Anesthesia for Craniopagus Separation Surgery

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Key Points Craniopagus twins (CPTs) are rare forms of the very uncommon conjoined twins; surgical separation of CPTs is one of the most complex neurosurgery procedures.

- The surgery includes the multi-staged separation of shared vasculature, interdigitating brain parenchyma, and reconstructive plastic surgery; the procedures are meticulously planned with a multidisciplinary team's participation.
- Multiple anesthesia episodes are required, such as for neuroimaging, tissue expander placement, multi-staged separation, and reconstructive surgeries.
- Neuroimaging procedures for CPTs can be carried out under sedation as well as general anesthesia.
- For separation surgery, clear communication is required among the anesthesia team members with specific attention to possible difficult airway, careful positioning, appropriate management of massive blood loss and fluid shifts, and anticipation of complications.
- With advances in medical technologies and surgical expertise, more separation attempts are expected despite the associated high cost and procedural complexities.

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19.1 Introduction

Joined in utero, the conjoined twins are known as "Siamese twins." There are different theories related to aberrant embryogenesis proposing why conjoining occurs [1]. One such theory suggests that a single fertilized egg may not split fully during the process of formation of identical twins, and the zygotic division occurs 2 weeks after the development of the embryonic disc, resulting in the formation of conjoined twins (*fission theory*). The other theory suggests that two fertilized eggs fuse in the early part of the development process (*fusion theory*); subsequent splitting of primitive nodes and streak partially may lead to this rare phenomenon. Conjoined twins occur in not more than 1:50,000 to 1:200,000 births [2].

The conjoined twins are typically classified based on the part of the body where they join (Table 19.1). The incidence of different types also varies [3]. The most common types encountered are thoraco-omphalopagus, thoracopagus, omphalopagus, parasitic twins, and craniopagus; thoraco-omphalopagus is considered as the commonest type. Forty percent of the conjoined twins are still-born, and 60% are live-born; one-third of the live-born may die within 24 hours after birth due to congenital anomalies. Hence, the actual occurrence of conjoined twins is very rare, and only about 25% of these twins survive long enough to be candidates for surgical separation. Additionally, conjoined twins are genetically identical and are of the same sex. Conjoining is more common in females with a male/female ratio of 1:3 [4]. No association with heredity, race, maternal age, or environmental factors is established.

Table 19.1 Classification of conjoined twins

Types of conjoined twins	Description(s)
Common types	
Thoraco-omphalopagus	Fused bodies from the upper to the lower chest Heart is shared; may share liver or partly the digestive system
Thoracopagus	Bodies fused at the chest Heart is invariably shared
Omphalopagus	Fused bodies at the lower abdomen May share a liver, digestive system, and diaphragm; but never share a heart
Parasitic twins	Asymmetrically conjoined twins; one twin is small and less formed and is dependent on the other (larger twin) for survival
Craniopagus	Joined at the head, but not on the face or the base of skull
Rare types	
Xiphopagus	Fused at xiphoid process; no other organ involved except the liver
Ischiopagus	Joined at the ischium; the lower half of the two bodies are fused with spines
Pygopagus or iliopagus	Bodies fused at the pelvis (buttock)
Rachipagus	Fused at the back of their bodies with the fusion of the vertebral arches

When such twins are fused at the skull, they are known as craniopagus conjoined twins. Craniopagus twins (CPTs) are one of the rarest forms of conjoined twins and account for 2–6% of all conjoined twins, with an incidence of approximately 1 in 2.5 million live births [4, 5]. In these twins, cephalic fusion may occur at any part of the calvarium except the face, foramen magnum, skull base, and vertebrae.

19.2 Historical Aspects of Conjoined Twins

The uniqueness and uncertain origin of conjoined twins have inspired many myths and legends in ancient literature for centuries. Art forms of different examples of conjoined twinning can be seen in museums throughout the world. They were feared as bad omens and, hence, were abandoned or even killed. Later, they were viewed with curiosity; they became sideshow acts, performed in circuses, or even became stage performers. They were also worshipped as gods due to their unusual appearances.

The Biddenden Maids (Mary and Eliza Chulkhurst), conjoined pygopagus twins, were born in the year 1100 in the Kentish village of Biddenden, England, and they lived as long as 34 years. They are considered the first documented case of conjoined twins [6]. Similarly, the first documented case of craniopagus twins (CPTs) of Bavaria, Germany, was mentioned as a monster (Ein monstrum) and was considered as a warning signal from God [6]. The twins born in 1491 were frontal CPTs and remained alive for 10 years. The other historical aspects of CPTs were well documented by the famous French surgeon Ambroise Paré in the sixteenth century, and his works were reprinted in 1840 (*On Monsters and Marvels*) as monsters who were a "warning from God" [6, 7]. In the eighteenth century, Sir Everard Home (1756–1832) reported a case of craniopagus parasiticus known as "Two-Headed Boy of Bengal," the CPTs born in India in the 1770s and whose skull is preserved at the Hunterian Museum at the Royal Society of Surgeons [6, 8]. Until the late 1800s, the CPTs

were considered "monsters." However, in the nineteenth century, August Förster (1822–1865) defined the twins joined at the head and introduced the term "craniopagus" in his works on the science of teratology [6]. Thenceforth, the adjective of "monster" was gradually replaced with different types of conjoined twins.

Chang and Eng Bunker, the original "Siamese twins" who were synonymous with conjoined twins during the early nineteenth century, participated in "freak shows" and became financially successful. Together, they fathered 21 children and died at the age of 62. Yvonne and Yvette McCarther, the American CPTs (born in 1949), were considered inoperable and lived up to 43 years. Despite their rarity, several CPTs lived into adulthood; nevertheless, more than 90% died by the age of 10 [6].

