A Retrospective Claims Study Evaluating Mortality in Patients with Lennox-gastaut or Dravet Syndromes in the United States

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Rationale: Lennox-Gastaut (LGS) and Dravet syndromes (DS) are rare developmental and epileptic encephalopathies characterized by pharmacoresistant seizures and cognitive impairment. Mortality rates in LGS and DS have been reported as 15.84 and 6.12 per 1000 patient (pt)-years, respectively (Sullivan J, et al. *Epilepsia*. 2024). Real-world mortality data in LGS and DS is limited. Here, we used real-world claims data to analyze mortality rates, suspected sudden unexplained death in epilepsy (SUDEP) as a contributing factor to mortality rates, and comorbidities associated with mortality for pts with LGS or DS in the US.

Methods: This retrospective claims-based analysis identified pts in the US diagnosed with LGS or DS from Oct 01, 2015, to Dec 31, 2023, using Real Chemistry's open claims database. Pts were included if they had ≥2 LGS or DS claims in the study period, a date of death (DOD) after Oct 01, 2015, and ≤60 days from their last claim to DOD. Outcomes in pts with LGS and DS included annual all-cause case fatality and crude mortality rates (primary), comorbidities in pts with vs. without a mortality event, and SUDEP-related fatality rates. SUDEP was suspected if no non-SUDEP cause of death was observed in the database. Fatality and mortality rates were adjusted to account for incomplete pt capture in claims data. Standardized mortality ratios (SMRs) and 95% confidence intervals for comorbidities were also reported.

Results: In total, 29,625 pts with LGS and 2259 pts with DS were included; of those, 2509 (8.5%) and 52 (2.3%) had a DOD during the study period, respectively. The yearly adjusted case fatality rate for LGS ranged from 4.2% to 6.2% from 2016 to 2023; the SUDEP-related rate for LGS ranged from 1%-2%. The LGS SMR was 4.98 (95% CI: 4.40-5.64) to 6.14 (5.82-6.47) times higher than the average person (**Figure 1**); adjusted crude mortality rates ranged from 41.8 to 62.5 per 1000 pt-years. In pediatric and adult pts with LGS, feeding difficulties and cardiovascular/respiratory

complications were most significantly associated with mortality at each 3-year interval examined (**Table 1**). In pts with DS, the 5-year adjusted case fatality rate was 8.6% from 2019-2023; the SUDEP-related rate for DS was 2.1%. The DS SMR was 2.11 (95% CI: 1.08-4.12) to 3.96 (3.14-5.00) times higher than an average person from 2020-2022 (**Figure 1**); the 5-year adjusted crude mortality rate was 17.28 per 1000 pt-years. From 2019-2023, encephalopathy (OR: 4.59 [95% CI: 1.84-11.47]) and feeding difficulties (OR: 4.30 [1.35-13.71]) were most significantly associated with mortality in pediatric and adult pts with DS, respectively.

Conclusions: As in the reported literature, this real-world analysis showed that the crude and standardized mortality rates for LGS are higher than for DS. Cardiovascular and respiratory complications were significantly associated with mortality in pts with LGS only. Future studies will examine healthcare resource utilization and antiseizure medication use and their association with mortality

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