

Managing Adult PWS Behavior

Same song, different verse

“No one knows about adults with PWS, people never thought they would live so long!”

mom of 28 year old daughter with PWS

Adults with PWS are living longer than ever with good health and a better outlook than ever before- Parents need to plan for support throughout their life.



Vanderbilt PWS Research Study

We have seen 74 adults with PWS

- Some have been followed since 1995.
- We have seen them grow up and know what does and doesn't change over time.
- All need some level of support- but level varies.

Little known about physical/mental changes



Often asked questions

- **How long is life expectancy?**
 - It used to be 25 years, but can be average if weight and food access are maintained
- **Can they live independently?**
 - All need some level of support
- **Does behavior get better as they age?**
 - For some, they get much better.
- **Does hyperphagia get better with age?**
 - For some, it does-but not many.

Parenting an Adult with PWS

- Birthday Party Task

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

- Interview

- Self-control: <http://youtu.be/ZfcHWdcBQsg>
- Changes: <http://youtu.be/GklcrLUcZlc>





Adults with PWS

- We assess cognitive functioning
- Adaptive functioning
- Maladaptive behavior
- Hyperphagia and food-seeking
- Psychiatric features: anxiety, depression and psychosis with interviews and other measures
- Autism features
- Independence/family issues
- Family stress and functioning
- Psychiatric medications

Cognition

- Average IQ around 65-70 (our sample ranges from 40-125)
- Relative strengths in visual-spatial, reading, weaknesses in auditory short-term memory/math
- Interference from food seeking and maladaptive behaviors lead to higher levels of care than predicted by IQ alone





Compulsive symptoms

- High rates of compulsive behaviors not related to food, increased risk of OCD
- Hoarding (paper, toiletries, stuff)- 80% of group
- Ordering and arranging by color, size, shape
- Symmetry, exactness (e.g., cutting coupons, stacking, excessive erasing and rewriting)
- Long showers-using a ton of shampoo/body wash
- Some food-related rituals-plates, eat in order
- Skin-picking, hair pulling

% Problems in 185 persons with PWS

| | | | |
|----------------|----|-----------------|----|
| • Skin-picking | 97 | • Impulsive | 76 |
| • Argues a lot | 95 | • Steals food | 72 |
| • Stubborn | 95 | • Compulsions | 71 |
| • Obsessions | 94 | • Anxious | 70 |
| • Tantrums | 88 | • Talk too much | 65 |
| • Underactive | 87 | • Isolated | 60 |
| • Overtired | 81 | • Hoards | 55 |
| • Disobedient | 80 | • Sad | 51 |



Compulsions in PWS

- Can be unusual preoccupations-pregnancy, love interest, slights, obsessions with others
- These may turn into something else-Psychosis?
- Seem to be difficult to curtail-strategies
- Can be fueled by changes, anxiety, too much time alone
- Medications like Celexa can help
- Can sometimes be transformed creatively
- Katherine



What helps?

- Cueing- timers, visual schedules, limits on topics
- Validating feelings and then moving on
- Time outs for Cooling Down
- Being proactive-not reactive
- Behavioral approaches combined with medication, especially those that target compulsive behavior (e.g., SSRI's)
- Applied Behavioral Analysis



Treatment Implications

- Clarify genetic subtype of PWS:
 - Deletions more likely to have increased frequencies of skin-picking, tantrums
 - Those with UPD have higher verbal skills, but poorer visual-spatial functioning
- Young adults with UPD prone to psychotic episodes (ASD also more common in UPD)
- We know very little about Imprinting Defects or Translocations. (psychiatric symptoms)



Prevalence of Psychosis

- Some studies say 80-90% of those with UPD will become psychotic- we saw several people with psychosis who are *not* UPD
- We have had 2 people with active psychosis and 9 with past hx-now either fully or partially resolved.
- Lingering odd thoughts/more common in those with Autistic features
- Medications and changes in environment can be crucial to recovery- Risperdal, Depakote, some SSRI's work well in PWS.



