orphanaesthesia

Anaesthesia recommendations for

Conjoined twins

Disease name: Conjoined twins

ICD 10: Q 89.4

Synonyms: Siamese twins

Conjoined twins are not only an extremely rare congenital malformation in humans but also one of the most difficult one to treat. This defect is an example of one of the most complex organizational and ethical issues encountered in medicine. The incidence of conjoined twins is estimated to be 1 in 50,000 to 1 in 200,000 births. Most conjoined twins are stillborn or die shortly after birth due to accompanying developmental defects. The defect is three times more frequent in girls and its occurrence is higher in Africa and Southwest Asia. There are two contradicting theories to explain the formation of conjoined twins. The most accepted theory is fission; when a single fertilized egg fails to split completely between the 13th and the 15th day after fertilization. This theory is supported by clinical observations. Conjoined twins are always of the same gender, are genetically identical, and are always joined symmetrically at the same body part. The second theory is secondary fusion of two originally separate embryos.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

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Disease summary

Conjoined twins are classified according to the site of union, with the suffix pagus meaning fixed. The most clinically useful, albeit highly simplified, classification of conjoined twins divides them into symmetrically conjoined (the same size, symmetrical to each other) and asymmetrically conjoined (one is always smaller, a parasite), and dependent on the other.

Symmetrically conjoined twins include children joined at the following regions:

- Chests (*thoracopagus*, *xiphopagus*) with possible fusion involving the heart, liver and upper gastrointestinal tract,
- Abdominal cavity (*omphalopagus*) - united at the region of the liver and gastrointestinal tract where children are facing each other,
- Sacral bones (*pygopagus*) - children with their backs to each other, usually joined by the pelivic organs, anus and pelvic nervous system,
- Heads (*craniopagus*) - usually fused medially sharing the nervous tissues, large vessels and sinuses,
- Pelvis (*ischiopagus*) - most frequently sharing the urogenital system, rectum and liver. These children can have either 4 fully developed lower limbs or only 3 with 1 fused and that is deformed; this defect may be also accompanied by omphalocoele.

The group of **asymmetrical conjoined twins** includes cases in which the body parts differ in size - one foetus, called a parasite, is smaller, partially developed and dependent on the other, fully developed foetus, called an autosite.

Typical surgery

Anaesthesia for conjoined twin's surgery, whether prior to, or for separation, is an enormous challenge to the anaesthesiologist. It must be emphasised that each twin should be treated as a separate entity. Anaesthetics for both diagnostic and the final separation procedures are always conducted by two independent anaesthetic teams.

Anaesthesia or sedation may be required prior to separation for diagnostic tests (CT, MRI, angiography, correction or palliation of congenital defects that require urgent surgery (laparotomies for intestinal obstruction, colostomies, necrotizing enterocolitis) and the insertion and removal of tissue expanders.

Conjoined twins may require anaesthesia for emergency separation surgery, but this is most frequently performed as elective procedures. The anaesthesia and surgical procedure may range technically simple to complex, depending on the point of fusion and the internal organs that are shared. In most cases separation is extremely risky and may be life-threatening. Pharmacokinetics and pharmacodynamics can be inconsistent between each twin and cross-circulation is of particular significance in the thoracopagus and craniopagus twins, and drug responses may be altered and unpredictable.

Type of anaesthesia

There are no limitations to drugs that can be used for this type of anaesthesia. Where conjoined twins are joined at their heads, chests or abdomen, shared circulatory systems of both children should be expected. This is an important consideration for induction of anaesthesia, especially an intravenous one.

Drugs administered to one twin may have unexpected effects on the other. The anaesthesia management plan should take into account the type and route of premedication, induction and maintenance drugs, as well as the extent and type of monitoring required for the procedure. Anaesthesia for diagnostic examinations and pre-separation procedures may only require non-invasive a different range of monitoring.

When selecting the anaesthetic technique, it is always preferable to wake the children immediately after the diagnostic or pre-separation procedure. Those with abnormal cardiopulmonary function should be referred to the ICU. Separation surgery requires different anaesthesia management. Planning for separation surgery must take into account the induction of general anaesthesia, intubation strategy, provision of peripheral and central venous accesses, direct arterial access, positioning of patients on the operating table and protection against heat loss. Anaesthesia is time-consuming and can take even several hours. The operating theatre should be equipped with two work stations, full monitoring for each child and individual coded equipment.

Necessary additional diagnostic procedures (preoperative)

The detailed planning for anaesthetic and surgical correction of conjoined twins requires a multidisciplinary team of specialists who have at their disposal detailed information regarding the malformation. To obtain this information, conjoined twins will require a range of investigative work-ups, including CT scans, MRI scans, ultrasonography, perfusion studies, echocardiography and blood tests. Anaesthetic management of conjoined twins must be based on current information obtained from these diagnostic tests and should be modified depending on the availability of new information.

Particular preparation for airway management

Endotracheal intubation may be difficult, particulary when twins are joined at chests.

