Conjoined Twins

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Conjoined twins are among the rarest developmental anomalies with incidence estimates ranging from 1 in 50,000 to 1 in 200,000 births. Most of the sets identified prenatally die either during pregnancy (25 %) or within 24 h of birth (50 %). While it is claimed that the incidence among stillborns is equal between boys and girls, girls predominate 3:1 among the live-born sets.

The twins are categorized by the location of the joining (thoraco-, omphalo-, cranio-) combined with the Greek term pagus ("that which is fixed") (Table 14.1). The attachment of an incompletely developed twin to the body of a fully developed twin is extremely rare and has been alternatively labeled heteropagus or parasitic or asymmetric conjoining. Twins joined at the chest and abdomen represent almost three quarters of the reported sets. There are two theories of the etiology of conjoined twins, the fission and the fusion theories. Historically, it has been assumed that conjoined twins resulted from incomplete separation of a monozygotic twin embryo between the 13th and 15th day after fertilization. An alternative theory (fusion) is that two embryos fuse after initially being separate. There is no association with previous conjoining, maternal age, or parity. While conjoined twins fit into the common classification categories with many similarities among them, it is best to consider each set a unique pair of individuals requiring careful anatomic evaluation and possessing separate moral and ethical identities.

Diagnosis

The diagnosis can be established by ultrasound as early as 12 weeks gestation by identifying constant relative positions of the fetuses, a single placenta with no separating membrane, or a single umbilical cord with more than three vessels. Follow-up scanning at 20 weeks provides reliable visceral detail and should include echocardiography. While fetal echocardiography is quite accurate, it tends to underestimate the degree of cardiac malformation and cannot reliably exclude myocardial fusion. Three-dimensional echocardiography provides greater detail. Since the chance of survival and separability are largely dependent on the extent of the cardiac anomalies, it is essential to obtain accurate cardiac imaging. In some instances, the imaging windows available are better prenatally than postnatally. If the pregnancy continues into the third trimester, a fetal MRI should be performed as it provides excellent soft tissue resolution and a larger field of view.

The frequency of associated anomalies and the site of fusion dictate the need for postnatal diagnostic studies. All sets should have plain radiographs of the chest and abdomen to identify associated anomalies such as diaphragmatic hernia, vertebral malformations, and cardiac lesions. Echocardiography and cranial ultrasound should also be performed in all cases. Additional studies are dictated by the location of the conjoining; the timing of these studies depends on their clinical condition. If the twins are stable, a few limited studies are obtained with the more complex imaging awaiting a period of transition and growth. If the twins' clinical condition is tenuous or discordant, suggesting that urgent separation might need to be considered, diagnostic studies should proceed with thoughtful multidisciplinary input.

In thoraco-omphalopagus twins, the bowel gas pattern might appear to be separate on plain radiographs but more specific studies such as GI contrast or CT with contrast should be obtained. Ultrasound of the liver, hepatic veins, and abdominal viscera can provide valuable information about separability and can be accomplished with portable equipment if the infants are unstable. CT (Fig. 14.1) and MRI of the head, chest, and abdomen are obtained under general anesthesia and should be planned with sequences and timing of contrast injections optimized to provide as much dynamic information as possible while limiting the duration of the studies.

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Table 14.1 Types of conjoined twins

Category	Fusion	Percentage (%)
Thoracopagus	Chest/abdomen	20–40
Omphalopagus	Abdomen	18–33
Pygopagus	Sacrum/buttocks	18–28
Ischiopagus	Pelvis	6–11
Craniopagus	Cranium	2
Parapagus	Ventrolateral	New term