Psychosis in PWS

- Occurs in late teen years to early 20's-some later
- Precipitated by stress- loss of friend/boyfriend, diet/school home changes, worries, transitions
- End of school is a big loss- engaged and active
- Looks disorganized- changes in sleep, diet and thinking-delusions that get bigger
- Need to be on the lookout and monitor for these symptoms when dieting/other stressors occur
 - Act quickly and be relentless



Psychosis in PWS

- Trust your gut/look for changes-don't chalk it up to PWS behavior
- Look for losses/other stressors
- Schedules and consistency can help a lot to recovery
- Sleep/eat and regular scheduled activities
- Supervision/outside help/respice
- Psychiatrists/psychiatric hospitals/inpatient treatment



Anxiety in PWS

- Anxiety is pretty common in children and adults with PWS.
- Many have free floating anxiety which worsens when schedules or expectations change.
- It is important to consider meds when environmental changes aren't enough. Some to consider- Celexa, Lexapro are very effective in PWS.
- Anxiety may look different in this population.



Depression in PWS

- Not as common in PWS-anxiety a lot more common
- Those who become depressed:
 - Females
 - Higher cognitive functioning
 - Those with fewer social connections
 - Family history of depression

It can look very different in PWS- irritable, easily annoyed, no motivation, displaced aggression, sleep changes, or more repetitive behavior

