Puberty in **Girls** with Dravet Syndrome

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Welcome to the Teenage Years!!!
Objectives

• To recognize health problems common in Dravet teenagers.
• To describe anticipatory guidance and health supervision needs specific for adolescent girls with Dravet.
• To discuss ways of improving hygiene through the use of visual schedules and newer alternatives.
• To illustrate body changes and the menstrual cycle in puberty.
• To differentiate methods of period management, including non-hormonal and hormonal treatments.
Health Problems Dravet Teen

• Behavior issues
• Movement and balance issues
• Growth and nutrition issues
• Sleeping difficulties
• Respiratory infections
• Dental problems
• Autonomic nervous system
Crouch Gait
Scoliosis
Anticipatory Guidance and Health Supervision

- Physical: acne, hair growth, gum problems, thyroid, scoliosis, gait, sleep apnea, growth, bowel/bladder continence
- Diet and Exercise: Calcium, Vitamin D, Iron intake
- Drugs, Alcohol, Smoking
- Vaccines: catch up, Tdap/Meningitis/HPV, Influenza yearly
Anticipatory Guidance and Health Supervision

- **Subspecialists and therapies:** Neuro, Ortho, Genetics, Psychiatry, Nephrology, Optho, Endocrinology, Cardiology, ENT, Psychologists, ST/PT/OT

- **Safety:** helmets/headbands, safety belts, medical ID bracelet or shoe tags, temporary tattoos, technology tracking aids, social media safety
If I am lost, please call
240-751-4900
www.tattoos.org
Anticipatory Guidance and Health Supervision

- School placement/vocational
- Psych: social/emotional/psychological maturity, aggression, challenging behaviors
- Strategies to prevent meltdowns, plan ahead soothing measures
- Life skills: self-care, hygiene, public/private, deodorant
Encouraging Good Hygiene – Showering Schedule Visuals

1. Fill tub with warm water
2. Turn on shower
3. Take off clothes
4. Get in tub
5. Get in shower
6. Wash whole body
7. Rinse soap off
8. Put shampoo on my hand
9. Rub into hair
10. Rinse out shampoo
11. Turn off the water
12. Dry off with towel
13. Put on deodorant
14. Put on clean clothes
15. I did a good job
**Public/Private Behaviors – Story**

**Private Parts**

*Public* places are where other people can see me. *Private* means away from other people, like in my bedroom or bathroom with the door closed. Everyone has *private* parts of their body. I can tell what parts of my body are *private* because I cover them with my underwear. I don’t touch my private parts in public where other people can see me. I don’t ever put my hands inside my pants in public. I can help myself remember not to touch by putting my hands by my side, crossing my arms, or folding my hands. Sometimes I need to touch my private parts, like when I itch or my underwear is uncomfortable. I can ask to go to the bathroom. When I am alone in my bedroom or bathroom, I can touch my private parts.
Anticipatory Guidance and Health Supervision

• Sexuality/supervision/contraception and knowledge, potential for abuse, ability to consent

• Future planning: guardianship, financial planning, group homes, workshop/community supported employment, insurance coverage and SSI, advance directives, adult medical care
Comprehensive care of children with Dravet syndrome
Body Changes

• Teaching correct language if possible
• May not be emotionally/psychologically mature
• Onset:
  – Girls: 8-12, usually around 10.5 years
  – Boys: 9-14, usually around 11.5 years
• May occur before, same or after peers
• Growth spurt
<table>
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<th>Tanner stages Girls</th>
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Period management

• Age onset usually 18 months after breast development

• Period/menstruation policy at school

• Period hygiene: pads, pad with diaper use, underwear (Thinx, Anigan, Luna)

• Tampons not recommended
Use of THIX during the days of the month when you need them most provides a leak and stain resistant support matrix that’s got you covered.
Menstrual Cycle and Seizures

- Follicular phase
- Luteal phase

1. Gonadotropin hormone levels:
   - FSH
   - LH

2. Ovarian cycle:
   - Preovulatory phase
   - Ovulation
   - Postovulatory phase

3. Ovarian hormone levels:
   - Estrogens
   - Progesterone

4. Uterine cycle:
   - Phases of the uterine cycle:
     - Menses
     - Proliferative phase
     - Secretory phase

DAYS: 0 7 14 21 28
Non-hormonal period management

- Period and mood diary, evaluate impact on life
- Evaluate if change in behavior influenced by pain/discomfort
- Try to evaluate seizure frequency and relationship to periods
- Pain management: NSAIDs (Ibuprofen, Naproxen) decrease pain and flow
During her period, your daughter may feel tired and moody. Her stomach may swell or cramp. Using a pain scale like this can help her tell you how much she hurts or feels uncomfortable.
Hormonal Management

Consider:
- Weight
- Bones/ fracture risk/mobility
- Risk of clots
- Family history of malignancy
- Need for contraception
- Teen’s understanding/compliance
Hormonal management

- Birth control pills
- Patch, ring: possible removal by patient
- Depo shots: given every four months, concern for bones, 49% bleeding
- IUD with progesterone
- Implants
Birth control pills
Extended/continuous birth control pills

A. Seasonale, Jolessa, Quasense: 84 active pills, then 1 week of inactive
   Period: week 13 (every 3 months)
B. Seasonique, Camresse: 84 active pills, then 1 week low dose estrogen
   Period: week 13, less side effects
C. Quartette: 84 active pills, with gradual increase estrogen
D. Amethyst: continuous for 1 year, no breaks
Mirena

• Contains progesterone levonorgestrel released slowly over 5 years

• Risks: infection, coming out on its own, usually general anesthesia for placement
Other concerns:

- Phenobarbital, Primidone, Topamax: usually require higher doses of hormonal treatments, frequent spotting
- Early puberty: onset of body changes before age 8
- Delayed puberty: no body changes by age 14 or > 5 yrs between breast and pubic hair development
- Periods should occur by age 16 yrs if body changes took place
Considering options

- Family situation and beliefs, cultural needs
- Daughter: intellectual/social level, impact on daily life, medication interactions
- Ultimately choosing what will be best for your daughter's health and quality of life
References


References (2)


Thank you!