Feeding difficulties and gastrostomy in Dravet Syndrome: A UK-wide caregiver survey

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Dravet Syndrome

- One of the most common monogenic epilepsies (incidence ~1/15,000)
- Associated with variants in SCN1A in 90% of individuals
- The core phenotype is of fever-sensitive, drug-resistant epilepsy, developmental delay, and intellectual disability.
- Comorbidities are common, and have a negative impact on quality of life
- Difficulties with feeding have been reported in surveys of Dravet syndrome caregivers, necessitating gastrostomy in some.

Current antiseizure medications:

<table>
<thead>
<tr>
<th>Medication</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Topiramate</td>
<td>20%</td>
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<tr>
<td>Cannabidiol</td>
<td>15%</td>
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<tr>
<td>Levetiracetam</td>
<td>15%</td>
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<tr>
<td>Fenfluramine</td>
<td>7%</td>
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<tr>
<td>Clobazam</td>
<td>75%</td>
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<tr>
<td>Valproate</td>
<td>69%</td>
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<tr>
<td>Stiripentol</td>
<td>62%</td>
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Questions

- How common are feeding difficulties and gastrostomy in Dravet syndrome?
- What do DS caregivers think about gastrostomy post-insertion?
- What are the post-gastrostomy outcomes in people with DS?

Methods

A cross-sectional survey regarding feeding difficulties and gastrostomy was distributed to Dravet Syndrome UK registered families via email and Facebook.

The cohort

Caregiver responses from 65 individuals with SCN1A-related Dravet Syndrome were included.

Current antiseizure medications: Clobazam (75%), Valproate (69%), Stiripentol (62%), Topiramate (20%), Cannabidiol (15%), Levetiracetam (15%), Fenfluramine (7%), Ketogenic diet (6%), Other (25%)

Results

Feeding difficulties

<table>
<thead>
<tr>
<th>Difficulty</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Yes (92%)</td>
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<tr>
<td>No (8%)</td>
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</tbody>
</table>

Most common feeding difficulties in people with Dravet syndrome (n=60)

- Poor appetite
- Weight loss/poor weight gain
- Dysphagia
- Poor appetite
- Weight loss/poor weight gain
- Dysphagia

Gastrostomy

- Yes (46%)
- No (60%)

Post-gastrostomy outcomes

- Convulsive seizures: 27%
- Walking: 31%
- Cognition: 42%
- Emotional problems: 19%
- Per-Farmerian: 19%

Conclusions

- Feeding difficulties are common in Dravet syndrome, and gastrostomy is often required
- More information about feeding difficulties and gastrostomy in Dravet syndrome is needed to support caregivers
- Difficulties should be proactively sought during clinical review of people with Dravet syndrome, and the potential need for gastrostomy discussed with caregivers
- Outcomes post-gastrostomy are positive, with many benefits reported by caregivers
- Gastrostomy complication rates are in keeping with those reported in other conditions related to gastrostomy

Caregiver opinion

- Strongly agree
- Agree
- Neutral
- Disagree
- Strongly disagree

Pre-gastrostomy

- 88% were worried about the gastrostomy
- Many concerns across a wide range of issues related to gastrostomy
- 54% had no access to relevant information
- 77% wanted information specifically about gastrostomy in Dravet

Post-gastrostomy

- Quality of life has improved since gastrostomy
- Overall health is better since the gastrostomy
- Ensures medications are received on time
- Ensures adequate nutrition
- Mealtimes are less stressful
- We have support from healthcare professionals
- The feeding regime is easy to follow
- The gastrostomy is easy to use
- I regret they had the gastrostomy
- I’m happy they had the gastrostomy

Cognition

- Comorbidities that affect cognition
- Difficulties with learning and memory
- Language and communication concerns

Post-gastrostomy complications

- Peristomal skin infection
- Granulation tissue
- External tube splitting
- Pain
- Improvements in quality of life

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