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Perioperative Management of Conjoined Twins

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THE BIRTH OF CONJOINED twins remains an extremely rare event that continues to fascinate both physicians and lay public. During the 1960s and early 1970s, the last reviews on perioperative management of conjoined twins stressed preoperative management discussions, dress rehearsals, and invasive intraoperative monitoring for blood and fluid losses. Although these earlier recommendations remain valid today, recent advances in diagnosis and management of conjoined twins have been introduced in the 1980s. Improvements in obstetrical ultrasound, fetal echocardiography, contrast tomography, magnetic resonance imaging, nuclear volume and flow studies, pulse oximetry, capnography, and extracorporeal membrane oxygenation now permit precise neonatal diagnosis, better evaluation of joining, and more extensive separation operations. Conjoined twins may now be diagnosed prenatally, have their degree of joining delineated, and have a safe, elective delivery, usually by Cesarean section. Planning for eventual surgical separation may also begin before delivery of the twins.

Some 600 sets of conjoined twins have now been reported worldwide.[1] Historians cite the earliest reference to conjoined twins as Janus, the mythical Roman god of beginnings, depicted as two bearded heads back to back.[1] In 1100 A.D., the earliest known written record of conjoined twins was made in Latin of the Biddendon girls, born in England, and joined ventrally from shoulders to hips with only one pair of arms and legs.[2] With the Renaissance, several detailed medical reports and illustrations of conjoined twins were published.[3] In his 16th century text, *Of Monsters and Prodigies*, the French surgeon Ambroise Paré accurately described conjoined twins and proposed 11 etiologic factors now known to be invalid.[3] Today, conjoined twins are said to result from an aberrant twinning process with incomplete fission of the zygote's primitive streak during the second week of gestation or 20 days post-ovulation.[4] This hypothesis has been supported by observations of identical sex in almost every reported case of conjoined twins.[5] There is little scientific support for suggestions that conjoined twins may result from the fusion of two separate fertilized ova.[4]

Conjoined twins are classified by their most prominent site of connection. Such sites may include thorax (thoracopagus), upper abdomen (xiphopagus), lower abdomen (omphalopagus), pelvis (ischiopagus), sacrum (pygopagus), or skull (craniopagus). Compound classifications may be used to describe all possibilities for joining. For example, omphalo-ischiopagus tripus twins are fused at the lower abdomen and pelvis and have two arms and one common leg. The major types of conjoined twins, their frequency of occurrence, sites of musculoskeletal attachments, and possible organs shared are described in table 1.

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TABLE 1. Classification of Conjoined Twins (Adapted from Filler and Crocker[3])

Type of Twin	Frequency	Site of Joining	Possible Organs Shared: Heart and Great Vessels	Biliary Tract	Upper GI Tract	Lower GI Tract	GU Tract	CNS
Thoracopagus	40%	Sternum, diaphragm, upper abdominal wall	Yes	Yes	Yes	No	No	No
Xiphopagus or Omphalopagus	34%	Xiphoid, anterior abdominal wall, umbilicus	No	Yes	Yes	Yes	Yes	No
Pygopagus	18%	Buttocks, coccyx, sacrum	No	No	No	Yes	Yes	Yes
Ischiopagus	6%	Lower abdomen, pelvis, sacrum, lower extremity	No	Poss.	Poss.	Yes	Yes	Poss.
Craniopagus	2%	Cranial vault, dural venous sinuses	No	No	No	No	No	Yes

Konig‡ performed the first successful surgical separation of conjoined twins in 1689. He separated omphalopagus twins joined at the umbilicus by slowly tightening an encircling tourniquet, constricting and necrosing the connecting bridge.‡ This technique is still in use today, particularly in the staged separation of craniopagus twins.[6]‡ Now, over 60 reports documenting successful separation of various types of conjoined twins have been published, with most surgical separations being performed in the past 25 yr.[3] Craniopagus[7] and pygopagus[8] twins were first separated in 1952 and 1953, respectively. Ischiopagus twins were first separated successfully in 1966.[9] Thoracopagus twins with joined hearts were first separated in 1979, with the single survivor being a twin with a normal heart who shared only an interatrial vascular bridge.[10] There have been no long-term survivors of separation procedures in thoracopagus conjoined twins with shared common myocardium, also referred to as "cardiopagus" twins.[11]

By far, the world's best known and among the longest lived conjoined twins were never separated.[12] Chang and Eng Bunker were born in Siam (now Thailand) in 1811, were exhibited worldwide by circus ringleader P. T. Barnum in the mid-1800s, married two sisters at age 31, fathered 21 children, and died at age 63.[12] The Bunkers' birthplace, Barnum's showmanship, and top circus billing gave the world the lay term, "Siamese" twins, still commonly used today to described conjoined twins.

