Techniques for a Staged Separation

James Tait Goodrich, Andrew Joshua Kobets, Kamilah Dowling, and Oren Tepper

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## Introduction

Craniopagus twins represent a rare disorder and remain a highly fascinating accident of nature. Craniopagus twins joined at the head occur about one per 2.5 million live births worldwide and represent just 2–6% of all types of conjoined twins. Approximately 40% of craniopagus twins are stillborn and an additional one-third dies within 24 h of birth. The common cause of early death is related to congenital organ anomalies with the most common related to pulmonary, heart (failure, hyper-and hypotension), and renal failure. This leaves about 25% of craniopagus twins for consideration of a surgical separation. Over the last three decades, it has been our experience that only 2–3 sets of craniopagus twins occur yearly that are possible candidates for separation. In the last half-century, with the many advances in medicine including brain imaging, 3D CADCAM imaging, 3D modeling, and advanced neuroanesthesia and neurosurgical techniques, successful outcomes are now possible following separation of even total craniopagus twins.

Successful separation of craniopagus twins where both have come out of surgery without neurological deficit historically has been very

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C. Di Rocco et al. (eds.), *Textbook of Pediatric Neurosurgery*,
https://doi.org/10.1007/978-3-319-72168-2_10

rare. With the introduction of a staging technique, devastating neurologic outcomes or death of one or both twins has become much less common. The Center for Craniofacial Disorders at the Children's Hospital at Montefiore has now been involved in 27 cases of craniopagus twins from around the world. Each set has had very unique medical and surgical issues that needed to be addressed. For a majority of the cases, surgical separation was not possible due to medical, surgical, financial, ethical, and religious reasons. These important factors that often preclude or limit the chances of successful separation need to be carefully weighed against statistics that suggest an 80% death rate by 2 years of age in untreated craniopagus twins due to medical complications (Browd et al. 2008, 2011, 2017; Goodrich and Staffenberg 2004, 2005; Roberts 1984; Stone and Goodrich 2007; Sugar et al. 1953; Winston 1987).

In undertaking the care of a set of craniopagus twins, the clinical and surgical pathology is key in understanding and essential in making any surgical plans for separation. The degree of brain, dura, and vascular sharing determines the degree of complexity, which can be mild to moderate in some cases and severe in others. Assessing the interfaces between the brains in our experience varies quite widely. In some cases, the degree of compression and brain distortion was so severe making it impossible to do a safe separation. In assessing these children it will be the degree of brain angulations, the amount of shared structures, and finally brain distortion that will allow the surgical team to decide on a separation surgery. In a 2006 paper, we reviewed 64 well-delineated craniopagus twins cases, including 41 operative separation attempts in children since the first successful craniopagus twins separation in 1952 (Stone and Goodrich 2007; Sugar et al. 1953). From this review, we learned to appreciate the complexity of these craniopagus twin cases. At the time of that review, 30% of craniopagus twins had shared or fused brain tissue. Partial forms were separated at an earlier age (6 vs. 11 months) leading to a lower mortality and better outcome compared with total forms. We have now shown that a multi-staged surgical separation for craniopagus twins has a significantly reduce mortality and morbidity when compared to a single-staged separation.

The surgical team also needs to be prepared to deal with some cases of craniopagus twins where there will be a need for an urgent or semi-urgent surgical separation. We have encountered several sets of craniopagus twins where early intervention was needed and not all had good outcomes. The causes were due to persistent aspiration and/or airway issues, cardiac failure, cor pulmonale, or the impending death of one twin. In the surgical planning, other potentially life-threatening aspects of craniopagus twins' separation include adequate dural closure with autologous, cadaveric, or synthetic dural substitutes. Also critical is scalp coverage with adequately designed and delayed pedicle flaps provided with the use of subgaleal tissue expansion. When skin flaps are adequate, this will significantly reduce the potential for cerebrospinal fluid (CSF) leakage and meningitis. CSF shunting for hydrocephalus or to promote wound healing may be also necessary. In our experience focal or generalized seizures are likely underreported and are not uncommon during the staging and at the final separation. Therefore, adequate anticonvulsant therapy should be considered, especially in cases of known brain fusion. The details of the above will be further discussed in the chapter.

## Clinical Workup

In a review of the surgical literature, it became apparent the type and angle of conjoining significantly influences the operative decision to separate. In our series, over half the children – due to the nature of the conjoined brains and vessels – could not be separated. The angular and angular occipital types have proven to be the most challenging and have the highest surgical mortality. The vertical or "stovepipe" configurations, however, lend themselves to better outcomes (Staffenberg and Goodrich 2010) (Fig. 1).

These children often come from very poor and medically underserved areas of the world. As a result, it is not uncommon to see a history of

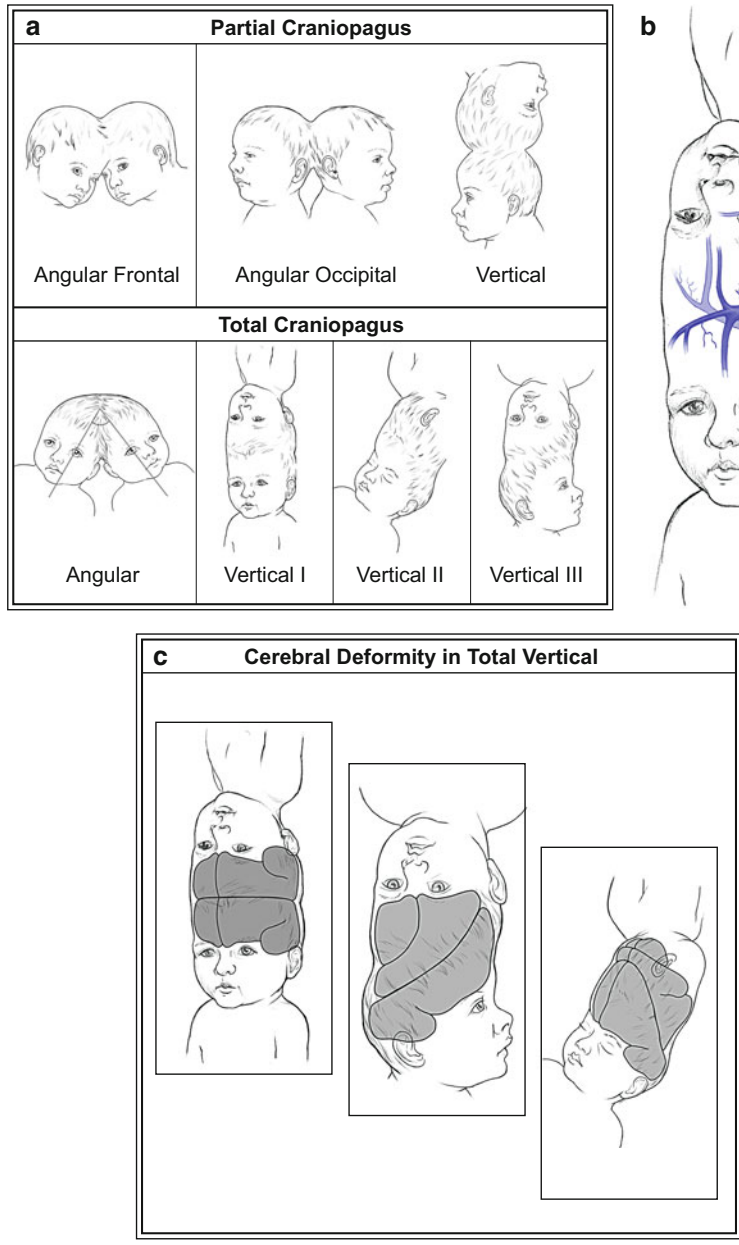


**Fig. 1** A review of the various types of craniopagus conjoining. These images were compiled from an extensive review of the literature. Each type of conjoining has its own unique surgical separation issues. The angular type craniopagus tend to be the most complex in the conjoined vascular anatomy (Stone and Goodrich 2007)

aspiration pneumonia, cardiac and renal issues along with other chronic medical conditions. A common problem is linked to their anatomy, i.e., typically lying on their backs, which leads to chronic aspirations in both children with oral feeding. These children typically present at about 60–80% of their normal body weight, with weight loss due to the chronic aspirations, pneumonia, and heart failure, among other illnesses. It is critical to get the pediatric subspecialties and intensive care teams involved at the very beginning to resolve as many of the medical issues as

possible prior to surgery. The most serious medical problems in our experience have been right-sided cardiac output failure, cor pulmonale, pneumonia, and renal issues.