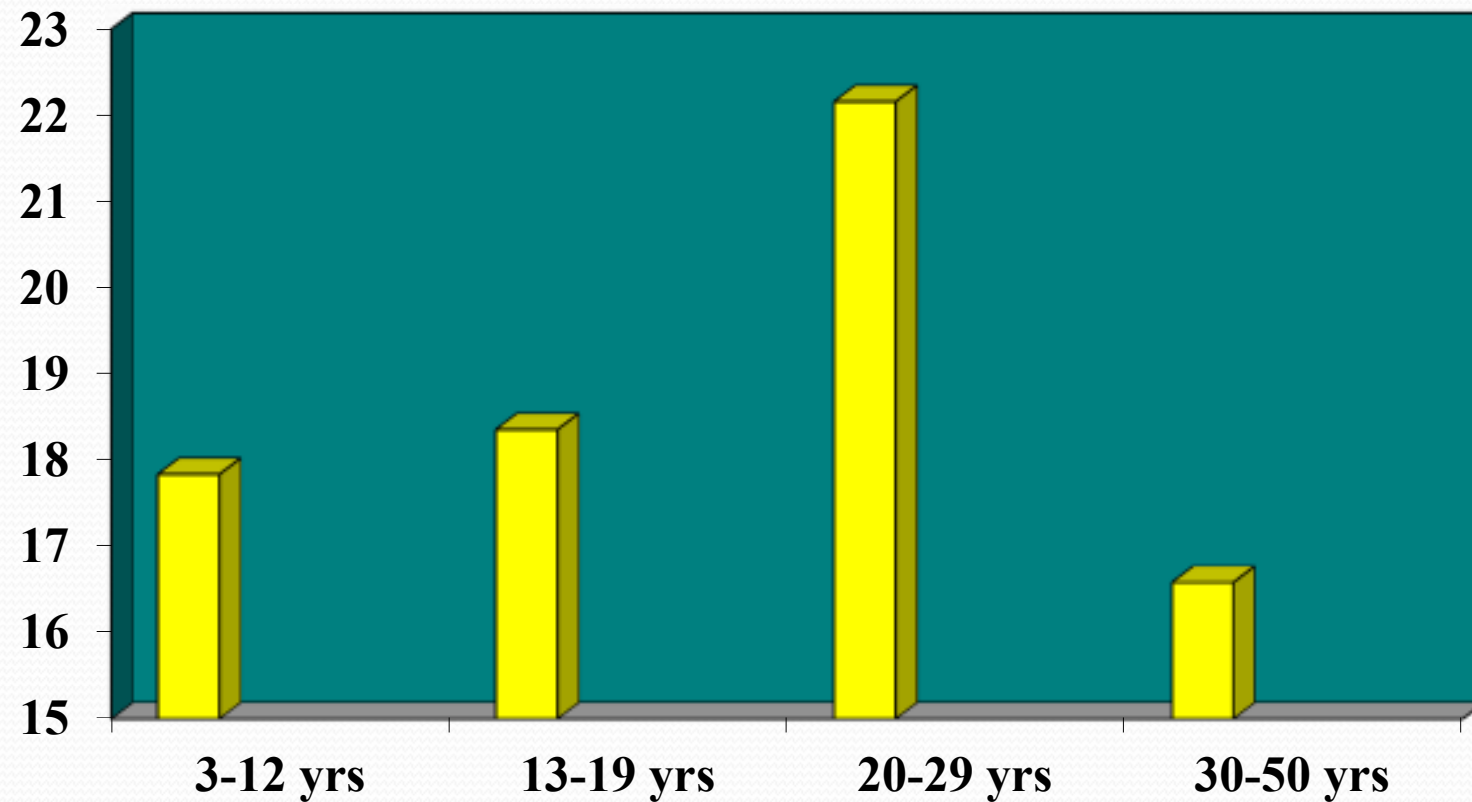


Age-Related Shifts & Mellowing

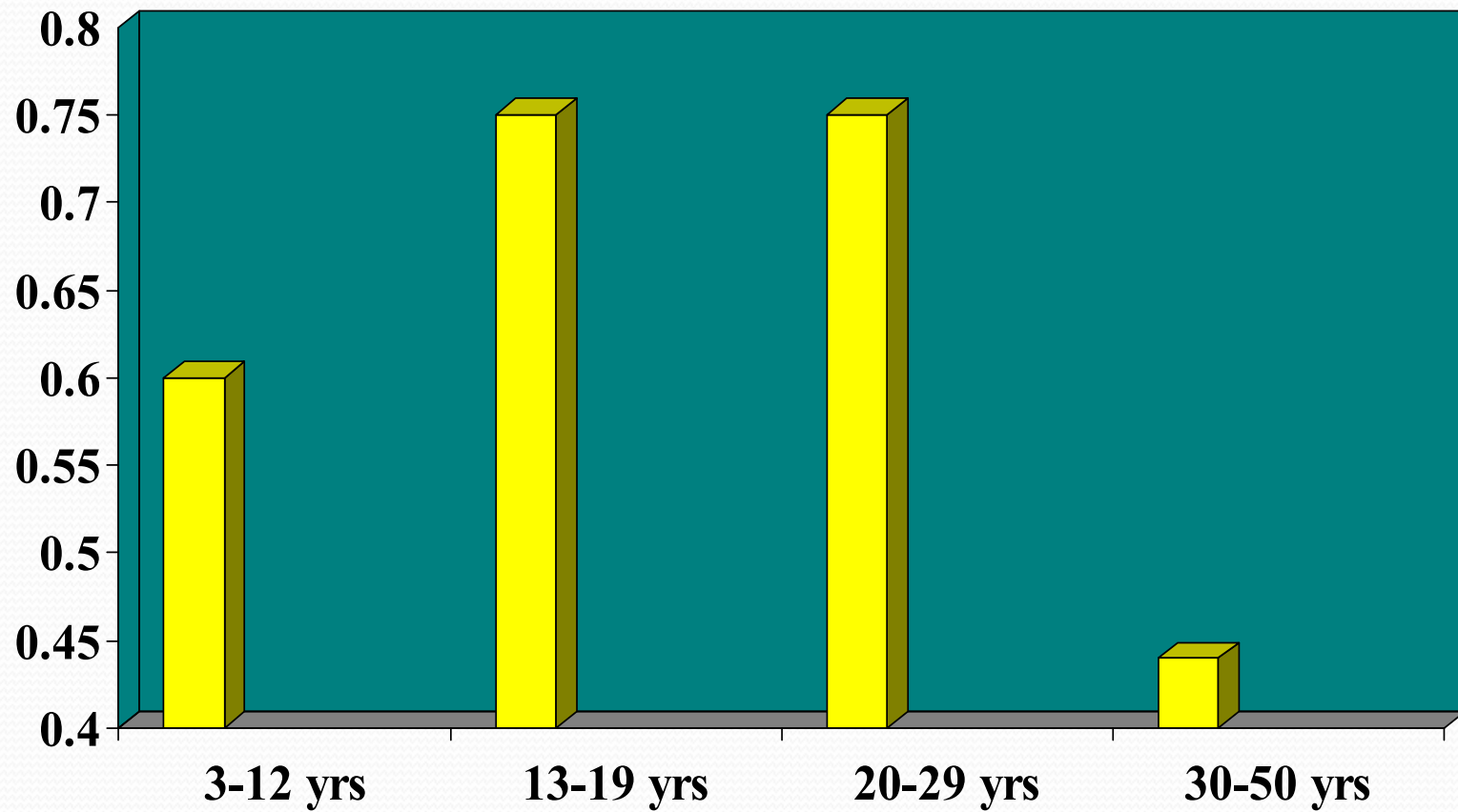
- Age-related increases in many symptoms, those in their twenties are consistently highest
- Remarkable drop in severity and frequency of symptoms in middle adulthood