Epidemiology

Clear relationships among maternal factors, environmental exposures, and conjoined twinning are lacking.[13-15] There appears to be, however, an increased incidence of stillbirths in mothers of conjoined twins.[5] Chronic malnutrition and intrauterine hypoxia have been suggested as possible etiologic factors in conjoined twinning, and could explain higher incidences of conjoined twinning in underdeveloped countries.[15] Family history, paternal age, maternal age, and multiparity do not appear to influence the occurrence of conjoined twins.[3]

The exact incidence of conjoined twins is not known. Estimated incidences in the United States range from 1 in 50,000 births to 1 in 200,000 births.[3,9] An increased incidence of conjoined twins ranging from 1 in 14,000 to 1 in 25,000 births has been observed in southeast Asia, especially India, Pakistan, and Thailand, and in Africa, especially East Africa, Nigeria, and South Africa.[13,14] In the United States, from 40% to 68% of conjoined twins are stillborn, and nearly 35% of the live births do not survive the first 24 h of life.[15] As in separate twins, the high perinatal mortality may be partially attributed to prematurity and low birth weight.[16] Shared organs are often anomalous as well, and also contribute to high perinatal mortality among conjoined twins.[15] Nearly 70% of conjoined twins are females.[17]

Prenatal Evaluation

Prior to the development of radiography, the prenatal diagnosis of conjoined twins was impossible.[17] Maternal polyhydramnios, twin gestation, unusual presentation, and labor dystocia with an adequate maternal pelvis suggested conjoined twins.[17,18] Xiphopagus, omphalopagus, pygopagus, and craniopagus twins could often be delivered vaginally because of the pliability of the connecting bridges.[17,18] Thoracopagus and ischiopagus twins had bony unions that caused severe dystocia and necessitated either abdominal delivery by Cesarean

‡ Konig G: Sibi invicem adnati feliciter separati, Ephemerid. Natur Curios Dec II, Ann VIII, OBS 145, 1689.

TABLE 2. Recommended Diagnostic Studies during Preoperative Evaluation of Conjoined Twins.

Type of Twin	Diagnostic Studies										
	CXR	ECG	ECHO	CARDIAC CATH	UGI	BE	LFT/SCAN	IVP	ANGIO	CT	MRI
Thoracopagus	+	+	+	+	+	+	+	+	−	+	+
Xiphopagus	+	+	+	+	+	+	+	+	−	−	+
Omphalopagus	+	−	−	−	+	+	+	+	−	−	+
Ischiopagus	−	−	−	−	+	+	+	+	+	−	−
Pygopagus	−	−	−	−	−	+	−	+	−	−	−
Craniopagus	−	−	−	−	−	−	−	−	+	+	+

+ = highly recommended; − = optional; CXR = chest X-ray; ECG = electrocardiogram; ECHO = echocardiogram; CARDIAC CATH = cardiac catheterization; UGI = upper gastrointestinal series; BE = barium enema; LFT/SCAN = liver function tests/liver-spleen scan; IVP = intravenous pyelogram; ANGIO = angiogram; CT = computerized tomogram; MRI = magnetic resonance imaging.

section or fetal dismemberment with vaginal extraction.[17,18]

With the introduction of pelvic radiography in obstetrics, the prenatal suspicion of conjoined twins could often be confirmed, especially in thoracopagus, ischiopagus, and craniopagus twins with bony unions.[18,19] The limited degree of sternal fusion, incomplete ossification, and the extent of rotation permitted by flexible connecting bridges prevented the prenatal radiographic diagnosis of xiphopagus and omphalopagus twins, in most cases.[18-20] Prenatal diagnosis was accomplished only twice (6%) in 36 sets of xiphopagus twins reported by Harper *et al.* in 1980.[18]

