Understanding the Incidence, Prevalence, Characteristics, and Healthcare Resource Utilization for Patients With Dravet and Lennox-Gastaut Syndromes

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Introduction

- Dravet syndrome (DS) and Lennox-Gastaut syndrome (LGS) are lifelong developmental and epileptic encephalopathies characterized by treatment-resistant seizures, developmental delays, cognitive impairments, and behavioral impairments[1-3]
- A recent systematic literature review of global studies estimated that the incidence and prevalence of LGS is generally greater than DS[4]
  - LGS and DS incidence ranged from 14.5-28 and 2.2-6.5 per 100,000 people, respectively
  - LGS and DS prevalence ranged from 5.8-60.8 and 1.2-6.5 per 100,000 people, respectively
- Greater insights into the disease states of DS and LGS may be useful to understand the overall burden of illness and potential gaps for future research[5]

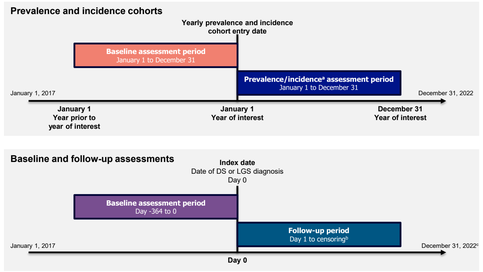
Objective

- To describe the real-world incidence, prevalence, patient and disease characteristics, as well as healthcare costs and healthcare resource utilization (HCRU) of patients with DS and LGS in the United States

Methods

- This retrospective cohort analysis used real-world, administrative claims data from the Merative® MarketScan® US database from January 1, 2017, to December 31, 2022
- Patients were required to have: a confirmed DS or LGS diagnosis using ICD-10 codes (DS ICD-10 codes G40.83, G40.833, and G40.834 were effective October 2020; LGS ICD-10 codes G40.81, G40.811, G40.812, G40.813, and G40.814 were effective October 2015), complete enrollment for the year of interest, and one calendar year baseline enrollment (30-day allowable gap) before January 1 of the year of interest (**Figure 1**)
- The date of first observable diagnosis was set as the index date of each respective cohort
- Patient characteristics were described during the baseline period (12 months pre-index)
- Comorbidities and treatments were assessed in the follow up period (Day 1 to censoring [730 days after Day 0, end of enrollment, end of data, or death])
- Healthcare costs and HCRU were assessed 6 months pre- and post-index
- Outcomes assessed included: yearly prevalence and incidence rates (primary), baseline patient characteristics
  - Clinical seizure characteristics, antiseizure treatments, all-cause and epilepsy-related healthcare visits, and all-cause and epilepsy-related healthcare costs, including emergency, inpatient, pharmacy, and outpatient costs, are described at baseline and follow-up
- Descriptive statistics were used along with 95% CIs

Figure 1. Study Design



[a]Incidence assessment included a washout window from the study start to Day -1 to cohort entry date.
[b]Patients were censored at the first instance of a maximum of up to 730 days after Day 0, end of enrollment, end of data, or death.
[c]Index date was on or before December 31, 2022, and follow-up period used censoring rules.
DS, Dravet Syndrome; LGS, Lennox-Gastaut Syndrome.

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Overview

QUESTION

- What are the current real-world data surrounding incidence and prevalence, patient characteristics, healthcare costs, and healthcare resource utilization in patients with Dravet syndrome (DS) and Lennox-Gastaut syndrome (LGS) in the US?

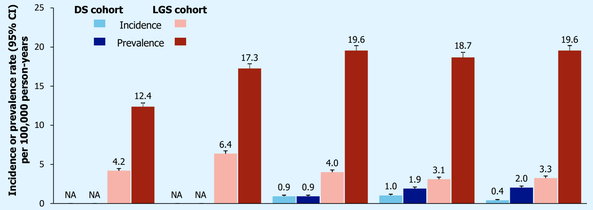
INVESTIGATION

- Using real-world, administrative claims data (January 01, 2017, to December 31, 2022) from the Merative® MarketScan® US database, we examined incidence and prevalence, patient characteristics, healthcare costs, and healthcare resource utilization in patients with DS or LGS

RESULTS

- In total, 579 patients with DS and 7846 patients with LGS were identified
- The incidence and prevalence of DS from 2020-2022 ranged from 0.4-1.0 and 0.9-2.0 per 100,000 person-years, respectively (**Figure**)
- The incidence and prevalence in LGS from 2018-2022 ranged from 3.1-6.4 and 12.4-19.6 per 100,000 person-years, respectively (**Figure**)
- Most patients with DS or LGS experienced focal seizures or seizures of unknown onset
- Clobazam was the most prescribed antiseizure medication (ASM) for patients with DS and LGS
- Mean 6-month total all-cause costs and total epilepsy-related costs increased from baseline to follow-up for both DS and LGS

Figure. Incidence and Prevalence of DS and LGS



[a]The ICD-10 codes for DS were not in use until 2020.
DS, Dravet Syndrome; LGS, Lennox-Gastaut Syndrome; NA, not applicable.

