orphanaesthesia

Anaesthesia recommendations for patients suffering from

Cystic fibrosis

Disease name: Cystic fibrosis

ICD 10: E84.0

Synonyms: Mucoviscidosis

Cystic fibrosis (CF) is an autosomal recessive, systemic disorder that is primarily characterized by chronic pulmonary infections, bronchiectasis, exocrine pancreatic insufficiency and elevated concentration of both sodium and chloride of the sweat.

In about 70% of cases, the disease is caused by deletion of the codon for phenylalanine at position 508 on chromosome 7, commonly referred to as DeltaF508. This leads to a defective biosynthesis of the cystic fibrosis transmembrane conductance regulator (CFTR) protein at the apical side of epithelial cells of most exocrine glands. Consequently, abnormal exchange of chloride, sodium and water generates the accumulation of viscous mucus secretions in the pancreas, small intestine, bronchial tree, biliary tract, and gonads. In addition, there may be an excessive loss of both sodium and chloride through sweat glands [1,2].

Within European derived populations, CF is generally estimated to have an annual incidence of 1 in 2,000-3,000 Caucasian newborns [3,4].This makes CF the most common inherited metabolic disease among Caucasians. Advances in treatment methods have led to increased average life expectancy of approximately 40 years. This is accompanied by a steady increase in the prevalence of these patients [5]. Undoubtedly, this epidemiological shift leads to increased contacts of CF-diseased patients at non-specialized centres [6].

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

Typical surgery

- Nasal polypectomy
- ENT procedures
- Investigation or treatment of gastrointestinal disorders
- Feeding tube (PEG) placement
- Bronchoscopy, pulmonary lavage
- Gastrointestinal endoscopies
- Sclerotherapy of oesophageal varices in portal hypertension
- Lung transplant

Type of anaesthesia

See “Anaesthesiologic procedure”.

Necessary additional diagnostic procedures (preoperative)

Due to their long-term commitment to specialized centres, CF patients have a pronounced understanding of their disease. The affected patients and their parents show a significantly increased rate of depressive symptoms (children: 29%, mothers 35%, fathers: 23%) [7]. Respecting their experiences and concerns is an important factor to gain confidence from these patients [8].

Medical history concerning diagnosis and disease progression should emphasize respiratory manifestations, pulmonary function and the exercise tolerance test. The investigation of cough, quality and quantity of mucus production, respiratory infections and bronchial hyper-reactivity are essential components of preoperative evaluation [8]. The physical activity level of the patient, as well as the number of previous hospitalizations, may serve as prognostic markers and may indicate an increased risk of perioperative pulmonary complications [9,10]. Contacting the patient’s pneumologist should be considered during preoperative evaluation.

In emergency situations, the essential information should center around the time course of pulmonary manifestations, current physical stress tolerance, recent hospitalizations, lung infections and antibiotic history. For the intra- and postoperative treatment, further questions focussing on diabetes mellitus, exocrine pancreatic insufficiency and liver diseases are recommended [5].

Diagnostic Radiology: As a consequence of CF (e. g. bronchiectasis), obstructive pulmonary disease with flattening of the diaphragm and a prominent retrosternal space may frequently appear on the chest x-ray [11]. Computed tomography can help gain insight to the extent of bronchiectasis. However, this does not always correlate with actual exercise capacity [12].

The findings on radiology are therefore appropriate to determine the amount of "air trapping" and the description of disease progression. Nevertheless, further studies to measure lung function are required to plan a stage-adjusted anaesthetic management [5].

Spirometry: The typical picture of the classic CF phenotype is an obstructive airway disease on the basis of bronchiectasis. Accordingly, changes in FEV (1) may indicate the disease severity and progression. Average reduction of FEV (25-75) in young patients before the preschool years is 7.5% (CI 0.9 to 13.6) [13]. A proceeding course of the disease may be characterized by a further decrease in dynamic lung parameters [5].

Other complications such as expiratory wheezing, cough and infections with Pseudomonas aeruginosa can independently provoke an additional reduction in pulmonary function. Also, elevated liver function tests, pancreatic insufficiency, female gender, and lower body weight are associated with a reduced FEV (1) [13,14].

Blood gas analysis: A highly reduced ventilation-perfusion ratio can elicit partial to global respiratory failure. This may result in an increased pulmonary resistance, right ventricular hypertrophy and the development of cor pulmonale with right heart failure.

Blood gas analysis (ABG) is recommended to avoid postoperative complications in suspected cases with advanced or decompensated course of CF. ABG should be particularly considered in patients with home mechanical ventilation. However, ABG might be omitted in patients with mild or stable respiratory disease [5].

Transthoracic cardiac echography: Pulmonary hypertension develops in a significant proportion of patients with CF and is strongly correlated with hypoxemia, independent of pulmonary function [15].Transthoracic cardiac echography can be used to look for signs of pulmonary hypertension, such as cardiac hypertrophy or dilatation of the right ventricle.

Liver function tests: Patients with Cystic Fibrosis Liver Disease (CFLD) have a more severe CF phenotype than do CF patients without liver disease.16 CFLD may advance to progressive biliary fibrosis and cirrhosis with coagulopathy, altered drug metabolism and a decrease in FEV (1). Elective cases with an > 1.5 fold increase of liver enzyme levels should be considered for further examinations such as ERCP.