Surgery is considered successful when both twins survive more than 30 days after separation [6, 9]. The first craniopagus separation surgery was attempted unsuccessfully in 1928 [10]. Until 1950, several CPTs underwent urgent yet unsuccessful separation surgeries. The first successful surgery (Roger and Rodney Brodie) was carried out in stages by Oscar Sugar (1952–1953). In this case, one of the twins died after 1 month of craniopagus separation, while the other survived until 11 years of age [9]. During the last 50 years, advancements in medical science, surgical strategies, anesthesia, and intensive care have encouraged approaching craniopagus cases with renewed interest. In this context, the contributions of renowned pediatric neurosurgeons like Dr. Ben Carson and Dr. James T. Goodrich are worth mentioning. Dr. Goodrich was the single most experienced surgeon for this complex craniofacial disorder and performed seven cases of craniopagus separation [7].

19.3 Classification of Craniopagus Twins

Various classifications have been proposed for describing CPTs; the most common is the O'Connell classification (1976). O'Connell broadly divided CPTs based on the extent of the

union as well as extracranial versus intracranial involvements [11]. CPTs could be partial with smaller extracranial union limited to the dura mater or total with the large intracranial union and extensively shared cranial cavities. Bucholz et al. subclassified the total craniopagus into four types: frontal, parietal, temporoparietal, and occipital [12]. O'Connell further subclassified the total (vertical or parietal) craniopagus into three types (I, II, III) based on the orientations of faces of the twins due to the long axis of one head that is rotated over the other through different angles (Table 19.2). Winston proposed another classification based on the "deepest shared structures" [13]. In the same year, Gaist and colleagues expanded the O'Connell classification with the inclusion of a transitional category apart from partial and total categories; deformities of the brain and cerebral venous connections were also described [14]. Stone and Goodrich proposed a simple classification [9], reviewing 64 cases based on *shared venous sinuses* and the extent of brain compression as either partial or total craniopagus. There were two subtypes, angular or vertical, for each category. The vertical craniopagi, like O'Connell classification, are

Table 19.2 Common classifications of craniopagus twins

Classifications	
Partial craniopagus	Total craniopagus
O'Connell classification	
Extracranial, usually frontal Sharing of minimal surface area	Intracranial Extensive surface area with wide connectivity of the cranial cavity • Type I: Facing same direction (<40°) • Type II: Facing opposite direction (140–180°) • Type III: Intermediate (40–140°)
Stone and Goodrich classification	
Less significant shared dural venous sinus • Angular • Vertical	Significant shared dural venous sinus Pronounced brain compression • Angular: <140° intertwined longitudinal axis • Vertical: I, II, III based on O'Connell classification

subclassified into three types based on intertwined axial facial rotations (Table 19.2). Browd et al. proposed a comprehensive grading system based on the issues related to separation surgery to evaluate the CPTs for surgical risks and possible success in separation [6].

19.4 Shared Vascular System in Craniopagus Twins

Cerebral venous system abnormalities are common in CPTs [11, 13]. Their complex nature may affect the separation surgery outcome due to significant intraoperative blood loss and postoperative venous infarcts. The superior sagittal sinus (SSS) may be absent in both the twins to be replaced by a single-shared SSS or circumferential venous sinus (CVS) (Fig. 19.1). Other than CVS, venous sinus lakes and separate SSS with interconnections between them have also been described [15]. These abnormalities may cause significant mixing of the venous circulation, and blood may drain to one twin preferentially. That leads to high blood pressure and cardiac output in one twin and low in the other [16, 17].

The cerebral arterial supply is usually separate in CPTs. Less commonly, there may be shared arterial supplies such as branches of one middle cerebral artery feeding both twins [18]; and large arteries may cross from one twin to another.

19.5 Management of Craniopagus Twins

Conjoined twins can be diagnosed during midpregnancy with a standard ultrasound examination; diagnosis also can be made by fetal magnetic resonance imaging (MRI). Delivery of these twins is commonly performed by a cesarean section a couple of weeks before the expected date; some twins have been reported to be delivered with normal vaginal delivery [19]. Proper evaluation needs to be carried out after delivery since CPTs may be associated with systemic comorbidities such as cardiovascular (hypo/hypertension, coarctation of the aorta, and patent ductus arteriosus), neurologic (hemiparesis, delayed milestones of development), and craniofacial (cleft lip and palate), genitourinary abnormalities, and anorectal agenesis. Cerebral blood flow (CBF) constitutes 15–20% of the cardiac output and in CPTs may present with unidirectional shunting of blood flow. Hence, the twins could present with features of cardiac straining as well as hypo/hypertensive episodes [6]. One of the twins may develop left ventricular hypertrophy secondary to chronic hypertension [20].

