Temporoparietal craniopagus

Case report and review of the literature

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A case of craniopagus twins joined in the temporoparietal area is presented, along with a review of the literature on craniopagus. A large area of brain was shared between the neurologically normal infants, with defects in the scalp, skull, and dura. The twins were separated in a three-step procedure. First, areas of shared brain were divided and separated with silicone sheets. The second procedure consisted of the insertion of scalp expanders to allow primary skin closure. In the third procedure complete separation was performed which was complicated by severe hypotension in one infant that was due to dural sinus hemorrhage. Cerebrospinal fluid leak was the most difficult problem encountered in the postoperative period; this was treated with lumboperitoneal and ventriculoperitoneal shunts. After 2 years, one twin is neurologically normal; the other is severely developmentally delayed, possibly related to the severe hypotension experienced during the third procedure. A review of the literature on craniopagus is presented. Analysis of data in the literature suggests that the area involved in the craniopagus as well as the venous connections are closely related to survival following separation of craniopagus twins.

KEY WORDS • craniopagus • conjoined twins

Conjoined twins represent a rare congenital abnormality, occurring at a frequency of 10.25 per million births in the United States. Of the various forms of this condition, cephalic conjoining or craniopagus comprises 6% of the total, making the appearance of this disorder 0.6 per million births. Attempts to separate such twins have had mixed results, with 24 of the 48 reported patients dying in the postoperative period, and an additional two patients left with severe neurological dysfunction.

A case of twins joined at the temporoparietal area is described in this report. They shared a common tentorium and cerebellar tissue. The plastic surgical aspects of the case have been described previously. The neurosurgical aspects of the separation are reviewed here, and a statistical analysis of 20 cases in the literature is presented, leading to a discussion of the differences between the forms of craniopagus.

Case Report

Craniopagus was diagnosed prenatally by ultrasound in a 20-year-old woman who had previously had three pregnancies and one delivery. This case has been presented previously, and only the neurosurgical aspects will be discussed here.

The temporoparietal area of Twin B was attached to the left temporoparietal region of Twin A, with the long axis of the twins forming a 75° angle (Fig. 1). The circumference of the attached region was 34 cm. The ears of both infants on the affected side were set low. The anterior fontanels were soft and the junction was rigid. Neurologically, the infants were normal with no focal deficit.

Computerized tomography (CT) scanning on the 1st day of life (Fig. 2) revealed apparently normal brain tissue extending through the skull defect area. Two falces cerebri were identified, connected by an abnormal dural fold running perpendicular to the line of connection. On either side of the fold, brain tissue protruded through the defect. This dural fold merged inferiorly onto a common tentorium, and additional brain tissue protruded through a defect inferior to this common tentorium.

Angiography performed on Twin B at 3 days of life provided evidence of joining of the vertebral arterial...
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Fig. 1. Photographs of the infants following birth. Left: Twin A is shown on the left and Twin B on the right. Right: Twin B is shown on the left, and Twin A on the right.

Fig. 2. Computerized tomography scans showing two falces cerebri (arrows, left) and the attachment of the falces cerebri onto a common tentorium (arrows, right).

supply of the twins (Fig. 3 left). The left vertebral artery of Twin B had abnormal branches that crossed the plane of junction and supplied tissue of Twin A. The left carotid artery of Twin B was also abnormal with an aberrant middle cerebral artery (Fig. 3 right); however, there did not seem to be cross circulation between the infants in the supratentorial area as seen in the anteroposterior projection. The dural sinuses between the infants could not be seen well even with subtraction views, probably due to dilution of dye by blood flowing from the uninjected twin. Cross circulation between the twins was subsequently confirmed by injecting technetium-99 diethylenetriaminepenta-acetic acid into Twin B and detecting excretion of the tracer by both infants. An electroencephalogram performed on both infants was normal.