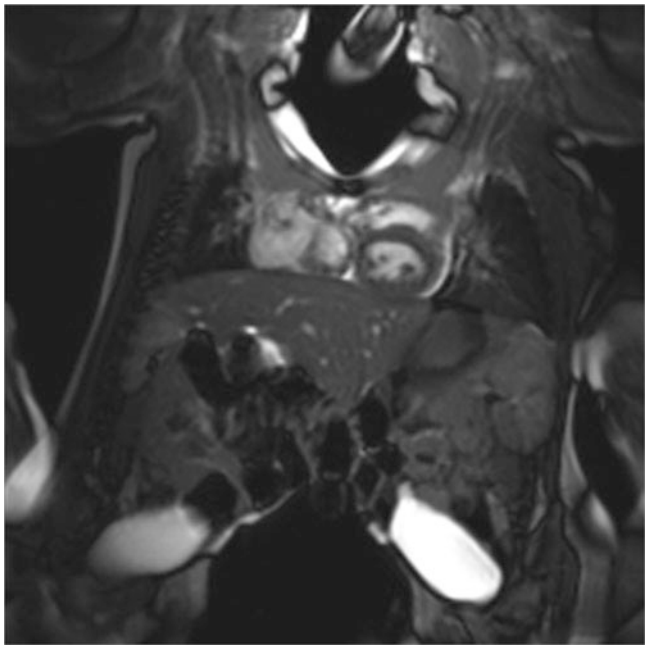

Fig. 14.1 Sagittal CT of thoraco-omphalopagus conjoined twins

Modern imaging software that allows three-dimensional reconstruction provides amazing detail and visualization of the proposed separation. The cross-sectional images of the viscera allow further planning both for the separation and the reconstruction and calculations of the anticipated defect in the body wall resulting from the separation. In addition, the imaging allows flow estimations about twin–twin shunting at the cardiac and visceral levels. In cases of possible separation with structural cardiac anomalies, cardiac catheterization should follow the same indications as those of a singleton infant and might also reveal pulmonary hypertension or substantiate cross-circulation. Imaging of the biliary trees should be accurate from the MRI but in some cases should be supplemented by nuclear imaging. Laboratory studies should include basic metabolic studies as well as oxygen saturations, arterial blood gases, and electrocardiograms. Twins with even small myocardial connections will usually have synchronous heart rates.

Twins joined at the pelvic (ischiopagus) and sacral (pygopagus) regions often have complex vertebral, orthopedic, and genitourinary abnormalities; CT and MRI are required to identify the bony and visceral anomalies as well as possible fusion of the spinal cord. Multiplanar MRI is helpful in cataloging the pelvic viscera (uterus, bladders, fallopian tubes). Cross-circulation is sometimes significant in these twins, and, as in thoraco-omphalopagus, the contrast injection is done in one twin only and the scanning timed to obtain arterial and venous information. Delayed images are helpful in determining renal function. Complementary information is obtained by performing contrast studies of the genitourinary and gastrointestinal tracts. Cloacal anomalies and single rectum are common and accurate definition of the anatomy that is critical to planning the surgical separation.

Twins joined at the head are classified as craniopagus or cephalopagus. Cephalopagus twins are usually also fused at the chest and have generally been thought to be nonviable, although a set of girls who are now 20 years old have chronicled their lives in short video clips on the Internet. Craniopagus twins account for approximately 2 % of all conjoining though they are more heavily reported in the lay press. Some cases have separate duras but most have significant connections of cerebral cortex and share at least a portion of the sagittal sinus.

Twins joined side to side (parapagus) can have extensive connections with complex pelvic anatomy. They usually have a shared leg, a single symphysis pubis, and one or two sacra. Unions that include the chest have complicated cardiac anomalies similar to the thoraco-omphalopagus twins and need extensive cardiac evaluation. The blood supply to the shared pelvis and lower extremity can be outlined with CT and MRI and rarely requires angiography.

Treatment

Multidisciplinary planning should begin prior to delivery. Counseling regarding viability and the possibility of separation should be accomplished with input from specialists with experience in the appropriate areas. Hospitalization is frequently indicated late in the pregnancy with a planned cesarean section although obstetrical complications are frequent and often necessitate an urgent delivery. Stabilization in the neonatal ICU should include standard neonatal care with multidisciplinary evaluation and attention to privacy. While in the NICU, public and media exposure is usually well controlled, but excessive or unnecessary examination by medical and hospital personnel is a risk and must be controlled. The optimal situation is stabilization of the infants such that they could be discharged home to return for further evaluation.