It is also essential to involve the anesthesiology team upon arrival of the twins for assessment and management. The angulations of the conjoined brains will have an enormous impact on positioning and intubation and these angulations can range from simple to very complex. Therefore the anesthesiology team will need to determine how to position and intubate the children from the very beginning. In two cases, the intensive care team decided it was more effective to place tracheotomies at the beginning then remove them after the final separation. In teaching institutions, where physician turnover can be relatively common, it is extremely important to provide each twin with its own anesthesia and neurosurgical team which remains constant throughout all stages of separations. Changing team members, including residents and fellows makes no sense in these complex and multiple procedures (Girshin et al. 2006; Goodrich and Staffenberg 2004).

From the beginning, the contribution of plastic surgery is critical. Each of these twin sets produces its own unique problems when it comes to skin and scalp coverage. The key goal for the plastic surgeons is to provide a complete and secure scalp covering of both brains. The flap design is critical and the typical style we use is a "yin and yang" type as these are safer and easier to expand at the final stage. The placement of these incisions and elevation of scalp flaps set the foundation for eventual scalp closure once separation is complete, and therefore should be carefully planned and performed with plastic surgery colleagues. Suboptimal execution of scalp flaps will significantly impact tissue viability and options for expansion, and thus diminish the chance of successful coverage in these types of cases. Keeping the incision within the hair baring area is aesthetically more desirable and should be considered if possible. Extremely important are the skin flaps, which should always include a reliable blood supply. The most suitable design is a sinusoidal pattern that allows one flap to turn over across the vertex of one twin, while the other does the same to cover the other twin. A technical pearl in such cases is to raise the scalp flaps in a subgaleal plane, and move to a subperiosteal dissection only in areas where cranium needs to be exposed for a craniotomy to be performed. The advantage of leaving galea over the remainder of the cranium is that it allows for well-vascularized tissue directly beneath high-risk areas such as the apices of the scalp flaps. This "insurance" proved beneficial in our most recent case of twin separation where the distal portions of our scalp flaps underwent necrosis, but granulated in well due to the underlying pericranium that had been present. Without this, we would have been faced with exposed bone, dura, or brain parenchyma beneath.

Keeping the forehead hairline in normal anatomical position is key in the design. If the hairline is at the eyebrow line the outcome for the patient and parents will be undesirable. As craniopagus twins present with multiple variations in angles of conjoining, these skin flaps become particularly unique for each case (Staffenberg and Goodrich 2005, 2008) (Figs. 2, 3, and 4).

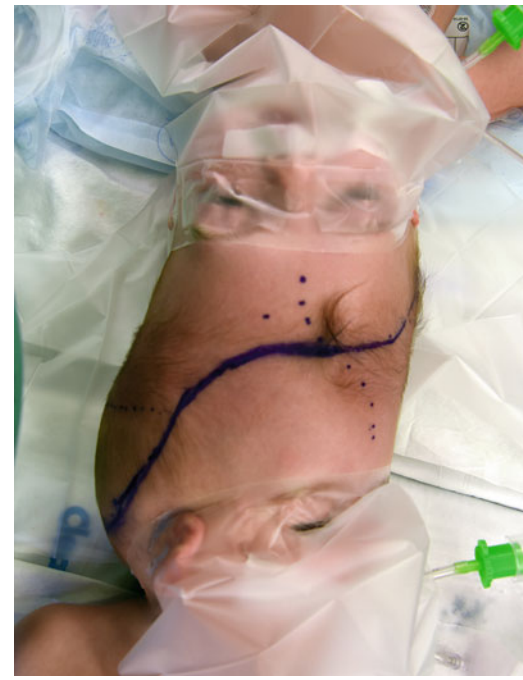

**Fig. 2** Frontal view showing the marked "Yin & Yang" style incision

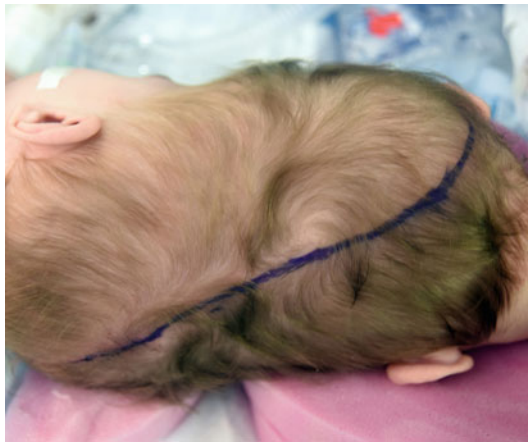

**Fig. 3** View of the backside of the twins showing the continuation of the "Yin & Yang" style incision

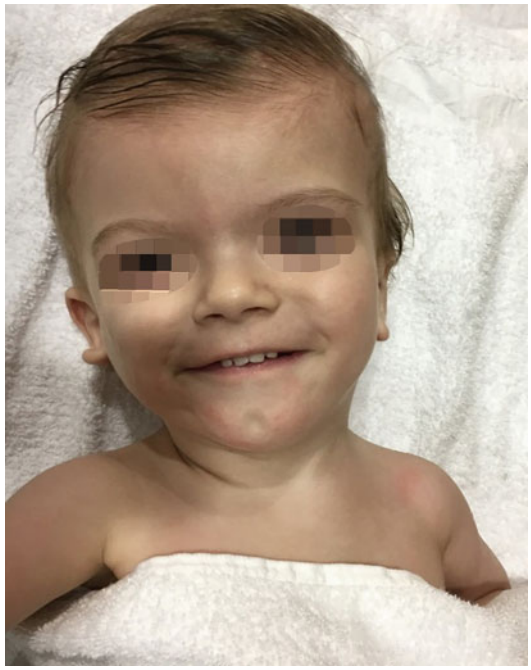

**Fig. 4** The surgical design of this incision allows the patients to maintain a decent aesthetic appearance of the face and hairline

Additionally, we have found the middle "circular" type incision less desirable as it reduces planes of expansion and has a higher risk of the tissue expander extrusion. Furthermore, this leaves the entire length of the incisions above the most high-risk areas once the twins are separated.

Another important point is the tissue expansion, which is not done until the penultimate surgical case; placing them sooner just leads to an increase in infection and extrusion. Another critical problem in tissue expanders is soft tissue necrosis and pressure sores over the expanders so the nursing team has to constantly be repositioning these children to avoid this issue. This can be a very difficult task as children typically are at an age where they are non-mobile and susceptible to direct pressure from positioning. For this reason, along with that of achieving maximal expansion, we believe it is best to place multiple expanders rather than a single or pair of large sized expanders. With multiple expanders in place, the surgical team has flexibility in terms of degree of expansion, and also can risk loss of a single expander without compromising the entire process. The critical goal if scalp coverage is to reduce CSF leaks to zero and avoiding meningitis, both of which significantly increase morbidity. For the team, success clearly requires an understanding of the complex interrelationship between the "separation" and the "reconstruction," and that decisions made for one aspect of the surgery will have a profound impact on another aspect of the surgery; this impact can be disastrous, or if planned well, can be advantageous. An important example requires that the plastic surgeon team is prepared for what can be significant cerebral edema at the final stage and be prepared to provide good expansion and adequate coverage. Herein lies the strength of the staged technique, as well as the importance in recognizing the nuance of planning.