A bicoronal incision was made at the initial operation performed at 9 days of life. Twin A was on the right as viewed from the surgeon's perspective. An abnormal suture in the bone could be seen running between the sagittal sutures of each infant (Fig. 4 upper). A diamond-shaped craniotomy was made anterior to this suture, and the dura was linearly incised. Immediately under the abnormal suture lay a sinus joining the two sagittal sinuses of both infants. Brain could be seen extending across the skull defect, and a cerebrotomy was performed to expose the common tentorium. A piece of silicone sheeting, folded to double thickness, was sewn into place to provide a plane for cleavage during the final separation. In the process of attaching the silicone sheeting, we realized that the dural fold was continuous with the left lateral wall of the intracranial compartment, and that there was no connection in this compartment between the brains of the infants. The isolated portion of the brain to the left of the cerebrotomy was removed, and the suturing of the silicone sheeting was completed (Fig. 4 lower). Posteriorly the silicone sheet was attached to a dural fold extending beneath the abnormal sinus connecting the two infants. The bone flap was replaced to provide good support for the scalp expanders which would be used later. One child developed hypotension, in spite of a relatively low blood loss of approximately 200 cc; this precluded performing the second part of the craniotomy over the second compartment in the joined region. During closure, it was noticed that the hypotension encountered was simply a result of the slight elevation of the hypertensive child above the normotensive twin, which allowed pooling of blood in the dependent twin. This hypotension could be easily controlled by adjusting the lateral tilt of the operating table. In subsequent procedures, blood pressure was maintained by using the side-tilt control in this fashion.

A second procedure performed 2 days later was a mirror image of the first with Twin B on the right. With knowledge gained from the first procedure, a single thickness sheet of silicone was sewn in place after retracting Twin B's parietal and temporal lobes to the right. The intracranial compartment found was identi-
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Fig. 3. Left: Left vertebral angiogram, anteroposterior projection, showing an aberrant course of shared branches of the posterior cerebral artery (arrows). Right: Left carotid angiogram, anteroposterior projection, showing an aberrant left middle cerebral artery (arrows).

cal to the one exposed in the first operation. There was no postoperative neurological deficit; a small subgaleal effusion slowly resolved without cutaneous leak.

A third operation was performed at age 36 days. By modifying the initial incision from the previous procedures, the bone ridge over the connecting dural sinus and the sinus itself were both severed. The primary purpose of this procedure was to insert Silastic tissue expanders* discussed in the previous report.18

The final separation occurred at 106 days of age. The bone flaps were elevated, and the planes between the silicone sheets were easily identified, allowing separation of approximately two-thirds of the shared area without significant blood loss. When the common tentorium or horizontal dural fold was opened between the infants, conjoined brain consisting of abnormal cerebellar tissue was found (Fig. 5). At the junction of the dural folds and at their reflection onto the dura of the cranial vault, large abnormal dural sinuses were identified. These sinuses consisted of the lateral sinuses of each infant and their junction with the left sigmoid sinus of both infants. Bleeding was well controlled until the end of the operation when these sigmoid sinuses were encountered. Hemorrhage from these sinuses could not be controlled with simple bipolar coagulation due to the acute angle formed by the infants' skulls. The final portion of the separation was carried out blindly, and severe hypotension immediately developed in both twins. Hypotension lasted approximately 10 minutes in Twin B and 45 minutes in Twin A. Closure was performed as an emergency procedure during resuscitation in Twin A, who was taken immediately to the intensive care unit. Bleeding was more easily controlled in Twin B, and closure was performed with placement of silicone sheeting over the exposed brain. In both twins, scalp closure was performed primarily.

Twin A was returned to the operating room the next day because of swelling underneath the flap and decreased level of consciousness. A large subgaleal hematoma was removed and a silicone sheet was sutured to the dural edges to separate the brain from the scalp flap.