In 1950, Gray *et al.*[19] established the first set of radiographic criteria for antepartum diagnosis of conjoined twins. In the 1960s, radiographic contrast studies and amniocentesis were introduced to corroborate pelvic roentgenography in the antepartum diagnosis of potential conjoined twins.[20] Indigo carmine or methylene blue dyes were injected into the amniotic sac at one site and amniocentesis was performed at another site to confirm monoamniotic twins, either identical or conjoined.[20] By the 1970s, the risks of repeated fetomaternal irradiation and amniocentesis were fully recognized, and noninvasive ultrasonographic evaluation replaced radiographic evaluation in the prenatal assessment of conjoined twins.[21] In 1985, Sanders *et al.*[22] correctly determined the extent of cardiac sharing in four sets of thoraco-abdominally conjoined twins studied prenatally by echocardiography. Postmortem findings in this study confirmed the usefulness of fetal echocardiography in the prenatal evaluation of thoracopagus twins with shared hearts and identified only two cardiac abnormalities commonly missed at echocardiography, anomalous pulmonary venous drainage and shared coronary circulation.[22] The authors suggested that conjoined twins could be more easily evaluated by echocardiography before birth than after birth, since more three-dimensional views could be obtained *in utero* with less hindrance by commonly associated omphaloceles.[22]

Clearly, early prenatal diagnosis of many types of conjoined twins is now possible by ultrasound with later echocardiography accurately predicting the extent of cardiac sharing in thoraco-abdominally conjoined twins.[21,22] Early plans may now be made for first or mid-trimester termination of conjoined twin pregnancy in cases with extensive cardiac sharing and poor prognosis.[21,22] Alternatively, early plans may also be made for surgical separation after Cesarean delivery of conjoined twins with little or no cardiac sharing at echocardiography.[21,22]

At delivery, we and others recommend that space and personnel be available to provide immediate neonatal resuscitation, especially if separation is being considered for thoracopagus twins with a shared heart.[17,18] With a limited number of trained personnel to attend each twin simultaneously, the less vigorous twin must be resuscitated first.[17,18] Urgent operations shortly after birth may be indicated in conjoined twins with ruptured omphaloceles, damage to the connecting bridge, perforated viscus, or intestinal obstruction.[3]

Preoperative Evaluation

The preoperative management of conjoined twins should be unhurried to permit growth, thorough diagnostic evaluation of visceral sharing, and creation of a multidisciplinary separation team from groups undertaking such operations for the first time. A variety of diagnostic studies may be necessary to define organ sharing and demonstrate coexisting congenital anomalies depending on the type of conjoined twins being separated (table 2).

The training of a multidisciplinary separation team should include classroom discussion to delineate duplicate duties, define degree of circulatory admixture, diagram extent of organ sharing, calculate blood and fluid requirements, and determine allowable blood losses.[11,16,23] The team should include, in addition to anesthesiologists, circulating and operating room nurses, neonatal or pediatric intensive care nurses, respiratory therapists, pediatricians, and surgeons. Laboratory and blood-banking personnel should be notified of the separation and informed that numerous laboratory tests and large volumes of blood and its components may be necessary.

Simple but accurate diagrams of organ sharing and circulatory admixture based on radiographic and radioisotopic evaluation can be found in recent case reports on separations of thorapagus[10,11,24] and craniopagus[6] twins. To estimate circulatory time and degree of admixture in conjoined twins, radioactive albumin, methylene blue, or indigo carmine dyes can be injected intravenously into one twin and its first appearance monitored in the other.[3] The rate of blood exchange can then be calculated.[25] Radioisotopic blood volume studies are also very helpful in evaluating circulatory volumes and calculating allowable fluid and blood losses at separation.[25] Circulatory admixture is greatest in thoracopagus twins with a shared heart who may demonstrate complete blood exchange every minute.[3,25]

During preoperative discussions, equipment operators should be designated, and placement of personnel and machinery precisely determined to avoid operating room crowding. Necessary medical equipment will include operating tables, warming devices, infusion pumps, anesthesia machines, ventilators, a variety of electronic monitors, and, possibly, heart-lung machines. Some authors have suggested scheduling separation procedures on Sunday to minimize operating room traffic and congestion.[11] Besides operating room crowding, other major intraoperative problems to be discussed include choice of anesthetics, fluid management, cardiovascular monitoring, temperature preservation, and surgical techniques, especially skin closure.

