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Biotinidase deficiency

Conjoined twins

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 2 | 2021

OrphanAnesthesia –

ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –

a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

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orphananesthesia

Anaesthesia recommendations for

Conjoined twins

Disease name: Conjoined twins

ICD 10: Q89.4

Synonyms: Siamese twins, conjoint twins

Disease summary: Conjoined twins are not only an extremely rare congenital malformation in humans but also one of the most difficult ones to treat. This defect is an example of one of the most complex organisational 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 fertilised egg fails to split completely between the 13th and the 15th day after fertilisation. 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.

Conjoined twins are classified according to the site of union, with the suffix 'pagus' meaning 'fixed'. The clinically most useful, although 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 pelvic 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.

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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'.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

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Typical surgery

Anaesthesia for surgery on conjoined twins, 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, necrotising 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 from technically simple to complex, depending on the point of fusion and the internal organs that are shared. In most cases, the 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 require a different range of monitoring, only a non-invasive one.

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 individually coded equipment.

Necessary additional pre-operative testing (beside standard care)

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

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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, particularly when twins are joined at chests.

Emergency intubation can also be extremely challenging. Whenever possible, an intubation strategy should be planned and the twins should be monitored so that intubation might be done in a controlled and fast way when the necessity is beginning to show. Ideally, deterioration and the need for intervention should be anticipated.

In cases where twins are joined at their chests, the maintenance of upper airways patency with 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. The 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 by the measurements of the blood volume suctioned from the surgical site, the weight of the surgical 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 cross-matched 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 post-operative venous thrombosis.

Particular precautions for positioning, transportation and 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 stabilise each child prior to transportation. Where necessary, airways must be secured to ensure adequate ventilation. Normothermia, adequate fluid supply, and safe positioning is important. A separate transport team should be assigned to each individual child. Cardiopulmonary resuscitation, should it be

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necessary, is very difficult and has a poor prognosis, particularly 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 achieve 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. At each stage of the separation, the order of the positions required has to be defined and the placement of lines, airways and monitoring must be considered. The table should be large enough to accommodate 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.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

When planning anaesthesia, it must be decided which twin will be anaesthetised 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 when 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 if indicated.

Muscle relaxation may be achieved with standard non-depolarising muscle relaxants such as rocuronium, vecuronium and atracurium, but since most separation procedures last 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 the use of caudal and epidural anesthesia in conjoined twins during the establishment of central vascular accesses or plastic surgery for post-operative 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 as every 1–2 hours.

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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 available methods to protect these children against heat loss in the operating theatre. Many authors emphasise 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, 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 surgery and avoid mistakes.

Most separation surgeries require several changes of the 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.

Post-operative care

Post-operative care and invasive monitoring on ICU is mandatory in most cases.

Disease-related acute problems and effect on anaesthesia and recovery

See descriptions above.

Ambulatory anaesthesia

Not reported.

Obstetrical anaesthesia

Not reported.

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References

1. Omokhodion SI, Ladipo JK, Odebode TO. The Ibadan conjoined twins: a report of omphalopagus twins and a review of cases reported in Nigeria over 60 years. Ann Trop Paediatr 2001;21:263–270
2. Pearn J. Bioethical issues in caring for conjoined twins and their parents. Lancet 2001; 357:1968–1971
3. Spitz L, Kiely EM. Experience in the management of conjoined twins. Br J Surg 2002; 89:1188–1192
4. Spitz L, Kiely E. Conjoined twins. JAMA 2003;289:1307–1310
5. Seefelder Ch, Hill DR, Shamberger RC, Holzman RS. Awake caudal anesthesia for inguinal surgery in one conjoined twin. Anesth Analg 2003;96:412–413
6. Thomas JM, Lopez JT. Conjoined twins - the anaesthetic management of 15 sets from 1991-2002. Pediatr Anesth 2004;14:117–129
7. Thomas JM. Anaesthesia for conjoined twins. Childs Nerv Syst 2004;20:538–546
8. Abdullah Al Rabeeah. Conjoined twins - past, present and future. J Pediatr Surg 2006;41:1000–1004
9. McHugh K, Kiely EM, Spitz L. Imaging of conjoined twins. Pediatr Radiol 2006;36:899–910
10. Chalam KS. Anaesthestic management of conjoined twins separation surgery. Indian J Anaesth 2009;53:294–301
11. Parameswari A, Vakamudi M, Raghupathy V, Siddhartha R: Anaesthetic management of total ceaniopagus twins for magnetic resonance imaging and cerebral angiography. Brain J Anaesth 2010;105: 368–370.
12. Jaya Lalwani, Dubey KP, Pratibha Shah. Anaesthesia for the separation of conjoined twins. Indian J Anaesth 2011;55:177-180
13. Chelliah KK, Faizah MZ, Dayang AA, Bilkis AA, Shareena I, Mazli M: Multimodality imaging in the assessment of thoraco-omphalopagus conjoined twin: lessons to learn. Case Rep Radiol 2012;2012:564036
14. Schleelein LE, Perate AR, Ganesh A. Supraclavicular block as a sole anesthetic technique in craniopagus twins. Reg Anesth Pain Med 2013;38:456–458
15. Aoun Tannuri AC, Pereira Batatinha JA, Prieto Velhote MC, Tannuri U. Conjoined twins – twenty years' experience at a reference center in Brazil. Clinics 2013;68:371–377
16. Denardin D, Telles JA, da Silveira Betat R, Fell PR, et al. Imperfect twinning: a clinical and ethical dilemma. Rev Paul Pediatr 2013;31:384–391
17. Kobylarz K. Anaesthesia of conjoined twins. Anaesthesiol Intensive Ther 2014;46:124–129
18. Kobylarz K. Anaesthesia of conjoined twins – case series. Anaesthesiol Intensive Ther 2014;4665–4677
19. Misook S, In-Sun C, Myong-Hwan K, Ji Mi O, Won-Jung S. Anesthetic management for separation of thoracopagus twins with complex congenital heart disease – a case report. Korean J Anesthesiol 2015;68:295–299.

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