From first case in 2004, we have found 3D printed models to be extremely helpful in both flap design and tissue expansion volume. We have been using ClearView Translucent Models (3D Systems Littleton, CO, USA) for building the skin surface from neck to neck and including the ears. The data for these models is generated from both MR and CT imaging studies. The early generation models were primarily limited to bone anatomy as well as gross brain parenchyma, and were printed as "stereolithographic models." Over the last decade, this technology has made significant advancements in terms of 3D printing

**Fig. 5** The tissue expanders are seen in position. During the expansion time, care has to be done to avoid any pressure necrosis or breakdown of the skin. The surgical team needs to work closely with the nursing team to maintain routine changes in position and avoid prolong pressure against the tissue expanders

capability. The detail of the stereolithographic 3D prints of bone and brain parenchyma far exceeds what was available previously. Moreover, full color prints with high fidelity are now possible, which can print (and color coordinate) various structures such as brain, dura, arterial vasculature, and the venous system. Soft tissue models that capture the external surface are also available and can be obtained with standard 3D photography captures systems based on stereophotogrammetry. The models represent a representation of "skin" covering which the plastic surgeon can use to design and modify well ahead of the first stage of surgery. Volumetric analysis is also done to determine the amount and size of the tissue expanders (Figs. 5, 6, and 7).

As 3D printing technology is increasingly being applied for cases of complex reconstructive surgery, so too has the notion of virtual surgical planning (VSP). VSP enables the surgical team to virtually plan operative steps in conjunction with biomedical engineering. In our most recent experience with craniopagus separation, this technology proved to be an invaluable part of the process. Through VSP, we were able to pre-determine a number of key elements, including our scalp flaps

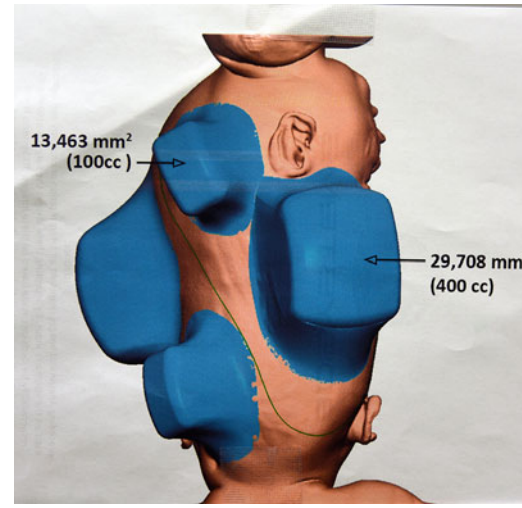


**Fig. 6** Graphics generated from the 3D CADCAM program showing how much tissue expansion will be needed for adequate coverage. Frontal view

design, the expected defect, extent of tissue expansion required, optimal size and placement of tissue expanders, and appropriate shape of the cranial reconstruction. Surgical guides can also be 3D printed based off of the VSP and sterilized for use during surgery.

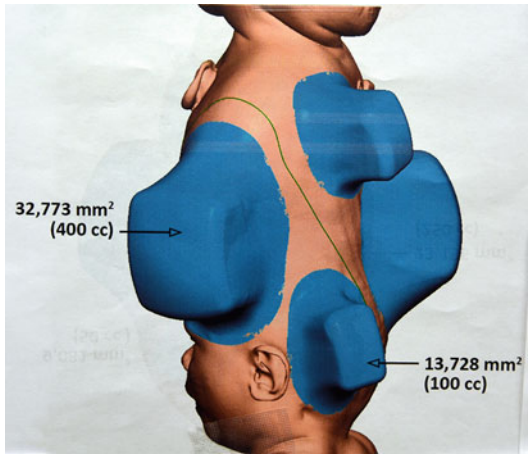


**Fig. 7** Graphics generated from the 3D CADCAM program showing how much tissue expansion will be needed for adequate coverage. Posterior view

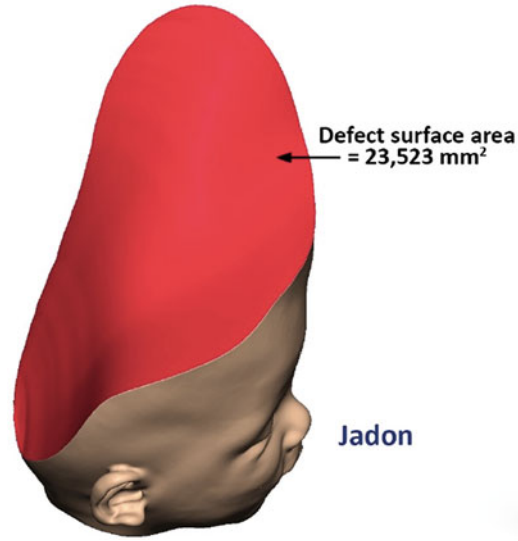


**Fig. 8** This graphic shows the calculations for the skin flap on one twin and the amount of expansion needed

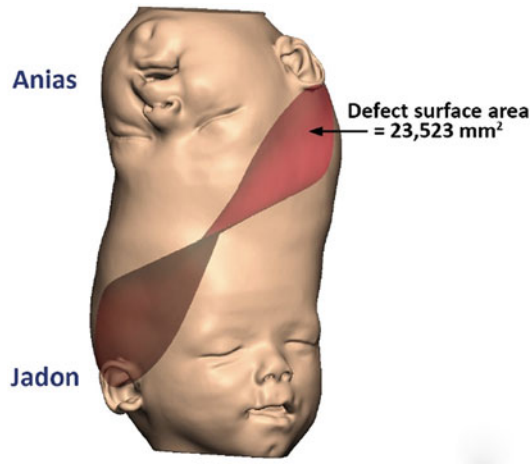


**Fig. 9** This graphic shows the calculations for the defect that will be present, if tissue is not expanded

Surgical teams in the past have considered the use of vascularized free tissue transfer flaps. However, in an infant we have found this style of flap far less useful than in an adult. The main reason why is the relative surface area of an infant's scalp is twice that of an adult, and vascularized free flaps, such as latissimus dorsi and rectus abdominis muscles are comparatively smaller than in adults and hence will not provide adequate coverage (Figs. 8, 9, and 10).

A medical problem often not considered is oral hygiene. As these children often come from poorer regions, the dental issues including abscess and infection are important factors. Our oral-maxillo-facial team will do an initial assessment so any dental restorations that are required can be completed in order to minimize infection and bacteremia during the various surgical stages.

## Neuroradiology and Imaging Studies

Imaging studies have proven to be critical for surgical planning. We routinely order MRI, MRA, MRV along with CTA and CTVs. Since our first case in 2004, we have routinely used CADCAM programs to develop 3D models. The 3D models can be produced with various anatomical views such as venous anatomy; these models have been crucial in developing surgical planes between the two patients. 3D modeling has been very helpful in determining the amount of fused brains and has been used to demonstrate "maps" of the shared venous anatomy. In some cases, fusion has been shown to be too severe therefore separation was not recommended (Alokaili et al. 2015).

In reviewing previous single staged separations, the incidence of hydrocephalus requiring

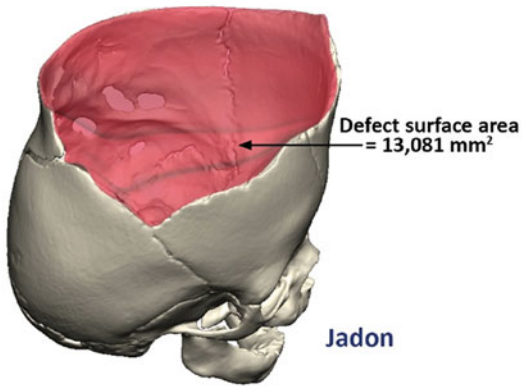


**Fig. 10** This graphic shows the calculations for the skull defect of one twin once separated

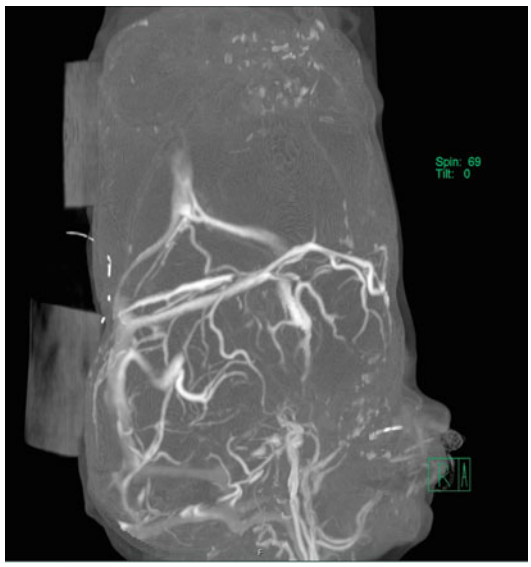


**Fig. 11** MRV showing venous perfusion of the lower twin, crossover to the upper twin is seen. The CVP is seen at the interface of the two brains

shunting was nearly 100%. With staged separations and adequate scalp closures, we have reduced shunting to zero (Figs. 11, 12, and 13).