Rehearsal of the separation procedure in the operating room can be very helpful in teaching personnel how to arrange needed equipment and coordinate their activities.[10,11,16] Rehearsal is especially necessary for a team performing its first separation of conjoined twins. A brief rehearsal is also recommended for experienced teams. We have also recommended that all personnel, equipment, monitoring leads, vascular lines, and records be color coded to identify at a glance with which twin they are associated.[23] Vascular lines, breathing circuits, and thermistor probes can be wrapped together with team-colored masking tape to form a single tangle-free "umbilical cord" for each infant.[16] The best dress rehearsal for proposed separation of conjoined twins is a palliative or diagnostic procedure that requires a general anesthetic, such as exploratory laparotomies or diverting colostomies.[6]

Due to circulatory admixture, drugs administered to one infant may have unpredictable effects on the other; therefore, light premedication, if any, of conjoined twins is recommended.[11,16,23] Intravenous drug administration is preferred over intramuscular administration because of unpredictable drug absorption.[11,16,23,25] The usual recommended intravenous dosages of premedicants, anesthetics, and adjuvants may be reduced by one-half and divided once again into equal doses for each twin. Using reduced incremental doses minimizes the dangers of compounding drug effects in one twin. The less vigorous twin should be premedicated first with incremental doses titrated to effect.[2,11,17] Steroid premedication has also been recommended before separation of conjoined twins as prophylaxis against adrenal insufficiency with hypotension in one or both twins.[26] Computerized tomograms have detected absence of adrenal tissue in xiphopagus, omphalopagus, and ischiopagus twins (table 2).[16] Magnetic resonance imaging may prove more helpful than computerized tomography in detecting presence or absence of adrenals in conjoined twins (table 2). Intravenous hydrocortisone (1.5 mg/kg) may be administered to each twin at induction of anesthesia, again at separation, and tapered postoperatively. Thoracopagus, craniopagus, and pygopagus twins have all been separated successfully without steroid prophylaxis and without evidence of adrenal insufficiency or hypotension unrelated to blood loss.[16] Prophylactic steroid administration to conjoined twins before separation remains controversial, except in twins with demonstrated absence of adrenal tissue.[16,26]

Some authors recommend establishment of airway and vascular access before transport of the twins to the operating room.[24] Others think that dislodgement of tubes and lines is more likely during transport, and prefer to wait until the twins are in the operating room to place monitors, intravenous lines, and endotracheal tubes.[16] We recommend that minimal transport monitors include electrocardiographs, skin or rectal thermistors, and pulse oximetry probes.

Normothermia can be maintained by transporting the twins in heated incubators or in radiant warmers, which will permit access and even surgery. We recommend transporting the twins to surgery (through cleared corridors) in the same radiant warmer in which surgery is to be performed to reduce risks of hypothermia or line dislodgement. As an option, two radiant warmers can be placed side-to-side or end-to-end in the operating room to permit the dual surgical teams to separate easily with their patients once the twins are disconnected. The twins may be positioned parallel or at right angles to the long axes of operating tables to provide for greatest surgical access. The problems of heat loss and surgical access must be individualized to the infants and to the separation operations, and they may be managed successfully with a variety of warming devices and operating tables.

The use of the same airway, breathing, and monitoring devices for transport and surgery can simplify transport duties and reduce confusion. To permit a continuum of care, certain anesthesia team members should also be designated as a transport team responsible for the twins' well-being, airways, vascular accesses, and monitors during both transport and surgery.