The optimal time for elective separation is undetermined but has been suggested at between 4 months and 2 years. We have noted that even with separation at 4 months of age, there are already significant musculoskeletal changes that require remodeling or physical therapy. On the other hand, larger size, more time for tissue expansion, and more “durability” of vessels and tissue are advantages of a delayed separation.

Emergency separation needs to be considered when one twin is unstable or if both physiologically deteriorate due to their connection. If one twin dies, the other will succumb within 4–6 h from disseminated intravascular coagulation. In the absence of complete preparation for separation, emergency separation should only be considered when the death of one twin is imminent and the goal is salvage of the healthier twin. The specific management of each set of twins will depend on their physiologic status and specific constellation of conjoining and associated anomalies.

Thoraco-omphalopagus is the most common type of conjoining. Twins joined at the pelvis will require involvement of orthopedic, urologic, and neurosurgical colleagues, and the operative plan will obviously be determined by the nature of their connections. The anesthetic and team coordination management of the twins begins with their diagnostic studies, as most will require general anesthesia. Experience with the twins' reaction to specific drugs, the degree of cross-circulation, and their recovery patterns are helpful in planning the separation procedure. We combine studies whenever possible, having obtained CT, MRI, and cardiac catheterization under a single anesthetic. Even with limiting of data acquisition, this can take 6–8 h.

Tissue expansion is usually required to obtain adequate skin coverage of the large body wall defect created by the separation. We have used tissue expanders in twins as young as 2 months of age and have placed the expanders either on the connecting bridge itself or parallel to it. The expanders can be filled fairly rapidly, with weekly injections, usually either with topical anesthetic or a brief general anesthetic. Care must be taken to avoid excessive pressure, as the expanders are placed on both lateral surfaces of the twins. The timing of the insertion and expected expansion needs to be coordinated with the separation date.

Younger twins or those requiring a preoperative bowel preparation should be admitted the day prior to separation, though we have admitted older twins with separate gastrointestinal tracts on the day of surgery. Some centers insert all monitoring lines under a separate anesthetic on the day prior to separation. The induction; insertion of central and peripheral venous, arterial, and urinary catheters; temperature probes; and positioning with careful padding of pressure points usually requires at least 2 h (Fig. 14.2).

The initial incision is centered at the midpoint of the connecting skin bridge, and the tissue expanders on the "up" side of the twins are removed. The abdomen is easily entered at the umbilicus, which frequently has a small omphalocele membrane that is usually epithelialized by the date of separation. The abdominal portion of the body wall connection is opened, and the peritoneal cavity of each twin is entered and the viscera inspected. The fused sternum on the "up" side is then carefully entered, which can be done without entering the common pericardium or separate pleural spaces. The pleurae of each twin can be bluntly dissected free of the sternal edges

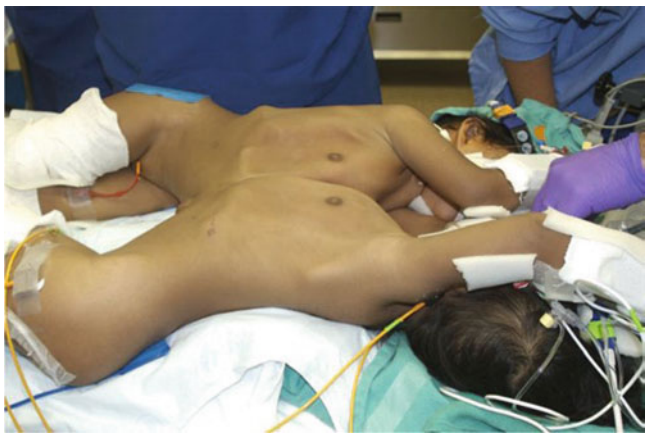

Fig. 14.2 Twins positioned with monitoring in place

to expose the pericardium, which is entered again at the midpoint of the connection. The degree of any cardiac connection can now be assessed, and preparation for potential cardiac bypass or pacing begun.