Subtype differences?

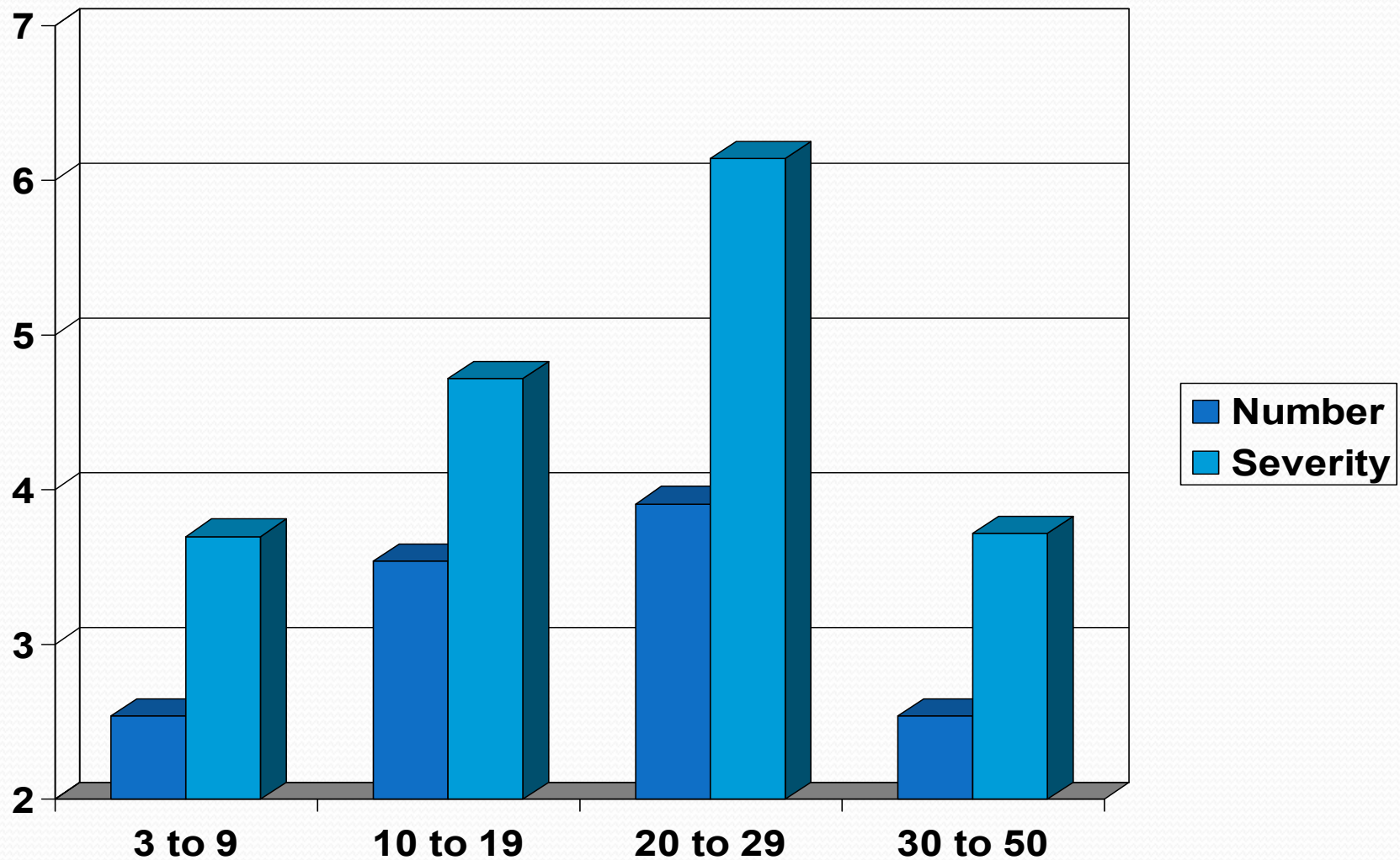
Externalizing Problems in PWS



Skin-Picking in PWS



Compulsive Behavior in PWS





Intervention implications?

- Tailor interventions accordingly?
- Being prepared when problems do occur-teens and early 20's
- Use when counseling families and considering placement choices, including with
 - Siblings
 - Specialized PWS group homes
 - Other group homes
 - Independence

Independence in Adults with PWS



- We have seen some of the oldest adults with PWS.
- They seem to be doing very well both physically and mentally.
- Due to environment and other supports.

Adult Challenges

- Finding a girlfriend:

<http://youtu.be/A1LQYAPpLe4>

- Living independently:

<http://youtu.be/WXzmcQvDSIg>



Can my adult child with PWS live on their own?

- We do not follow anyone with PWS living on their own
- Some live semi-independently and have help
- Biggest predictor is few maladaptive behaviors-not cognitive ability
- Those with best adaptive behavior skills



The Story of Joe

Dx'd with PWS at age 8



Now, 34 years old and is married





Joe's Challenges

- Few behavior problems
- Graduated with help from a private school
- Had several jobs –Walgreens and ARC
- Didn't qualify for any services because his IQ was higher than 70.
- Learned to drive and married his HS sweetheart (also has a disability) and lived in an apartment.
- But his weight kept increasing.....

Joe's Intervention



- Joe had to move home and lost his home and car.
- Joe lost 100 lbs at home and then went to Pittsburgh and lost 65 more.
- Now maintains a weight of 285 lbs.
- Contingency/contract plan

Adults with PWS Need Lifelong Help



- Money access
- Food access
- Job/volunteer placement
- Residential services
- Religious/spiritual life
- Friendships/social outlets
- Exercise/leisure



Residential Services for PWS

- Oconomowoc in Wisconsin
- Alachua County in Florida
- Latham Center in Mass.
- Others in NC, CA and TX
- Use them as a model of comprehensive services throughout life
- Need to plan for that transition
- Need to prepare your child for it, too
- Need input from families and other professionals
- Lobby your state
- Make your voices(collectively) heard to your state

Lifelong Support



- Continue to change as needs change
- Parent towards independence- focus on what they can do for themselves.
- Focus on life skills/ social skills
- Try to let others step in and help you

You are not alone!!



- You have to take care of your needs- both physical and mental
- You have to let go a little bit
- You have to trust others
- You have to trust yourself to do what is best.
- Oh, and you will have to fight like hell to get services.



Consultation and Help

- PWSA has some wonderful resources and crisis help.
- Pittsburgh is very helpful with inpatient hospitalization for PWS.
- We can help with behavioral consultation, even if you are not in the study.
- State organizations and lobbies are the best way to get residential services for your adult child.
- Disability and other legal steps need to be taken early in order to qualify.

It is important to reach out to others to get the information.



Thanks to all of our families!

We couldn't do it without you. We have learned so much about what helps, and what you need, and we admire and respect you so much.