Cerebrospinal fluid (CSF) leaks were persistent problems in both infants. Attempts to control the leaks by revision of the skin margins were unsuccessful. Repeat CT scans revealed slit ventricles in both infants. The leaks were initially controlled with daily lumbar taps, and subsequently both twins underwent lumboperitoneal shunting, which resulted in cessation of the CSF leaks. These shunts eventually had to be removed after the CSF was colonized by *Candida and Staphylococcus*. Twin A subsequently developed hydrocephalus and had a standard ventriculoperitoneal shunt placed after an intravenous course of amphotericin. Twin B has been shunt-independent since the removal of his lumboperitoneal shunt, and repeated CT scans have shown no evidence of ventriculomegaly.

At 2 years, Twin A continues to be institutionalized with poor feeding, respiratory problems, and severe neurological deficit, including a right facial droop and a left gaze preference. He grasps objects but does not transfer and has only fair head control; he manifests a severe developmental delay. Twin B at 2 years old is normal with excellent ambulation. His developmental milestones are normal; his last CT scan showed normal cerebral anatomy except for an area of hypodensity in the left occipital region.

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* Expanders manufactured by Dow Corning Corp., Midland, Michigan.
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**FIG. 4.** Upper Left: Intraoperative photograph, with scalp flaps retracted, showing the sagittal sutures (arrows) and an abnormal suture running between two sagittal sutures (arrowheads). Upper Right: Drawing demonstrating areas of craniotomies performed at the first and second operations. The dural incision is marked by a dotted line, and the venous sinuses found at surgery are indicated by shading. The abnormal sinus is shown running between the two sagittal sinuses of the infants. Lower Left: Operative drawing of the deep suture line attaching the silicone sheeting to the common tentorium between the infants. The retractor is on the brain of one infant; the other wall of the cavity is formed by the falk cerebri of the other twin. Lower Right: The superficial suture line closes the dural defect in the twin to the left. Dural closure in the twin on the right is formed by the falk cerebri of that twin.

**Review of Cases**

Surgical separation of craniopagus twins was most recently reviewed by Todorov, et al., in 1974. They identified 14 reported cases of craniopagus in sufficient detail to allow secondary analysis. Since the publication of that paper, seven additional cases have been described, including the current report. These 21 cases are summarized in Table 1. For purposes of simplification, they have been grouped by the area of junction into frontal, parietal, temporoparietal, or occipital junctures as depicted in Fig. 6. In frontal craniopagus, the twins face each other and the axis of the bodies forms an acute angle. In parietal craniopagus, the junction is at the vertex with the axis of the twins forming an extreme obtuse angle. Temporoparietal and occipital craniopagus twins have an axis of junction forming an acute angle. In temporoparietal craniopagus, the ipsilateral ears tend to be set low and the area of junction is immediately above the external acoustic meatus. In occipital craniopagus, the area of junction lies immediately over or involves the external occipital protuberance, and the infants face away from each other. The amount of venous drainage interrupted...
TABLE 1