Fig. 19.1 Schematic diagram of a variant of shared venous architecture in craniopagus twins. A common venous sinus may drain the twins' cerebral cortices (Twin 1 and Twin 2)
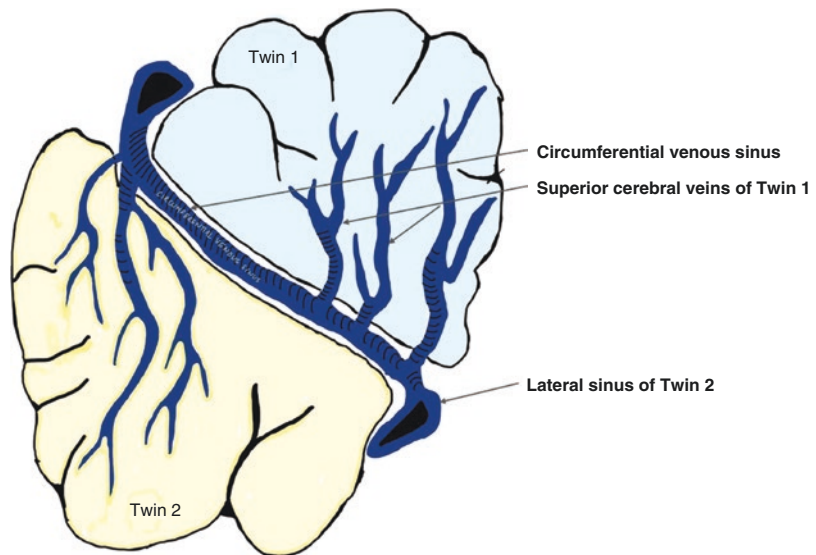
Separation surgery for craniopagus twins is a very complex procedure. It requires appropriate planning before separation and reconstruction of different layers of tissue such as the skin, skull bone, dura mater, brain parenchyma, and vasculature. The separation surgery can be carried out as a routine procedure to allow the twins an independent life. Their separability is determined based on the extent of sharing of intracranial structures. Separation at a younger age (~1–2 years) is recommended; it is presumed the brain plasticity may help the early recovery of brain insult after separation surgery [9, 12, 21]. It is also reported that mortality is higher with such an extensive surgery at a younger age [22]. Besides this, CPTs are separated on an emergent basis due to one child's death, increasing the other's risk of death. Anesthetic and surgical concerns remain the same whether separation is planned as a routine or emergency procedure. Multiple procedures are carried out to achieve a successful separation. Neurosurgical separation includes the separation of shared vasculature, interdigitating brain parenchyma, white matter connections between the thalamic regions [23], and other structures [16]. Reconstructive surgery includes cranial and soft tissue coverage, plans for duroplasty, cranioplasty, and tissue flaps.

The final tissue defect in cases of total vertical CPTs is expected to be quite large in surface area. It is challenging to cover, exposing the twins to further complications despite a successful separation of brain tissue and vasculature. Much before the final separation surgery, tissue expanders can be placed to create the extra amount of skin required for coverage. The skin expanders are placed anteriorly followed by posteriorly and expanded with aliquots of 10 mL of saline at regular intervals [24]. Saline injection for expansion into the expanders is usually associated with severe pain and requires analgesic supplementation. At times, thinning of the skin following rapid expansion may lead to implant extrusion due to skin ulceration, commonly seen posteriorly. The total time taken for adequate expansion of the scalp may range from a few months to 1.5 years. The tissue expander use may be deferred until the final

separation surgery (expanders kept for 4–6 weeks) to reduce infection risk [6]. However, the use of microvascular skin flaps obviates the need for skin expanders.

19.5.1 Staged vs. Non-staged Separation

The staged concept of surgery is based on the risk of massive blood loss and the twins' ability to tolerate the surgery. The presence of a shared SSS or CVS is the most challenging aspect of craniopagus separation surgery. In a single-stage separation surgery, the CVS is given to one twin, while the sinus is reconstructed in the other. It increases cerebral venous pressure during the separation; hence, it may lead to a cascade of events favoring failure of the procedure rather than success [25]. In a multi-stage approach, one twin (dominant) receives the CVS. Simultaneously, the other (non-dominant) develops the venous drainage system over a period during which serial surgical ligation and detachments of draining veins are carried out [26]. Staged separation offers a more graded approach to change the venous drainage in both twins. It improves venous collateral circulation and, hence, venous drainage, thereby preventing increased venous pressure and the possibility of brain edema. During final venous separation, the channels in each brain are adequate to allow complete separation. This process also favors the integrity of dura and flap repair that would reduce the risk of cerebrospinal fluid (CSF) leak. The staged approach is also intended to minimize intraoperative hemorrhage and transfusion of blood products. It may allow the twins to recover from each stage before progressing to the next stage with a gap of 4–6 weeks or more while continuing supportive therapy. Nevertheless, the other potential advantages of the staged separation surgery are the reduced duration of general anesthesia (GA), less bleeding, fewer transfusion requirements of fluids and blood, less probability of brain edema and infarction, and lesser fatigue of the operating team [24]. Technological advances in endovascular therapy also help prevent the draining/bridging

veins with coil embolization and avoid open surgery and associated complications [23]. Such procedures may play an essential role for staged surgeries in CPTs in the future.

After separation, the dominant twin may have robust vitals, whereas the non-dominant twin may present with low cardiac output, hypotension, oliguria, low weight, or failure to thrive. The staged approach is not required for partial CPTs.

19.6 Preoperative Evaluation and Preparation

There should be adequate preparations for manpower, equipment, and monitoring tools apart from a prior mock-drill before the CPTs are planned for separation. Multiple team meetings are necessary for appropriate planning and preparation.

19.6.1 Multidisciplinary Team (MDT)

A well-equipped multidisciplinary team (MDT) of surgical, anesthesia, and medical specialties, comprehensive radiological (anatomical) evaluation, addressing ethical concerns for separation, and parental participation are important prerequisites for the separation surgery of CPTs. MDT should ideally be formed under the leadership of a senior pediatric neurosurgeon, comprising of two pediatric neuroanesthesia leads (one for each twin), specialists from neuroradiology and imaging, pediatric intensive care, pediatric surgical specialties including plastic and reconstructive surgery, pediatric medicine specialties including cardiology and nephrology, and many other professionals [20]. The rarity of this complex, expensive, and lengthy yet technically challenging surgery for separation encourages international collaborations, particularly when planned in a developing country [23]. Moreover, multidisciplinary teleconferences are recommended in countries without extensive separation experiences in conjoined twins [27].

