

## Anaesthesia recommendations for patients suffering from

### **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 dyskinesias (PCD) and characterized with a course including the triad of sinusitis, bronchiectasis and situs inversus. It was first described by Siewert in 1904, but Kartagener recognized in 1933 the triad of disorders as a distinct congenital syndrome. More than 35 genes mutations that causing in disorder of ciliary morphology and function, are known to cause 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.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

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## **Disease summary**

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Dysfunction or lack of the dynein arms enabling ciliary motion, normally 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 transplant 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 cilium has a rotary motion, which drives a vectorial movement, therefore, laterality of organ lateralization during embryogenesis. Organ lateralization is 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 a diagnosis of PCD however that cannot be used to diagnose of patients with PCD (15-20%) with normal ultrastructure. Currently, all genes mutations that cause to PCD, have not been discovered yet. Immunofloresance analysis to be highly specific for PCD but sensitivity is currently limited. Ciliary beat pattern (CBP) and frequency (CBF) measurement has been recommended as a first-line diagnostic test for PCD.

## **Typical surgery**

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Adeno-tonsillectomies, tympanostomy tube insertions, sinus surgery, nasal polypectomy and pulmonary surgery due to bronchiectasis, infertility investigations, cardiac surgery are of the cited surgeries performed.

## **Type of anaesthesia**

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On account of the comorbid respiratory disorders, local or regional anaesthesia are the best choice whenever feasible as against 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 to prevent upper and lower respiratory tract infections are recommended.

In 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.

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### **Necessary additional diagnostic procedures (preoperative)**

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Lung function tests: FEV1 (Forced expiratory volume at 1 second), FVC (Forced vital capacity), FEV1/FVC, total lung capacity, diffusing capacity of lung for carbonmonoxide (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 functioning.

Cardiac, thoracic and abdominal anatomies have to be determined preoperatively. 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.

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### **Particular preparation for airway management**

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

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### **Particular preparation for transfusion or administration of blood products**

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

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### **Particular preparation for anticoagulation**

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

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### **Particular precautions for positioning, transport or mobilisation**

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Not reported.

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

Not reported.

## **Anaesthesiologic procedure**

Whereas bronchodilators, steroids, antispasmodic 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 for situs inversus. If the endotracheal tube is extended far, 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 and vice versa. Similarly, the normally right internal jugular vein catheterization should be regarded as the left jugular vein catheterization 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 fiberoptic 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 hypoxemia. Hydration of the inspiratory gasses 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 anesthesia with the volatile anesthetic agents is useful because of drug-induced bronchodilatation and rapid elimination thereby reducing the respiratory depression in the early postoperative period. For minimizing the postoperative respiratory depression, preference of short-acting opioid use during the intraoperative period, and making 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 catheterization, and endobronchial tube aspiration, should be carefully maintained.

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

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### **Particular or additional monitoring**

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

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### **Possible complications**

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Not reported.

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### **Postoperative care**

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Respiratory physiotherapy, which eases the elimination of the secretions, is required in the postoperative period. If the requirement of mechanical ventilation continues in the postoperative 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 go under general anaesthesia during surgery, they must be placed under intensive-care unit postoperatively for close observation and treatment.

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### **Information about emergency-like situations / Differential diagnostics**

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*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.:*

Not reported.

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### **Ambulatory anaesthesia**

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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 and if especially to be without depressive effects on the respiratory system, or if the surgery is compatible with regional anaesthesia.

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### **Obstetrical anaesthesia**

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

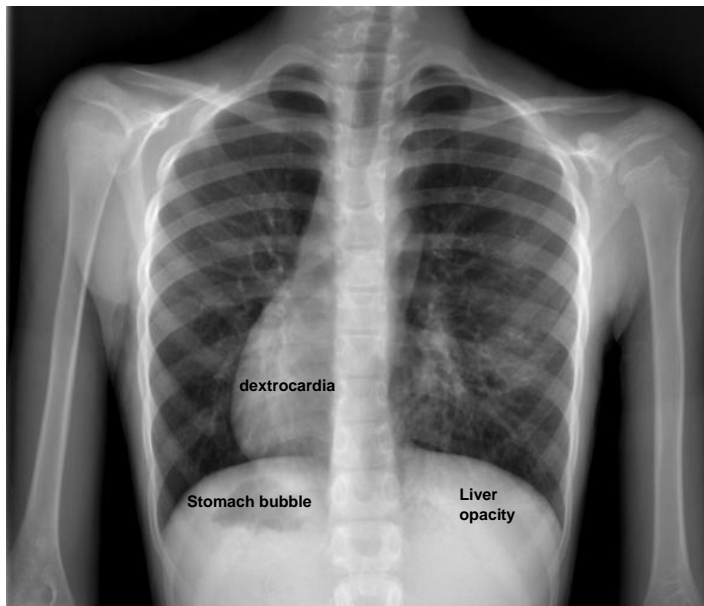


Figure 1. Chest X-ray

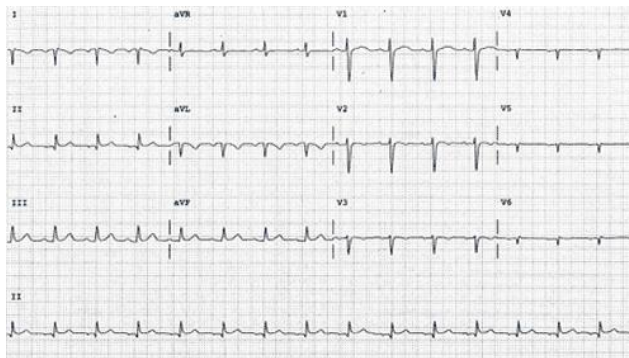


Figure 2. Atrial depolarization is initiated by a left sinus node, so P waves are inverted in leads I and aVL and upright in lead aVR. Reversed ventricular activation and reversed repolarization. Lead I QRS negative and the T wave inverted, lead aVR resembles aVL and vice versa, right precordial leads resemble leads from corresponding left precordial sites. Septal Q waves appear in right precordial leads because septal depolarization 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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**Peer revision 1**

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**Peer revision 2**

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