

MANAGING YOUR CHILD'S PWS BEHAVIOR

What you wish they told you years ago!

VANDERBILT PRADER-WILLI SYNDROME LONGITUDINAL RESEARCH STUDY

- *Aims:*
- *To phenotype behavior in 166 children and adults with PWS over a span of 5 years to see what behaviors emerge and how these relate to age, gender and genetic subtype*
- *Reality:*
- *Have evaluated 207 children and a lot of older adults with PWS to understand which behaviors we should focus on for possible change and to best help parents and professionals manage these behaviors in many settings.*



THE STUDY COVERS **MANY** DIFFERENT AREAS

- *Cognitive functioning (IQ)*
- *Adaptive functioning-what your child does independently*
- *Maladaptive behaviors*
- *Psychiatric features*
- *Autism features*
- *Interest in food/food seeking*
- *Physical features of PWS- weight, height, bp/hr*
- *Family history functioning (stress) relationships*
- *Medication history*
- *Child daily life characteristics- diet, exercise, screen time*
- *EEG for food and social interests*
- *Blood draw for detailed genetics, measure of genes, and neurochemicals related to behavior*
- *Cortisol measure for stress response.*

PSYCHIATRIC FEATURES IN PWS OF MOST INTEREST

Anxiety

Generalized Anxiety Disorder

Obsessive Compulsive Disorder

Out of 200+ children and adults with PWS

- *About 25% meet criteria for generalized anxiety disorder*
- *Feels anxious or uptight*
- *Can't relax*
- *Intense /lasts for several hours a day*
- *Affects school and home*

Psychosis

Autism Spectrum Disorder

ADHD

Vocal and Motor Tics

- *More (42%) have Obsessive Compulsive Disorder*
- *Have to repeat actions*
- *Arranging or ordering*
- *Hoarding*
- *Have to have things a certain way*
- *Very few have obsessive thoughts-hard to tell*

ANXIETY IN PWS

- *Anxiety is not related to hyperphagia or food seeking behaviors.*
- *Many children with PWS feel anxious most of the time. Dx'd younger age*
- *Manifests with lots of questions*
- *Constant need for reassurance*
- *Insistence on sameness*
- *Meltdowns when expectations aren't met*
- *Shutting down-completely nonresponsive*

PSYCHOSIS IN PWS

HOW COMMON IS IT REALLY?

- *Some have said as high as 80-90% of those with UPD*
- *Very few cases seen with deletion*
- *Genetic predisposition – UPD*
- *Very few descriptive features:*
 - *Hallucinations*
 - *Delusions*
 - *Disorganized thinking*
- *We have seen 74 people with UPD and only 11 have **had** true psychotic features*
- *Delusions/magical thinking*
- *Some hallucinations*
- *Some with Deletion, some with Imprinting Mutations*
- *Precipitating events/stressor*
- *Standardized interviews with parents/ interview with person with PWS*

AUTISM IN PWS

- *More common in those with UPD and boys*
- *Poor social and communication skills*
- *Restricted interests*
- *Rigidity and repetitive behavior*
- *Sensory issues*
- *Poor eye contact*
- *Lack empathy and ability to read social cues*
- *We have found cases in those with Deletion and Imp Mutation*
- *Some have classic Autism and PWS phenotype is overshadowed*
- *Most of cases that meet criteria don't seem to have "true ASD"*
- *We use the Gold standard and multiple raters to ensure accurate diagnosis*

ADHD IN PWS

More common in boys with PWS

Less hyperactivity, but some in younger children

Impulsive, inattentive, distracted

- *Common medications may help, but have some side effects: nervousness, irritable*
- *Non-stimulants can be a better choice*
- ***You have to separate anxiety from inattention- difficult to do***
- *Lots of behavioral intervention work well with meds*

