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Situs inversus totalis

Spinal muscular atrophy

orphan**a**nesthesia

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

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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.

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 recommendations 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.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

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orphananesthesia

Anaesthesia recommendations for Situs inversus totalis

Disease name: Situs inversus totalis

ICD 10: Q89.3

Synonyms: Situs inversus, Situs inversus viscerum, Complete situs inversus, Complete situs inversus viscerum

Disease summary: Situs inversus is a congenital abnormality in which organs in the thorax and the abdomen are opposite to their normal positions. The normal arrangement of organs is known as situs solitus. Situs inversus is the mirror-imaged arrangement of the organs. (The anatomical left lung is in the right hemithorax and the anatomical right lung is in the left hemithorax. The stomach and spleen are on the right, the liver on the left. The heart is on the right.) Situs inversus is categorised under heterotaxy syndromes, which results from failure of the developing embryo to establish normal left-right asymmetry and is associated with a wide range of cardiac and extracardiac congenital anomalies. During the embryological development, a 270 degree clockwise rotation instead of normal 270 degree anti-clockwise rotation of the developing thoraco-abdominal organs results in mirror image positioning of the abdominal and thoracic viscera.

Situs inversus is a hereditary syndrome, which is estimated to occur in 1 in 8,000 to 25,000 individuals. It was reported as one in 5,000–20,000 births. Situs inversus totalis is usually an autosomal recessive disorder, and may sometimes be X-chromosome-related. There is no established gender or ethnic difference in its incidence, but genetic predisposition and familial occurrence point towards multiple inheritance patterns. Most cases are caused by ciliary abnormalities (the primary ciliary dyskinesia disorders). Cilia also are responsible for determining proper visceral laterality in utero.

Thus, during embryogenesis in patients with primary ciliary dyskinesia, visceral laterality will be determined at random, and half of these patients will have situs inversus totalis. Situs inversus totalis is associated with right-sided heart (dextrocardia). Situs inversus may be abdominal, thoracic, or both. Most patients are asymptomatic and maintain their normal life.

The association of situs inversus totalis with syndromes such as Kartagener syndrome (i.e. situs inversus, primary ciliary dysfunction (PCD), chronic sinusitis and bronchiectasis), cardiac anomalies, spleen malformations, mucociliary dysfunction, and airway anomalies makes the clinical management difficult for the anaesthesiologist.

There is a significantly higher incidence of heart defects in this group compared to normal hearts. Situs inversus totalis is associated with numerous cardiac anomalies such as atrial septal defect, ventricular septal defect, transposition of great vessels, abnormal atrioventricular valves, absent coronary sinus, conduction abnormalities, double-outlet right ventricle, total pulmonary venous defect and pulmonary valve stenosis. Dextrocardia with complete situs inversus occurs in approximately 2 of 10,000 births. The most prevalent cardiac defects

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associated with situs inversus are ventricular septal defect and transposition of the great vessels. There are limited reports of sick sinus syndrome diagnosed in patients with dextrocardia. In general, heart rhythm disturbance is not a significant finding in patients with situs inversus compared to situs solitus.

The extracardiac anomalies that most frequently challenge the anesthesiologist are gastrointestinal abnormalities, hepatic dysfunction, splenic dysfunction, respiratory abnormalities, and associated midline defects.

Situs inversus is associated with primary ciliary dyskinesia (Kartagener syndrome) in 20–25 % of cases. Kartagener syndrome occurs with the triad of bronchiectasis, chronic sinusitis (as symptoms of PCD) and situs inversus and should be excluded by a carefully obtained medical history. Situs inversus totalis is present in 50 % of individuals with Kartagener syndrome. The patients are prone to an increased probability of developing respiratory complications. Aggressive pre-operative optimisation and treatment of infection is required. A history of intermittent respiratory tract infections, especially during the winter season, should be watched.

The coexistence of situs inversus and congenital duodenal obstruction is an extremely rare situation. A relationship between ectopic thyroid and situs inversus totalis was reported. Most patients with ectopic thyroid show clinical features of euthyroid, while a few patients show clinical features of hyperthyroidism. Thyroid function tests are useful for the pre-operative diagnosis.

Spinal dysrapism, which includes all forms of neural tube defects from thickened filum terminalis to meningocele, is an important abnormality that is associated with situs inversus regarding neuraxial blocks.