## Contraindications

Often not appreciated in these cases are the contraindications of when not to do a separation surgery. It has been our experience that in the

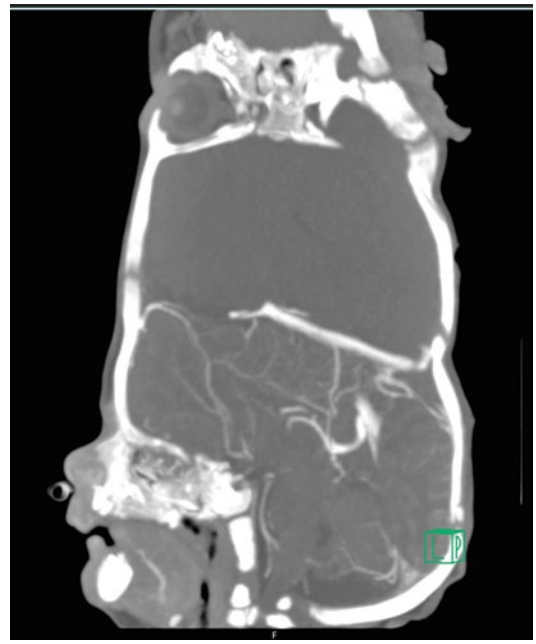

**Fig. 12** CTV sagittal view showing the brain interface and the CVP enhances at the interface

majority of cases of conjoined brains, the brains have proven to be too complex to separate. Leading the list of complexities is the degree of conjoined brain. We have found that this can range from zero (rare in true craniopagus twins) to severe meaning there is a greater than 8 cm area of conjoined brain. We have had two sets of angular conjoined twins in which the anatomy was such that there was a diencephalic bridge between the two twins. This anatomy precluded any separation effort. In another set of twins, the angle of rotation of the conjoined brains was too severe to do the separation and therefore would require removal of the occipital lobe of one child to access the conjoined brain. In addition, one of the twins was born without kidneys and was using the other twin for kidney function. The angle of conjoining, amount of fused brain, and the inability to develop a good anatomical plane between the two brains without severe morbidity and mortality precluded this set from separation. We are also finding that to some degree, radiographic studies may underestimate the degree of conjoined brain, only to find a greater neural connection between the twins than initially anticipated. We have also illustrated a set

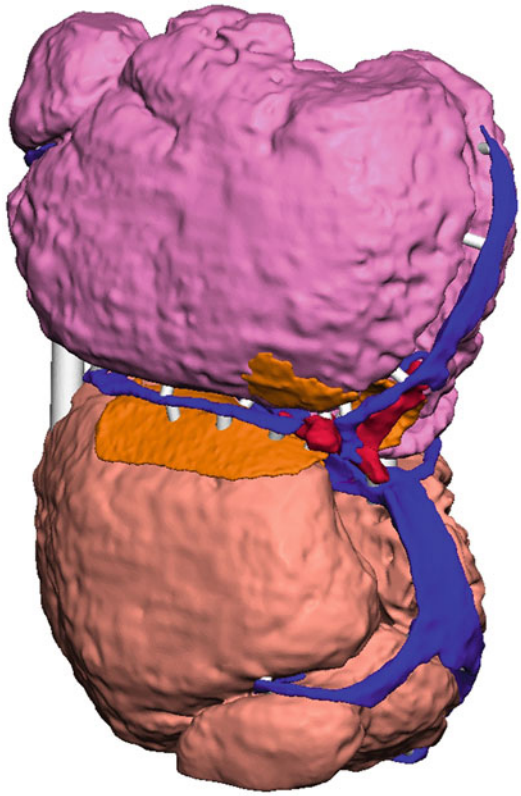

**Fig. 13** An example of one of the 3D models showing the brain interface, the venous outflow, and the silicone sheets (colored orange) placed between the brains, done at each stage

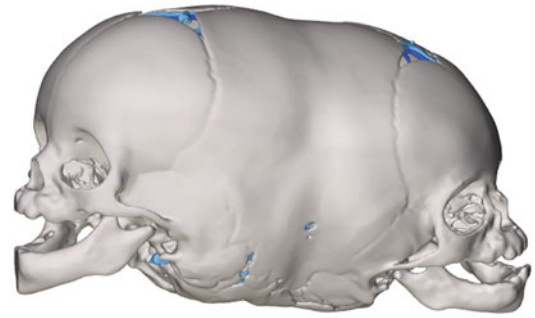

**Fig. 14** The occipital and angular types of conjoining can lead some quite complex vascular anatomy. In this case of an occipital angular type, the 3D skull anatomy is shown. Using a pdf reader, the anatomy can be further detailed by removing layers of scalp and skull

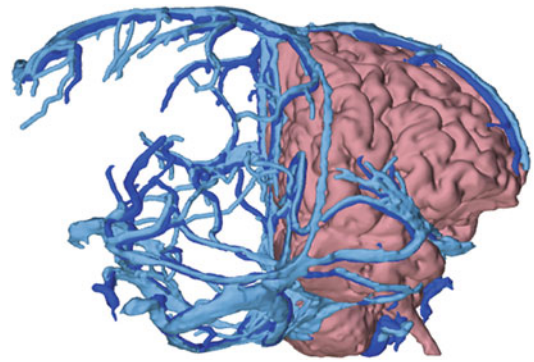

**Fig. 15** Using a pdf reader, the anatomy can be further detailed by removing layers of scalp and skull. In addition of that, brain of one or both twins can be removed. In this case, the brain has been left on the right and removed on the left. These images can be rotated around in any direction providing a 3D perspective

of occipital conjoined twins with a very dense vascular plexus at the conjoined site. A significant portion of brain was to be fused. We have had only one case where interventional neuroradiology has been of use and that was a case of vertical conjoining with a 180° rotation. This led to the sagittal sinus of one child to directly end in the torcula of the other child. In this case, the sagittal sinus of each twin was taken before the third and fourth stages. We have also found that the older the twins get the more complex the vasculature and conjoining becomes. Ideally, most separations should be done under 2–3 years of age (Figs. 14, 15, 16, 17, and 18) (Winston et al. 1987).

While not often thought of as a contraindication, religious and social issues can be quite significant. We have been involved in several sets of craniopagus twins where the family, for mostly religious or sometimes social reasons would not consider a surgical separation. The concept that these children were "an evil warning from God," i.e., monsters or *monestere* (Latin), was an important part of the family beliefs and subsequent reasoning.

## Preoperative Preparation

Common in craniopagus twins is a medical issue related to the complex vascular connections of the brain leading unequal cardiac outputs. Our

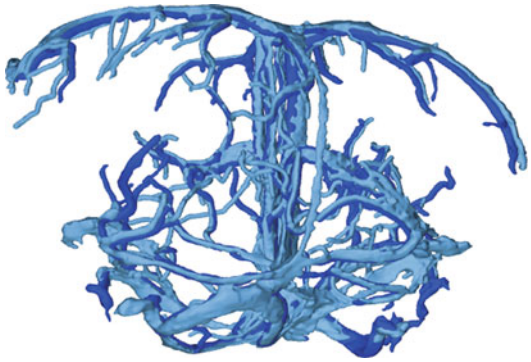

**Fig. 16** With the two brains removed and just looking at the venous anatomy, it becomes quite apparent the complex conjoining along with a large CVP between the two brains. This type of complex anatomy normally precludes any intervention due to complex interweaving of vessels. In the few cases in which separation has been attempted, the surgical mortality was 100%