Particular preparation for airway management

During the last 50 years, the specific morbidity and mortality rate of CF patients has been dramatically reduced by modern techniques, now reaching the levels of healthy subjects [17-19]. Although pulmonary complications of CF patients remain problematic during anaesthesia, the anaesthetic risk has been minimized even for complex interventions [20].

Especially in young CF patients the severity of disease frequently correlates with bronchial hyper-responsiveness. In addition, infections of the sinuses are known to occur in a majority of patients with CF. These infections are considered potential triggers for bronchospasms because of the post-nasal secretions that often accompany sinus infection [21]. Beta-agonists are usually sufficient to treat these bronchospasms. Accordingly, these drugs are recommended for patients with significant airway obstruction before induction-, during maintenance- and before reversal of anaesthesia [5]. However, relaxation of the respiratory muscles might contribute to paradoxical airway obstruction in about 10-20% of CF patients. In these patients the damaged bronchiectactic airways may require airway smooth muscle tone for patency [22,23].

Due to the high incidence of nasal polyps, nasopharyngeal airways should be avoided.24

The distinct production of bronchial secretions may cause increased airway resistance during mechanical ventilation and consequently enhance the risk for ventilation-induced pulmonary barotrauma [25]. Preoperative respiratory physiotherapy may help to reduce secretions and to maintain adequate oxygenation and ventilation during induction, and just after intubation. If this is not sufficient, frequent segmental or subsegmental bronchoalveolar lavage may be beneficial [5].

Particular preparation for transfusion or administration of blood products

Many patients with cystic fibrosis are considered candidates for lung transplantation [26]. Therefore, transfusion should be avoided to reduce the risk of any antibody reaction and production before a potential transplantation.

Particular preparation for anticoagulation

See “Necessary additional diagnostic procedures (preoperative)”, “Liver function tests”.

Particular precautions for positioning, transport or mobilisation

CF patients are often underweight. Special care should be taken on compression points for long surgical procedures.

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

Not reported.

Anaesthesiologic procedure

The anaesthetic approach to CF-patients should be performed with special attention to achieving minimal respiratory depression and optimal recovery of pulmonary function at the end of surgery [8]. Special attention should be paid to the postoperative pain therapy. The potential risk of opioid-induced respiratory depression needs to be balanced against the risk of constrained spontaneous ventilation as a consequence of insufficient analgesia. Amongst other therapeutic strategies, the perioperative infusion of i. v. lidocaine, the administration of ketamine or application of NSAIDs might be appropriate to reduce postoperative opioid consumption.

To avoid the risks of mechanical ventilation, regional- or neuroaxial methods are adequate alternatives for general anaesthesia. Due to their long-lasting analgesic effects, these techniques not only reduce the postoperative need for opioids [27], but also prevent airway manipulation. Thereby they also reduce the rate of subsequent complications such as respiratory depression and pulmonary infections [28-32]. In case of unavoidable general

anaesthesia, CF patients may benefit from the bronchodilatative effects of volatile anaesthetics, e. g. Sevoflurane [33].

Volume deficiency may trigger the collapse of mucous membranes in CF patients because of their limited capacity for bronchiolar lubrication. Accordingly, this makes adequate fluid resuscitation essential to minimize the risk of mucosal obstruction of the lower airways [34,35].

To avoid residual blockade after neuromuscular relaxation, a train-of-four fade ratio of more than 0.90 should be aimed [36].

Pulmonary recruitment manoeuvres and physical procedures can be performed preceding the end of anaesthesia. The spontaneously breathing patient should be extubated as soon as possible in waist-high position [37].

Particular or additional monitoring

The perioperative monitoring is based on the aforementioned requirements for airway management and anaesthesia, as well as on the current guidelines of professional societies.

Particular attention should be paid to major intraoperative complications such as airway obstruction by viscous secretions, bronchial hyper responsiveness and hypoxemia. Furthermore, altered drug metabolisms due to decreased pseudocholinesterase activity as well as the possible lack of clotting factors might be considered in cases of hepatic dysfunction.

Ketoacidosis and symptomatic hyperglycaemia are rarely observed in CF-affected patients. However, anaesthesia and surgery induce perioperative stress and therefore potentially affect blood glucose levels. Accordingly, glucose should be closely monitored every 2 hours approximately and maintained at levels below 180 mg / dl.38,39.

Possible complications

Not reported.

Postoperative care

Postoperative pain may constrain respiratory function especially after abdominal- and thoracic surgery.40 Affected patients might have a decreased tidal volume, an insufficient cough and an increased susceptibility to postoperative respiratory complications [41]. Multimodal pain concepts are beneficial in postoperative pain management, and these concepts are demonstrated to result in accelerated recovery and shortened hospital stay in CF patients [42,43]. In addition, anxiety is often seen in the postoperative recovery area and can be managed with explanations, hypnosis or medication.

Non-invasive ventilation can be of great help during the immediate postoperative period and should be available. Patients with home mechanical ventilation should be granted immediate access to their personal devices.

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease

Not reported.

Ambulatory anaesthesia

Ambulatory anaesthesia should be discussed with the pneumologist and the patients. This option depends on the surgical procedure, their respiratory status and the distance between the hospital and the patient's home.

Obstetrical anaesthesia

Not reported.

Literature and internet links

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