Partnering with your Pediatrician

Tips for families and providers to maximize primary care visits for Dravet Syndrome Patients
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*Pediatric Hospitalist, University of Tennessee (Chattanooga) 4 years
  *Started clinic for medically complex patients, many with epilepsy
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  *4,000 medically complex patients, many with epilepsy. 12 Faculty.
  * I am PCP for several patients with Dravet Syndrome
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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
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)  

- 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-a response, decreased IL-10 and IL-13 response).  

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

- Avoid times when seizures are already above baseline
- Bridge Therapy (Clonazepam or other)
- Space if desired
- Keep working on it!
...and for those not so good days
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