19.6.2 Neuroradiology and Imaging

Neuroimaging modalities such as computed tomography (CT), MRI, and digital subtraction angiography (DSA) contribute a very important role in diagnosing shared vasculature, interdigitating brain parenchyma, dura mater, and skull in the CPTs [26]. A comprehensive evaluation of the shared venous and arterial anatomy helps anticipate perioperative complications such as hemorrhage, air embolism, thrombosis, and infarction. Digital and 3D modeling of CT and MRI data is very useful for surgical planning and intraoperative guidance for the neurosurgeon. CT angiography (CTA) and venography (CTV) provide information on vascularity. CT venography is particularly utilized for planning at every stage of separation. MRI gives a detailed anatomical and developmental assessment of the shared cerebral cortex, ventricles, venous sinuses, and other anomalies. CTA and CTV are superior for studying the vasculature, whereas magnetic resonance angiography (MRA) and magnetic resonance venography (MRV) are superior in studying soft tissues, including brain parenchyma. In older or adult CPTs, functional MRI (fMRI) is used to define the hemispheric dominance of the language function and surgical planning [28].

DSA is used preoperatively for anatomical clarity of the vasculature, and venography helps to identify the twin with fully formed sinuses. A temporary balloon occlusion test may be carried out at the shared vinous sinus to test changes in the venous drainage and collaterals, hemodynamics, and requirements of a bypass [24, 29, 30]. Venous rerouting and promotion of collateralization can be achieved by using endovascular venous coil embolization [30, 31]. Full segment endovascular occlusion of the shared venous system has been utilized for successful separation [23]. The endovascular approach and separation could be a preferred procedure, since preoperative and intraoperative clipping and/or bypass creation are associated with very high risks. Coil embolization of shared arterial supply has also been utilized as a part of endovascular therapy in CPTs [18].

3D reconstruction of CT data helps in evaluating the extent of bone fusions in CPTs, which can be further reconstructed to create a life-sized 3D model of transparent acrylic and ceramic types as well as holograms for depicting vasculature in relation to the other tissues [26]. The 3D models better depict the surgical anatomy and are used for surgical reference at various steps along with intraoperative neuronavigation. It is also used to plan scalp tissue expanders' placement for adequate coverage and craniotomy and to design bone grafts for subsequent cranial reconstruction. Newer techniques such as computer-aided design and modeling, custom-made devices that used distraction osteogenesis and soft tissue molding, and intraoperative neuronavigation help successful separation among young twins [32].

19.7   Anesthetic Management

The CPTs may undergo many procedures under GA before the final separation surgery. Since most patients are young, GA is a prerequisite even for diagnostic imaging. Broadly, techniques may include (1) neuroradiologic imaging for planning and prognostication, (2) tissue expander placement at least in two stages, (3) single or staged separation, and (4) reconstructive plastic surgeries. Sometimes, an attempt to obtain central venous access for any reason other than surgery may also require GA.

During the preanesthetic evaluation, IV access and airway should be properly assessed along with a thorough general and systemic examination of the twins. It is advisable to restrict **vascular access** only to peripheral lines for minor procedures so that central veins could be utilized for the separation surgery. CPTs could likely present with **difficult airways** owing to distorted necks, conjoined heads with restricted mobility of head and neck, and congenital oropharyngeal anomalies. Possible problems with mask ventilation and endotracheal intubation should be anticipated. The signs of an increased airway obstruction would need nasopharyngeal airway in certain situations; a planned tracheostomy prior to separation sur-

gery would prevent loss of airway under challenging conditions.

Anesthetic concerns depend on the procedure planned (Table 19.3). Prior confirmation of logistic support, MDT meetings, as well as mock-drill at different anesthetic areas are mandatory to prevent confusion and possible mismanagement. While all imaging and surgical procedures would focus simultaneously on both twins, anesthetic management would require their management as different individuals since they are physiologically different [20]. Hence, there should be **two anesthesia teams** led by two pediatric neuroanesthesiologists. They should be supported by human resources (assistants and staff), equipment (anesthetic workstations with monitors, infusion pumps, and other equipment), and materials (drugs with specific color code for each twin, blood, and products), all in duplication.

The anesthetic management for CPTs can be broadly described under two headings: (a) anesthesia for neuroimaging procedures and (b) anesthesia for separation surgery. However, there are possibilities that CPTs could present for other procedures on an elective or emergency basis [33, 34]. In such a CPT case, one twin (dominant) successfully underwent adenoidectomy for obstructive sleep apnea under anesthesia [34]. Such types of emergency surgeries in CPTs, imaging and neurointerventional procedures, and different staging surgeries before the final separation may be described under *nonseparation anesthesia* (Table 19.4). The detailed discussion on this topic is beyond the scope of this chapter.

