

Chapter 11

Conjoined Twins



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The patients are 7-month-old conjoined twin females born at 32 weeks' gestation. They are joined at the sacrum. Planned surgery is separation.

Preoperative Evaluation

Questions

1. What is the rate of conjoined twin birth? What is their chance of survival? How urgent is the surgery?
2. How are conjoined twins classified? How does their classification impact their prognosis?
3. Describe the nature of the preoperative evaluation for conjoined twins.

Answers

1. Conjoined twins are a relatively rare occurrence with an incidence of approximately 1 in 50,000 to 1 in 200,000 births. They are always monozygotic and monochorionic twins. Forty percent of these pregnancies end in stillbirth. Of the 60% that are born, only about 20% live to be eligible for separation. The defect leading to conjoined twins is likely a fusing of overlapping or closely contiguous twin embryonic axis formative fields within a single embryonic disc. It is thought that these factors are responsible for the failure of twins to separate after the 13th day after fertilization. Conjoined twins have been created in amphibians by simply constricting the embryo so that two embryos form, one on each side of the constriction. Most often this surgery is not urgent, and time can be taken to optimize the planning for the environment as well as the underlying health of the twins. Many of these cases are performed when the twins are a year or more of age. On rare occasions, based on the shared anatomy, the health of one or both twins may be adversely impacted by their conjoined nature – in which case the need for surgery becomes more time-sensitive.
2. Conjoined twins are classified according to the region by which they are joined. Craniopagus twins are joined at the head. Thoracopagus twins are joined at the upper half of the trunk. (This is the most common form of conjoined twins making up 35–40%.) Omphalopagus twins are joined at the chest or abdomen – the second most common form of the anomaly at 30% of all cases. Most often these patients share a liver and prognosis for separation is generally good. Pygopagus are joined at the sacrum and constitute 19% of all cases. Separation is most straightforward in these cases since they do not generally share vital organs and survival is high.
3. Preoperative evaluation will vary with the site at which the twins are conjoined and the nature of shared organs. In this case, it would include routine blood and urine analysis, coagulation screen, plain X-rays, and ultrasound evaluations of the abdomen and pelvis. Computed tomography and digital subtraction angiography (DSA) delineate anatomic and bony detail including organ position, shared viscera, and vascular anatomy. In this case, a CT myelogram or MRI would be critical to determine the extent to which neural elements and spinal cord anatomy are shared. We preferred the use of the CT myelogram since that did not involve anesthetizing the patients where an MRI generally would require sedation/anesthesia. The determination of the blood supply to these elements would also be critical. Preoperative assessment and planning with interdisciplinary communication and cooperation is vital to the success of this type of procedure. A dedicated team of anesthesiologists for each twin is required. There must be two sets of all monitoring equipment and resuscitation equipment. All equipment and monitors need

to be labeled for each twin. It is often advised to color code the equipment that is to be used and separate between the two patients (one is green, the other yellow, etc.). It is helpful to color code the teams as well. It is advised to “trial” the operating room setup and simulate the operation and environment at least once prior to surgery. Plans for the surgical, anesthesia, OR technical, and nursing aspects of the case must be fully reviewed and trialed.

Prior to surgery, it may be desirable to test for the presence of cross-circulation. At times, this is very clear from the MRI or CT scans that are obtained for surgical planning. If the degree of shared circulation is in doubt, one simple way to determine this is by administering an anticholinergic such as atropine to one twin and monitoring if heart rate change is observed in the other twin. Alternatively, other agents such as Tc-99m, microcolloidal human serum albumin (HSA), and Tc-99m HIDA can be injected in one twin and detected in the other. Finally, indigo carmine can be injected into one twin with the examination of the other twin’s urine for indigo carmine excretion. This is critical when considering how to safely induce anesthesia for these patients. If the patients share circulation, anesthesia must be induced simultaneously since any drug administered to one twin will necessarily affect the other. Conversely, if circulation is not shared, induction could be achieved separately.