There is some association between situs inversus and renal abnormalities. Kidney function should be tested pre-operatively if situs inversus with Kartagener syndrome is suspected. Biliary atresia may occur in 28 % of infants born with situs inversus as compared to the 0.01 % of the general population. The anaesthetic concerns include managing the abnormalities related to liver pathology (deranged metabolism, coagulation problems). Situs inversus is also a very rare feature of Bardet-Biedl syndrome.

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

Typical surgery

Thoracic surgery; cardiovascular surgery; gastrointestinal surgery; liver and spleen surgery; sinus and ear surgery.

Type of anaesthesia

There is no definite recommendation for either general or regional anaesthesia.

There are reports of spinal, epidural, combined spinal-epidural, caudal anaesthesia and nerve blocks without any complications. The spinal deformities like split cord, spina bifida, meningocele, and scoliosis has been described in the literature; thus, the patient should be evaluated carefully if any surgery is planned under neuraxial anaesthesia.

A regional block may be the method of choice if feasible. It should be provided that the motor block does not spread high into the thorax and respiratory muscle tone is maintained. Successful axillary brachial plexus blockade was reported under midazolam sedation. The primary advantage of regional technique is non-interference with respiratory muscle function, especially in patients with Kartagener syndrome. It also allows for improved clearance of respiratory secretions and early post-operative ambulation. An important consideration is the association of situs inversus with spinal dysraphism and other spinal malformations. Regional anaesthesia is considered a safe method for any infra-umbilical surgery compared to general anaesthesia if no spinal anomaly or dysraphism is confirmed.

Thoracic segmental epidural anaesthesia was reported in cases of situs inversus associated with Kartagener syndrome for laparoscopic cholecystectomy. This may be considered due to chronic airway disease and to prevent prolonged mechanical ventilation. However, the definitive advantage of this approach may be debated and the safest in the respective hospital best established anaesthesia technique should be preferred to techniques that are infrequently used, especially in patients with rare diseases.

General anaesthesia can be administered as total intravenous anaesthesia or with volatile anaesthetics. Nitrous oxide can also be used, but should be avoided in case of manifest cardiac involvement such as an atrial septal defect or a ventricular septal defect.

There is no contraindication for premedication such as sedation or analgesia for non-symptomatic patients. In patients with symptoms of the disease, the administration of sedative and analgesic drugs should only be performed after considering the individual risks carefully.

Necessary additional pre-operative testing (beside standard care)

The precise diagnosis of situs inversus and a thorough pre-operative evaluation can minimise the difficulties and the various potential challenges. The most preferred diagnostic technique involves chest and abdominal radiographs. A chest X-ray helps to assess cardiac situs and size, pulmonary blood flow, bronchial anatomy, liver symmetry, and stomach. The patient's chest X-ray reveals dextrocardia and a right-sided stomach bubble.

Ultrasound can determine the situs of abdominal organs, particularly abnormal liver symmetry and presence of the spleen and gallbladder. Magnetic resonance imaging is

helpful for defining extra-cardiac and cardiac anomalies. An upper gastrointestinal contrast study screens intestinal abnormalities and other digestive tract disorders.

Computed tomography (CT) imaging shows pancreatic and airway anomalies and the morphology of the right atrium. CT may show lung overinflation, bronchial wall thickening, and sinus malformations in patients with primary ciliary dyskinesia. Electron microscopy may be used to analyse respiratory mucosa specimens.

Evaluation of cardiovascular anatomy by echocardiography requires careful evaluation of each cardiovascular segment. An electrocardiogram may help to assess atrial situs and conduction abnormalities. Cardiac catheterisation and magnetic resonance imaging may be necessary in some patients to completely delineate cardiac anatomy. Transoesophageal echocardiography (TEE) has also been described as a useful imaging modality to diagnose dextrocardia. TEE examination in situs inversus may be challenging and requires vigilance when differentiating morphologic and anatomic left/right structures.

Lung function test including lung volumes and blood gas analysis may be done to evaluate the grade of pulmonary involvement. Respiratory testing usually reveals little deviation from normal parameters.

Before anaesthesia, cardiology consultation is suggested. Physical examinations of the heart, such as percussion and palpation during routine exams, are important.