Opening of the abdomen and chest has thus allowed complete assessment of the visceral connections. Our strategy has then been to complete the separation of the abdominal viscera and the abdominal body wall of the "down" side prior to any cardiac procedures – although cannulation for bypass is possible in the lateral position, this would allow for expeditious separation of the "down" sternum should either twin deteriorate.

The majority of thoraco-omphalopagus twins have a fused liver, usually with separate biliary and vascular supply but with significant intraparenchymal vascular connections. On occasion, the livers are completely separate though touching. Bowel connections are separated with stapling devices and reconstruction deferred until separation is complete. Splitting of the diaphragm allows exposure to the contralateral surface of the liver connection, which can be encircled with umbilical tape or a Penrose drain. We have had good results dividing the liver with a variety of devices including the harmonic scalpel, hydro-dissector, bipolar and monopolar coagulators, and direct suture ligation. At the completion of the separation, the raw surface can be sealed with the argon beam coagulator with little risk of a bile leak.

Attention is then turned to the cardiac separation. The cardiovascular strategy depends on the degree of connection, structural integrity of each heart, and the physiologic status of each twin. Sometimes the hearts are completely separate within a common pericardium, in which case the posterior body wall is separated and tissue expanders on the "down" side removed. Myocardial connections can be small or large and are frequently atrial. A significant ventricular connection is usually identified preoperatively and precludes separation. The myocardial connections are test clamped to identify the physiologic consequences of their separation. While preparations for pacing or bypass are made ready prior to the division of the

connection, they have not been necessary. Once the myocardial connection is severed and closed, the posterior body wall is completed, and the twins rotated to the supine position. In the absence of structural cardiac anomalies, one twin is moved to a separate operating room with his or her entire team so that reconstruction of the body wall can proceed simultaneously.

Structural cardiac malformations can be repaired or deferred depending on the magnitude of the corrective surgery, the need for cardiopulmonary bypass, and the physiologic status of the twin. Our most recent separation outlines the cardiac options and strategies. The twins shared a large atrial connection approximately 6 cm in cranio-caudal dimension. One twin had double-outlet right ventricle, while the other had left main pulmonary artery stenosis with significant pulmonary hypertension. During test clamping of the atrial connection, both twins remained stable, and separation was uneventful. The twin with pulmonary artery stenosis underwent patch angioplasty and definitive body wall reconstruction and closure. The twin with double-outlet right ventricle underwent definitive closure of the abdomen with skin closure of the chest. She was stabilized for 48 h and then underwent cardiac repair with reconstruction and closure of the chest. While bypass was available, the ability to avoid its use immediately after the liver separation appears to have contributed to the uneventful recovery of both twins.

Following separation, the abdominal and thoracic viscera are inspected and repaired (Fig. 14.3). Hemostasis is ensured, and the abdomen is closed with minimal tension, which usually means placing a soft tissue patch in the upper fascial closure. Prosthetic material is used to provide a stable bridge between the sternal halves. We prefer sheets of material as opposed to struts and have had good experience with lactic acid polyglycolic acid copolymer products. The skin flaps are then generously mobilized and closed over drains placed in the mediastinal and subcutaneous spaces. If closure of both twins is completed at the same time, their return to the critical care area should be staggered.