19.7.1   Anesthesia for Neuroimaging Procedures

The nil per oral (NPO) status and normal routine blood and urine need to be ascertained beforehand [35]. Sedative **premedication** should ideally be avoided in the twins; however, oral anxiolytics in the presence of parents help alleviate apprehension before subsequent activities. Induction should be carried out preferably in a place with appropriate arrangements, if not the operating room (OR), before neuroimaging. The

Table 19.3 Anesthetic concerns during the different procedure for craniopagus separation

Procedure(s)	Anesthetic concern(s)
Neuroimaging (CT, MRI, angiography)	1. Non-operating room anesthesia and other logistical issues 2. Effective communication between two anesthesia team members and other supporting staffs 3. Difficult airway and vascular access 4. Positioning patient, personnel, and equipment in different neuroimaging setups with less optimal conditions for the twins 5. Cross-transfer of administered drugs and fluids 6. Hemodynamic disturbances 7. Prolonged anesthesia time 8. Hypothermia 9. Contrast-related issues 10. Anesthetic neurotoxicity at younger age group 11. Issues with transportation to different neuroimaging suites
Tissue expander placements[a]	All concerns as above except 1, 4, 7, 9, 11 12. Surgical positioning for placement of expanders first anteriorly, and then posteriorly, in two stages
Separation surgery	All concerns as above except 1, 4, 7, 9, 11 13. Surgical positioning; preferably prone separation in the first stage followed by supine separation 14. Severe bleeding and massive transfusion 15. Massive fluid shift 16. Intraoperative tight brain 17. Venous air embolism (VAE) 18. Intraoperative cardiac arrest in one or both the twins and resuscitation 19. Long-duration surgery 20. Shifting of one twin immediately to the adjacent OR, kept ready, after separation, along with man, monitor, and machine 21. Infection control
Reconstructive surgery (duroplasty, cranioplasty, skin and tissue flaps/rotational flaps)	• Massive fluid shift • Hemorrhage and exsanguination of blood • Brain bulge during the cranioplasty • Infection control
Follow-up surgeries (ventriculoperitoneal shunt surgery for hydrocephalus and CSF leak, skin grafting)	• The absence of skull bone may lead to accidental pressure on the brain during the surgical manipulation causing hemodynamic perturbations (e.g., bradycardia) • Infection • Wound dehiscence

[a]This procedure may be combined with neuroimaging to reduce the number of anesthetics.

Table 19.4 Anesthesia encounters in craniopagus twins

Anesthesia for nonseparation surgery	Anesthesia for separation surgery	Anesthesia for reconstructive surgery and additional follow-up procedures

1. Neuroimaging procedures:
  • CT, MRI, angiography
  • Endovascular separation
2. Placement of tissue expanders to increase skin area necessary for coverage after final separation surgery
3. Multi-staged separation procedures before the final separation
  • Number of procedures depends on the separation planning
4. Emergency surgical procedures not related to separation: For example, adenoidectomy, colostomy, etc.
	1. Surgery during the final separation
	  • Occurs in the last and single planned attempt
		1. Plastic and reconstructive procedures
		  • For example, duroplasty, cranioplasty, skin grafting, etc.
		  • Number of anesthetics depends on the requirements for individual twins
		2. Additional procedures:
		  • Ventriculoperitoneal (VP) shunt surgery for hydrocephalus
		  • Placement of lumbar drains

difference in hemodynamic parameters (heart rate and blood pressure) gives a rough idea about the presence of physiological interdependence (cross circulation) in CPTs before neuroimaging. Cross-circulation between the twins may also be ascertained with IV injections of anticholinergic agents (atropine or glycopyrrolate) if an IV access is present. The variations observed in hemodynamics (heart rate) at different time points suggest the presence of cross-circulation [16, 17, 35]. Premedication with anticholinergics also helps reduce oropharyngeal secretion and may be useful, particularly when intramuscular ketamine is used for induction before IV access is secured [36]. Anesthesia may preferably be induced with sevoflurane if IV access is not secured in both the twins, else propofol may also be used. However, the procedure such as MRI can be carried out under sedation with oral triclofos, intermittent boluses of propofol along with O_2 supplementation [35].

The anesthetic **induction** may be carried out simultaneously in both twins or one after another at an interval of few minutes. Crossover of the anesthetic agents may induce anesthesia in the other twin simultaneously. Hence, oxygenation with mask ventilation should be carried out in both accordingly. Ideally, two anesthesia machines are utilized for anesthetizing CPTs. The anesthetic locations outside the OR are unlikely to have spacious accommodation for two sets of equipment and professionals. Hence, it requires adaptation to the available facility, which could be ensured during mock-drills prior to the procedures. In this context, the use of a single anesthetic machine with two breathing circuits attached to the common gas outlet with a Y-connection may be useful [37, 38]. In fact, arrangements of MRI-compatible anesthesia machines in duplication could be of logistic issue. Nevertheless, all other gadgets used in duplication should be MRI safe [38]. Heart rate, blood pressure (noninvasive and/invasive), oxygen saturation (SpO_2), ECG, and end-tidal carbon dioxide ($EtCO_2$) are to be monitored continuously from two monitors.

Many anesthesiologists prefer induction of the twin with hypertension and antihypertensive therapy first. However, anesthesia-induced hypotension may cause a further decrease in blood pressure in the other (hypotensive) twin. Pharmacologic measures utilized to control hypo/hypertensive episodes in these twins might not yield optimal results in the presence of crossover [37]. Hence, physiological control by placing the hypertensive twin higher up than the other twin with a pillow's help has been attempted. It helps to counter gravity-dependent shunting of the blood between the twins.

Transportation of the anesthetized twins to different locations outside the OR is required for preoperative neuroimaging. The twins under GA may have to be transported in a trolley with ventilatory and monitoring support. An optimal communication among supporting staff is desirable to prevent disconnection and kinking of breathing circuits, catheters, and lines. The twins could be at increased risk of adverse cardiac and respiratory events. The majority of them are preventable; if they occur, they may adversely affect the outcome [39]. Similarly, positioning during the procedures is problematic as none of the neuroimaging patient tables are specifically made for CPTs with conjoined heads. Combining all neuroimaging procedures as a single procedure may reduce the number of anesthetic attempts and, possibly, anesthesia toxicities, but it would increase the anesthesia time. The twins are prone to hypothermia due to prolonged anesthesia time and the low temperature of MRI and DSA suites. Appropriate precautions such as the use of warm crystalloid infusion and convection warmers and wrapping twins with warm blankets throughout the process may help prevent significant hypothermia.