Answers

1. Surgery to separate conjoined twins is always a long process with the potential for significant blood loss and fluid shifts. A full set of ASA monitors is required for both patients. In addition, it would be appropriate to place a radial arterial line for both twins. The need for central access can depend on the exact nature of the conjoining of the twins. As a general rule, it is preferable to have central access for each patient in order to provide fluids, blood products, and vasoactive drugs and obtain some measure of central filling pressure. Urinary catheters should be placed.
2. The induction of anesthesia will depend on the nature of the airways involved and the presence of shared circulation of the twins. Conjoined twins may be positioned in such a way that their airways are difficult to access even though they may be anatomically normal. In addition, they may have craniofacial anomalies that could make their airway management challenging regardless of positioning. In this case, the twins were normal in appearance and facing away from each other. They underwent inhaled induction (with a single IV catheter in place on each twin) simultaneously, muscle relaxant was given after mask ventilation was established, and intubation was accomplished without difficulty. If the circulation were shared, it would be particularly important to induce anesthesia simultaneously since both twins would receive some effect from any drug admin-

3. Are there any other medications or precautions that are particularly important for this procedure?

Postoperative Course

Question

1. Would you extubate these babies postoperatively?

Additional Question

Question

1. A set of 9-month-old thoracopagus conjoined twins requires a CT myelogram to delineate an uncertain area of possible shared spinal anatomy. How would you accomplish sedation for the injection and CT scan?

istration. In any case, if twins are induced separately, it is critical to observe the nonanesthetized twin for any reaction or effect.

3. As these are very long procedures and blood loss is likely to be several blood volumes, it is important to plan for blood conservation and transfusion. The administration of tranexamic acid (TXA) or epsilon aminocaproic acid (Amicar®) would be reasonable to attempt to minimize fibrinolysis. If the children were large enough, blood salvage techniques should be in place. Fluids should include a maintenance fluid of D5 1/2NS and an isotonic solution such as lactated Ringers to account for third spacing and blood loss. Colloidal fluids such as albumin could be added when fluid replacement exceeds 50 mL/kg, but there is little indication that this changes outcome. The blood bank should be notified of this case, and appropriately typed, screened, and (if necessary) cross-matched blood should be available. The blood bank should be at least one to two units “ahead” at all times for each twin. The need for platelets and fresh frozen plasma should also be anticipated. If thromboelastography (TEG) is available, it can be very helpful in these cases to determine the nature of anticoagulation in the face of massive transfusion needs.

Answer

1. No. After any separation procedure, the duration of the procedure and the administration of large amounts of fluid and blood require a prolonged recovery. Airway edema and neurological status are not going to be appropriate for extubation. ICU beds should be available, and sedative/opiate infusions will be needed for (usually) a significant period while the twins recover.

Answer

1. We believe that in these cases the least amount of anesthesia that accomplishes the goals of the procedure would be most desirable. It is important to remember that if the circulation is shared, sedating one twin may suffice for both. Also, it is important to note that only one twin needs to undergo contrast injection. In this case, we chose the twin whose spine offered the easiest exposure and place EMLA cream at L3–L4. Sucrose pacifiers were given to both twins. While their nurse held them on her chest, a 22 G spinal needle was placed and clear CSF was accessed. The contrast was then administered. The twins were then positioned for the CT scan. One twin was administered 0.05 mg/kg of midazolam and 0.25 mg/kg of ketamine. Both became quiet. With standard ASA monitors in place, the scan was completed.

Suggested Reading

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- Thomas JM. Chapter 36: Anesthesia for conjoined twins. In: Davis PJ, Cladis FP, editors. *Smith's anesthesia for infants and children.* 9th ed. Philadelphia: Elsevier; 2017.
- Zhong H-J, Li H, Du Z-Y, Huan H, Yang T-D, Qi Y-Y. Anesthetic management of conjoined twins undergoing one-stage surgical separation: a single center experience. *Pak J Med Sci.* 2013;29(2):509–13.