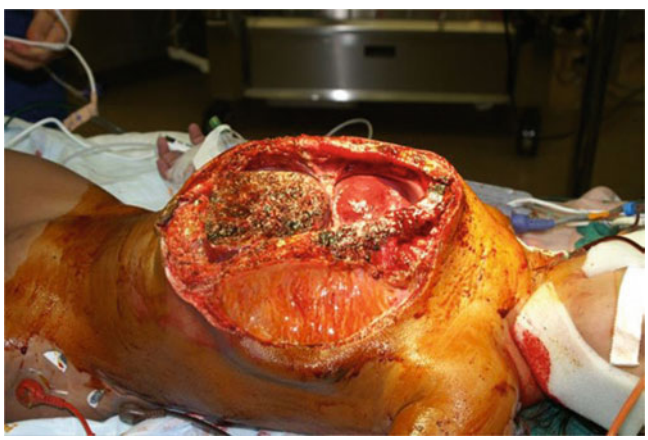

Fig. 14.3 One thoraco-omphalopagus twin after separation. The sternum is still split, revealing the heart and the cut edge of the divided liver bridge that is visible below the diaphragm

Postoperative Care

A written plan for the postoperative care with individuals from each discipline identified and specific responsibilities spelled out in detail minimizes confusion postoperatively. Preprinted order sets that have been reviewed and agreed upon by all relevant disciplines are also helpful. Initial care is directed at optimization of respiratory and hemodynamic status. Careful fluid and ventilator management predominates in the first days, but careful monitoring of liver function, fluid drainage, and the viability of skin flaps is also important. Early revision of any problems with the chest wall stabilization and skin flaps facilitates weaning from mechanical ventilation. Nutrition is critical, and a period of tube feeding should be anticipated. As recovery progresses, a physical therapist should address the musculoskeletal issues imposed by the conjoining and the separation. Hospitalizations of 2–4 weeks should be anticipated for relatively uncomplicated recoveries and longer if any complications intervene. Long-term care is directed at any underlying structural anomalies that required correction and the body wall reconstruction.

Planning Process

Almost every series or case report about conjoined twins stresses the need for careful and intense planning for a successful outcome. A strategy that we have employed is regularly scheduled (weekly or every other week) meetings including a representative from every involved medical and surgical discipline, hospital operational departments, and nursing and hospital leadership (Table 14.2). A flow diagram of each step of the process with a responsible individual identified is helpful. This working group anticipates all outcome scenarios and develops strategies for each with information gathered along the way from diagnostic studies and the twins' responses to anesthesia and the environment. Mock-ups of the operating room with a specified location for each individual and piece of equipment are done on paper and then tested in person with walk-throughs in the designated operating room (Fig. 14.4). All equipment should be

Table 14.2 Planning team—thoraco-omphalopagus

Anesthesia	Pediatric surgery
Cardiology	Cardiothoracic surgery
Radiology	Plastic surgery
Laboratory medicine	Critical care
Operating room director	Operating room nursing
Critical care nursing	Social services
Physical therapy	Admitting/registration
Medical records	Hospital administration
Security	Public relations/media