Simultaneous **mask ventilation** after anesthetic induction of the twins might be difficult due to the paucity of space between them; the angular CPTs may lead to further difficulties in terms of mask ventilation as well as intubation. In the case of difficult mask ventilation, muscle relaxants should be avoided, and a check laryngoscopy (preferably videolaryngoscope) helps assess the situation. Supraglottic airways can be used in older CPTs undergoing neuroimaging under GA [38], whereas very small twins need tracheal intubation.

19.7.2 Anesthesia for the Separation Surgery

It is desirable to have multiple meetings of MDTs apart from few separate meetings among the team members for anesthetics. All proper investigations, including the imaging studies, are to be reviewed thoroughly. The anesthesia team should have a thorough understanding of the sequence of planned surgical steps and perioperative care. Such surgeries should be carried out in a referral medical center where the facilities can carry out such a complex procedure. The major concerns of separation include massive intraoperative hemorrhage, cerebral edema, venous infarcts, swelling of the skin flaps, and dehiscence of the repairs with CSF leak, meningitis, and exposed brain [25].

The basic **preanesthetic preparation** is more or less similar to neuroimaging procedures. One twin may be physiologically dominant, and the calculated anesthetic drug doses can be unpredictable. The planning should be done individually for each twin; drugs should be prepared in duplications in a dose appropriate for each twin. The body weight may be calculated by dividing the twins' total weight into two parts, assuming both are of similar weight. Each twin can handle a particular drug dose differently from the other. It would depend on the crossover of the circulation and hepatorenal function; the changes would be unpredictable after separation [40].

The laboratory investigations should include a complete hemogram, fasting blood sugar, hepatic and renal function tests, coagulation studies, blood grouping, and cross-matching for each twin individually. A chest X-ray and echocardiography for each twin should also be done.

The OR should be prepared beforehand, and all necessary drugs and equipment should be arranged meticulously with drugs labeled with color coding for each twin. The OR personnel should be counseled about their specific roles for this procedure. Two tables may be joined to create a single operating table considering the possible positioning of the CPTs (Fig. 19.2). A prior **simulation or rehearsal (mock-drill)** helps organize the OR and procedural planning [41], such as identifying space for the teams involved, anesthesia equipment, ventilator, and monitor in duplication. It also decides optimal patient positioning. Simulations on common intraoperative scenarios and emergencies are also suggested [42].

Both IV and inhalational induction with sevoflurane are considered suitable for the CPTs depending on the presence of an IV access [20]. Anesthesia for both twins may be induced simultaneously; however, one may take a longer time for the induction. Opioids such as fentanyl or remifentanil and muscle relaxants are to be given at a dose appropriate for each twin. Both nasotracheal and orotracheal intubation can be done [38]. While nasotracheal helps during continued postoperative mechanical ventilation after the separation, however, the possible increase in infection and meningitis incidence prevents its practice in neurosurgical patients. Hence, the orotracheal method is preferred as the endotracheal tube (ETT) can be secured relatively away from the surgical site. There may be difficulties in securing the airway when intubation is attempted in both twins simultaneously [35], or when there is restricted neck movement owing to conjoined and fixed heads. Twins with prolonged conjoining may develop cervical lordosis that inhibits mandibular growth causing further difficulties [43, 44]. The difficult airway cart should be kept ready in each case of conjoined twins. Direct laryngoscopy, as well as videolaryngoscope, are preferred during intubation. While appropriately sized cuffed PVC tubes may be used for anesthesia during neuroimaging procedures, it is preferable to use reinforced ETTs for separation surgery. There could be considerable manipulation of the head and neck during the intraoperative period leading to kinking of the ETT. In some of the CPTs, it may be necessary to lift and rotate one twin during laryngoscopy and intubation in the other and vice versa.