CONCLUSIONS

- This real-world study of US claims data confirmed that the incidence and prevalence of LGS is higher than for DS
- Compared to pre-diagnosis baseline, increases in healthcare costs were observed during follow-up in both DS and LGS cohorts

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Results

Baseline Characteristics

- 579 and 7846 patients with DS and LGS were identified, respectively, and mean (SD) age at index was 13.2 (10.9) and 20.7 (15.4) years, respectively (**Table 1**)

Table 1. Baseline Characteristics

Characteristic	Dravet Syndrome (N=579)	Lennox-Gastaut Syndrome (N=7846)
Age, mean (SD), years	13.2 (10.9)	20.7 (15.4)
Age category, n (%)		
<18	422 (72.9)	3964 (50.5)
≥18	157 (27.1)	3882 (49.5)
Male, n (%)	300 (51.8)	4402 (56.1)
Race, n (%)		
White	226 (39.0)	3149 (40.1)
Black	48 (8.3)	883 (11.3)
Hispanic	36 (6.2)	323 (4.1)
Other	35 (6.0)	324 (4.1)
Missing	234 (40.4)	3167 (40.4)
Payer plan type, n (%)		
Commercial	165 (28.5)	2098 (26.7)
Medicare	2 (0.3)	14 (0.2)
Medicaid	412 (71.2)	5734 (73.1)

SD, standard deviation.

Incidence and Prevalence

- The incidence of DS (from 2020-2022) and LGS (from 2018-2022) ranged from 0.4-1.0 and 3.1-6.4 per 100,000 person-years, respectively (**Overview Figure**)
- The prevalence of DS (from 2020-2022) and LGS (from 2018-2022) ranged from 0.9-2.0 and 12.4-19.6 per 100,000 person-years, respectively (**Overview Figure**)
- Median follow-up times in DS and LGS were 1.7 and 2.0 years, respectively

Seizure and Treatment Types and Comorbidities

- In DS, focal seizures were reported in 81.2% and 68.0% of patients at baseline and follow-up, respectively (**Table 2**); seizures of unknown type were reported in ≥86% of patients with DS
- In LGS, focal seizures were reported in 100% and 88.3% of patients at baseline and follow-up, respectively; seizures of unknown type were reported in 80% of patients
- In DS and LGS, clobazam (DS, 56.5%; LGS, 42.8%), cannabidiol (DS, 42.0%; LGS, 35.3%), and levetiracetam (DS, 32.0%; LGS, 33.5%) were the most prevalently prescribed antiseizure medications (ASMs)
- In the DS and LGS cohorts, comorbidities that occurred in ≥5% of patients at baseline and follow-up were anxiety (DS, 5.2-8.8%; LGS, 10.4-13.1%) and autism (DS, 28.8-32.5%; LGS, 22.9-24.9%); LGS also included depression (6.5-7.6%), and infantile spasms (9.2-8.6%)

Table 2. Clinical Seizure Characteristics and Associated Medications Utilized From 2018-2024[a]

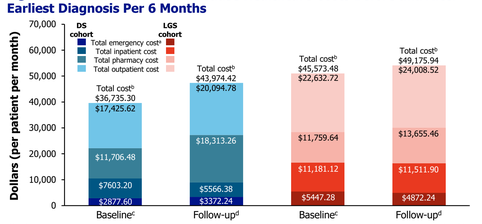
Treatments, n (%)	Dravet Syndrome (N=579)		Lennox-Gastaut Syndrome (N=7846)	
	Baseline	Follow-up[b]	Baseline	Follow-up[c]
Seizure type, n (%)				
Focal	470 (81.2)	394 (68.0)	7846 (100.0)	6923 (88.3)
Generalized	163 (28.2)	136 (23.5)	3201 (40.8)	3115 (39.7)
Unknown	542 (93.6)	501 (86.5)	6289 (80.2)	6245 (79.6)
Status epilepticus, n (%)	82 (14.2)	70 (12.1)	811 (10.3)	943 (12.0)
Rescue medications, n (%)	307 (65.5)	342 (72.9)	3896 (55.6)	4352 (62.1)
Other antiseizure treatments or associated medications, n (%)				
Psychotropic medications	389 (82.9)	403 (85.9)	5219 (74.5)	5375 (76.7)
Neurosurgery	11 (2.3)	35 (7.5)	365 (5.2)	646 (9.2)
Epilepsy surgery	9 (1.9)	19 (4.1)	187 (2.7)	326 (4.7)

[a]Last index date was December 31, 2022 and maximum follow-up time was 2 years. [b]Mean (SD) follow-up time was 493.17 (254.33) days. [c]Mean (SD) follow-up time was 575.19 (229.66) days.
SD, standard deviation.