Fibre-optic bronchoscopy may be necessary for the evaluation of the bronchial tree for one-lung ventilation. The mirror image airway anatomy demands meticulous planning for the placement of lung separation devices.

The above mentioned diagnostic options should be performed in patients with suspected Situs inversus totalis or Kartagener Syndrome. In patients already diagnosed, the anaesthetist should have access at least to a contrast-enhanced CT scan and the echocardiography results, including pressure gradients. A pre-operative heart and lung ultrasound should be part of any elective procedure in patients with situs inversus.

Particular preparation for airway management

The association of situs inversus with other syndromes and diseases such as Kartagener syndrome, mucociliary dysfunction, airway anomalies (narrowing of trachea) may predispose the patient to numerous varieties of airway difficulties.

Airway anomalies are rarely seen. Anomalies of relevance include choanal atresia, micrognathia, cleft lip and palate, tracheo-oesophageal fistula, tracheal bronchus and aglossia/hypoglossia. When performing nasal intubation, difficult airway management is required due to nasal cavity narrowing by chronic sinusitis, if present. It is best to avoid nasal intubation due to narrowing of the nasal cavity by chronic sinusitis. Inspired gases should be humidified to avoid thick secretions. Lung transplant candidates have limited pulmonary reserve and may desaturate quickly, so airway management should be performed as expeditiously as possible. During intubation, left endobronchial placement of the endotracheal tube may occur since the left bronchus is more in line with the trachea.

Particular preparation for transfusion or administration of blood products

Requirement for blood products in patients with situs inversus totalis may be higher during high invasive cardiac or thoracic surgery.

Particular preparation for anticoagulation

Patients with situs inversus totalis and Kartagener syndrome may be predisposing factor for thromboembolism. Impaired mobility may suggest a higher risk of post-operative thrombosis.

Particular precautions for positioning, transportation and mobilisation

The right gastric place should be kept in mind in regard of positioning of the patients.

Interactions of chronic disease and anaesthesia medications

No special considerations.

Anaesthetic procedure

Successful management requires appropriate understanding of the special anatomy. Considering the mirrored position of the thoracic viscera is especially important when placing a central line. As opposed to patients with situs solitus, the left internal jugular vein provides the most direct access to the morphologic right atrium. In central venous catheter and pulmonary artery catheter placement, a left internal jugular vein approach may be more prudent. Ultrasound guidance should be utilised. Total pulmonary venous anomaly and right superior vena cava are reported to accompany situs inversus. Thus, the right internal jugular vein is a preferable site for central venous cannulation in such a case.

Premedication drugs that depress ventilation or ciliary activity should be avoided in situs inversus associated with Kartagener syndrome. Nasal airways and/or nasal intubation should be avoided in these patients due to sinusitis.

If the patient has a diagnosis of Kartagener syndrome, certain pre-operative pulmonary issues should be identified or addressed before proceeding with surgery. These patients are predisposed to multiple airway anomalies and infections. Pulmonary percussive therapy, bronchodilators, incentive spirometry, postural drainage, steroids, and antibiotics are beneficial to optimise the pulmonary system before surgery.

Induction with inhalational agents is slow in case of pulmonary hypertension, but faster with intravenous agents in right to left shunts.

The abnormalities related to the impaired motility of neutrophils require cautious aseptic techniques during anaesthesia. This includes placement of a needle and catheter insertion for epidural block.

Auscultation sites must be changed for confirmation of tracheal intubation. It should be kept in mind that the left lung will be ventilated better in case of over-advanced endotracheal tube, not the right one.

A case of prolonged paralysis after administration of succinylcholine has been reported in a patient with situs inversus totalis. This may show a possible link between situs inversus and atypical cholinesterase but routine testing for atypical cholinesterase is not recommended at this point.

The mirror-image anatomy of the bronchi makes lung isolation challenging. Bronchoscopic inspection of the bronchial anatomy through a large single-lumen endotracheal tube (ETT) before double-lumen ETT is advantageous. Use of a right-sided double lumen ETT in the left lung of the patient was reported to maintain the ventilation of the apical segment of the left lung. The use of a bronchial blocker can also be considered. Confirmation with a fibre-optic bronchoscope is a reliable option. In children, one-lung ventilation using a single lumen tracheal tube has also been reported in situs inversus.