Vascular access may be difficult in younger CPTs; it may be complicated by a prolonged pre-separation period with multiple punctures of peripheral veins for different procedures. Central venous access for each twin with a triple-lumen catheter may be planned under ultrasound guidance. The access is needed for fluid management,
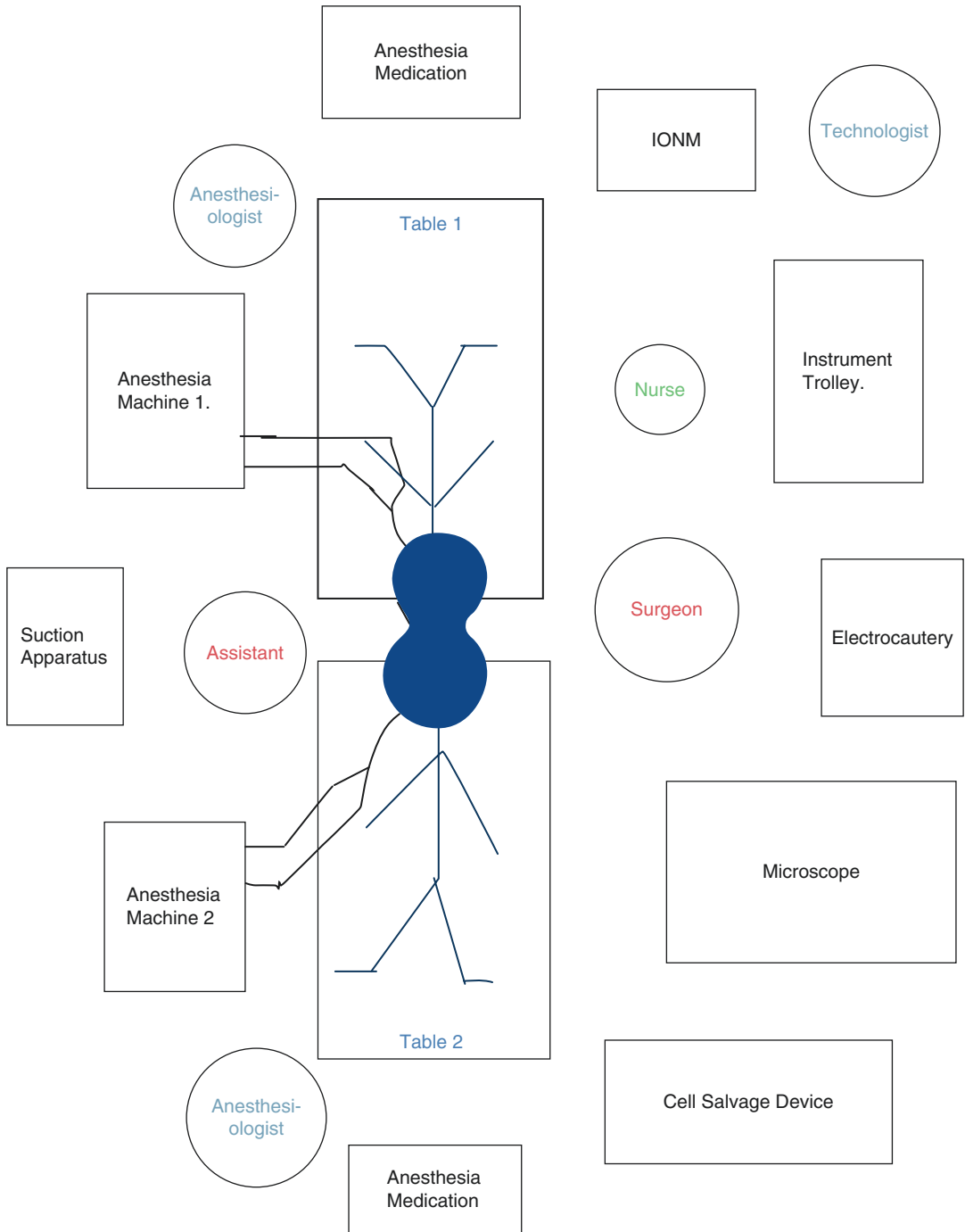
Fig. 19.2 Proposed operating room (OR) arrangements during craniopagus separation surgery

drug infusions, and measuring trend of central venous pressure (CVP) and as part of treatment in case of intraoperative venous air embolism (VAE) to aspirate air [17]. The site of preference for central venous catheter placement may vary among the clinicians. The authors would prefer

femoral or subclavian veins; however, an internal jugular vein can also be utilized after separation surgery whenever indicated. The goal is not to allow any of the central lines to come into the surgical field. The peripheral venous catheters can be placed in the limbs, preferably in the upper limbs. Arterial catheters should be placed in both the twins for continuous monitoring of blood pressure and arterial blood gas (ABG) analysis.

Monitoring should include all the routine parameters such as ECG, SpO_2, $EtCO_2$, arterial pressure, CVP, airway gases, blood loss, blood and fluid transfusion, and urine output [38]. Temperature can be recorded both from the core (esophageal and rectal temperature) and peripheral sites. Apart from these, neuromuscular monitoring (NMT), neurophysiologic monitoring (somatosensory and motor evoked potentials), and regional cerebral oxygenation (NIRS) may be measured.

Maintenance of anesthesia is done with inhalational agents (isoflurane/sevoflurane/desflurane) in oxygen and air. The use of nitrous oxide may be avoided in view of its effects on intracranial pressure and the possible occurrence of VAE [38]. Opioid supplementation at regular increments or as infusions can be given also help to achieve immediate postoperative analgesia. Total intravenous anesthesia (TIVA) with or without muscle relaxant may be utilized in case of intraoperative tight brain and neurophysiologic monitoring, respectively. However, continuous infusion of propofol for a prolonged period should be avoided because of its potential adverse effects.

The positioning of the CPTs requires careful consideration and appropriate planning based on the available gadgets. The upper ends of two OR tables are joined together to the conjoined heads on a headpin or a specially prepared headrest [17]. Precautions must be taken to keep the twins surgically accessible while ensuring that the lines and circuits are not dislodged, twins are accessible to anesthesia teams, and the pressure points are well-padded. There is no particular recommendation available regarding the twins' surgical position for the initiation and completion of separation surgery. It may be started with the twins in

prone; final separation should be carried out with the twins in a supine position. It could be beneficial for last-minute turning (supine) of the twins after separation. On the contrary, if separated in prone, the twins may have to be transferred to separate tables in the prone position with fully exposed brains [38].

Management of fluid and blood loss needs utmost attention. Crystalloids and colloids are administered in a titrated manner to both children. During this prolonged surgical period, 1–2% dextrose solution may be preferred in younger twins to avoid intraoperative hypoglycemia. The insensible losses from the surgical site are unmeasurable. The near-ideal replacement of fluid and blood loss is difficult to achieve but should be guided by arterial waveform analysis, CVP trends, urine output, electrolytes, point-of-care determinations of ABGs, and thromboelastography. Blood loss estimation is complicated during the separation surgery. Even a moderate loss would have significant clinical problems, particularly in young twins. The bleeding from the common surgical would is different for each twin and is usually estimated from blood collected from the suction chamber, amount and weight of gauze pieces used, and hemoglobin and hematocrit values measured at regular intervals. Half of the estimated volume should be transfused to each twin [45]. Close monitoring of the hematological and coagulation status of each patient needs to be ensured throughout. The blood loss also could be managed with staged separation and preserving the cleavage plane between the separated brains with silicone sheets.

