

Common Diagnostic Reasons for Hospitalizations and Emergency Department Visits Among Females with Rett Syndrome in the United States

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BACKGROUND

- Rett syndrome (RTT) is a rare neurodevelopmental disorder that primarily affects females and is associated with a range of mild to severe symptoms.¹⁻³
- The substantial concomitant disease burden associated with Rett syndrome (RTT) has been established.
- However, to date, there is limited research on comorbid conditions leading to healthcare resource utilization in this population.

OBJECTIVES

To describe the top diagnostic reasons for hospitalizations and emergency department (ED) visits among females with RTT.

METHODS

Data Source:

- This study used pre-adjudicated administrative medical claims from the IQVIATM Medical Claims Data (Dx) database (11/1/2016–10/31/2019), to identify patients with RTT, and describe the top diagnostic visits for health resource utilization.

Study design:

- A retrospective longitudinal cohort study design was used to address the study objectives, with the index date defined as the date of the first-observed diagnosis of RTT and the observation period.

Study population:

- Eligible female patients were required to have the index date as the first-observed diagnosis of RTT (International Classification of Diseases, 10th revision, Clinical Modification [ICD-10-CM]: F84.2) in any position of the medical claim and patients were followed until earliest of end of data availability or enrollment.
- Diagnostic reasons for hospitalizations and ED visits during follow up were identified based on ICD-10-CM diagnosis codes in the primary position of a medical claim for a hospitalization or ED visit.
- The most common reasons for hospitalizations or ED visits were reported using frequencies with proportions.
- All analyses were conducted on the overall population and in subgroups of pediatric (age <18 years) and adult (age ≥18 years) patients.

Study outcomes and statistical analysis

- Patient demographics evaluated on the index date were summarized using means, standard deviations (SDs), and medians for continuous characteristics, and frequencies and proportions for categorical characteristics.
- The unique ICD-10-CM diagnosis codes occurring on the admission date of a hospitalization or ED visit and in the primary position were identified as potential reasons for the corresponding visit.
- Among patients with multiple types of visits on the same day, inpatient stays were prioritized over all other types of visits, followed by ED visits.
- Consecutive days of inpatient stays and ED visits were considered one visit.

RESULTS

- There were 5,940 females with RTT identified for this analysis.
- Of 5,940 eligible females with RTT there were pediatric: 3,078 [52%] and adult: 2,862 [48%].
- 1603 (27%) had ≥1 hospitalization and 2289 (39%) had ≥1 ED visit over a median follow up of 2.4 years.
- Overall, the most common reasons for a hospitalization or ED visit included:
- RTT syndrome (22% and 13%, respectively), convulsions (15% for both visit types), and epilepsy (11% and 10%, respectively). Pneumonia (15%) and sepsis (12%) were more common reasons for hospitalizations, whereas cough (11%) and fever (10%) were more common reasons for ED visits.

Table 1. Top Ten Reasons for Hospitalizations and ED Visits Among Patients with RTT (N=5,940)

ICD-10-CM ¹	Description	Frequency of diagnoses, n (%)
Patients with ≥1 hospitalization, ^{2,3}		
F84.2	Rett syndrome	347 (21.6)
R56.9	Unspecified convulsions	236 (14.7)
J18.9	Pneumonia, unspecified organism	235 (14.7)
A41.9	Sepsis, unspecified organism	198 (12.4)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	169 (10.5)
R91.8	Other nonspecific abnormal finding of lung field	157 (9.8)
J69.0	Pneumonitis due to inhalation of food and vomit	134 (8.4)
R50.9	Fever, unspecified	124 (7.7)
J96.01	Acute respiratory failure with hypoxia	120 (7.5)
M41.45	Neuromuscular scoliosis, thoracolumbar region	80 (5.0)
Patients with ≥1 ED visit, ^{2,3}		
R56.9	Unspecified convulsions	338 (14.8)
F84.2	Rett syndrome	295 (12.9)
R05	Cough	249 (10.9)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	226 (9.9)
R50.9	Fever, unspecified	223 (9.7)
J06.9	Acute upper respiratory infection, unspecified	159 (6.9)
J18.9	Pneumonia, unspecified organism	155 (6.8)
K59.00	Constipation, unspecified	153 (6.7)
R10.9	Unspecified abdominal pain	148 (6.5)
K94.23	Gastrostomy malfunction	146 (6.4)

