

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

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QUESTION

- What are the current real-world annual all-cause Lennox-Gastaut syndrome (LGS) and Dravet syndrome (DS) all-cause and sudden unexpected death in epilepsy (SUDEP)-related case fatality rates?
- What comorbidities are associated with higher mortality in patients with LGS and DS?

INVESTIGATION

- Using real-world data from the Real Chemistry open claims database and the Veritas mortality database, we examined case fatality rates, standardized mortality ratios (SMRs), and comorbidities associated with mortality in pediatric and adult patients with LGS or DS from October 01, 2015, to December 31, 2023

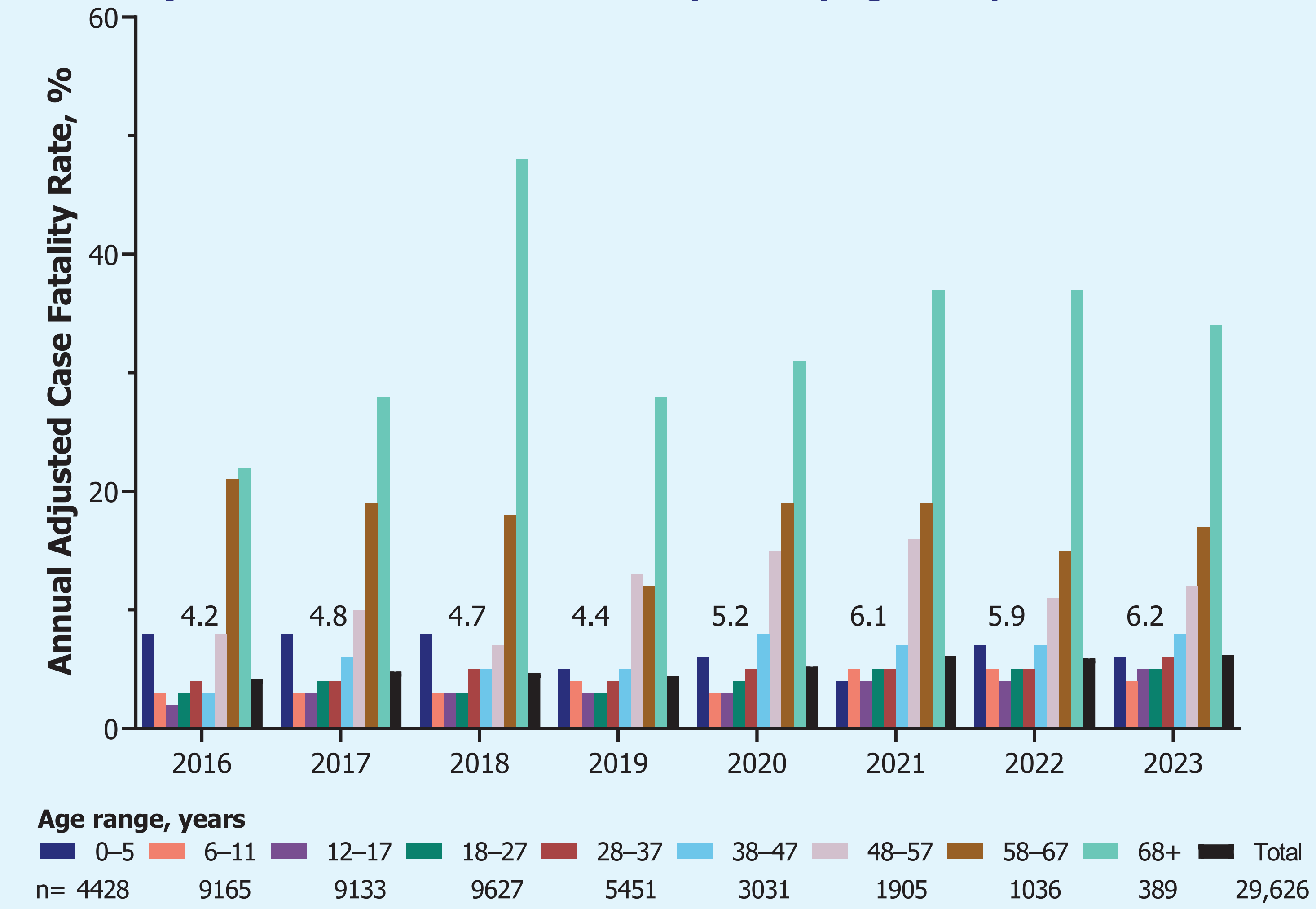
RESULTS

Patient Characteristics and Payer Channel

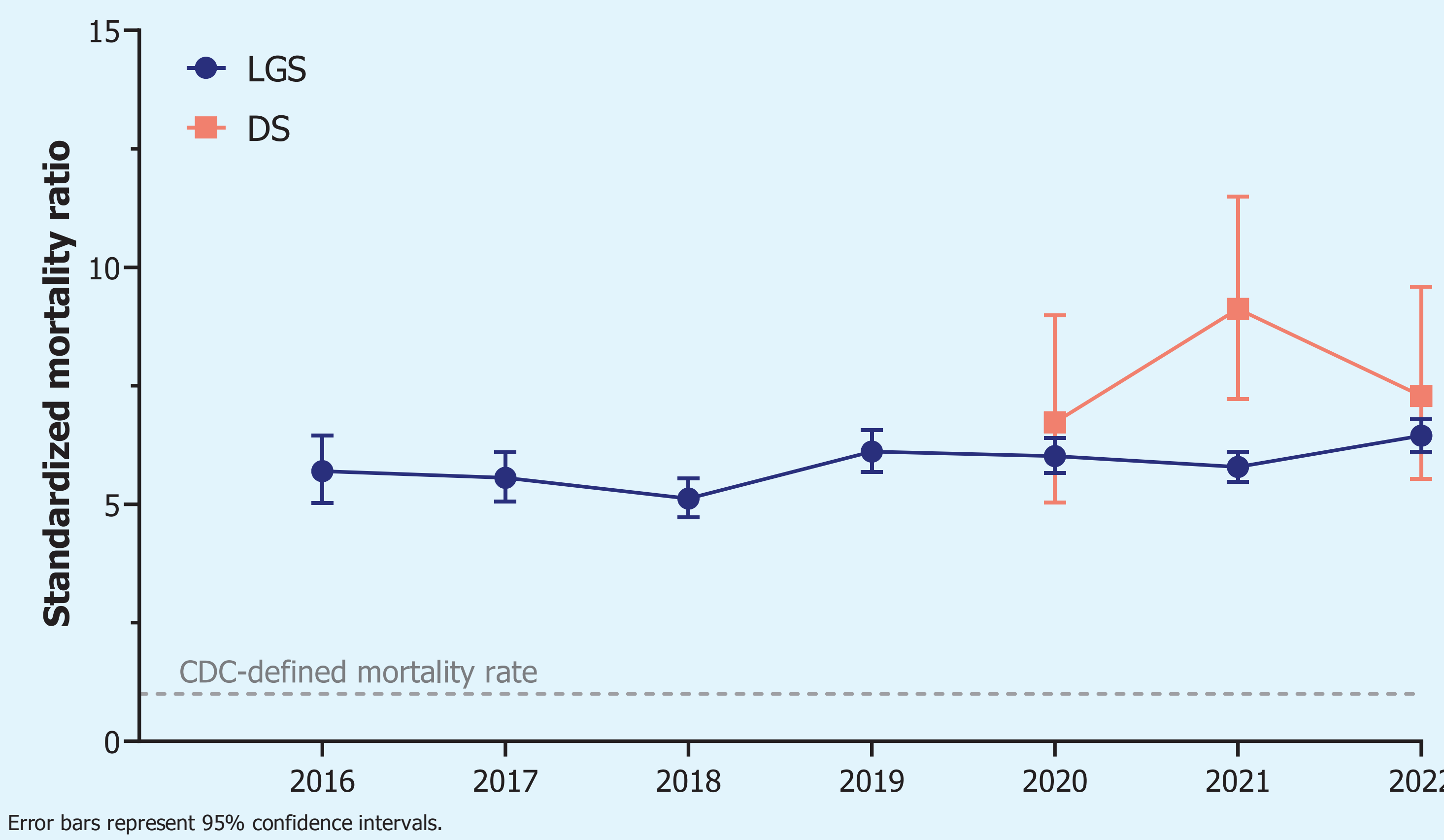
	Patients With Lennox-Gastaut Syndrome		Patients With Dravet Syndrome	
	Deceased n=2509	Living n=27,117	Deceased n=52	Living n=2207
Median age ^a (IQR), years	22 (34)	16 (18)	10 (18)	10 (12)
Male sex, n (%)	1420 (57)	15,293 (56)	28 (54)	1140 (52)
Payer channel, n (%)				
Medicaid	728 (29)	8775 (32)	16 (31)	715 (32)
Commercial	574 (23)	7650 (28)	20 (38)	844 (38)
Medicare	511 (20)	3010 (11)	5 (10)	70 (3)
Other	141 (6)	1736 (6)	5 (10)	175 (8)