Summary of reported cases of craniopagus*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors &amp; Year</th>
<th>Cerebral Connection</th>
<th>Staged Separation</th>
<th>Separation Age (mos)</th>
<th>Twin A</th>
<th>Twin B</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Area Joined</td>
<td>Venous Drainage</td>
</tr>
<tr>
<td>1</td>
<td>Cameron, 1928</td>
<td>no</td>
<td>no</td>
<td>0.3</td>
<td>P</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>Leiter, 1932</td>
<td>yes</td>
<td>no</td>
<td>0.3</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>Barbosa, 1949</td>
<td>no</td>
<td>no</td>
<td>0.1</td>
<td>P</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>Grossman, et al., 1953</td>
<td>no</td>
<td>yes</td>
<td>13.1</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>5</td>
<td>Robertson, 1953</td>
<td>yes</td>
<td>yes</td>
<td>5.0</td>
<td>P</td>
<td>3</td>
</tr>
<tr>
<td>6</td>
<td>Wilson &amp; Storer, 1957</td>
<td>no</td>
<td>no</td>
<td>5.0</td>
<td>F</td>
<td>2</td>
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<tr>
<td>7</td>
<td>Voris, et al., 1957</td>
<td>yes</td>
<td>yes</td>
<td>6.0</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>8</td>
<td>Baldwin &amp; Dekaban, 1958</td>
<td>yes</td>
<td>yes</td>
<td>3.0</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>9</td>
<td>Nolot, 1963</td>
<td>no</td>
<td>no</td>
<td>0.3</td>
<td>O</td>
<td>3</td>
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<tr>
<td>10</td>
<td>O'Connell, 1964</td>
<td>yes</td>
<td>yes</td>
<td>11.0</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>11</td>
<td>O'Connell, 1964</td>
<td>yes</td>
<td>yes</td>
<td>11.0</td>
<td>P</td>
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<tr>
<td>12</td>
<td>O'Connell, 1968</td>
<td>no</td>
<td>yes</td>
<td>12.0</td>
<td>P</td>
<td>2</td>
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<tr>
<td>13</td>
<td>Wolfowitz, et al., 1968</td>
<td>no</td>
<td>no</td>
<td>3.0</td>
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<td>1</td>
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<tr>
<td>14</td>
<td>Kohama, et al., 1972</td>
<td>yes</td>
<td>yes</td>
<td>4.0</td>
<td>P</td>
<td>3</td>
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<tr>
<td>15</td>
<td>Pertuiset, 1975</td>
<td>no</td>
<td>no</td>
<td>2.0</td>
<td>P</td>
<td>1</td>
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<tr>
<td>16</td>
<td>Marcinski, et al., 1978</td>
<td>no</td>
<td>no</td>
<td>0.2</td>
<td>P</td>
<td>1</td>
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<tr>
<td>17</td>
<td>Jain, 1979</td>
<td>no</td>
<td>no</td>
<td>1.3</td>
<td>P</td>
<td>3</td>
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<tr>
<td>18</td>
<td>Wong, et al., 1980</td>
<td>yes</td>
<td>yes</td>
<td>15.0</td>
<td>P</td>
<td>2</td>
</tr>
<tr>
<td>19</td>
<td>Villarejo, et al., 1981</td>
<td>no</td>
<td>no</td>
<td>0.8</td>
<td>P</td>
<td>1</td>
</tr>
<tr>
<td>20</td>
<td>Stanley, et al., 1983</td>
<td>no</td>
<td>no</td>
<td>1.2</td>
<td>O</td>
<td>1</td>
</tr>
<tr>
<td>21</td>
<td>Bucholz, et al., 1987</td>
<td>yes</td>
<td>yes</td>
<td>3.0</td>
<td>P</td>
<td>3</td>
</tr>
</tbody>
</table>

* Area joined: P = parietal; F = frontal; O = occipital; TP = temporoparietal. Venous drainage: 1 = no venous drainage interrupted; 2 = only cortical veins divided; 3 = major dural sinuses encountered and ligated. Outcome scale: 1 = normal; 2 = focal neurological deficit; 3 = severe neurological deficit; 4 = dead.

TABLE 2

Grouping of cases by type of junction*

<table>
<thead>
<tr>
<th>Area Joined</th>
<th>Outcome Scale</th>
<th>No. of Cases</th>
<th>Cerebral Connection</th>
<th>Mean Venous Scale†</th>
</tr>
</thead>
<tbody>
<tr>
<td>frontal</td>
<td>1 2 3 4</td>
<td>6</td>
<td>33%</td>
<td>2.0 ± 0.89</td>
</tr>
<tr>
<td>parietal</td>
<td>4 6 2 17</td>
<td>29</td>
<td>48%</td>
<td>2.4 ± 0.78</td>
</tr>
<tr>
<td>temporoparietal</td>
<td>1 0 1 2 4</td>
<td>50%</td>
<td>50%</td>
<td>2.7 ± 0.58</td>
</tr>
<tr>
<td>occipital</td>
<td>1 0 0 2 3</td>
<td>4</td>
<td>0%</td>
<td>2.0 ± 1.0</td>
</tr>
<tr>
<td>total</td>
<td>11 6 3 22</td>
<td>42</td>
<td>43%</td>
<td>2.4 ± 0.79</td>
</tr>
</tbody>
</table>

* Outcome and venous scales are defined in Table 1. † Mean ± standard error of the mean.

