

Did You Bring Two Isolettes? Transport of Conjoined Twins

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Case Study

Air medical transport was requested for full-term conjoined twins who were born by C-section for failure to progress in labor. The fact that the twins were conjoined was unknown until delivery, when the real cause for the failure to progress was revealed. The referring staff eagerly anticipated the arrival of the transport team, whose en route preparations were both physical and psychologic. Two full sets of monitoring and resuscitation equipment were necessary because the extent of organ sharing was unknown. Psychologic preparation was perhaps the most difficult because no member of the flight team had ever transported conjoined twins.

The flight team found omphalopagus (joined at the abdomen) twins waiting on their arrival. One infant was intubated with ventilatory support; the other required only oxygen by head-hood. The membranous sac joining the infants was covered with sterile saline-soaked gauze and plastic wrap to help prevent hypothermia and insensible water loss. As the crew "packaged" the twins, issues involving the physical logistics of placing these children in one isolette became evident and were addressed.

At approximately 1 month old, after an extensive workup and multiple surgeries, the girls were discharged home, alert and playful.

Historical Perspectives

A search through history reveals that perhaps the first descriptions of con-

joined twins were from Greek mythology. Janus, the mythical Roman god of beginnings, was depicted as having two bearded heads back to back.¹ The first documented conjoined twins were the Biddenden Maids, who were born in 1100.¹ Perhaps the most famous conjoined or "Siamese" twins were Eng and Chang Bunker, born in 1811 but never separated. Later, both married and fathered a total of 22 children between them. Chang's death was quickly followed by Eng's, an occurrence thought to be a result of the inability of one twin's cardiac output to continue to perfuse both bodies.

The Bunker twins story became legendary from the promotions of P.T. Barnum and his traveling circus side show.¹ Throughout history, as with many genetic anomalies, conjoined twins have been viewed as "freaks of nature" with little respect to the psychologic and emotional sequelae of their condition.

Epidemiology

Worldwide the incidence of conjoined twins ranges from 1 in 14,000 to 25,000 in southeast Asia to 1 in 50,000 to 200,000 in the United States. Approximately 50% of these children are aborted spontaneously or suffer an intrauterine death. An additional 25% to 30% die within the first 24 hours after birth. Of the remaining 20% of twins that live more than 24 hours, only 5% actually survive to leave the hospital after definitive separational surgery.^{2,4} Numerous embryologic research studies are ongoing to determine

the etiologic factors of twinning. Currently, however, the events that precede formation of conjoined twins remain merely speculative.⁵

Types of Conjoined Twins

Conjoined twins are classified with the suffix *pagus*, which comes from the Greek meaning "that which is fixed." The most common site of connection is at the thorax (thoracopagus); other areas of attachment include head (craniopagus), spine (rachiopagus), abdomen (omphalopagus), pelvis (ischiopagus), and sacrum (pygopagus).^{5,6}

Transport Considerations

In the transport setting, the full extent of shared blood vessels, organs, and tissues between the twins usually is not known. Therefore the crew must assume that whatever medication or fluid is given to one twin eventually will be shared with the other. Depending on the site of connection, each twin may have a complete set of life-sustaining organs, such as the brain, heart, or liver; other twins may share all or a portion of these organs.

Given the infrequency of live conjoined twins, the opportunity to manage these potentially critically ill infants is uncommon. However, transport teams' chances of interactions with these children are increased because of the need for rapid transport from the delivery site to a neonatal ICU for stabilization and resuscitation or to a tertiary medical center for possible separational surgery.

