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Kartagener syndrome

Kennedy disease

orphananesthesia

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

SUPPLEMENT NR. 10 | 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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Anaesthesia recommendations for

Kartagener syndrome

Disease name: Kartagener syndrome

ICD 10: Q89.3

Synonyms: sinusitis-bronchiectasis-situs inversus (triad) syndrome

Disease summary: Kartagener's syndrome (KS) is a rare autosomal recessive genetic disorder with a prevalence of 1:32,000, constituting about 50% of the primary ciliary dyskinesia (PCD) and characterised with a course including the triad of sinusitis, bronchiectasis and situs inversus. It was first described by Siewert in 1904, but Kartagener recognised the triad of disorders as a distinct congenital syndrome only in 1933. More than 35 gene mutations causing the disorder of ciliary morphology and function are known to be involved in PCD. Most areas of the upper airways including the nasal mucosa, paranasal sinuses, the middle ear, Eustachian tube and the pharynx, and of the lower airways, from trachea to down to the respiratory bronchioles, are lined with ciliated epithelium.

Dysfunction or lack of the dynein arms enabling ciliary motion (normally they are attached to the structural elements making up the cilium) results in the disruption of the coordinated ciliary movement and the propulsion of the mucus. Retention and accumulation of mucus leads to a variety of recurrent infections in the chest, ears, nose, throat and the sinuses. Also observed is male infertility due to immotile spermatozoa.

Typical symptoms of chronic sinusitis, bronchitis, bronchiectasis are severer in the first decade of life, moderating within the second decade. Severe cases of KS could be fatal unless lung transplantation is carried out. A small percentage of the KS patients present with hydrocephalus. The ependymal cells in the lining of the brain ventricles involved in CSF production are also ciliated. Impaired ciliary function may involve prevention of CSF reabsorption resulting in development of communicating hydrocephalus that causes chronic headache. Situs inversus is the congenital condition in which the major intrathoracic and/or intraabdominal organs are reversed or mirrored from their normal positions, possibly resulting from lack of ciliary control of the organ positioning in the embryo with primary ciliary dyskinesia. Normally, a cilium has a rotary motion which drives a vectorial movement, which accounts for laterality of organ lateralisation during embryogenesis. Organ lateralisation will be random if the ciliary function is absent. Incomplete/complete situs inversus is seen in about a half of the PCD syndrome cases.

During the embryonic stage, ciliary dyskinesia may lead to other organ anomalies such as biliary atresia, intestinal malrotation, asplenia, and polysplenia. Although frequent development of upper and lower airway infections after birth, comorbidity of situs inversus and a family history may suggest KS, but the definitive diagnosis depends on ciliary ultrastructural analysis or molecular genetic testing. There is no "gold standard" diagnostic test for PCD. Electron microscopy (EM) has been the traditional test used to confirm a diagnosis of PCD; however, it cannot be used to diagnose patients with PCD (15-20%) with

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normal ultrastructure. Currently, not all gene mutations that cause PCD have yet been discovered. fluorescence analysis is said to be highly specific for PCD, but its sensitivity is currently limited. Ciliary beat pattern (CBP) and frequency (CBF) measurement have been recommended as a first-line diagnostic test for PCD.

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

Adeno-tonsillectomies, tympanostomy tube insertions, sinus surgery, nasal polypectomy and pulmonary surgery due to bronchiectasis, infertility investigations, cardiac surgery have been cited in the literature.

Type of anaesthesia

On account of the comorbid respiratory disorders, local or regional anaesthesia is the best choice, whenever feasible, as compared to general anaesthesia.

In order to optimise the respiratory functions prior to surgery, respiratory physiotherapy, postural drainage to reduce the secretions, bronchodilator treatment to prevent airway reactivity, and antibiotic therapy to treat or prevent upper and lower respiratory tract infections are recommended.

In cases of KS with congenital heart disease, comorbidity, endocarditis prophylaxis and measures to prevent air embolism should be performed along with its treatment.

Central or peripheral nerve blocks should be preferred with the aim to minimise the need for opioids, which bring about respiratory depression.

Necessary additional pre-operative testing (beside standard care)

Lung function tests: FEV1 (forced expiratory volume at 1 second), FVC (forced vital capacity), FEV1/FVC, total lung capacity, diffusing capacity of lung for carbon monoxide (DLCO) should be determined when a normal or obstructive pattern emerges. Chest radiography is needed to exclude pneumonia and atelectasis. Determination of the arterial blood gases may be useful for evaluating respiratory function.

Cardiac, thoracic and abdominal anatomies have to be determined pre-operatively. Situs inversus can be diagnosed with radiography (Figure 1). In cases with comorbid congenital cardiac disorders, cardiac anatomy and functions must be evaluated by taking history, physical examination, ECG and echocardiography. The ECG is the mirror image of the normal due to dextrocardia (Figure 2). Also, chronic bronchiectasis may have progressed to cor pulmonale.

Particular preparation for airway management

KS does not have any direct correlation with difficult tracheal intubation. Nasotracheal intubation is relatively contraindicated because of the possibilities of chronic sinusitis, nasal polyp and chronic otitis media.

Particular preparation for transfusion or administration of blood products

In severe cases of KS in lung transplantation candidates , transfusion should be avoided to reduce the risk of any antibody reaction.