TABLE 3

Grouping of cases by outcome scale*

<table>
<thead>
<tr>
<th>Out-</th>
<th>Mean Age (mos) at Separation</th>
<th>Cerebral Connection</th>
<th>Staged Separation</th>
<th>Mean Year Separated</th>
<th>Mean Venous Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2.9 ± 1.6</td>
<td>36%</td>
<td>36%</td>
<td>1969 ± 10.7</td>
<td>1.5 ± 0.69</td>
</tr>
<tr>
<td>2</td>
<td>11 ± 5.3</td>
<td>67%</td>
<td>83%</td>
<td>1971 ± 11.3</td>
<td>2.0 ± 0.63</td>
</tr>
<tr>
<td>3</td>
<td>7.0 ± 3.6</td>
<td>100%</td>
<td>100%</td>
<td>1969 ± 14.6</td>
<td>2.6 ± 0.38</td>
</tr>
<tr>
<td>4</td>
<td>3.5 ± 4.3</td>
<td>32%</td>
<td>27%</td>
<td>1959 ± 17.5</td>
<td>2.8 ± 0.50</td>
</tr>
<tr>
<td>total</td>
<td>4.7 ± 4.8</td>
<td>43%</td>
<td>43%</td>
<td>1964 ± 15.7</td>
<td>2.4 ± 0.79</td>
</tr>
</tbody>
</table>

* Outcome and venous scales are defined in Table 1. Means are given ± standard error of means.

by surgery is graded in Table 1 by a three-point "venous scale" — 1: no venous drainage was interrupted, 2: only cortical veins were divided, and 3: major dural sinuses were encountered and ligated in the course of separation. The outcome of surgery was graded by a four-point scale shown in Table 1. The presence of a cerebral connection is also noted in this table along with the age at final separation and whether the separation was staged.

Table 2 displays the cases of craniopagus classified by area joined. Parietal craniopagus is twice as common as all other forms of craniopagus combined, whereas frontal junction is associated with the lowest mortality and best outcome. This is in contrast to temporoparietal and occipital craniopagus, where only one twin has survived following separation prior to the current report. The frequency of cerebral connection is highest in temporoparietally joined twins and high in frontally joined infants, but it is rare in parietal or occipital craniopagus. An average venous scale was calculated for each form of craniopagus. This value is lowest for frontally connected infants and highest for temporoparietal craniopagus. Surprisingly, it is also low for occipital craniopagus which, because of the area joined, would be expected to have a relatively high incidence of venous interruption due to the close proximity of the junction to the torcular. However, this value may be spuriously low. There are only three cases of occipital craniopagus recorded in the literature. In one case reported in 1983 by Stanley, et al., a very abnormal
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FIG. 6. Schematic drawings showing (from top to bottom): frontal craniopagus, parietal craniopagus, temporoparietal craniopagus, and occipital craniopagus.

twin was joined only partially to an otherwise normal infant. This case was not so much an example of craniopagus, where the infants involved are symmetrical in size, but rather of a parasitic monster attached to the occipital area of an otherwise normal infant. If this case is excluded, occipital craniopagus is associated with the highest frequency of interruption of major dural sinuses, and of perioperative mortality.

In Table 3 the cases of craniopagus are grouped by outcome. The improvement in mortality following separation is apparent in that the mean venous scale, which increased from 1.5 for outcome Grade 1 to 2.8 for outcome Grade 4. It is interesting to note that the presence of a cerebral connection does not seem to play a role in survival following surgery, but does play a role in quality of neurological function in those who do survive the operation. Of those infants surviving separation, 36% with a normal neurological examination following separation had cerebral connections, whereas 100% of those with a massive neurological deficit postoperatively had a cerebral connection.