^aAge at first Lennox-Gastaut syndrome or Dravet syndrome claim during study period.

Annual Adjusted LGS All-Cause Case Fatality Rate by Age Group



Standardized Mortality Ratio in Patients With LGS or DS by Year

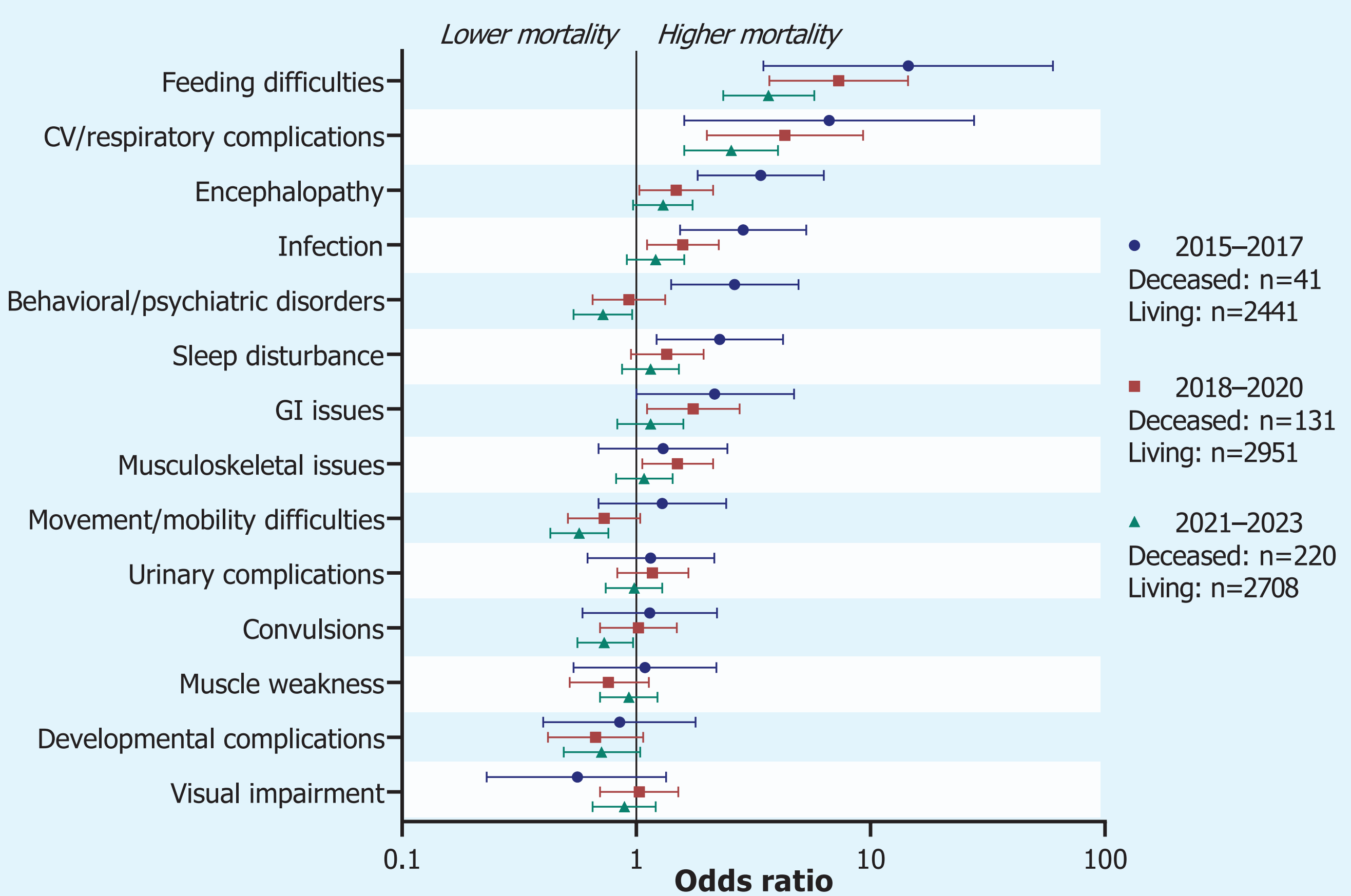


- In patients with DS, the 5-year adjusted all-cause case fatality rate was 