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Kearns-Sayre syndrome

Kikuchi-Fujimoto disease

orphananesthesia

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

SUPPLEMENT NR. 9 | 2017

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 common 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.

A survey of until now in A&I published guidelines can be found on:

www.ai-online.info/Orphsuppl
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Anaesthesia recommendations for patients suffering from

Kikuchi-Fujimoto disease

Disease name: Kikuchi-Fujimoto disease

ICD 10: I88.1 Lymphadenitis cervical, non specified site, chronic or subacute

Synonyms: Histiocytic necrotizing lymphadenitis, Kikuchi disease, Kikuchi-Fujimoto disease, Kikuchi lymphadenitis, lymphadenopathy, KFD

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

▶ **Citation:** Rossetti E, Appierto L, Bianchi R, Picardo S: Kikuchi-Fujimoto disease. Anästh Intensivmed 2017;58:S482-489. DOI: 10.19224/ai2017.S482

Disease summary

KFD is a rare lymphohistiocytic disorder due to cervical inflammatory lymphadenitis (or axillary region and even other location seldom) with an unknown etiopathogenesis, that is most commonly seen in young Asiatic people (male/female=1:1, despite some data suggest it is more frequent in women). Some HLA class II genes are more frequent in patients with KFD. In particular, the incidence of DPA1*01 and DPB1*0202 alleles is significantly higher in patients with KFD than in healthy control subjects. It is mainly characterized by lymphadenopathy, fever, nocturnal sweats, myalgia, weight loss, and arthralgia, and commonly follows a self-limited course. Less frequently symptoms observed include cutaneous lesions, hepatosplenomegaly, central nervous system impairment and haemophagocytic syndrome. The laboratory and radiologic tests available for the diagnosis are nonspecific. The common laboratory abnormalities are leukopenia, usually neutropenia; anaemia; thrombocytopenia; elevated C-reactive protein and erythrocyte sedimentation rate; impaired liver function; and atypical lymphocytes on peripheral blood smear.

This disease is misdiagnosed as malignant lymphoma in up to one-third of cases and is associated with the development of systemic lupus erythematosus (SLE) and other autoimmune diseases.The differential diagnosis is challenging as many other conditions such as malignant lymphoma, metastatic disease, tuberculosis and infectious lymphadenopathies can present in a similar way. KFD is considered a self-limiting disease: spontaneous regression may occur between 1–6 months (4). In more severe cases, nonsteroidal anti-inflammatory drugs (NSAIDs) and/or steroids treatment has proven beneficial.

Typical surgery

Lymph node excissional biopsy, bone marrow aspirate and biopsy, short term central venous catheter positioning, liver biopsy, upper airway endoscopy or urgent tracheostomy.

Ultrasonographically guided biopsy is a profitable strategy, because it allows select the lymph nodes with less necrotic tissue to reach a histological diagnosis.

Other incidental surgeries apart from disease or for diagnosis may also be required in such children.

Type of anaesthesia

There is no definite recommendation for either general or regional anaesthesia. A moderate sedation, associated with local or regional anaesthesia, may avoid difficult upper airway management due to cervical lymphadenopathy. Spontaneous breathing or CPAP-assisted ventilation are suggested.

In order to limit airway management, any neuromuscular blocker should be avoided, as much as deep sedation or general anaesthesia.

To date, a role of anaesthetic drugs as trigger factor for KFD has not been reported in literature.

Necessary additional diagnostic procedures (preoperative)

Cardiac function tests (electrocardiography, echocardiography) according to general conditions (prolonged fever, malnutrition, dehydration, and severity of phlogosis indices' alteration).

Blood examinations, enlarged metabolic or coagulation tests.

BNP blood level is useful to monitor cardiac failure, if suspected.

X-ray of the thorax, lung ultrasound, and a CT scan to rule out other pathologies, define the extent of the lesion and to locate the most accessible adenopathy for the biopsy.

Consultation of a specialist to document for juridical reasons already existent deficits, e.g. of neurological nature.

Particular preparation for airway management

Patients with significant cervical lymphadenopathy could benefit from steroids therapies before and after surgery.

Since cervical lymphadenopathy of KFD has often been important, it requires to be trained on 'cannot intubate/cannot ventilate' protocol and ready with supraglottic devices, video laryngoscopy, airway endoscopy device, and cricothyrotomy set.

Consultation of an ENT specialist for more detailed examination if required.

Planning of a safe clinical pathway for induction of anaesthesia if airway management cannot be avoided.

Particular preparation for transfusion or administration of blood products

There are no particular recommendations for blood products or transfusion administration.

Particular preparation for anticoagulation

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

Particular precautions for positioning, transport or mobilisation

Not reported.

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

Not reported.

Anaesthesiologic procedure

Induction of anaesthesia either administering endovenous or inhaling volatile anaesthetics are allowed. In case of serious upper airway narrowing due to cervical lymphadenopathy and expected airway management, an ENT specialist support is required. Therefore, a non-invasive airway management is supposed as much as possible.

Perform steroid replacement therapy in anaesthesia induction where appropriate.

Particular or additional monitoring

No particular monitoring is required.

In case of KFD with neurological involvement, BIS monitor or intraoperative EEG monitoring may avoid or prevent neurologic state worsening.

Possible complications

Upper airway swelling and obstruction related to difficult airway management for severe cervical lymphadenopathy.

Rare complications:

- Cardiac complications such as tamponade.

- Interstitial lung disease and pleural effusion.

- Hepatitis is another rare complication.

- Haemophagocytic syndrome.

Postoperative care

Possible ICU admission for postoperative monitoring or weaning of mechanical ventilation in expected difficult airway management.

Information about emergency-like situations / Differential diagnostics

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

Related to actual upper airway obstruction.

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Ambulatory anaesthesia

As described above for type of anaesthesia.

Obstetrical anaesthesia

As described above for type of anaesthesia.

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