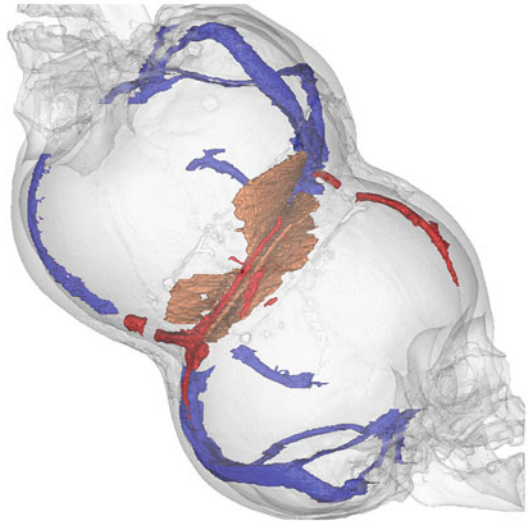

**Fig. 17** A complex set of twins with a vertex conjoining and a 180 degree rotation. On each side, the sagittal sinus drained into the torcula of the other twin. The sagittal sinus is marked in red where each child was embolized. The silicone sheets are outlined in brown

research tells us that it is common to see one child in high cardiac output failure with the other child acting as a "parasite" to the twin. The other twin is typically hypotensive and oliguric. It is also common to find one child with high blood pressures of 220/110 mmHg while the other was 60/40 mmHg. Just visually accessing the twins, one typically

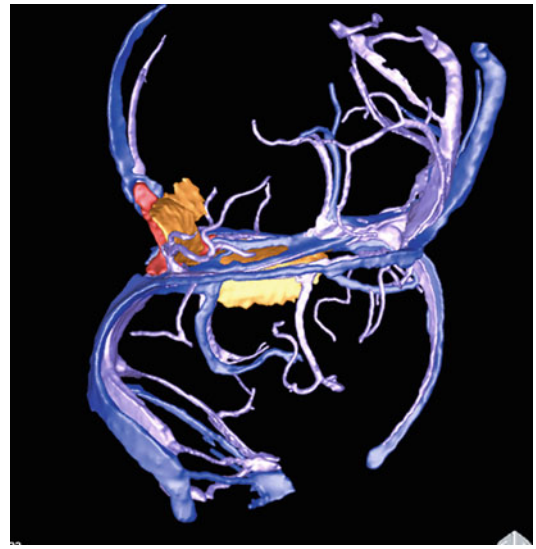

**Fig. 18** The same case as Fig. 17 but here detailing the venous outflow anatomy and the site of the first embolization done at stage three. (Images courtesy of Ahmed Al-Feranyan, embolization by Riyeh)

tends to be larger in size than the other with a healthier appearance. Dealing with and treating the various cardiac, pulmonary, and renal issues are done immediately upon arrival by the various subspecialties. Critical to the treatment is not to over-treat as one child is often dependent on the other for blood pressure and dialysis; too drastic a reduction in particularly blood pressure can lead to dire consequences. The hypotensive twin is often in right side cardiac output failure at presentation. A critical concern in treatment is if you give one twin a medication, how does it affect the other due to crossover blood flow – the answer can vary greatly and be very unpredictable! For the anesthesia team, medication sharing becomes quite critical during any sedation procedures and the various surgeries (Fig. 19) (Winston et al. 1987).

The neurosurgical preoperative workup is designed to sort out three important anatomical concepts. The first is the shared vascularity of the two brains and in all cases it is the venous anatomy that is the most important. The second anatomical issue is the presence of fused or anatomically conjoined brain. The third anatomical anomaly is not seen in normal brains and that

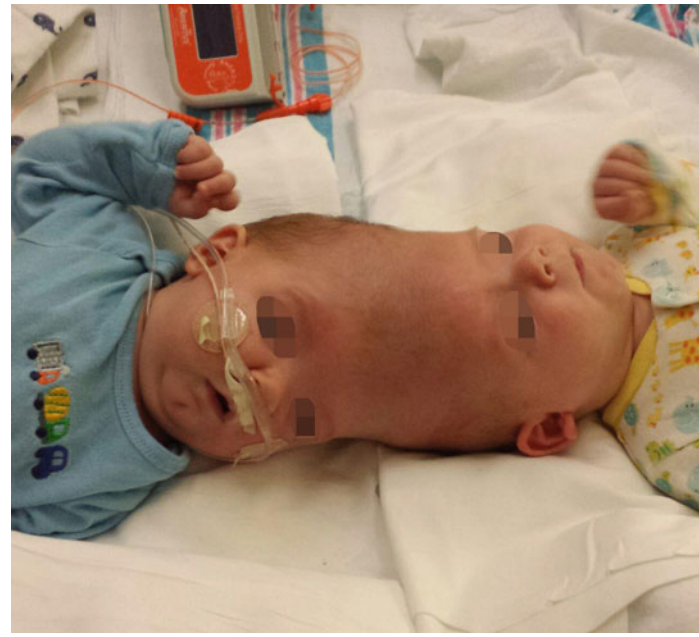

**Fig. 19** Preoperative photograph of a set of vertical conjoined twins. The difference in body habitus is clearly evident. The twin on the left was hypotensive with chronic oxygen desaturations which at times went down into the 30s. This child also had chronic aspirations and gastric reflux. Due to poor feeding nasogastric feeds were needed. The twin on the right was more hypertensive, feed orally with no issues, and medical issues were less than the other twin

is a "circumferential venous plexus" (CVP), an anatomical lake or "pool" of venous blood between dural leaves situated between the two opposing brains. In our experience, this shared venous lake cannot be easily separated or bypassed. A preoperative surgical decision has to be made regarding which twin will get the CVP. In our experience, this plexus typically goes to the dominant twin, meaning the one that receives most of the venous outflow. The other twin with typically poor venous outflow will develop its own deep draining venous anatomy as the children goes through each of the staged separations (Figs. 20 and 21).

We have found that the variability of conjoined brains in craniopagus twins can vary quite a bit. Once the vascular anomalies have been sorted out, we evaluate the two brains looking for surgical pathways of separation and also for fused or conjoined brain. For this evaluation, our workup includes cerebral angiography, both venous and arterial phases. In the surgical planning, it is the venous anatomy that becomes critical. Arterial connection is surprisingly rare and if present, so far have not been significant issues in the separations. In evaluating for conjoined brains we have found the MRIs to be surprisingly misleading! CSF planes between the two brains are often misleading giving the impression that there is a true interface. If the

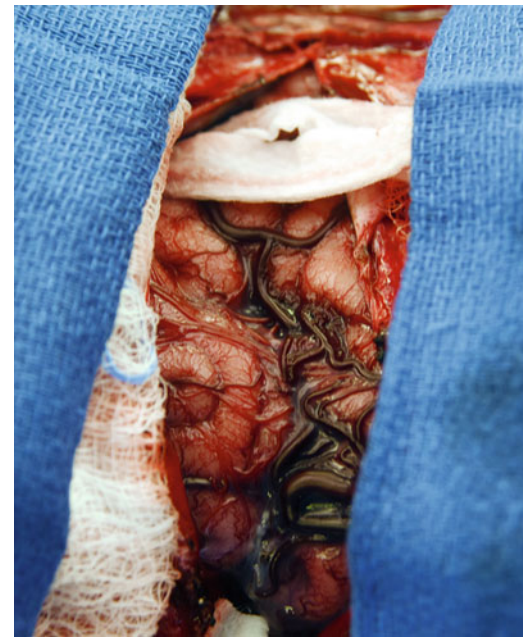

**Fig. 20** An interoperative photograph during the fourth of a four stages. The dense fusion and vascular interconnections are complex and have to be separated

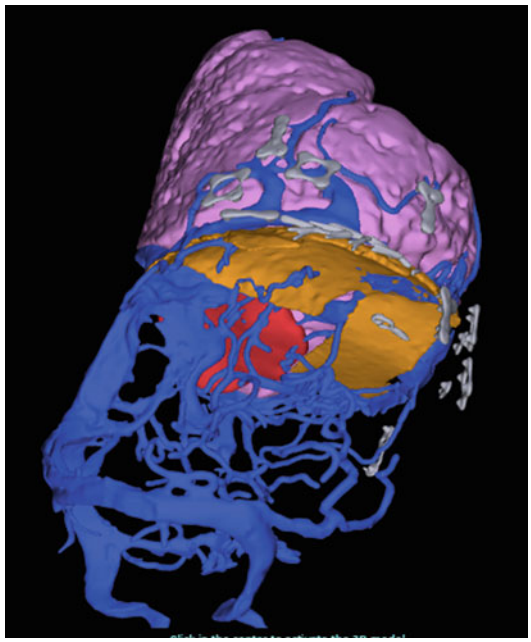

**Fig. 21** In this 3D reconstruction, the orange coloring is the area of the CVA. The area in red is the fused brain illustrated in Fig. 20. There is also a large torcula structure which in this case was separated with a Hunterian ligation