8.6% from 2019-2023; pediatric and adult fatality rates were 6.6% and 11.6%, respectively

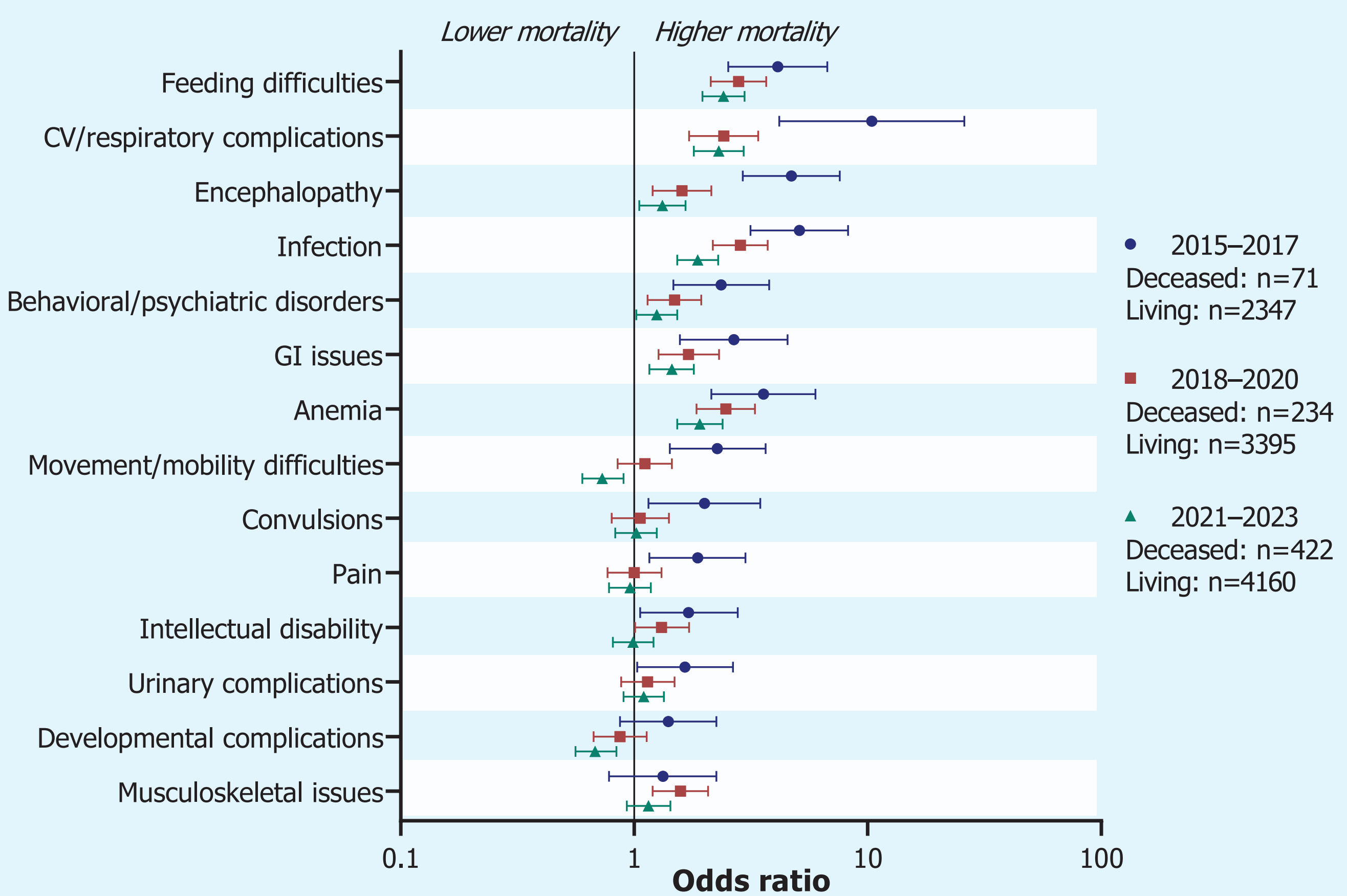
Abbreviations: ASMs, anti-seizure medications; CDC, Center for Disease Control; CV, cardiovascular; DS, Dravet syndrome; GI, gastrointestinal; IQR, interquartile range; LGS, Lennox-Gastaut syndrome.

