

FRIDAY, JULY 20, 2018

Partnering with your Pediatrician

Tips for families and providers to
maximize primary care visits for
Dravet Syndrome Patients



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- *Pediatrician, Certified by American Board of Pediatrics
- *Emory University (Atlanta, GA) for undergraduate and medical degrees
- *Emory University Department of Pediatrics for residency and chief year
- *Pediatric Hospitalist, University of Tennessee (Chattanooga) 4 years
 - *Started clinic for medically complex patients, many with epilepsy
- *2014 - Special Care Clinic, Children's Hospital Colorado
 - *4,000 medically complex patients, many with epilepsy. 12 Faculty.
 - * I am PCP for several patients with Dravet Syndrome
 - *Also serve as inpatient hospitalist and consult to our admitted patients
- *Member, AAP Council on Children with Disabilities

I have no financial disclosures



Goals of talk

- Primary care visits for patients with Dravet Syndrome
- Handling “sick day” visits with Dravet patients
- Maximizing the PCP/Neurology relationship
- Questions





Wellness Visits (Parents):

General:

- Prepare for the time
 - Distraction items
 - Visual schedule
 - Child life specialist if available
 - Allow time with nutritionist, lab, etc.
- List of concerns in hand, consider messaging to doctor in advance
- Bring forms needed for school, camp but can always send these later (1 year)
- Know that vaccines will come up



Growth/Weight Gain

Highly variable in Dravet, but generally smaller than average

CHCO Data (2017):

- Increasing drop from the mean each year in height > weight.

By age 8:

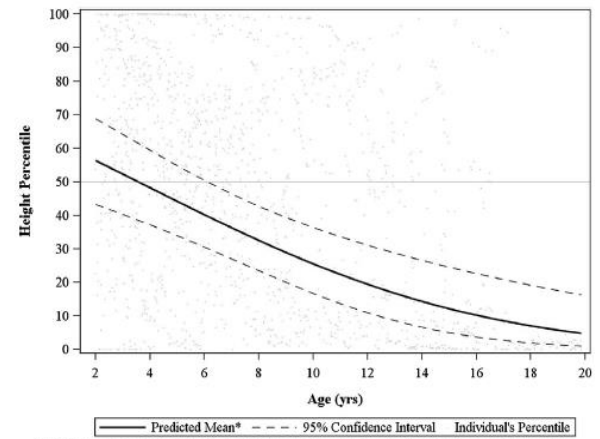
Height Z was -0.45 from mean and

Weight Z was -.09 from mean

-IGF-1 low in 7/15

-Testosterone low in 2/10

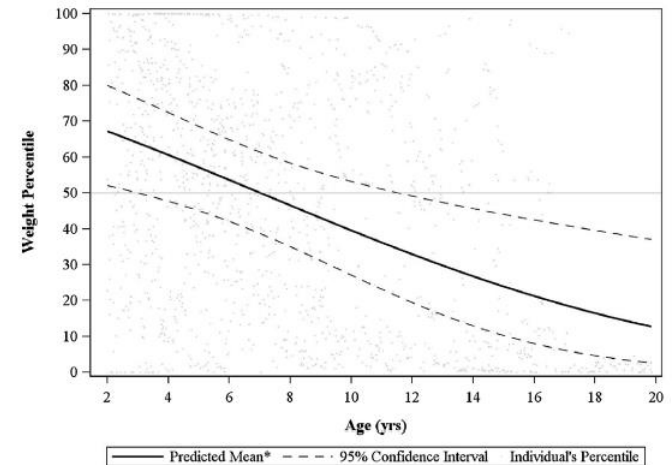
-Thyroid abnormal < 10% of this study 4



*Predicted mean and CI are calculated by transforming predicted Z-scores from a mixed linear regression model with a random intercept and slope.

Fig. 1. Predicted mean height percentile by age of children with Dravet syndrome.

K. Eschbach et al./Seizure 52 (2017) 117–122



*Predicted mean and CI are calculated by transforming predicted Z-scores from a mixed linear regression model with a random intercept and slope.

Fig. 2. Predicted mean weight percentile by age of children with Dravet syndrome.

Nutrition

Pay attention to overall adequacy of nutrition, variety in diet

- Check Vitamin D (30-100) (Teagarden, et al)
- Check Ferritin (link to restless leg)
(goal 20-50 mg/dL)
- Think of Carnitine supplementation to protect liver (if on Depakote)
- Hydration markers
- Refer to nutrition if not already followed in Neurology if concerned



Other items for well child check

- Check in on **Behaviors, Family impact**
- School plan** (IEP, Seizure Action Plan, Tube feeding plan, etc)
- Therapy plan/orders** (PT, ST, OT, ABA)

Physical Exam Pearls:

- Watch for postural changes, worsening ataxia (support devices)
- Dental concerns - make a plan!
- Watch Gtube length



Vaccinations

-**May** trigger seizures (Germany/Austria: 70 patients studied, 27% had seizure after vaccines, DTwP and MMR more prominent, 2/3 with fever) ¹

-Still recommended (**prevented diseases would also likely trigger seizures**)

-Can mitigate seizures with “**bridge**” therapy: Talk with Neurologist

-Seizures may be related to altered cytokine response (France: increased TNF- α response, decreased IL-10 and IL-13 response). ²


-Vaccinating does not change long-term developmental trajectory or onset of non-vaccine associated seizures. (Netherlands) ³



Practical approach to vaccinating

- Avoid times when seizures are already above baseline
- Bridge Therapy (Clonazepam or other)
- Space if desired
- Keep working on it!



A close-up photograph of a young child with light hair and black-rimmed glasses. The child is smiling broadly and sticking their tongue out. The background is a soft, out-of-focus yellow. A blue diagonal graphic element is on the left side of the image.

...and for those
not so good days



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Sick Visit Tips

- For epilepsy concerns, Epileptologist is primary
- Keep your PCP in the loop on big changes
- Consider the role of supplements (Flax seeds/oil, fish oil) and certainly cannabinoids in overall AED levels. Be open and honest with all your doctors.
- I include Lipase when on cannabinoids and vomiting is the concern due to increase in pancreatitis on high cannabinoid doses



Maximizing Parent/ PCP / Neurology relationship

- Direct messaging if available
- PCP: **Pick up the phone and Call when concerned**
- **Avoid splitting:**

Primary care is there to advocate for your child's health in partnership with specialists; it's a **Team Sport**.
- **Parents: Beware the Patient Portal**
 - Don't use this in emergencies
 - Don't use this when same-day answer is needed
 - Realize support staff screen these messages
 - Messages live in patient's chart; all can read.





Questions?

Thank you for the opportunity to speak today!



References

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