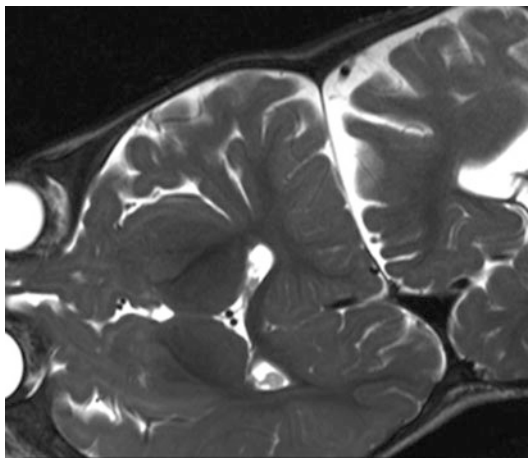

**Fig. 22** MRI T2 sequence showing an open nonfused interface between the two brains. Dura is present which is a very helpful finding in determining if there is conjoined brain

MRI shows CSF space at the brain interface but no dura then the two brains are likely fused. If the gyri interdigitate then this is always fused brain. To date, all of the surgically operated cases have had some, if not significant, degrees of brain connection. If the amount of fused brain exceeds 6–8 cm of brain, the morbidity and mortality increases significantly which is a critical factor in the decision to perform the separation procedure (Figs. 22, 23, and 24).

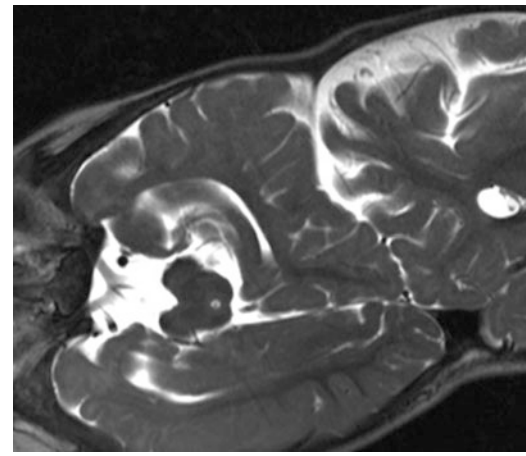

**Fig. 23** MRI T2 sequence with a good CSF space superiorly and the gyral patterns are clearly outlined. The inferior half, despite the presence of a CSF signal will be in reality fused brain. We have found situations like this in many of the studies

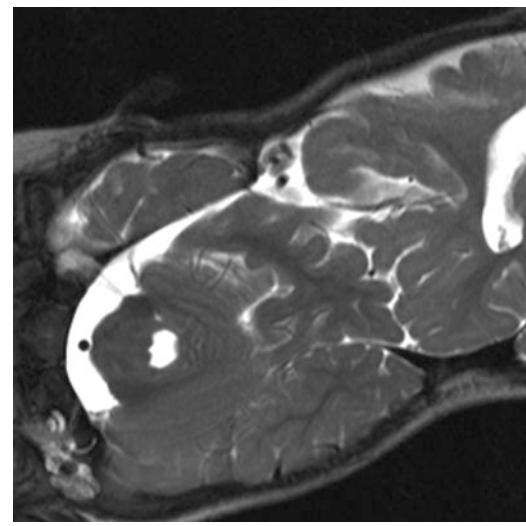

**Fig. 24** MRI T2 sequence which again shows a CSF space at the interface of the two brains. In reality this will be fused brain and the CSF nonexistent. The interdigitating gyri is also a strong clue the brains are conjoined

## Staging the Surgical Separation

The first surgical decision is to decide on which side of the CVP, the surgical plane is to be made. In all cases so far, we have given the CVP in toto to the dominant twin with the most vigorous venous outflow. That will mean the nondominant twin will have to develop its own deep venous circulation. This twin does not develop new veins, but instead makes use of preexisting veins that have not been dilated out. As the stage separations go forward, these venous patterns will become quite robust. Even more interesting, the hemodynamic issues will start to resolve as the children become more equal in their outflows. This is also the case with the renal systems. The oliguric child will typically increase the urine output so that by the final stage both will have nearly normalized urinary outputs (Figs. 25, 26, and 27).

The first stage is typically done at the area between the two brains where brains interface and there is a good CSF plane. Commonly one of the sagittal sinuses is taken at this time. A

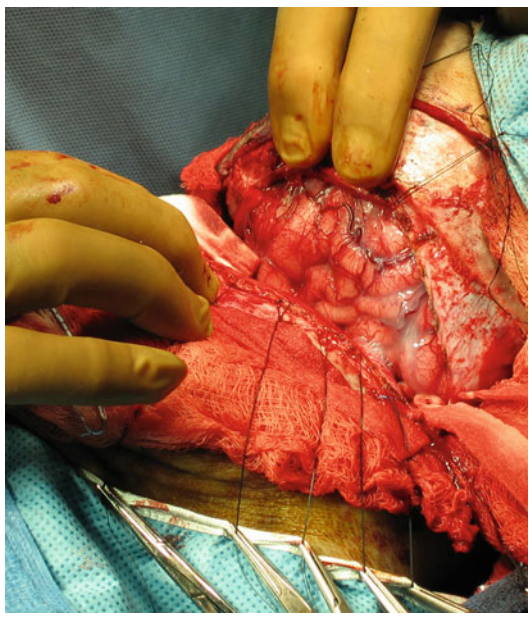

**Fig. 25** Stage one of a vertical set of conjoined twins. The first craniotomy has been done and the dura opened exposing the brain and arachnoid surface, which is under the fingers of the hand to the right. Under the left hand is the dural margin and the dural of the CVP

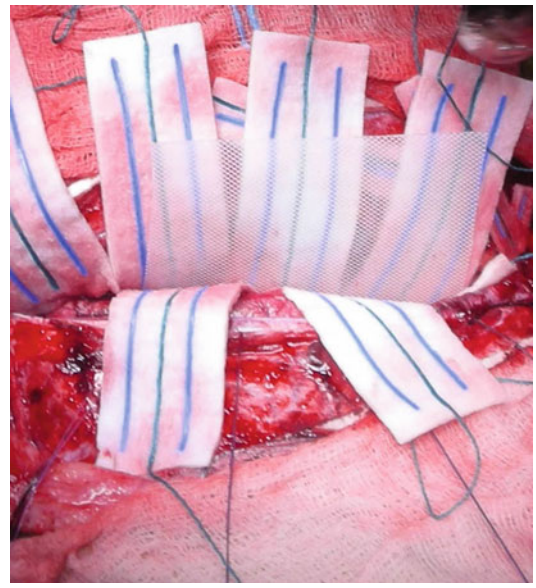

**Fig. 26** Illustrated here is the placement of a silicone sheet in between the dissected brains. This sheet prevents adhesions between the two brains while going through the various stages