Healthcare Costs and HCRU

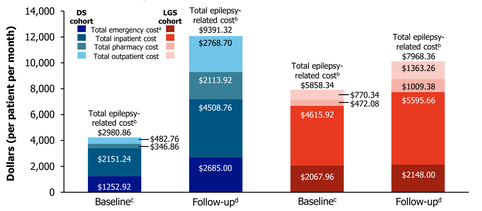
- Mean 6-month total all-cause costs increased from baseline to follow-up by approximately $7000 for DS patients and $3500 for LGS patients (**Figure 2**)
- Mean total epilepsy-related costs increased from baseline to 6-month follow-up by approximately $6500 for DS patients and $2000 for LGS patients (**Figure 3**)
- HCRU remained similar from baseline to follow-up for both the DS and LGS cohorts (**Figure 4**)
- Mean epilepsy-related outpatient visits increased from baseline (DS: 1.4; LGS: 1.7) to 6-month follow-up (DS: 6.3; LGS: 3.1) for both cohorts (**Figure 5**)

Figure 2. DS and LGS Mean All-Cause Healthcare Costs Before and After Earliest Diagnosis Per 6 Months



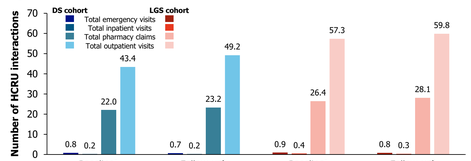
[a]Total emergency costs were calculated using inpatient and outpatients claims that were associated with emergency visits (could not be summed in addition to other total costs). [b]The calculation of total all-cause costs was performed by summing the costs from all inpatient, outpatient, and prescription claims. [c]Assessed over 365 days. [d]Assessed if began over follow-up.
DS, Dravet Syndrome; LGS, Lennox-Gastaut Syndrome.

Figure 3. DS and LGS Mean Epilepsy-Related Healthcare Costs Before and After Earliest Diagnosis Per 6 Months



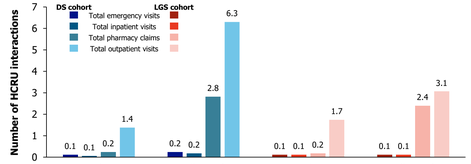
[a]Total emergency costs were calculated using inpatient and outpatient claims that were associated with emergency visits (could not be summed in addition to other total costs). [b]The calculation of total epilepsy-related costs was performed by summing the costs from all inpatient, outpatient, and prescription claims. [c]Assessed over 365 days. [d]Assessed if began over follow-up.
DS, Dravet Syndrome; LGS, Lennox-Gastaut Syndrome.

Figure 4. DS and LGS Mean All-Cause HCRU Before and After Earliest Diagnosis Per 6 Months



[a]Assessed over 365 days. [b]Assessed if began over follow-up.
DS, Dravet Syndrome; HCRU, healthcare resource utilization; LGS, Lennox-Gastaut Syndrome.

Figure 5. DS and LGS Mean Epilepsy-Related HCRU Before and After Earliest Diagnosis Per 6 Months



[a]Assessed over 365 days. [b]Assessed if began over follow-up.
DS, Dravet Syndrome; HCRU, healthcare resource utilization; LGS, Lennox-Gastaut Syndrome.

Limitations

- This study was limited by the capture rate of the database, which required the use of ICD-10 codes for DS, LGS, treatments, and comorbidities
  - ICD-10 codes for DS were not available until 2020, and any lag in providers' use or understanding of these codes may have led to inaccurate claims data
  - Treatments and comorbidities were not confirmed clinically
- The cohort size for DS was much smaller compared with LGS and limits the conclusions that can be drawn for DS

Conclusions

- This real-world study suggests that both incidence and prevalence of LGS are greater than for DS
- In both DS and LGS, all-cause and epilepsy-related healthcare costs increased in the 6 months after the earliest diagnosis (follow-up) compared with 6 months prior to diagnosis (baseline)
- The mean number of all-cause and epilepsy-related HCRU remained similar for all subcategories prior to diagnosis to after diagnosis for both the DS and LGS cohorts, except for an observed increase in total epilepsy-related outpatient visits
- Epilepsy-related healthcare costs showed a greater increase from baseline to follow-up in DS compared with LGS; this may be due to the greater proportion of pediatric patients in the DS cohort
- This US-based retrospective claims analysis adds to the existing literature regarding the current incidence, prevalence, patient characteristics, and healthcare costs and resource utilization for patients with DS and LGS

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