Lennox–Gastaut syndrome

Lennox-Gastaut Syndrome is a rare condition that begins in childhood as atonic seizures (“drop attacks”). Often develops into tonic seizures with mental retardation.

Treatment

Lennox–Gastaut syndrome treatment

Outcome

Seizures are often polymorphic, difficult to treat medically, and may occur as often as 50 per day. May also present with status epilepticus. Approximately 50% of patients have reduced seizures with valproic acid. Corpus callosotomy may reduce the number of atonic seizures.

Lennox–Gastaut syndrome (LGS) is a difficult-to-treat form of childhood-onset epilepsy that most often appears between the second and sixth year of life. LGS is characterized by a triad of signs including frequent seizures of multiple types, an abnormal EEG pattern of less than 2.5 Hz slow spike wave activity, and moderate to severe intellectual impairment.

Combined VNS and prior CC produced satisfactory seizure outcomes in LGS patients with different seizure types, including DAs. Even non-responders to prior CC responded to subsequent VNS for residual seizures, except for DAs. There is a greater likelihood that these procedures may be more feasible in patients who possess conversation ability prior to VNS [1)].

Case series

Patients (n = 16, mean age 11.4 ± 6.4 years, range 6-19 years) had a mean seizure frequency of 24.5 ± 19.8/days (range 1-60) and a mean intelligence quotient of 25.23 ± 10.71. All had syndromic diagnosis of Lennox-Gastaut syndrome, with the following etiologies: hypoxic insult (10), lissencephaly (2), bilateral band heterotropia (2), and microgyria and pachygyria (2). Surgery included complete callosotomy and the section of anterior and posterior commissure by microscopic approach through a mini craniotomy (11) and endoscopic-assisted approach (5). Complications included meningitis (1), hyperammonemic encephalopathy (2), and acute transient disconnection (5). There was no mortality or long-term morbidity. Mean follow-up was 18 ± 4.7 months (range 16-27 months). Drop attacks stopped in all. Seizure frequency/duration decreased >90% in 10 patients and >50% in 5 patients, and increased in 1 patient. All patients attained presurgical functional levels in 3 to 6 months. Child behavior checklist scores showed no deterioration. Parental questionnaires reported 90% satisfaction attributed to the control of drop attacks. The series was compared retrospectively with an age/sex-matched cohort (where a callosotomy only was performed), and showed better outcome for drop attacks (P < .003).

This preliminary study demonstrated the efficacy and safety of complete callosotomy with anterior, hippocampal, and posterior commissurotomy in Lennox-Gastaut syndrome (drop attacks) with moderate to severe mental retardation [2].

1)
Katagiri M, Iida K, Kagawa K, Hashizume A, Ishikawa N, Hanaya R, Arita K, Kurisu K. Combined surgical intervention with vagus nerve stimulation following corpus callosotomy in patients with Lennox-Gastaut syndrome. Acta Neurochir (Wien). 2016 May;158(5):1005-12. doi: 10.1007/s00701-016-2765-9. Epub 2016 Mar 15. PubMed PMID: 26979179.
2)
Chandra SP, Kurwale NS, Chibber SS, Banerji J, Dwivedi R, Garg A, Bal C, Tripathi M, Sarkar C, Tripathi M. Endoscopic-Assisted (Through a Mini Craniotomy) Corpus Callosotomy Combined With Anterior, Hippocampal, and Posterior Commissurotomy in Lennox-Gastaut Syndrome: A Pilot Study to Establish Its Safety and Efficacy. Neurosurgery. 2016 May;78(5):743-51. doi: 10.1227/NEU.0000000000001060. PubMed PMID: 26474092.