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Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation therapy.

Particular precautions for positioning, transportation and mobilisation

Not reported.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

Whereas bronchodilators, steroids, antisialagogue medication and short-acting opioids should be preferred in the premedication, drugs that suppress the cough reflex, respiration and ciliary activity should be avoided.

Attention should be paid to cardiac and respiratory anatomy. The ECG and the defibrillator electrodes should be placed in the reverse order for situs inversus. If the endotracheal tube is far extended, it often progresses into the left main bronchus. Attention has to be paid to the anatomic position of the bronchi when the double-lumen endobronchial tube is to be used. The normally used left endobronchial tube should be turned and used on the right side and vice versa. Similarly, normally right internal jugular vein catheterisation should be regarded as left jugular vein catheterisation and as an anatomical landmark. Procedural difficulties arising from anatomical differences can be overcome if central venous catheterisation and peripheral blockade procedures are guided by ultrasonography, and double-lumen endobronchial intubation is controlled by fibre-optic bronchoscopy.

Nasotracheal intubation is relatively contraindicated because of the probability of chronic sinusitis, nasal polyp and chronic otitis media.

After intubation, difficult ventilation due to secretions can lead to hypoxaemia. Hydration of the inspiratory gases would reduce the viscosity of the secretions and ease their cleaning up. Anticholinergic agents may decrease the amount of pulmonary secretions. Intraoperatively, frequent suctioning of the trachea may be required.

During induction and maintenance of general anaesthesia, volatile anaesthetic agents are useful because of drug-induced bronchodilatation and rapid elimination, thus reducing the respiratory depression in the early post-operative period. For minimising the post-operative respiratory depression, short-acting opioids during the intraoperative period should be preferred, and central or peripheral nerve blocks are recommended.

KS patients may have abnormal neutrophil chemotaxis and low levels of IgA. Even if incidences of infections other than those of the respiratory system are not increased, antisepsis during any intervention, mainly for epidural anaesthesia, central venous cathetherication and endobronchial tube aspiration, should be carefully maintained.

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When reversing the neuromuscular block, the use of sugammadex may be better for the patients instead of cholinesterase inhibitors such as neostigmine, which increase the secretions.

Particular or additional monitoring

Particular attention should be paid to major intraoperative complications such as airway obstruction by secretions, bronchial hyperresponsiveness, hypoxaemia and hypercarbia.

Possible complications

Not reported.

Post-operative care

Respiratory physiotherapy, which eases the elimination of the secretions, is required in the post-operative period. If the requirement of mechanical ventilation continues in the post-operative period, the inspired gases should be hydrated and systemic hydration should be maintained.

If the KS patients with severe respiratory or cardiovascular problems have to undergo general anaesthesia during surgery, they must be placed in an intensive care unit post-operatively for close observation and treatment.

Disease-related acute problems and effect on anaesthesia and recovery

Not reported.

Ambulatory anaesthesia

Decision for ambulatory anaesthesia depends on the severity of the disease and the surgical procedure to be performed. Ambulatory anaesthesia can be considered if the respiratory condition of the patient is good and the surgical procedure is expected to take a short time, especially if it is to be without depressive effects on the respiratory system or if surgery is compatible with regional anaesthesia.

Obstetrical anaesthesia

Depending on the general condition of the patient, regional anaesthesia may be preferred.

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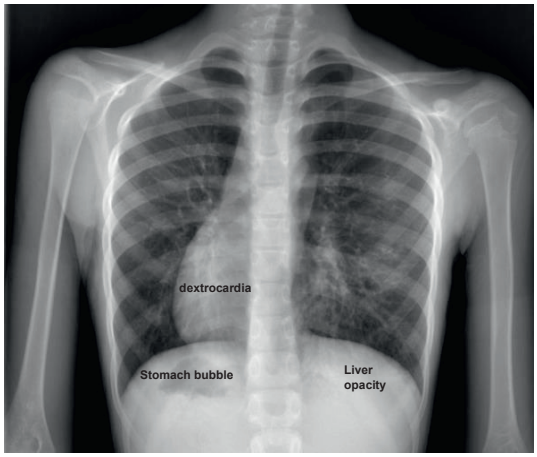

Figure 1. *Chest X-ray*

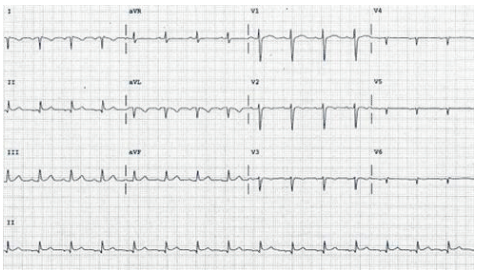

Figure 2. *Atrial depolarisation is initiated by a left sinus node, so P waves are inverted in leads I and aVL and upright in lead aVR. Reversed ventricular activation and reversed repolarisation. Lead I QRS negative and the T wave inverted, lead aVR resembles aVL and vice versa, right precordial leads resemble leads from corresponding left precordial sites. Septal Q waves appear in right precordial leads because septal depolarisation is from right to left. (The ECG can be "corrected" by reversing the limb leads and recording chest leads from the right precordium.)*

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