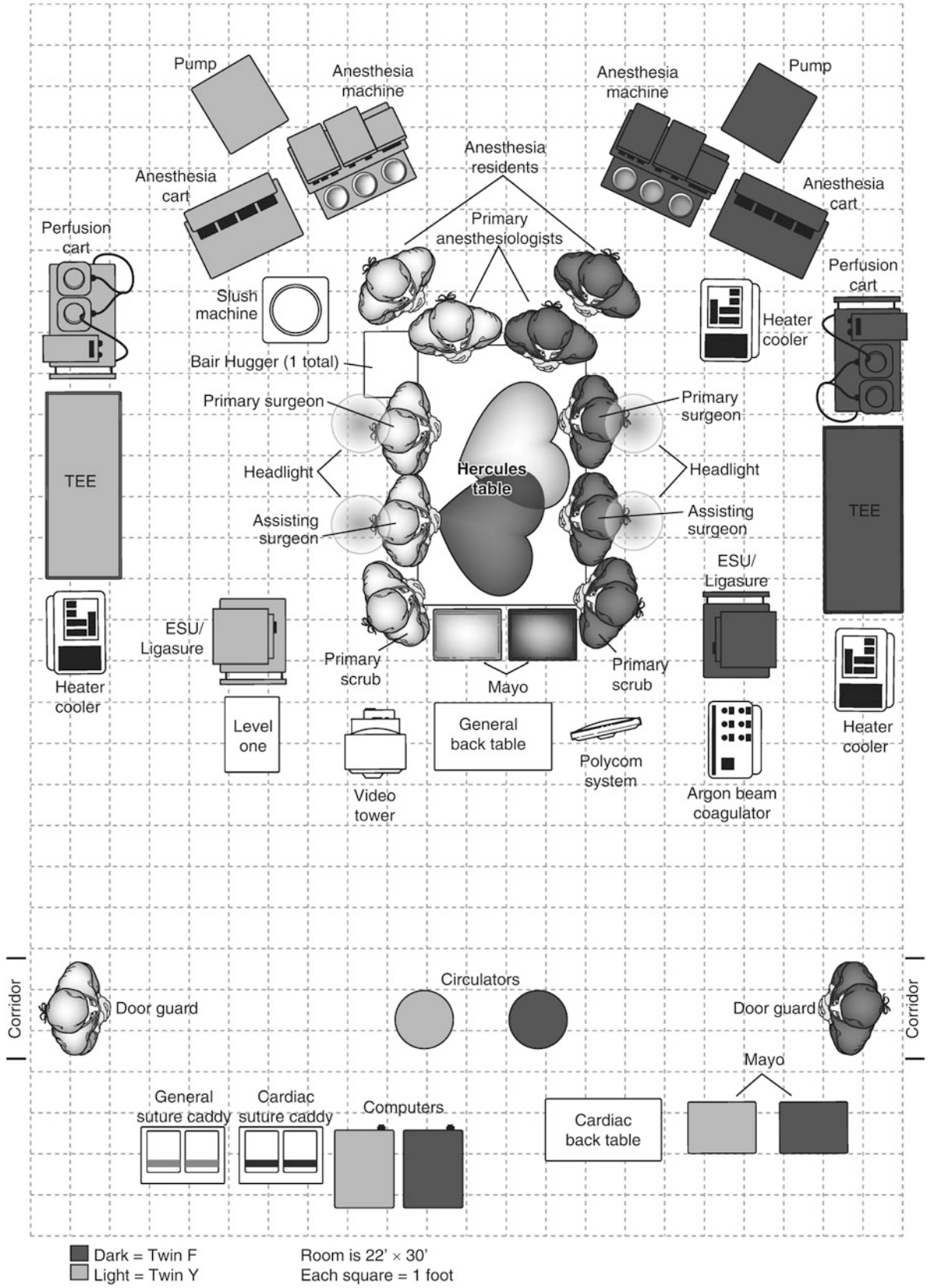

Fig. 14.4 Diagram of OR setup prior to separation

turned on to test the electrical capacity of the room, which frequently needs supplementation with temporary power (up to 100 Å or more).

On the day of separation, attention to security and crowd control is facilitated by having a room general who has no clinical responsibilities but has the authority to remove anyone from the room. Accommodations for legitimate educational and clinical interest can be accomplished with a video feed to designated secure viewing areas. For particularly lengthy procedures, planning should include rest periods for staff and a designated individual to relay progress reports to the family.

Editor's Comment

One still occasionally sees the obsolete and insensitive term "Siamese twins" in the lay press and in the medical literature (even from presidential candidates, who should know better). The proper term, for some time now, is "conjoined twins." Computer-enhanced three-dimensional imaging has allowed for much better preoperative planning for these often extremely difficult and tedious operations, but the assembling of a team of experts and meticulous planning of each minute detail, including contingency plans for every conceivable snag, is still the most important aspect of the care of these unique individuals. Given the intense societal interest in these cases, it is also advisable to involve a team of bioethicists, hospital administrators, and public relation experts from the very beginning so that medical personnel can concentrate on providing excellent care without being distracted.

Suggested Reading

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