Seems to better by 10-12 years of age, but impulsivity continues

Some are treated with meds, other with environmental means



VOCAL AND MOTOR TICS IN PWS

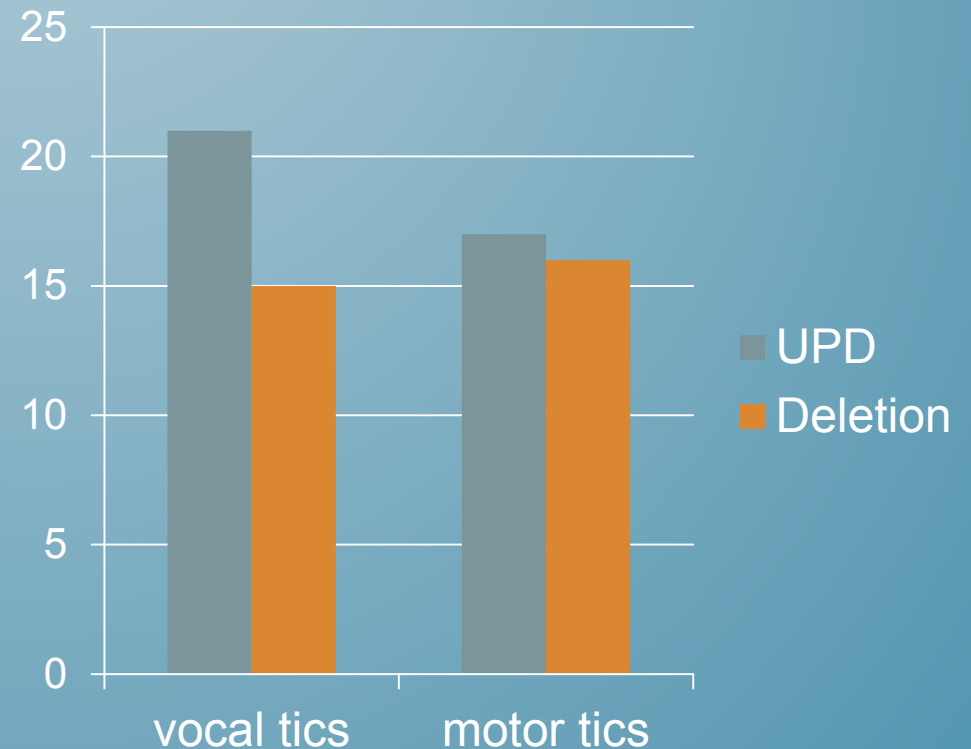
More common than we thought

About 30% of the sample has them-more boys

Vocal: Repeating sounds, words, phrases

Motor tics: movements like touching mouth, nose, or other things

More likely when nervous/excited



SOME OF THE SKILLS YOU NEED TO DO THIS FOR 18 YEARS:

- *Listening skills*
- *Patience*
- *Creativity*
- *Flexibility*
- *Develop trust and long term relationships*
- *What parents need:*
 - *Someone who can really listen and understand their journey*
 - *Skills and proven track record*
 - *Honesty*
 - *Deep understanding and compassion for their child with PWS*

HOW DO I KNOW WHAT IS PWS BEHAVIOR FROM REGULAR CHILD BEHAVIOR?

- **IT doesn't matter!**
- **You have to decide what you can live with and what you can't- different for every family**
- **Then you have to decide what can be changed**
- **Realistic expectations**
- **Developmentally appropriate**
- **How much time you can/will devote to this?**
- **Can you get everyone on board and if not- how much will that affect you?**

PARENTS ARE THE BIGGEST FACTOR IN POSITIVE OUTCOMES OF THEIR CHILD



- *Preston at 9 years*
Weighed 209# and did no exercise- no food locked, very poor diet
- *Preston now at 14 years*
ON GHT, weighs 174#
Food locked, good diet
Plays on several teams and exercises at least 2 hours a day.