Intraoperative Management

Conjoined twins may require general anesthesia for diagnostic and emergency procedures not involving separation, as well as for urgent or elective separation and reconstruction of tissue defects. Many of the anesthetic management problems are similar regardless of operative procedures, and include difficult airway establishment and maintenance, unpredictable drug responses from cross circulation, thermal instability, and risks of extraordinary intravascular volume shifts or losses. Diagnostic procedures for conjoined twins may include endoscopy, invasive radiographic studies, or exploratory laparotomies.[3] Palliative procedures may include amputations, diverting colostomies, or ileal conduits.[3] As noted, urgent separations are recommended only for catastrophes, damage to the connective bridge, or rapid deterioration of one or both twins.[3] Most separations can be carefully planned, organized, and executed by highly trained teams experienced in complex infant surgery. The intraoperative anesthetic management of conjoined twins requires a dual team of one or two anesthesiologists for each twin, with an additional anesthesiologist acting as anesthesia team coordinator.

Anesthetic Management

The intraoperative anesthetic management of conjoined twins should aim to provide appropriate analgesia, amnesia, and muscle relaxation with control of the airways, adequate ventilation, thermal stability, and maintenance of normal circulatory and renal function. Such a perfect anesthetic state cannot always be achieved. Frequent adjustments in levels of anesthesia and muscle relaxation may be necessary in each twin because of cross circulation, cardiovascular depression, and unpredictable drug absorption and response.[25]

Analgesia and amnesia can be provided with inhalation and intravenous anesthetics. Muscle relaxants may be required to avoid cardiovascular depression from high inspired doses of inhaled anesthetics. We recommend percutaneous insertion of a 22- to 24-gauge intravenous catheter into each twin while awake; larger vascular access lines can be established after induction of anesthesia. Compared to the nursery course, the intraoperative course of conjoined twins being separated is one of fluctuating cardiopulmonary and volume status, temperature instability, and loss of neuromuscular irritability and tone.

Monitoring

The intraanesthetic monitoring of conjoined twins must be at least as extensive as that employed during routine infant anesthesia, and may be more so, depending on the nature of the surgical procedure.[27] Twins being separated require additional, more invasive monitoring, such as direct blood pressure monitoring by radial artery cannulas, umbilical artery catheters, or combinations. Right atrial catheters are indicated in separation surgery to provide reliable cardiac filling pressures and central circulatory access for vasopressors and volume restoration.[10,11,16] A pulse oximeter attached to a limb of each twin for continuous monitoring of peripheral perfusion and oxygenation will often sound the first warning if either twin is in jeopardy. We and others also recommend that urine volume be monitored for separation procedures and other major procedures lasting more than 1–2 h.[11,16] Brown *et al.* have used low-dose continuous dopamine infusion (3.0 μg/kg/min) to augment renal perfusion and maintain a urine output of at least 0.5 ml/kg/h during separation surgery.[11] Blood loss is measured by suction traps calibrated to 200 ml and by weighing of sponges. Serial determinations of arterial blood gases, glucose, electrolytes, ionized calcium, hemoglobin, platelets, prothrombin time, partial thromboplastin time, and fibrinogen should be measured frequently during separations and procedures with major blood loss.

Airway Management

Endotracheal intubation is recommended for intraoperative airway management of conjoined twins and insufflation of O_2 and anesthetic gases during separation or non-separation operations. In neonatal twins, awake tracheal intubation is recommended to prevent pulmonary aspiration and airway obstruction which could complicate intravenous or mask inductions. Endotracheal intubation may be difficult in thoraco-abdominally conjoined twins of any age connected ventrally and facing each other. Spontaneous ventilation with 100% oxygen followed by awake tracheal intubation is also recommended in such cases.[28] Nasotracheal tubes may offer more stability than orotracheal tubes in infants who require prolonged postoperative ventilation.