As the team prepares to transport conjoined twins, all necessary equipment must be available to resuscitate two newborns, including equipment for airway management and ventilatory support, cardiorespiratory monitors, IV pumps, and other monitoring equipment. If time permits, color-coding all equipment is recommended. For example, "twin A" and his or her equipment can be designated by red labels. Similarly, blue stickers are placed on "twin B" and his or her equipment. This method has been highly successful in minimizing confusion in the transport and operative settings.^{4,6}

Conjoined twins can be difficult to manage even in the controlled setting of the neonatal ICU; therefore management during transport can be expected

to be a significant challenge.⁷ In neonatal transport, the team focuses on stabilizing and resuscitating both infants. As with any neonatal patient, areas of concern include airway management, positioning, normothermia maintenance, and fluid/medication administration. Airway management needs can range from simple positioning to intubating both infants. One neonate may require intubation while the other requires minimal supplemental oxygen. Intubation may be technically difficult because of the joining site, making supine positioning difficult if not impossible, or the presence of airway abnormalities associated with chromosomal abnormalities.^{5,6}

Any sedative medications given to aid in intubation will cross over to the other infant's circulation, which may result in the need for unexpected intubation of the second twin.^{8,9} During transport, blended oxygen must be sufficient to sustain two infants on 100% oxygen for the duration of the transport. If needed, one infant can be placed on the isolette ventilator, and the other infant can be manually ventilated.

Positioning depends on the site of connection and may require extensive "improvisation" by the transport team. Whether both infants are able to lie supine or are facing each other will affect the transport team's decisions, specifically regarding airway management during packaging and transport. Warmed blankets and padding should be used to maintain alignment in addition to preserving endotracheal tubes and all invasive lines. Ensuring both children are at the same physical level in the warmer or isolette will help prevent inadvertent hypovolemia because venous return to the nondependent twin may be compromised by gravity.^{8,10}

Normothermia maintenance is always a concern in the transport of patients of all ages. Conjoined twins are at particular risk for hypothermia because of their greater body surface area.^{4,7,11} Warming units, hats, and ear attenuators should be applied before placing the children in a prewarmed isolette. Lastly, if available, temperature monitoring should be used during transport to aid early detection and hypothermia management.

Crossover and shared circulations

must be assumed when administering IV fluids and medications to these twins. Intravenous access typically is obtained by large-bore peripheral IV line (PIV). *Each child will require a separate PIV.* Intravenous fluid rates should be calculated based on a total kg weight basis and then divided between the children.^{6,9}

As with IV fluids, medications should be given based on the children's total body weight in kilograms. Medications administered to one infant will affect the other; however, the rate and extent of distribution is not predictable.^{6,8-10} Each infant may be considered a separate "compartment," allowing the drug to distribute quickly within the first infant to create a peak concentration, while the second infant still is reaching a state of equilibrium. For this reason, *sedatives, analgesics, and paralytics should be administered to the smaller or "weaker" of the infants first.* Adverse reactions, such as respiratory depression, quickly will become manifest and can be addressed.^{5,6,9,11}

Cross-circulation may be unbalanced and may alter the rate of distribution to a steady state concentration.^{12,13} Hepatic biotransformation and clearance and renal elimination also may be altered because of the number of organs shared, blood flow through those organs, and the infants' clinical conditions. Serum drug concentrations may be helpful to determine pharmacokinetics and dosing of a given agent over time, but these data must be interpreted carefully in light of the lack of literature. Adverse reactions may be seen in one twin or the other, such as indomethacin administered for patent ductus arteriosus closure in one twin with anuria seen in the second twin only.¹⁴

Generally each infant should receive *half* the calculated total dose of a given drug, and *both* children should be carefully monitored. When sepsis is suspected, half the total calculated dose of antibiotics should be given to each infant. Gentamicin levels should be monitored for dosing purposes because the distribution kinetics and elimination rates probably will be altered. Pressors and resuscitation medications should be administered to both children, with close monitoring for potential effects on each infant.

The last aspect the team must consider is the psychosocial issues that transports of this type may evoke. Historically, the public has viewed conjoined twins as freaks of nature, and a great deal of media attention continues to be associated with these children. The

team should be prepared for the possibility of media presence at either end of the transport. Patient and family confidentiality should remain of utmost importance in protecting the privacy and dignity of everyone concerned.^{5,7,11}

In summary, conjoined twins are

transports that will not soon be forgotten. The care of these infants is in many ways very similar to routine neonatal transports. The key is remembering that two separate infants require the best care because they share a most intimate bond.

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