Abbreviations: ED: emergency department; ICD-10-CM: International Classification of Diseases, 10th revision, Clinical Modification; RTT: Rett Syndrome

Table 2. Top Ten Reasons for Hospitalizations and ED Visits Among Pediatric Patients with RTT

ICD-10-CM ¹	Description	Frequency of diagnoses, n (%)
Patients with ≥1 hospitalization, ^{2,3}		
F84.2	Rett syndrome	229 (26.8)
R56.9	Unspecified convulsions	158 (18.5)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	110 (12.9)
J18.9	Pneumonia, unspecified organism	109 (12.8)
R91.8	Other nonspecific abnormal finding of lung field	81 (9.5)
M41.45	Neuromuscular scoliosis, thoracolumbar region	77 (9.0)
R50.9	Fever, unspecified	64 (7.5)
J96.01	Acute respiratory failure with hypoxia	63 (7.4)
G40.919	Epilepsy, unspecified, intractable, without status epilepticus	58 (6.8)
R09.02	Hypoxemia	50 (5.9)
Patients with ≥1 ED visit, ^{2,3}		
R56.9	Unspecified convulsions	215 (18.0)
F84.2	Rett syndrome	184 (15.4)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	149 (12.5)
R50.9	Fever, unspecified	141 (11.8)
R05	Cough	140 (11.7)
J06.9	Acute upper respiratory infection, unspecified	109 (9.1)
K59.00	Constipation, unspecified	85 (7.1)
J18.9	Pneumonia, unspecified organism	79 (6.6)
R10.9	Unspecified abdominal pain	72 (6.0)
R91.8	Other nonspecific abnormal finding of lung field	71 (5.9)

Abbreviations: ED: emergency department; ICD-10-CM: International Classification of Diseases, 10th revision, Clinical Modification; RTT: Rett Syndrome

Table 3. Top Ten Reasons for Hospitalizations and ED Visits Among Adult Patients with RTT

ICD-10-CM ¹	Description	Frequency of diagnoses, n (%)
Patients with ≥1 hospitalization, ^{2,3}		
A41.9	Sepsis, unspecified organism	153 (20.4)
J18.9	Pneumonia, unspecified organism	126 (16.8)
F84.2	Rett syndrome	118 (15.8)
J69.0	Pneumonitis due to inhalation of food and vomit	88 (11.7)
R56.9	Unspecified convulsions	78 (10.4)
R91.8	Other nonspecific abnormal finding of lung field	76 (10.1)
R50.9	Fever, unspecified	60 (8.0)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	59 (7.9)
J96.01	Acute respiratory failure with hypoxia	57 (7.6)
N39.0	Urinary tract infection, site not specified	48 (6.4)
Patients with ≥1 ED visit, ^{2,3}		
R56.9	Unspecified convulsions	123 (11.2)
F84.2	Rett syndrome	111 (10.1)
R05	Cough	109 (10.0)
R50.9	Fever, unspecified	82 (7.5)
K94.23	Gastrostomy malfunction	78 (7.1)
G40.909	Epilepsy, unspecified, not intractable, without status epilepticus	77 (7.0)
N39.0	Urinary tract infection, site not specified	77 (7.0)
J18.9	Pneumonia, unspecified organism	76 (6.9)
R10.9	Unspecified abdominal pain	76 (6.9)
K59.00	Constipation, unspecified	68 (6.2)

Abbreviations: ED: emergency department; ICD-10-CM: International Classification of Diseases, 10th revision, Clinical Modification; RTT: Rett Syndrome

- Notes:
1. Unique ICD-10-CM diagnosis codes occurring on the admission date of a hospitalization or ED visit and in the primary position were identified as potential reasons for the corresponding visit.
 2. Among patients with multiple types of visits on the same day, inpatient stays were prioritized over all other types of visits, followed by ED visits.
 3. Consecutive days of inpatient stays and ED visits were considered one visit.

CONCLUSIONS

- Our findings corroborate the multisystem comorbidities experienced by patients with RTT and provide valuable insights to providers and caregivers on conditions leading to hospitalizations and ED visits in this population, including epilepsy, respiratory viruses, and infections.

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DISCLOSURES

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