To investigate further the relationship of factors involving survival after separation, a statistical analysis was performed on the 42 cases of craniopagus using the Statistical Analysis System (SAS).\(^\text{17}\) Correlations among the variables including year, presence of cerebral connection, staged separation, age at final separation, area joined, venous scale score, and outcome grade were computed. In addition, these factors were compared to whether or not the infant survived the separation. For the purposes of analysis, areas joined were coded as: 1 for frontal junction; 2 for parietal junction; 3 for temporoparietal junction; and 4 for occipital junction. Outcome is coded 1 to 4, corresponding to the outcome scale as given in Table 1. Survival was coded as 0 if the infant survived separation, and 1 if it did not.

The variable that correlated most highly with outcome and survival is the venous scale score with a correlation coefficient of 0.70 for outcome and 0.62 for survival (p < 0.0001 for both). The year of separation is negatively correlated with outcome and survival, with correlation coefficients of −0.32 for outcome (p < 0.04) and −0.35 for survival (p < 0.02), again confirming the improvement in outcome in recent years. Performing the separation in a staged fashion is also correlated significantly (p < 0.03) with a lower death rate.

A separate analysis was carried out for those cases separated since 1955, so as to control for temporal changes in surgical techniques. In addition, the unusual case reported by Stanley, et al.,\(^\text{19}\) in 1983 was excluded from this analysis for the reasons previously given. Results are similar to those for the full series. In this group of 31 infants, the venous scale is again highly correlated with outcome and survival, as is the area joined. The area joined is significantly correlated with outcome (r = 0.45, p < 0.02) and with death (r = 0.36, p < 0.05). Staging of the operation is also correlated with lower mortality (r = −0.38, p < 0.05), and the area joined is significantly correlated with venous scale (r = 0.40, p < 0.03).

Discussion

Todorov, et al.,\(^\text{20}\) compared 24-year-old craniopagus twins who had not been separated to the surgically separated cases he found in the literature. The comparison led him to suggest that conservative management might be an acceptable alternative to separation, which carried high risks of death and major neurological deficit.
This opinion needs to be reevaluated in light of a decade of improvement in surgical and anesthetic techniques. Perioperative mortality in cases of craniopagus in the past decade is 36% compared to 61% for twins separated before 1974. As shown by statistical analyses, the death rate has fallen and is correlated with the year of the separation. Twin A in this report is a clear example of a patient who would have died had it not been for excellent anesthetic support and improved surgical hemostasis. Furthermore, this decrease in the mortality rate has not been at the expense of increased morbidity. Patients surviving separations recently have not been left with severe neurological deficit. Rather, postoperative quality of life has been excellent with only one of nine survivors of the past decade suffering a profound neurological deficit.

An operative mortality of 34% suggests that it is important to identify those factors that predispose to a difficult separation. Todorov, et al., identified four problems routinely encountered during separation that determined the surgical outcome, and proposed two factors that had an impact on the long-term quality of survival of separated twins. These factors are reviewed in light of current experience.

The first two problems encountered during separation are the plane of cleavage and presence of cerebral connections. Almost all infants have only dura separating their brains; bone partitions are rare. The presence of shared brain tissue does not necessarily affect survival after separation, but does affect quality of outcome. All cases left with profound neurological deficits have had cerebral connections prior to separation. Cerebral connections, consisting of shared neural tissue, tend to be common only in frontal or temporoparietal junctions. The presence of such connections cannot be reliably predicted preoperatively even with modern imaging technology. The apparent sharing of brain tissue as seen on CT scanning can be misleading as in our case, in which a dural separation was found intraoperatively even though none was seen on the CT scan. As neurological outcome is affected by such cerebral connections, it would seem reasonable to perform separation of the conjoined areas as early as possible in development so as to minimize neurological deficit. This was one of the primary reasons for performing a staged separation in the current case.

