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Kleefstra Syndrome

Leprosy

orphanaesthesia

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

SUPPLEMENT NR. 20 | 2020

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

Leprosy

Disease name: Leprosy

ICD 10: A30

Synonyms: –

Disease summary: Leprosy is a chronic infectious disease caused by Mycobacterium leprae and affects the skin and nerves, often seen in developing countries. The prevalence of leprosy is 5.7 per 10,000 population. There are two forms included, tuberculoid and lepromatous leprosy. Pathway of the infectious disease is mainly through nasal droplet infection, contact with infected soil or insect vectors. Leprosy primarily affects the skin and the peripheral nerves, especially the mucosa, the upper respiratory tract, subcutaneous parts of the nerves and the eye. Neuropathy causes insensitiveness and myopathy such as plantar ulceration, foot drop and joint deformities. The autonomic nervous system, cardiovascular system, respiratory system, hepatobiliary system, and renal system are also affected. These manifestations lead to important complications such as baroreflex dysfunction, respiratory dysautonomia, leprous hepatitis, orchitis, glomerulonephritis, amyloidosis. With early diagnosis followed by an appropriate treatment with rifampicin, dapsone and, in case of lepromatous leprosy, additionally with clofazimine, patients can be cured without further disabilities.

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

Orthopaedic surgery, Caesarean section, nephrectomy, ophthalmic surgery, cataract, emergency surgical procedure.

Type of anaesthesia

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

Spinal and epidural anaesthesia management needs carefulness in patients with leprosy because of an increased risk of hypotension and an increased incidence of urinary retention. Neurologic deficits can be encountered before and after nerve blocks or regional anaesthesia as well. Regional anaesthesia is controversial in the case of bacteraemia and local infection. Aseptic meningitis has been reported as a complication after regional anaesthesia for elective Caesarean section. Combined spinal/epidural anaesthesia application for an emergency Caesarean section can be preferred in a patient with lepromatous leprosy.

Necessary additional pre-operative testing (beside standard care)

The following organ systems should be examined before surgery and anaesthesia:

- Cardiovascular system: disordered autonomic nervous function, impaired myocardial contractility and myocardial ischaemia. These can lead to cardiac arrest, hypo-reactive heart rate, dysrhythmias and sudden death during intubation, extubation and after the administration of various drugs. Therefore, ECG (arrhythmias and increased QT-intervals) and echocardiography are recommended.
- Neurological system: assessment of neurological status before regional anaesthesia.
- Respiratory system: increased risk of infection, aspiration, difficult intubation and delayed post-operative recovery. Pulmonary function tests, careful airway examination and aspiration prophylaxis are recommended.
- Renal system: amyloidosis, glomerulonephritis or interstitial nephritis may occur. Renal function should be assessed.
- Hepatobiliary system: leprous or drug-induced hepatitis can lead to altered metabolism of drugs. Transaminases should be determined.
- Haematological system: anaemia, methaemoglobinaemia, agranulocytosis and thrombocytopenia can be seen. Oxygen carrying capacity and clotting disorders can occur, risk of postoperative infection is increased. Haemogram and coagulogram should be checked.
- Skeletal system: osteomyelitis, bone resorption may develop. Radiological assessment can be useful.

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Particular preparation for airway management

There is a risk of aspiration and difficult intubation due to respiratory dysautonomia, nasal obstruction, vocal cord disorder, osteomyelitis and bone resorption especially in the cranio-facial area. Detailed airway assessment, indirect laryngoscopy and prophylaxis for aspiration are useful strategies.

Management of difficult airway is mandatory.

Particular preparation for transfusion or administration of blood products

There is some significant evidence for thrombocytopenia, agranulocytosis, anaemia and methaemoglobinaemia.

It has been reported that changes in the intraneural blood vessels can be found with impairment of the basement membrane of capillaries and oedema of vessel walls, which will result in occlusion of their lumina, potentially causing ischaemia to nerves.

Particular preparation for anticoagulation

There is some evidence for impaired clotting. Alterations in the activated partial thromboplastin time (APTT) have been observed [3,5].

Particular precautions for positioning, transportation and mobilisation

Patients with leprosy suffer from bone resorption and osteomyelitis. Pathological fractures during positioning are a risk.

Interactions of chronic disease and anaesthesia medications

For the therapy of lepromatous leprosy, a multi-drug therapy is recommended by WHO including dapsone, rifampicin and clofazimine for more than 30 years.

Dapsone, a folate antagonist, has important adverse effects including haemolytic anaemia, methaemoglobinaemia, agranulocytosis, hepatitis, peripheral neuropathy, psychosis and lepra reaction.

Rifampicin may be prone to following adverse effects; hepatitis and intermittent toxic syndromes such as flu syndrome, shock syndrome, and rarely, thrombocytopenic purpura and acute renal failure.

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Anaesthetic procedure

Leprosy is a highly infectious disease of low pathogenicity. The nasal mucosa of lepromatous cases harbour millions of M. leprae which are discharged during sneezing. The bacteria can also exit through ulcerated or broken skin of infected patients. These facts may leave implications for the anaesthesiologists when such patients come for surgery or for treatment in intensive care units. Fortunately, it has been found that the local application of rifampicin drops or spray destroys most of the bacteria within a short period.

A careful physical exam and organ function tests should be performed, especially in patients with long-lasting disease, but in some cases, organ manifestations already occur in beginning forms. Special attention should be given to the assessment of difficult airway situations.

General and regional anaesthesia can be performed. During general anaesthesia, an impaired metabolism of the drugs applied should be considered, e.g. prolonged effects of the hypnotic and analgesic drugs. Before regional anaesthesia, the neurological status and grade of sexual impotence should be recorded.

Spinal and epidural anaesthesia should be performed cautiously, because they may lead to profound hypotension and urinary retention in patients with an involvement of the autonomic nervous system.

Patients with leprosy have cardiac dysautonomia with impaired blood pressure responses, silent cardiac ischaemia and prolongation of the QT interval. These can lead to bradycardia, hypotension and cardiac arrest or various other arrhythmias. Especially anaesthesia induction, extubation or the use of anticholinergic drugs indicate special attention.

Avoid drugs which lead to prolongation of QT interval, like paracetamol or setrons.

Particular or additional monitoring

No special monitoring is necessary. Standard monitoring as ECG, SPO2, etCO2, temperature, urine output should be done.

Invasive blood pressure measurement is useful in case of high-risk surgery.

Possible complications

Patients with leprosy are at risk for respiratory and cardiac insufficiency.

Drug metabolism may be affected due to renal function disorders or liver dysfunction. Hypotension, arrhythmia and bradycardia may be seen.

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Post-operative care

Post-operative monitoring depends on the surgical procedure and the pre-operative condition of the patient. Intensive care is not mandatory, but should be available in certain conditions.

Respiratory dysautonomia leads to a decreased breath-holding time, depressed cough reflex and the risk of aspiration that may increase the incidence of post-operative complications or need for post-operative ventilation.

Avoid prolonged ventilation because the patients might have respiratory dysautonomia and an increased risk of pulmonary infection.

Disease-related acute problems and effect on anaesthesia and recovery

Disease-triggered emergency-like situations are not common.

Ambulatory anaesthesia

Not reported. Avoid ambulatory anaesthesia in patients with impaired autonomic nervous system, cardiovascular system, respiratory system, hepatobiliary system or renal system.

Obstetrical anaesthesia

The first clinical presentation of leprosy or aggravation of the existing disease can be seen in pregnancy.

There is one case report in which an emergency Caesarean section has been performed with spinal anaesthesia.

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