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A case of Lennox- Gastaut Syndrome in a child with Panhypopitutarism

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Introduction

Lennox-Gastaut syndrome is childhood epileptic encephalopathy characterized by intractable seizures resident to antiepileptic medications. Here we describe a child with this syndrome who presented for gastroduodenoscopy.

Case Description

A 9-year-old male with Lennox-Gastaut syndrome with refractory seizures to antiepileptic therapy presented for gastroduodenoscopy for evaluation of gastroesophageal reflux disease. His history was significant for cerebral palsy, agenesis of corpus callosum, neurodevelopmental delay, sleep apnea, panhypopituitarism and cortical blindness due to septo-optic dysplasia. Panhypopituitarism was secondary to congenital absence of pituitary gland. Inhalation general anesthesia was induced with nitrous oxide, oxygen and sevoflurane and after securing an intravenous line, anesthesia was maintained with intermittent doses of propofol. Stress dose of hydrocortisone was intravenously administered prior to the procedure. Perioperative course was uneventful.

Clinical Features[1]

- ✓ Epilepsy with frequent seizures
- ✓ Childhood epileptic encephalopathy
- ✓ Learning disability
- ✓ Delayed development of motor milestones
- ✓ Developmental delay

Case Discussion

- Lennox–Gastaut syndrome is a severe form of childhood epilepsy that typically begins at 3-5 years of age.[1]
- Mode of inheritance: Most of the cases are sporadic with no family history.
- From 3 to 30% may have a family history of epilepsy.[1]
- More common in males than females.[1]
- Frequency: Accounts for 4% of all childhood epilepsy.[1]
- Incidence: 1; 50,000-1:100,000.[1]

Seizures in Lennox-Gastaut Syndrome[1]

- Intractable seizures resistant to medications
- Usually brief, sometimes prolonged
- Seizures occur most often during sleep
- About 75% of the seizures are tonic-clonic
- Other types of seizures include atypical absence seizures
- Seizures are associated with complete or partial loss of consciousness
- Seizure leads to confusion and lack of alertness which may be prolonged
- Seizures may be associated with drop attacks with sudden loss of muscle tone

Pathophysiology[1]

- No specific genes are identified
- Associated with pre-existing neurological condition.
- May result from brain injuries occurring before or during birth
- May be associated with cortical dysplasia
- May be a part of the genetic disorder tuberous sclerosis

Anesthetic Considerations[2]

1. Preoperative anxiety, child may not be cooperative for induction of anesthesia
2. Administer regular pre-operative anti-epileptic medication
3. No specific anesthetic considerations
4. Inhalational induction sevoflurane well tolerated
5. Propofol total intravenous anesthesia (TIVA) is well tolerated (Propofol has anti-epileptic activity)
6. Hepatic microsomal induction may need higher dose of drugs

Evidence of anesthetic management is limited to case reports.

CONCLUSION

No specific modification in anesthetic technique is needed and inhalational induction and maintenance of anesthesia with volatile anesthetic agents and nitrous oxide/air or TIVA is well tolerated as per case reports.[2] We managed this case successfully with such technique uneventfully.

References

1. Lennox-Gastaut syndrome: Genetic Home Reference.
2. Kapoor I, Rath GP: Anesthetic management of a child with Lennox–Gastaut syndrome with intractable epilepsy posted for intracranial surgery! J Pediatr Neurosci. 2016; 11: 290–91.