VENTILATORY MANAGEMENT

Spontaneous ventilation is not recommended during anesthesia in conjoined twins because of anesthetic-induced depression of alveolar ventilation and widening of the alveolar to arterial O_2 gradient (A-aDO_2).[29] Apparatus for administering gas anesthesia and ventilating conjoined twins during surgery should have minimal dead space and little resistance to breathing, should eliminate CO_2 adequately, and should provide for assisted or controlled ventilation. We also recommend that anesthesia breathing circuits allow for heated humidification of inspired gases and be compact and lightweight because of frequent positional changes during skin preparation, separation, and wound closures. To meet these needs, various nonbreathing modifications of Ayre's T-piece system that require a fresh gas flow of at least twice minute ventilation to prevent rebreathing are recommended.[30-33] We prefer hand ventilation to mechanical ventilation during separation procedures, because hand ventilation affords better appreciation of subtle changes in lung compliance and lower airway resistance. Hand ventilation may also be timed with surgical techniques during delicate cardiothoracic dissections. The use of mechanical ventilation with these nonrebreathing circuits, or with an infant circle system and CO_2 absorption, is advocated for longer procedures and during tedious plastic reconstructions of tissue defects. Inspiratory and expiratory pressures must be carefully monitored and arterial blood gases measured at frequent (30-min) intervals during intraoperative mechanical ventilation.

The adequacy of oxygenation and ventilation may be assessed by evaluation of color, breath sounds, chest excursions, transcutaneous O_2 saturation, and arterial blood gases. Cutaneous pulse oximetry will provide more reliable noninvasive monitoring of oxygenation than transcutaneous O_2 tension because of frequent anesthetic-induced alterations in skin perfusion and fluctuating skin temperature.[34] Unlike transcutaneous O_2 tension, however, recent investigations have shown transcutaneous CO_2 tension to be a more reliable indicator of adequate ventilation than either intermittent arterial CO_2 sampling or end-tidal CO_2 monitoring by infrared or spectrometric analysis.[35] The small tidal volumes and rapid respiratory rates common to infants are now known to interfere with end-tidal capnograms and spectrometric analysis of both inhaled and exhaled gases.[35]

FLUID THERAPY

In newborn infants with critical energy requirements and unstable cardiorespiratory and metabolic systems, intravenous fluid maintenance with both dextrose-containing and isotonic solutions may be required throughout any surgical procedure. Acute care fluid requirements are 4 ml/kg/h for twins 10 days or older, and 2 ml/kg/h for the first 10 days.[36] Infants on appropriate preoperative intravenous therapy have no fluid deficits. Older infants and children accumulate deficits from the last oral intake. By assigning one-half the combined body weight to each child, the blood volume, fluid requirement, deficit replacement, and blood replacement can be determined.[23] We and others recommend that deficits be replaced 50% in the first hour of surgery and 25% in each subsequent hour until complete.[23,36] Exposure of bowel to high intensity lights causes fluid evaporation that can be replaced at a rate of 2 ml/kg/h, and exposure of lung requires 4 ml/kg/h of extra intravenous fluids.[36] Abdominoperineal separation often requires an extra 6 ml/kg/h above the 4 ml/kg/h basic replacement.[36] All fluid replacements may be made using balanced electrolyte solutions without dextrose, *e.g.*, plain Ringer's lactate.[36] Infants needing glucose to maintain blood levels may be maintained on a separate infusion containing the appropriate dextrose concentration.[23,36]

Blood loss and need for replacement are assessed by obtaining serial hematocrits every half hour throughout the procedure.[23] A predetermined low hematocrit level should be chosen during the preoperative conference, at which point red blood cells will be replaced. Up to that point, we recommend that losses be replaced 1 ml for 1 ml with clear fluids.[23] It is critical to maintain circulating volume and avoid shock. In premature and newborn infants up to 10 days of age, hematocrit of 40% is probably advisable, if for no other reason than to try to sustain closure of the ductus arteriosus.[36] In older infants, a hematocrit of 30% is probably more logical.[36] In infants older than 6 months, hematocrits as low as 25% may be acceptable.[36]

Packed red blood cells are used for replacement of lost blood when appropriate.[23] Exact quantities required to maintain a given hematocrit are injected *via* a syringe and stopcock using blood warmed to body temperature.[23,36] If massive blood loss occurs at separation, the anesthesia team coordinator must determine which twin is losing the volume and direct replacement accordingly.[23] The initial need is to replace volume—using balanced electrolytes.[23] The hematocrit is then adjusted—using red blood cells.