The third problem cited by Todorov, et al., is the presence of skull and scalp defects. Early reports espoused elaborate techniques to allow for skin closure which in some cases was very difficult. This problem has largely been resolved with the advent of subcutaneous skin expanders. Although CSF leakage was a problem in our case, the primary closure of the scalp and temporary diversion of CSF via a lumbo-peritoneal shunt were critical to the final resolution of this difficulty. The skull defect is not a major problem; we have found that these skull defects become smaller as the infants develop.

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The fourth problem, the extent of venous connec-

tions at the junction site, remains an extremely impor-
tant determinant of operative mortality and morbidity. The reduction of operative mortality and morbidity seen in the past decade is partially a result of better control of hemorrhage from these venous sinuses. However, as seen in this report and in the literature, it can be difficult to visualize dural venous sinuses in such infants on preoperative angiography. The good correlation between venous connections and area of junction pointed out in this paper suggests that the type of junction may be used as a surrogate for the amount of venous interruption that will occur during surgery.

Todorov, et al., proposed that the angle of junction was one of two important prognosticators of outcome from surgery. They hypothesized that an acute-angle junction led to a better outcome. The data do not support this notion. Temporoparietal and occipital junctions, which are always in an acute angle, are associated with a high mortality and morbidity. Rather, it may be that the type of craniopagus is a critical factor in determining outcome: temporoparietal and occipital junctions are related to the highest mortality and morbidity rates, followed by parietal junction, and then frontal craniopagus with the lowest mortality. This relationship is statistically significant when the cases reported prior to 1955 are removed from analysis.

The second prognosticator of long-term outcome identified by Todorov, et al., is the timing of surgery, but this is only somewhat important. Infants who have been separated immediately postnatally have the highest mortality of all age groups. However, this may be due to the fact that these cases represent the infants in greatest distress, requiring immediate separation in order to save one infant. Generally, surgeons have waited to separate craniopagus twins if their condition was stable enough to allow such delay. However, excessive delay with concomitant growth in skin and skull defects can make surgery more difficult. Our review did not find a significant correlation between timing of surgery and surgical morbidity and mortality. However, craniopagus twins who have been separated in stages uniformly did better than those separated at one-step procedures and have had a significant reduction in operative mortality. In the current case, blood loss was the major limiting factor to continued surgery, and this loss was minimized by performing a staged dissection and preserving the cleavage plane between operations with silicone sheeting. Staged separation is also beneficial in terms of allowing the use of skin expanders and in allowing the division of brain tissues early in development.

In 1976, O'Connell suggested classifying craniopagus twinning into partial or complete forms. He rejected the notion of classification based on location of junction as he thought it made no distinction between "unions of limited superficial extent and depth and the extensive unions in which the two cranial cavities are in wide connection." He concluded that parietal cranioptagous, which tended to have a wider area of junc-
Temporoparietal craniopagus

tion between the infants, was associated with the highest risk of mortality and morbidity. This hypothesis is not supported by the data in the current review. Parietal craniopagus carries a lower risk of mortality as compared to temporoparietal and occipital cases. In addition, O'Connell's distinction between partial and complete craniopagus is imprecise. Therefore, no attempt has been made to make this distinction in the current literature review.

It may well be that the amount of conjoined area involved in craniopagus infants will correlate best with the degree of fusion of venous structures as well as brain tissue. However, this information is rarely cited in the literature and therefore could not be factored into our current analysis. With the assimilation of more cases of craniopagus, the relationship between diameter of joined area and surgical outcome can be assessed, providing the surgeon with a greater ability to predict outcome following separation.

The current excellent outcomes in even difficult cases of craniopagus such as reported here suggest that separation should always be considered in craniopagus with the possible exception of the rare occipital form. A carefully planned staged procedure, approached with a team of neurosurgeons, plastic surgeons, and anesthesiologists, should have a high success rate in separating craniopagus twins who would otherwise lead a life of ridicule and deformity.

References

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