

A&I

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Lennox-Gastaut syndrome

Low syndrome

orphan **a**nesthesia

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

SUPPLEMENT NR. 10 | 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.

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 recommendations 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.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

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

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orphananesthesia

Anaesthesia recommendations for patients suffering from **Lennox-Gastaut syndrome**

Disease name: Lennox-Gastaut syndrome

ICD 10: G40.4

Synonyms: Not reported.

Lennox-Gastaut syndrome (LGS) is a severe form of epilepsy associated with intractable seizures and impaired cognition. On inter-ictal electroencephalography (EEG), LGS is characterized by generalized epileptiform discharges including slow (1.5-2.5 Hz) spike-and-wave complexes and bursts of paroxysmal fast activity. Onset typically occurs between the ages of 2 and 8 and affects predominantly males. Aetiologies underlying LGS vary widely across patients, and include both genetic and acquired causes (e.g., diffuse brain damage following asphyxia, tuberous sclerosis, encephalitis or metabolic disorders). Prevalence varies between 1-10% of all childhood epilepsy. In 15-20% of patients, the cause of LGS remains unknown. Around 20% of patients develop LGS following infantile West syndrome.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

► **Citation:** Fischer A: Lennox-Gastaut syndrome. AnästH Intensivmed 2017;58:S491-S496.
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Disease summary

Daily multiple seizures are found in LGS, often occurring nocturnally and thus undetected by parents or caregivers. Individual patients often show a variety of seizure types (including tonic, atonic, and atypical absence seizures). Tonic seizures, which cause patients to unpredictably stiffen and fall to the ground, are often associated with head injuries which can further worsen patients' cognitive impairment. Atonic seizures (also called drop attacks, lasting between 1 and 4 seconds) cause head nods, loss of posture and sagging at the knees; 50% of patients experience one or more episodes of non-convulsive status epilepticus.

Therapy is challenging, with more than 80% of patients experiencing lifelong seizures and intellectual disability. Potentially effective anti-epileptic medications include Valproic acid and Rufinamide. The ketogenic diet may be an effective treatment option in some cases. In patients with a focal epileptogenic lesion visible on MRI, resective surgery can occasionally lead to seizure improvements. Other surgical options include vagus nerve stimulation (VNS), corpus callosotomy, and deep brain stimulation (DBS). Wearing a helmet is often demanding.

Typical surgery

Epilepsy surgery (corpus callosotomy, resective surgery, vagus nerve stimulation, electrical stimulation of the centromedian thalamic nucleus, ESCM), dental and gingival operations, general surgery, trauma surgery (following drop attacks).

Type of anaesthesia

There are only few recommendations based on case reports. General anaesthesia will be the only option. In limb surgery or other painful procedures, carefully administered local or regional anaesthesia should only be considered as add-on, given the neurotoxic potential of local anaesthetic drugs.

Necessary additional diagnostic procedures (preoperative)

Current blood levels of anti-epileptic medications are recommended. Bleeding history (check list) should be taken, as some anti-epileptic drugs can cause von-Willebrand-Jürgens-like alterations. Further recommendations depend on the impact of surgery and follow general considerations (e.g. blood supply)

Particular preparation for airway management

Usual precautions of upper airway management are sufficient. Gingival alterations due to long-term antiepileptic medication may cause airway abnormalities. Patient should be free of upper respiratory infection. Neck injury after falling may lead to emergency surgery and difficult airway management.

Particular preparation for transfusion or administration of blood products

Not reported.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Not reported.

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

Several anti-epileptic drugs enhance cytochrome P450 enzymes stimulating the metabolism of anaesthetic agents and thus cause higher dosage demands to maintain depth of anaesthesia. Rufinamide has minor power to do so. Valproic acid inhibites P450 enzymes and thus elevates phenobarbital (thiopental not reported) levels. In patients with ketogenic diet, the application of amino acids and carbohydrates causes decrease in plasma ketons, which may interfere with the therapy. On the other hand, metabolic acidosis due to ketogenic diet represents a challenge during long-term surgery, and intensive care treatment and should be monitored carefully. Hypokalaemia may occur as consequence of therapy with topiramate.

Anaesthesiologic procedure

Dependence on caregivers and separation from them during induction may be a challenging psychological issue. Premedication with benzodiazepines is of conflicting evidence. Dose requirement may be elevated. However, there is a description on benzodiazepines precipitating non-convulsive status epilepticus. Oral ketamine should be safe given its potential to even stop non-convulsive status epilepticus.

Thiopental (5mg/KG) was used without adverse effect, due to general considerations propofol should be safe and has been successfully administered by the author of this article as induction agent and continuously during Total Intravenous Anaesthesia (TIVA). Opioids are safe, the effect of long acting opioids can be extended. Rocuronium bromide (1mg/kg) was safely administered, the same applies to the reversal with glycopyrrolate and pyridostigmine. There is no data addressing the use of succinylcholine. Isoflurane has been given as inhalational agent, as well as sevoflurane during inhalational induction. Increased concentrations during inhalational induction with sevoflurane are critical though, given the potential to cause epilepsy-like EEG alterations.

Regional or local anaesthesia is recommended for postoperative pain management and to reduce the demand of opioids. Dosage management should be conservative, and reduced patient compliance will not allow these procedures to be performed as stand-alones.

Particular or additional monitoring

Monitoring can be extended with EEG-based devices.

Note that the initial results before induction can mimic deep narcotic patterns as for the common slow wave alterations patients often bear.

Once typical epilepsy waves are extinguished by the induction agent. The continuous monitoring should be reliable (observation of the author of this article).

Possible complications

Seizures during operations may deteriorate the outcome and must be avoided especially in intracranial interventions. There is no evidence that the number of postoperative seizures would increase after anaesthesia.

Postoperative care

Falling in post-anaesthetic care units has to be taken into consideration. Seizures will mostly occur as consequence of the underlying disorder and require a protocol which should preoperatively be elaborated in cooperation with neurologists.

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

Not reported.

Ambulatory anaesthesia

The crucial issue is the competence of the parents or caregivers to distinguish between clinical events as existing before and new onset complications, which would require further action. The availability of the anaesthesiologist and a daily recall is mandatory. Type of surgery (e.g. dental interventions) should guide ambulatory approach whenever possible.

Obstetrical anaesthesia

Not known.

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