Most Common Comorbidities and Their Association With Mortality

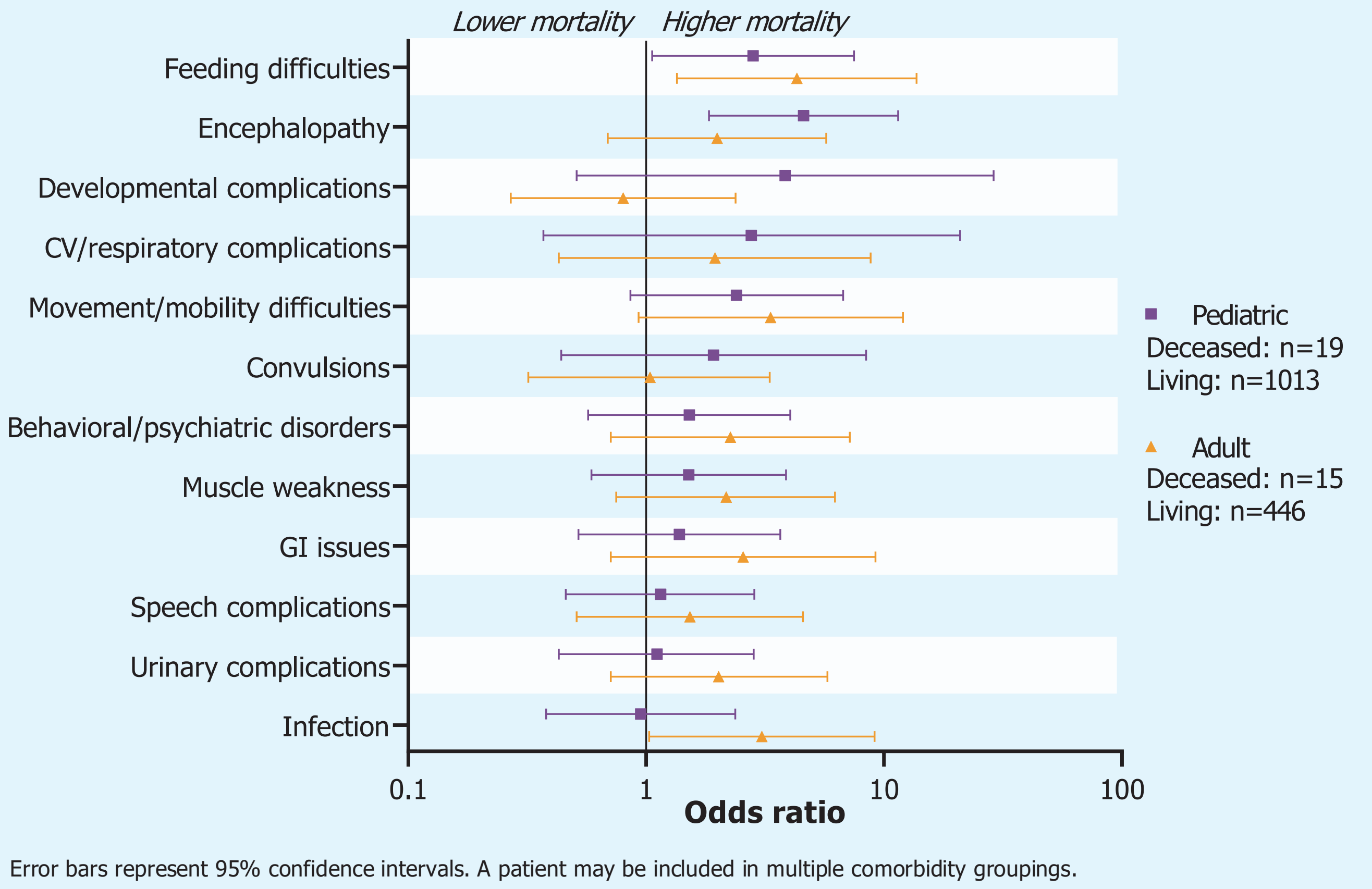
A. Pediatric Patients With LGS



B. Adult Patients With LGS



C. Pediatric and Adult Patients With DS



METHODS

- This retrospective claims-based analysis identified patients in the US diagnosed with LGS or DS from October 01, 2015, to December 31, 2023, using the Real Chemistry open claims database, and further classified as living or deceased if present in the Veritas mortality dataset
- Patients were included if they had ≥ 2 LGS claims (ICD-10: G40.81, G40.811, G40.812, G40.813, and G40.814) or ≥ 2 DS claims (ICD-10: G40.83, G40.833, G40.834) in the study period; patients with a mortality event were required to have a date of death (DOD) after October 01, 2015, and ≤ 60 days from their last claim to date of death
 - Because data from 2015-2018 were scarce, and DS ICD-10 codes were not in use until 2020, DS endpoints were analyzed from 2019-2023
- Outcomes in pediatric (<18 years old) and adult (≥ 18 years old) patients with LGS and DS included annual all-cause case fatality rates (primary), comorbidities in patients with vs. without a mortality event, and SUDEP-related fatality rates
- SUDEP was suspected if no non-SUDEP cause of death related claim was observed in the database within ± 60 days of DOD