Emergency intubation can also be extremely challenging. Where possible, an intubation strategy should be planned and the twins should be monitored so that intubation might occur in advance of it being required as an emergency. Ideally deterioration and the need for intervention should be anticipated and intubation carried out in a controlled way.

In cases where twins are joined at their chests, the maintenance of upper airways patency a face mask during induction is hindered by the fact that they are facing each other. It is also difficult to expose the larynx during intubation and rotation of children’s heads to facilitate intubation may in turn deform the upper airway.

The use of the LMA in conjoined twins depends on the type of conjunction and the procedure being performed. In thoracopagus twins where their heads are facing each other, the placement of an LMA equally is challenging. After intubation, it is essential to secure the endotracheal tube. Frequent position changes during surgery pose a risk of displacement, or

even accidental extubation. Nasal intubation secure taping and continuous monitoring are the most effective means of protecting from endotracheal tube displacement.

Particular preparation for transfusion or administration of blood products

Anticipate and monitor for blood loss and have blood products available. Blood loss may be massive in craniopagus or in thoracopagus twins. Estimation of intraoperative blood loss in conjoined twins is extremely difficult. Often surgical bleeding and transfusion requirements are different in each twin. Blood loss can be estimated bythe measurements of the blood volume suckioned from the surgical site, the weight of the surgica swabs used, and the haemodynamic parameters, haematocrit and haemoglobin determined in each twin.Each child should by reassesed at regular intervals. Blood products should be crossmatched in advance for each individual twin.

Particular preparation for anticoagulation

There is no evidence to support the need for anticoagulation. Central line placement may pose a higher risk of postoperative venous thrombosis.

Particular precautions for positioning, transport or mobilisation

Conjoined twins are more difficult to transport than individual newborns and the transport team should be suitably prepared and equipped. It is essential to stabilize each child prior to transportation. Where necessary airways must be secured to ensure adequate ventilation. Normothermia, and adequate fluid supply, and safe positioning is important. A separate transport team should be assigned to each individual child. Cardiopulmonary resuscitation, should it be necessary, is very difficult and has a poor prognosis, particulary in children joined at the thoracic and abdominal region. Due to defective anatomy, chest compressions may not be possible and may also cause damage to the intestines and liver.

The cardiac arrest or death of one twin is frequently an indication for emergency surgical separation, and failure to achive urgent separation will result in the death of the second twin. Positioning of the babies for separation surgery must be planned with each of the surgical specialties involved in the procedure. Each stage of the separation, an order of procedures the positions required and the placement of lines, airways and monitoring must be considered. The table should be large enough to accomodate both twins and meticulous care with the use of protective rests and padding is important. Simulation has been used to plan positioning, movements and transfers in advance of the separation procedure.

Probable interaction between anaesthetic agents and patient's long-term medication

Not reported.

Anaesthesiologic procedure

When planning anaesthesia it must be decided which twin will be anaesthetized first, the

method of induction of anaesthesia and an intubation plan. Inhalational induction with sevoflurane is the preferred method. In twins where no airway difficulties are anticipated, intravenous induction can be used. Anaesthesia is preferentially maintained by combining inhalational anaesthesia and fractionated doses of opioids. Intravenous anaesthesia may be used in accordance with the preferences and experience of the anaesthetists performing the procedure, but the technique is challenging where circulations are shared. The same technique should be used on each twin regardless. If nitrous oxide is used for inhalational induction, it should be discontinued where indicated.

Muscle relaxation may be achieved with standard non-depolarising muscle relaxants such as rocuronium, vecuronium and atracurium but since most separation procedures lasts several hours, pancuronium is preferable.

Perioperative antibiotic prophylaxis should be prescribed according to local hospital guidelines.

Local or regional anaesthesia may be used when possible. There are several reports on use of caudal and epidural anesthesia in conjoined twins during the establishment of central vascular accesses or plastic surgery for postoperative pain control.

Particular or additional monitoring

The separation of conjoined twins can last several hours. A full range of monitoring is necessary including invasive monitoring. Regular laboratory testing must be carried out during the surgery, when indicated or as often ae every 1 - 2 hours.

Possible complications

Hypothermia caused by the extensive surgical wound, which increases heat loss by evaporation, radiation and convection is a serious problem during separation surgery. It is necessary to use all the available methods to protect these children against heat loss in the operating theatre. Many authors emphasize that the preservation of normothermia during separation surgery is one of the most important factors affecting the outcome of surgical treatment In order to increase the safety of conjoined twins during anaesthesia and separation surgery for conjoined twins, all monitoring, infusion lines, ventilation systems and equipment are coded with a different colour for each child, which makes it easier to distinguish the twins during the surgery and to avoid mistakes.

Most separation surgeries requires several changes of twins' positions on the operating table and it is necessary to remain extremely vigilant of the airways and vascular access points. Careful observation for sudden heart failure caused by circulatory collapse at separation, unappreciated blood loss and undiagnosed cardiac abnormalities is vital.

Postoperative care

Postoperative care and invasive monitoring on ICU is mandatory in most cases.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.:

Emergency situations have been described previously.

Ambulatory anaesthesia

Not reported.

Obstetrical anaesthesia

Not reported.

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