Adequate fluid therapy produces urine at least 0.5 ml/kg/h of specific gravity 1.012 to 1.018.[11,23] Occasionally, when large quantities of clear fluid have been used, albumin may be required to restore the serum albumin level to 5 g/100 ml or greater.[23,36] This is achieved by infusing 5.0 ml aliquots of "salt-poor" 25% albumin in divided doses.[23] When large volumes of replacement fluids have been infused, examinations of platelet counts and coagulation activity may be repeated. It is advisable to use blood that is as fresh as possible from the blood bank for all transfusions.[23] Cumulative graphic summaries of fluid and blood product administration directed by fluctuating hemodynamic and clotting status can be found in many twin separation reports.[16,23,24]

ANESTHETIC PHARMACOLOGY

Increased anesthetic requirements and greater sensitivity to the cardiodepressant properties of both inhaled and intravenous anesthetics apply to conjoined twins as well as to normal infants.[37]§ In addition, limited reserves in shared systems help to account for the unpredictable responses to anesthetics in conjoined twins *versus* normal infants.[16,23,25]

Inhalation Anesthetics. For satisfactory surgical anesthesia, conjoined twins, like all infants, will require higher delivered concentrations of N_2O, halothane, enflurane, and isoflurane than older children and adults.[37,38]§ Interestingly, recent clinical investigations in newborn infants have shown isoflurane requirements to be less than anticipated in comparison with equipotent halothane doses, suggesting less inhaled anesthetic for satisfactory anesthesia with even faster emergence.[38] Isoflurane could prove a valuable inhaled anesthetic for surgery in conjoined twins. As with resuscitation and premedication, the less vigorous twin should be anesthetized first.[2,11,17] During N_2O-relaxant anesthesia, additional analgesia with narcotics will be needed.[30] Narcotics or barbiturates added to N_2O may promote more cardiovascular instability and, if used, should be carefully titrated as to intraoperative effects (hypotension, hypoventilation) and postoperative effects (delayed emergence, respiratory depression).[30] N_2O dilutes inspired O_2 concentrations, becomes a potent cardiovascular depressant when combined with narcotics, and rapidly expands air-containing cavities that are not being adequately decompressed, such as pneumothorax, congenital lung cysts, lobar and interstitial emphysema, or obstructed stomach or bowel.[39]¶ Such obstructed air-containing cavities often complicate preoperative evaluation and surgical management of conjoined twins with duplicated respiratory and alimentary tracts.[3] Compressed air or helium may also serve as O_2-diluting carrier gases in neonatal conjoined twins at risk of retrolental fibroplasia or in those in whom N_2O is contraindicated.§¶

Muscle Relaxants. Neuromuscular blocking drugs occupy a prominent place in infant anesthetic management for two main reasons.[29] First, muscle relaxants can provide optimal operating conditions and permit reduced doses of inhaled anesthetics with less cardiovascular depression.[29] Second, assisted or controlled ventilation of paralyzed, anesthetized infants provides better alveolar ventilation with less mismatching of ventilation and lung perfusion than spontaneous ventilation.[29]¶ Whether depolarizing or nondepolarizing, surgical muscle relaxants have distinct dosage requirements, cardiovascular effects, and elimination half-lives in normal infants that do not apply exactly to conjoined twins. As with all medications, dosage requirements, effects, and elimination are unpredictable, and usually differ with each twin.[23] Variable response and resistance to antagonism will be common in conjoined twins, and may indicate the need for postoperative mechanical ventilation.[16,23,24] Hypothermia, hypocalcemia, and concomitant aminoglycoside antibiotic therapy will potentiate nondepolarizing neuromuscular relaxants and interfere with adequate reversal in conjoined twins, as well as other infants.§¶

Intravenous Agents. Despite their increasing popularity and use in adult anesthesia, most intravenous agents have not been well studied in infants, and especially in conjoined twins. Narcotics and barbiturates produce more cardiorespiratory depression in newborns and infants than in adults.[40,41] Brown *et al.*[11] recently chose a single high-dose injection of fentanyl, 100 $\mu g/kg$, to induce and anesthetize throacopagus twins for separation. Such single injection techniques may reduce pharmacologic manipulation, allow the anesthesia team more time to concentrate on thermoregulation, fluid management, and blood administration, and provide better cardiovascular support during separation.[11] In experimental animals, high-dose fentanyl anesthesia combined with atropine has been found to provide more hemodynamic stability than either fentanyl alone or inhalation anesthetics.[42]

Ketamine has proved most useful as an intravenous anesthetic in the sickest infants with cyanotic congenital heart disease who will require prolonged postoperative ventilation.[43] Intravenous ketamine titrated to effect could provide for safer induction of anesthesia in conjoined twins needing urgent separation or surgery for abdominal emergencies.