- Non-SUDEP causes of death included brain swelling, coagulation defect, coma, COPD, COVID, CV, dementia, gastrointestinal hemorrhage, heart attack, hospice, infection (kidney, liver), neutropenia, other metabolic disorders, pneumonia, respiratory, respiratory failure, sepsis, severe malnutrition, shock, sleep apnea, and stroke
- Annual fatality rates were adjusted to account for incomplete patient capture in claims and mortality datasets
- Standardized mortality ratios (SMRs) and 95% confidence intervals for comorbidities were also reported
 - SMRs compare all-cause fatality rates to the US general population from 2016-2022 for LGS and 2020-2022 for DS

CONCLUSIONS

- In this study, all-cause fatality rates in children and adults with LGS or DS were high
- Mortality in patients with LGS peaked early in life, and again at 48 years and older
- In DS, 5-year fatality rates for adults were higher than for pediatric patients
- Feeding difficulties were associated with mortality at any age in both LGS and DS, whereas cardiovascular and respiratory complications were significantly associated with mortality in LGS only
- Future studies will examine healthcare resource utilization and antiseizure medication use and their association with mortality

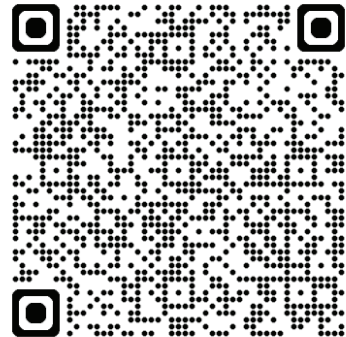
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Disclosures

AL: Employee of UCB with stock ownership.
Disclosures for all authors can be found in the full poster at the QR code.

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