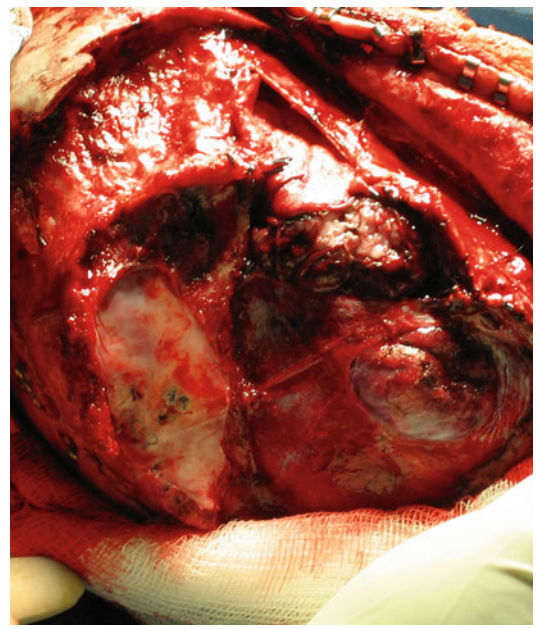

**Fig. 27** A case of a vertical craniopagus twins that have just been separated. The lower and left half of the brain is covered with dura. The other twin had deeply invaginated into his twin brother making the surgical planes somewhat challenging. The area to the top was all fused brain and included the right parietal lobe of one child fused to the left parietal lobe of the other twin

craniotomy is done; typically about 25% of the skull is elevated. Of note, the side of the skulls the children lay on commonly has very dense adhesions to overlying skull bone. This craniotomy can be quite tenacious to elevate, the other craniotomies not typically so. The width of the flap should be wide enough to include the interface of the two brains and then another 15–20 mm in additional width. It is quite common that the brains are not "flat" surfaces as they abut; rather they are very uneven and in parts can form deep invaginations into the conjoined brain. 3D modeling has been very helpful in working these areas out. Once the surgical plane has been dissected, a silicone sheet is cut and shaped and placed between the exposed two brains. By doing this, the brains will not adhere and thereby allow an easier separation at the final stage. We have found that four stages timed over 8–12 months have worked well for us. The craniotomy flap is fixated with a "micro" metal fixation system. This fixation is needed, as each craniotomy will be elevated again at the final surgery.

The dura is opened being careful to leave a 0.5–1.0 cm edge of dura on the dominant twin's side. This dural edge is important for closing, and one should be cautious about getting into the CVP, as the dura is extremely thin and not easy to repair. As you encountered the bridging veins, they are cauterized. We are also generous with the use of vascular clips provided in multiple sizes. If the veins are large we will put a temporary vascular clip on and observe for any brain swelling. If swelling occurs, then we will take the vein at slower pace. In the larger structures, like the sagittal sinus, especially those that come into the torcula, we will do a Hunterian ligation with silk sutures. If significant swelling develops, the team takes a break to give the brain time to reroute the venous blood, and this can take up to 15–20 min in some cases.

Fused brain is quite common in craniopagus twining. If the amount of fusion is significant, we will save this portion for separation till the last stage. In some cases, this is rare; you can tease the brains apart. In most cases, you will have to dissect through the fused portion so that you can go around this area circumferentially. On the surface of the fused brain, there is typically a sulcal pattern where the veins come out and go in opposite directions. This is the plane to follow for the final separation of the two brains. 3D modeling has been extremely helpful in defining these fused areas of brain (Figs. 28 and 29).

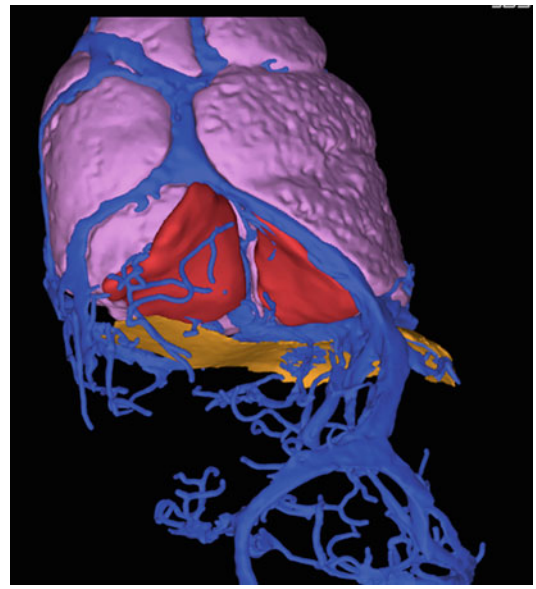

**Fig. 28** 3D reconstructions for the final fourth stage separation in a vertical set of twins. The area in red is the fused brain. The brain shown inferiorly has triangular projection of the parietal lobe up into the other twin. The 3D modeling was very helpful in mapping out the surgical planes and where to cut through the fused brain. The large torcula was given to the other twin, but each of the sinuses coming into it was ligated

For the final separation, we have the children on two separate OR tables. The heads are resting on separate horseshoe head holders or the "U" shaped cup holders. This positioning allows the two tables to be rotated 90° and each child then has its own independent space and surgical teams to begin the reconstructions and closure.

The dural closure will be different in each child. The child that took the CVP will have a full dura except at the area of conjoined brain. This is usually a straightforward closure with a dural graft needed for the defect. We have been

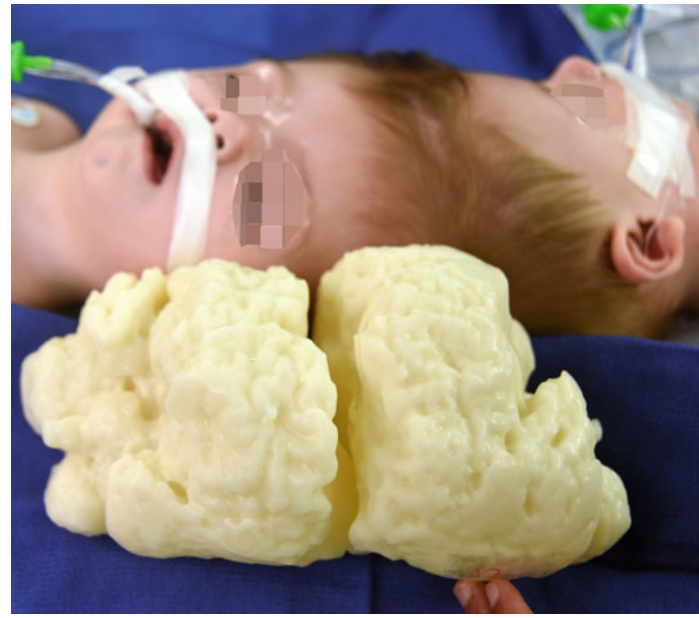

**Fig. 29** These reconstructed models have been helpful in detailing the interface between the two brains. Not easily appreciated from the radiological studies was the vertical projection of one twin's brain into the other twin. There was a 6–7 cm-wide bridge between the two parietal lobes. Post op, both children had significant hemiparesis – one on the left, the other on the right. One child recovered full function within 2 months. The other child also has had an excellent recovery though still has some neglect in using the hand


**Fig. 30** In the OR, the 3D printed models have been very helpful positioning, especially during the various stages. The children laying on two reversed abutting operating tables. Horseshoe head holders are used to support each twin. At the time of the final separation, the tables are pulled apart and rotated 90° so that each of the twin's teams can come in and complete the reconstructions

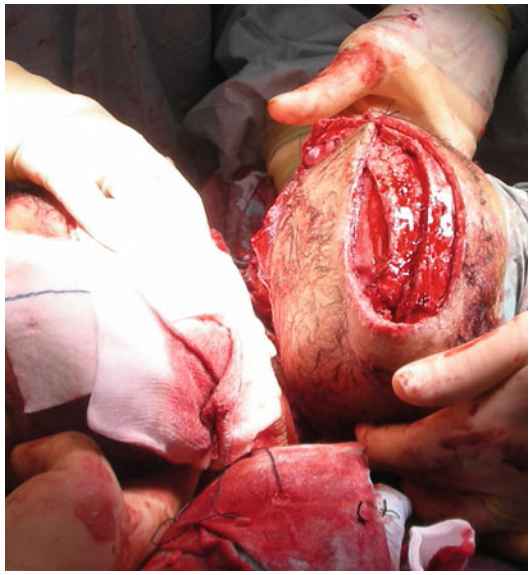

**Fig. 31** At the time of the final separation, each child is firmly supported to prevent the brain from herniating. This is especially important in the twin, which needs the dura reconstruction. Warm and wet lap pads soaked in a physiological saline solution are used for protection

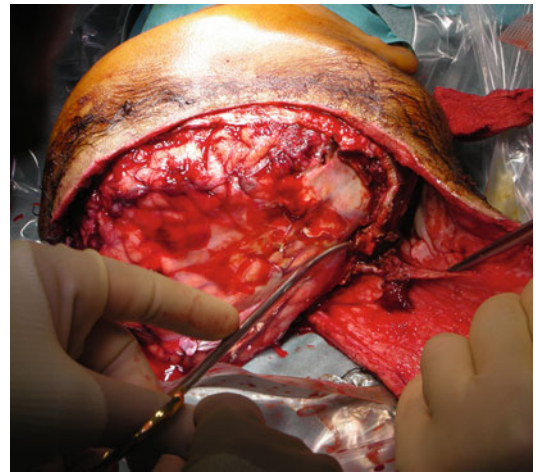

**Fig. 32** In this operative view, the amount of exposed brain with no dura is quite apparent. The expanded skin flap is off to the right and needs to be kept on stretch for the plastic surgeons while the neurosurgeons are reconstructing the dura

using a porcine graft (Durasys – Cook, USA) for our repairs. This graft sews nicely, can be made watertight, and forms an excellent dura matrix useful for any later reconstructions. The other child will have no dura and at the time of separation, a member of the surgical team will need a warm wet lap pad to hold the brain from slumping out of the skull. The dural repair here is much larger and again meticulous attention has to be devoted to the closure so that it is watertight. We have had discussions about using material like fascia lata, but this is a grainy material and difficult to sew in a watertight fashion. We always try to keep the pericranium as a separate layer at each surgery, but after four stages, it is not as viable and not large enough to close even in the best cases. (Figs. 30, 31, and 32).