In summary, all intravenous anesthetics can produce unpredictable effects in conjoined twins, and should be carefully titrated as reduced dosages at induction to avoid profound cardiovascular depression.[23] The re-

§ Downes JJ, Betts EK: Anesthesia for the critically ill infant, ASA Refresher Courses in Anesthesiology. Philadelphia, J. B. Lippincott, 1977, pp 58–60.

¶ Hinkle AJ, Alper MH: Anesthetic considerations for neonatal surgery, ASA Refresher Courses in Anesthesiology. Philadelphia, J. B. Lippincott, 1982, p 123.

versible agents, like narcotics and muscle relaxants, may offer more versatility and less risk of postoperative apnea than the longer-acting irreversible agents, like barbiturates, tranquilizers, and ketamine.

Termination of Anesthesia

At completion of surgery, several decisions must be made, including whether to reverse neuromuscular blockade, when to extubate the trachea, and what the course of postoperative pulmonary management is to be. Continued blood loss, systemic hypotension, hypothermia, and hypocalcemia are common after separation of conjoined twins, and contraindicate reversal of nondepolarizing muscle relaxants.[11,16,23] Endotracheal tubes that will remain in place postoperatively must be re-evaluated as to proper placement and patency, and may need changing at the completion of surgery. Only when the infants are conscious and vigorous may endotracheal tubes be removed, usually several days postoperatively.[11,16] Critically ill infants recently separated will certainly need controlled ventilation followed by a trial of spontaneous breathing with a T-piece and continuous positive airway pressure (CPAP) before tracheal extubation.[11,16,23] Reassessment of adequacy of oxygenation and ventilation will determine proper timing for tracheal extubation. Tight skin closures can restrict spontaneous ventilation and prolong the need for postoperative controlled ventilation.[44] Therapeutic pneumoperitoneum prior to planned separation or staged abdominal wall closures may reduce restrictive deficits following skin closure and permit earlier tracheal extubation postoperatively.[44]

Postoperative Management

Postoperatively, separated twins should be transported to intensive care areas in warmed isolettes or radiant warmers, and monitoring for hemorrhage, hypotension, hypothermia, hypocalcemia, hypokalemia, hypoxia, hypercarbia, and acidosis should be continued.[3,11,16] Invasive cardiovascular monitoring of systemic blood pressure and central venous pressure should also continue, as unrecognized volume losses and cardiogenic instability occur commonly in separated twins.[3] Cardiovascular and respiratory failure remain the most frequent causes of death in the immediate postseparation period.[3] Prolonged inotropic support and continued mechanical ventilation of each twin are often required.[3,11] Adrenocortical steroid therapy should be continued in infants without adrenal glands.[26] Further operations for wound dehiscences, secondary wound closures, and skin grafting of relaxing incisions are often necessary in the first few postoperative weeks. Prolonged hospitalization and multiple operations will quickly eliminate peripheral intravenous access sites. Central venous lines will allow for prolonged intravenous fluid and nutritional support and permit frequent intravenous anesthetic inductions. Wound infections, pneumonia, and catheter sepsis from long-term vascular or urinary catheters may also plague postoperative recovery of recently separated conjoined twins.[3]

Conclusions

Successful intraoperative management of conjoined twins undergoing surgical exploration or separation requires timing, teamwork, an appreciation of risks involved, and a firm understanding of conjoined pathophysiology. Prenatal diagnosis of conjoined twins is now possible, and permits careful planning for delivery, for preoperative work-up if separation is possible, and for early mobilization of a multidisciplinary separation team. Delayed separation for weeks or months will allow the needed time for growth, thorough preoperative evaluation of extent of joining, procedural planning sessions, and separation rehearsals. Urgent operations or separations may be necessary in conjoined twins. In these situations, rapid preoperative evaluation and management can be provided by precise noninvasive studies and cardiopulmonary stabilization before transport to the operating room.

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