In patients with Kartagener syndrome, humidification of gases should be added to the anaesthesia circuit and frequent suctioning of the endotracheal tube may be required.

During defibrillation, paddles should be placed in reverse position: i.e. on the upper left and lower right of the chest.

It is recommended to obtain informed consent for possible mini-tracheostomy prior to surgery from every patient with Kartagener syndrome.

Particular or additional monitoring

Pre-operative monitoring will be different because of dextrocardia. For each particular type of dextrocardia, the electric potentials may reflect differently, depending on the stereotypic position and situs type. The electrocardiogram lead placement should be reversed as a mirror image (left-sided limb leads and fifth precordial leads are placed on the right, and right-sided limb leads on the left). The sinus node is located to the left (mirrored) in case of situs inversus, so the P wave is inverted. This means that there is a negative P wave in leads I and aVL, and a positive P wave in aVR. QRS complex is negative in lead I, while the T wave is inverted if the leads are not reversed. Mirror positioning of the heart results in right-to-left septal depolarization, so Q waves will be present in the right precordial leads. Invasive monitoring may be useful for patients with cardiac disease.

Possible complications

Respiratory complications are the primary concern that the anaesthesiologist will face in case of association with Kartagener syndrome. The patient's pulmonary status should be optimised prior to the surgical procedure if general anaesthesia is planned. Antibiotic prophylaxis, bronchodilators, chest physiotherapy, steroids, postural drainage, and incentive spirometry may be required. Using disposable air equipment and performing smooth manipulations during intubation and extubation are required to avoid traumatising the mucosa. Bronchoconstriction can be avoided by using volatile anaesthetics. Corticosteroids may be useful prophylactically to prevent bronchospasm. The possibility of secretions blocking the endotracheal tube necessitates frequent endotracheal tube suctioning. Lung compliance and peak inspiratory pressures should be observed closely. Mucus plug obstructing the

endotracheal tube and causing desaturation was reported which required changing the tube. Attention should be paid to mucus that may obstruct the airways.

Situs inversus totalis patients are at risk for peri-operative cardiac insufficiency as well.

Post-operative care

Intensive care is recommended after cardiothoracic surgery with situs inversus totalis. High-risk patients should be observed in the intensive care unit for arrhythmias and cardiac ischaemia. In patients with situs inversus totalis and Kartagener syndrome, prolonged ventilation may occur.

Adequate analgesia should be provided for the patient. Inadequate analgesia may lead to respiratory insufficiency in presence of Kartagener syndrome. Pleuritic chest pain can occur up to 21% of patients associated with Kartagener syndrome. This kind of pain is usually seen in patients with bronchiectasis. Thoracic paravertebral block and electrical nerve stimulation has been successfully used in the management of intractable pleuritic chest pain. Improved ventilatory function is expected with relief of chest pain. Epidural analgesia should be considered in patients undergoing thoracotomy.

Disease-related acute problems and effect on anaesthesia and recovery

The diagnosis is usually found incidentally which can be occult in emergency cases. Patients should be evaluated carefully prior to surgery, especially paediatric patients, as they are more likely to present with hidden abnormalities. Informing the patient about his/her own situation and the patient informing the clinician will make the diagnosis easier. Documenting situs inversus totalis in a person is important after the signs of the disease occur. The absence of heart sounds on auscultation of the precordial area, confusion during the diagnosis of acute cholecystitis, acute appendicitis and splenic lesion on the basis of clinical examination raises the suspicion of situs inversus totalis. Radiologic evaluation, initially a chest X-ray, is suggested for diagnosis.

Ambulatory anaesthesia

Ambulatory anaesthesia (according to common guidelines) should only be done in situs inversus totalis patients with no cardiopulmonary symptoms and low risk surgery.

Obstetrical anaesthesia

Prevention of aorto-caval compression may be problematic. Due to the atypical location of the vena cava, it is difficult to determine whether left or right uterine displacement is needed. Uterine positioning is suggested to be to the right.

In obstetric patients with situs inversus and Kartagener syndrome requiring caesarean section, this can be successfully managed with spinal anaesthesia. In these cases, the CT scan of the spinal column, usually performed during the primary diagnostic roll-up, should be carefully examined.

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