During the surgery we keep the tissue expanders in place to reduce the amount of shrinkage that naturally occurs due to skin elasticity. If there has been too much shrinkage, then skin grafts can be placed and ideally these should be over areas where there is well vascularized tissue to maximize graft viability. Once the dura is closed, the plastics team completes the final skin closure. Meticulous attention is necessary for the closure so as to avoid over stretching and necrosis and even worse CSF leakage.

## Postoperative Care

Extubation is common at the end of surgery in the initial stages. These procedures have typically shorter operative times of 3–5 h. The final stage is quite long as the original craniotomies have to be redone for the final brain separation. The closure of dura followed by the reconstruction can take an additional 6–8 h, so in total the final case length can be up to 24-h. The children are then kept intubated anywhere from 3–7 days. This allows any brain swelling that has occurred to resolve. It is also a period where the children will often be quite agitated; therefore additional sedation helps reduce stress on the children and also on the scalp flaps.

Using staged surgical procedures, the development of hydrocephalus has not occurred in any of our sets of twins. But having said this, the team

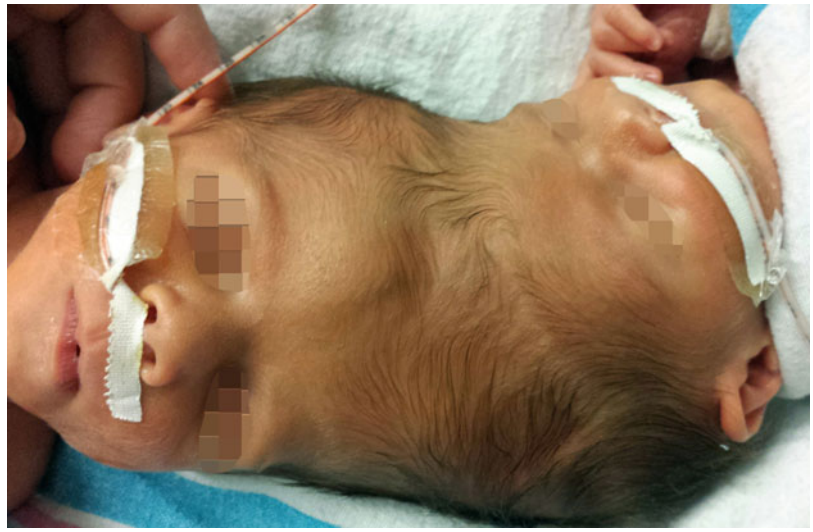

**Fig. 33** A set of vertical craniopagus twins shortly after birth

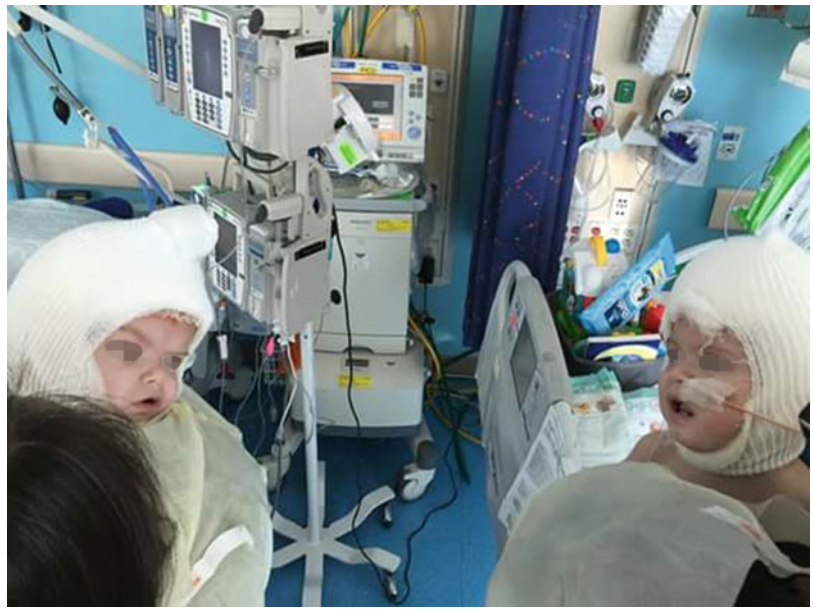

**Fig. 34** The same set of twins after a four stage separation seeing each other for the first time. This photograph was taken 3 weeks postop

should always be considering hydrocephalus a real risk. If hydrocephalus does develop, this will increase the probability of a CSF leak. In reviewing the earlier literature on single-staged separations, CSF leakage often led to infection, brain injury, and worse outcomes.

Fascinating to watch unfold during the staged separations are the changes in physiology in each child. As a result of the unequal sharing of vascular flow, it is common for one of the twins to be oliguric with minimal urine output. The dominant twin does the vast majority of the dialysis typically going through 5–8 diapers a day, while the other may use only one a day. As the staged surgeries advance, it is common to see the oliguric twin pick up on urinary output and

normalize. The same happens in cardiac output and correction of cardiac output failure. Widely varying blood pressures will normalize as the stages are done along with reduction of the cardiac medications.

We have been routinely using perioperative anticonvulsant medications (ACD). While there has been no set length of time for treatment, the average time for these children has been in the range of 4–6 months of postoperative treatment. In the 16 children now separated, four have require long-term ACDs.

## Conclusion

There are number of key points to be addressed in assessing a set of craniopagus twins. First and foremost is the amount of conjoined brain and the vascular venous anatomy. Single-stage separations should only be reserved for those cases of minimal conjoining, i.e., scalp and skull only. The decision as to which twin will get the CVP is key and typically this structure goes to the dominant twin, i.e., the twin with dominant venous outflow. A significant advantage of doing staged separations in craniopagus twins is the ability to stop the surgery at any point when critical issues such as excessive blood loss or venous swelling of the brain occur. Allowing the two brains to equilibrate by staging only is the wisest option. Instead of vascular gridlock, one allows the venous patterns to slowly reroute reducing infarction and swelling. We have learned to gauge the time between each stage based on how well the separation is going at the time and on how well each twin is recovering.

Surgical errors to avoid: (Alokaili et al. 2015) The team attempting get too much done in each stage leading to potentially severe edema and wound healing complications, (Browd et al. 2008) Not allowing enough time between each stage for healing and softening of the potentially tight indurated scalp (Browd et al. 2011). The timing between stages cannot necessarily be determined in advance and is best judged by the surgical team's examination of the patients healing and nutrition. The addition of OT/PT between stages is extremely helpful in the recovery. Monitoring nursing care is critical after the placement of the tissue expanders. Extruded or exposed expanders lead to the loss of scalp covering due to necrosis and can result in CSF leakage (Figs. 33 and 34).

Craniopagus twins are clearly a complex multidisciplinary problem when it comes to deciding on a surgical separation. The team's philosophy should always be teamwork; determining what will be triumphs, knowing when to walk away, and understanding that each is equally important. As seen in our experience, only one-third made it to the operating room. Hopefully those numbers will improve as more data and surgical technique come to the forefront.

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