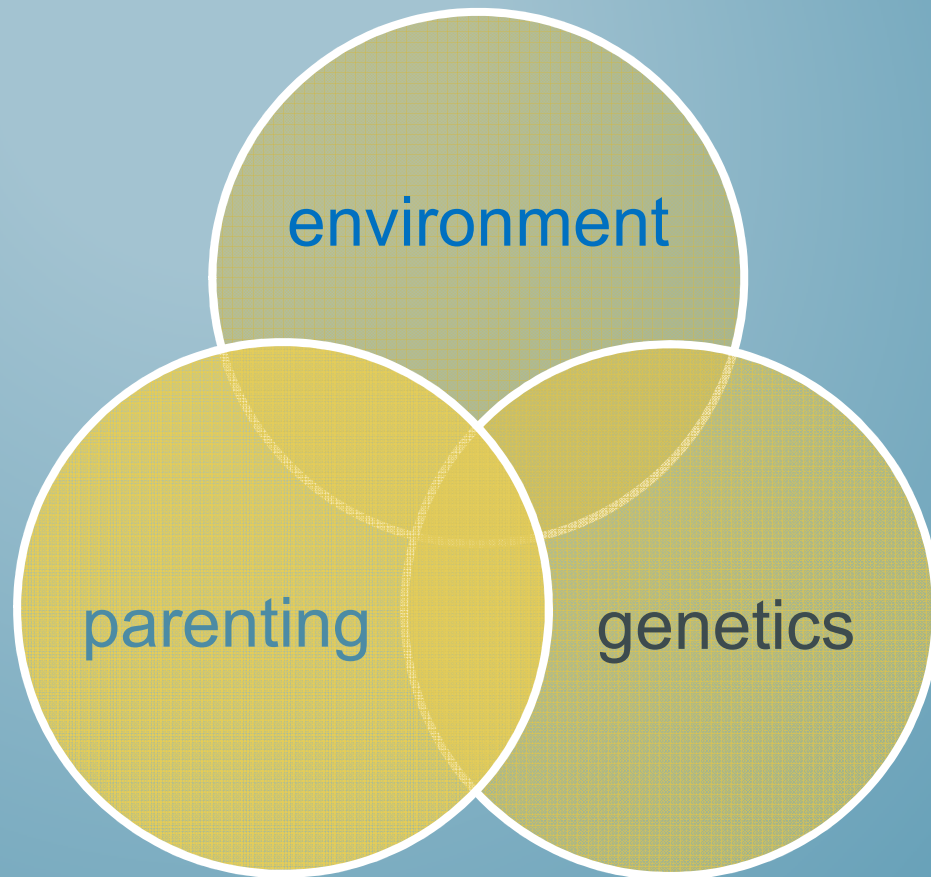
EXAMPLES OF PARENTING: BIRTHDAY PARTY TASK

- Celery: <http://youtu.be/Ww96K3w40PA>
- Salad: <http://youtu.be/LOAJdZTTT1A>
- Cookies: <http://youtu.be/A1UXcfmWNCK>



HOW MUCH
CONTROL
DO YOU
REALLY
HAVE OVER
YOUR
CHILD'S
BEHAVIOR?

Genetics of Behavior



CHARACTERISTICS OF SUCCESSFUL PARENTS



- *They nurture themselves*
- *They balance the needs of their child with PWS and other roles*
- *They continue to adapt to new situations*
- *They have clear, but realistic expectations*
- *They get new information and problem-solve when needed*

*So many worries and
concerns-too much
information- too little
information*

DIAGNOSED

NOW
WHAT?



COMMON BEHAVIOR ISSUES IN CHILDREN WITH PWS

Delays in understanding/motor skills/speech

Compliance issues

Demanding

Temper tantrums

Stubbornness

Emerging food issues

Odd, repetitive behaviors

Sensory seeking behaviors

Emotional outbursts

Shutting down

Aggression/biting and hitting

Avoidant behaviors

ANXIETY


COMPLIANCE
“HOW DO I GET MY
CHILD TO DO WHAT I
WANT?”

*It is vital **HOW** you ask-
SET them up to succeed*

VISUAL SCHEDULES AND NATURAL TRANSITIONS

- *Examples of how to use them effectively*
- *Consistency is the key*

Brush Teeth 

Get toothbrush,
toothpaste, and floss 

Wet toothbrush 

Put toothpaste on brush 

Brush teeth 

Brush front of teeth 

Brush back of teeth 

Brush tongue 

Spit in sink 

Floss teeth 

Throw away floss 

TRANSITIONS ARE TOUGH

- *How to make them easier-
timers, visual cues, verbal cues*
- ***Heads UP!** Have a mantra to
help soothe your child*
- *Don't get too used to a groove-
repetition leads to intolerance
to change*

HARD NOW-
EASY LATER

OR

EASY NOW-
HARD LATER

GETTING DRESSED OR READY FOR SCHOOL

*Emotional Outlook- bright affect,
confident and capable of getting
the best out of your child*



*PWS child- slow, repetitive
behavior, not motivated and has
to do things a certain way*



LOW
EXPRESSED
EMOTION
BUT... BRIGHT
AFFECT,
CALM, AND
CONFIDENT

IF YOU
BELIEVE,
THEY WILL
BELIEVE!



NEGOTIATING=ARGUING AND BARGAINING AND MANIPULATION

- *Be clear about expectations and even get them in writing*
- *Do these before the situation-not on the fly*
- *Use calm and bright affect*
- *If argument starts-DO NOT ENGAGE!*
- *How can they control their emotions if you can't?*



TEMPER TANTRUMS & SHUTTING DOWN- HOW THE FUR WILL FLY!



Don't let them rattle you. You have to stop talking and negotiating, and refuse to engage. When they get you arguing, they wear you down. You can't control their behavior, **ONLY YOURS**. But by controlling **YOURS**, their behavior will change.

SHUTTING DOWN IS COMMON IN PWS

- *What does it mean?*
 - *Overstimulated*
 - *Frustrated*
 - *Too upset to give a response*
 - *Too angry*
- *You cannot make them comply when they shut down- you want to avoid it by reading body cues, voice cues and situational cues*
- *ANTICIPATE Problems and respond- not*
REACT!!!!!!





WHAT WORKS?

Allowing them to calm down

Letting them decide when they are ready to return to situation

Not rehashing or demanding an apology

*You want to return to balance and NOT HAVE TO BE RIGHT! Right means someone is **WRONG!***

TIP SHEET CONSULTATION RESEARCH STUDY

Lauren Deisenroth, RAll in the lab

THANKS TO
THE
FAMILIES
WHO COME
TO SEE US
AND TEACH
US SO
MUCH
ABOUT
THEIR
CHILDREN