Complications such as hemodynamic perturbations, massive blood loss, brain edema, and hypothermia may occur during the perioperative period. **Hemodynamic disturbances** in the twins may occur due to blood loss and massive fluid shifts, and persistent differences in blood pressures throughout the surgery due to cross-circulation and a unidirectional vascular flow or unopposed shunting [20, 36, 38]. Therefore, one twin may remain hypotensive, whereas the other may present with hypertension; supplementation of fluids and inotropes in the hypotensive child

can worsen hypertension in the other twin without any obvious benefits [36]. The fluid volume requirements could be more significant in one twin, whereas the urine output is significantly more in the other [20], suggestive of a unidirectional intracranial flow pattern. The twin may become hypervolemic enough to develop cardiac failure. In contrast, the other twin might be oliguric/anuric, leading to renal failure and requiring continuous renal replacement therapy (CRRT) and possibly renal transplantation [16]. This complication of unopposed shunting may occur at any time during the multi-staged separation during intraoperative or postoperative period requiring vasoactive managements and even withdrawal of blood in the twin with impending cardiac failure; it generally resolves after final separation [38]. **Massive blood loss** should be anticipated during the stage of venous separation. The loss has to be replaced with blood and products. It is unclear which twin is more affected by the blood loss. It is also not uncommon for one twin to present with more significant clinical symptoms of hemorrhage or volume overload following transfusion, despite the blood loss shared by both [40]. The loss can be extensive and may lead to hypovolemic shock, bradycardia, and even cardiac arrest requiring resuscitation [36]. **VAE** is a strong possibility at every stage of the separation surgery. **Hypothermia** may be attributed to the extensive surgical wound as well as prolonged surgical time, which causes heat loss by evaporation, radiation, and convection. All available measures should be utilized to prevent it and to maintain normothermia as intraoperative hypothermia affects the surgical outcome [43, 45]. Post-separation, the twins may develop brain edema due to the formation of venous infarct and deranged cerebral autoregulation after surgical manipulations. **Infection** at the surgical sites, meningitis, CSF leak, and ventriculitis may occur, requiring constant vigilance and preventive measures. The high potential for infection in such cases is due to the presence of an indwelling catheter, shunts, tissue expanders, and drains. Hence, antibiotics are used during the different periprocedural periods.

After final separation, one twin has to be transported to the adjacent OR with the anesthesia team assigned. The ventilator, monitor, and infusion of drugs accompany the twin to further reconstruct the dura, calvarium, and scalp with artificial dura, absorbable plates, and split skin grafts. Both the twins at this stage develop physiological changes manifested mainly with hemodynamic perturbations.

19.8 Postoperative Intensive Care

After separation, the twins develop a lot of physiological changes, develop hypotension, and often need inotropes to maintain blood pressure. One (non-dominant) twin may develop seizures due to venous infarcts' formation and require prophylactic antiepileptic drugs (AEDs). Continued ventilation with optimization of hemostasis, hypoxia, hypercarbia, hypotension, hypothermia, hypotension, and electrolyte abnormalities is mandatory. Seizures may also occur in any of the twins due to postoperative complications such as meningitis, hydrocephalus, and metabolic derangements [17]. Fluid shift and blood loss during the perioperative period need intensive fluid management and blood component therapy. Extensive venous infarcts may cause cerebral edema and brain bulge. Many exposed areas are prone to develop severe infection and septicemia; aggressive antimicrobial treatment helps to attain a better outcome.

The twins may need multiple wound dressing episodes, skin grafting in case of wound dehiscence, lumbar drainage for CSF leaks, and VP shunt insertion for hydrocephalus under sedation or GA as and when indicated. The individual status of the twins needs consideration for anesthetics. The surgical outcome is significantly better with the separation of partial CPTs, whereas with total CPTs, both mortality and morbidity are more [9]. One or both of the twins may have to undergo rehabilitation for neurologic and cognitive disabilities due to developmental reasons and surgical complications. The twins may have a prolonged hospital course even after the final separation surgery.

19.9 Legal and Ethical Concerns

Legal and ethical questions are always raised concerning the separation of conjoined twins irrespective of phenotypes. The issues encountered with CPTs are based on principles of autonomy (respect decision of patient), informed consent in young children, principles of beneficence, and nonmaleficence (act in the benefits of the patient and not harm the patient), and justice [46]. The ethics are made even more complex when one twin is dependent on the other for survival as the latter twin may have to be sacrificed for the former's survival. In this context, a review by the hospital ethics committee opinion may help. It has also been suggested to have a prior legal opinion before separation [42].

19.10 Conclusion

The separation surgery for CPTs is one of the most complex procedures undertaken in neurosurgery. Of late, successful separation has become more common with the advances in neuroimaging, neuroanesthesia, and neurosurgical techniques. Experience and expertise with this surgical procedure are limited. Hence, the MDT may prefer having inter-institutional or international collaborations, if required, to close the learning gap. The success of surgery depends on early separation (less than 1 year), the shared vasculature nature, and multi-staged surgery. Anesthetic management requires meticulous planning and clear communication among the team members with particular attention to the difficult airway, adequate intravascular access, careful positioning, appropriate intraoperative fluid and blood management, maintenance of normothermia, and effective management of the perioperative complications.

Conflict of Interest The authors were part of 'AIIMS Craniopagus Team' responsible for the first successful craniopagus separation surgery in India.

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