orphanaesthesia

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

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

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 is recommended. Bleeding history (check list) should be taken, as some anti-epileptic drugs can cause Von-Willebrandt-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 anesthesia. 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 anesthesia 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.

Literature and internet links

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