
Another ACE: Growing Up with a Sibling with a Severe Developmental Disability (PWS)

Laura Bennett Murphy, PhD
Associate Clinical Professor of Pediatrics
University of Utah School of Medicine

Emma Thornton
Jane Thornton
Utah State University

DISCLOSURES

- Today's speakers do not have a relationship or affiliation with a commercial interest or organization that would create a conflict or potential conflict of interest for this presentation.
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OBJECTIVES

1. To briefly describe PWVS
2. To describe the current literature on how living with a sibling with developmental disabilities affects psychosocial functioning in healthy siblings
3. To identify ways to assess for PTSD and other psychological concerns among healthy children and adolescents
4. To assess the role of exacerbating and maintaining factors, resilience factors, and suggest appropriate interdisciplinary interventions for families



PRADER WILLI SYNDROME



PWS

- Genetic condition with pervasive effects
 - Chromosomal deletion in a particular region of chromosome 15
 - In about 25-50% of children, it a uniparental disomy at 15
- Early development
 - Often spend some time in the NICU
 - Hypotonic, poor growth, feeding difficulties, delayed motor and developmental skills, mild to moderate cognitive impairment
- Childhood
 - Delays continue, especially speech, developmental of hyperphagia
 - Emergence of tantrums, rigidity, skin picking, compulsive interests and behaviors, sleep abnormalities, stubbornness
- Adolescence
 - Incomplete/delayed puberty, emergence of psychiatric concerns



BACKGROUND

- Children with PWS have multiple medical and developmental needs and those needs change across the landscape of development.
- This devastating disorder affects not only the individual but the entire family.
- Very little research exists to address the ways in which families and siblings adapt

Chronic Illness Literature

Siblings of children with chronic illness have higher rates of distress than the general population

- Siblings have higher rates of depression & anxiety than peers with healthy siblings
- Peer activities and cognitive development scores are lower
- Illnesses and treatment regimes that affect daily functioning have the most negative impact
- However, these findings are modified by lots of factors (family cohesion and control, flexibility, age, birth order, gender).
- Parent reports show worse functioning than child reports

In some groups, parents and siblings are more vulnerable than the ill child

- Dunn et al. (2012) found that 41% of mothers and 30% of fathers report PTSS exceeding clinical cut-offs
- Kaplan et al. (2012) found 60% of siblings scored in the moderate to severe range of PTSS and 22% met full criteria for PTSD

- The Millennium Cohort Study (MCS; UK) found that 14% of older siblings of children with intellectual disability had adjustment problems compared to 9% of those whose siblings did not have ID (Hayden et al., 2019)
- In general, siblings of children with intellectual delays are at greater risk for psychological and social difficulties (Kovshoff et al., 2017)
- Results are typically small and modified by factors like gender, SES, single parent household, and whether or not sibling took care of the child.

- Not surprisingly, siblings of children with autism have more psychological, behavioral, and social problems than their peers where all family members are developing typically.
- Ross & Cuskelly (2009) report that about 40% of siblings developed clinically significant challenges, usually of the internalizing type (anxiety, depression)
- The more aggressive the child with ASD, the more the typically developing sibling struggled, *EVEN THOUGH, they typically attributed the aggression to the illness, not child or self*

What might we expect with PWS?

- Because the child with PWS often has intense and complex health, developmental, and behavioral concerns, siblings are chronically confronted with challenges in multiple domains of life
- The needs and the demands change over time with development – Both the development of the child with PWS and the sibling.
- Two studies suggest that siblings of children with PWS suffer more than those with Down's Syndrome or non-specific delays, even as adults. This is not modified by parenting style or attachment to parents (O'Neill & Murray, 2016)
- Mazaheri (2013) compared 13 siblings of children w/ PWS to those with siblings of chronic illness. 92% of siblings of children with PWS had moderate to severe symptoms of PTSD and quality of life was markedly lower (47% vs. 81 or 62.5% (inpt, outpt))

ACES

American Academy
of Pediatrics



DEDICATED TO THE HEALTH OF ALL CHILDREN™

POLICY STATEMENT

Early Childhood Adversity, Toxic Stress, and the Role of the Pediatrician: Translating Developmental Science Into Lifelong Health

ACES – CYW Questionnaire

Section 1. *At any point since your child was born...*

- 10 ■ ■ Your child's parents or guardians were separated or divorced
- 10 ■ ■ Your child lived with a household member who served time in jail or prison
- 10 ■ ■ Your child lived with a household member who was depressed, mentally ill or attempted suicide
- 10 ■ ■ Your child saw or heard household members hurt or threaten to hurt each other
- 10 ■ ■ A household member swore at, insulted, humiliated, or put down your child in a way that scared your child OR a household member acted in a way that made your child afraid that s/he might be
- 10 ■ ■ Someone touched your child's private parts or asked your child to touch their private parts in a sexual way
- 10 ■ ■ More than once, your child went without food, clothing, a place to live, or had no one to protect her/him^[SEP]
- 10 ■ ■ Someone pushed, grabbed, slapped or threw something at your child OR your child was hit so hard that your child was injured or had marks^[SEP]
- 10 ■ ■ Your child lived with someone who had a problem with drinking or using drugs^[SEP]
- 10 ■ ■ Your child often felt unsupported, unloved and/or unprotected^[SEP]

CURRENT STUDY

Quantitative

- 86 parents, 56 siblings closest in age
- Linked data on 27 dyads
- Parent
 - Family Environment Scale
 - 14 item General Well-Being Scales
 - Youth Outcome Questionnaire (healthy sibling)
- Child
 - Positive and Negative Affect Scale
 - Connor Davidson Resilience Scale
 - Child Report of PTS Symptoms (CROPS)

Qualitative

- 31 participants between the ages of **-18
- Answered 17 open ended questions which were then coded into common themes

INTERVIEWS

What is it like for you being a sibling to your brother/sister with PWS?

Do you have friends over at your house?

What are your feelings toward your brother/sister with PWS?

What are some good things about being a sibling to someone with PWS?



RESULTS



OVERVIEW OF FINDINGS

- Resilience lower 25 (9-38)
- Sibling Distress High 44 (-4 - 135)
 - 20% of parents report moderate to severe levels of distress in their child (>70)
 - 5% of parents recognized suicidal ideation in their child
- PTSD **59% clinically significant**

Predicting PTSD & Sibling Distress

The only variable to predict child trauma in siblings is the ratio of reported positive to negative emotions

Family control and organization predicted overall distress, ie anxiety and depression



FINDINGS FROM THE QUALITATIVE STUDY



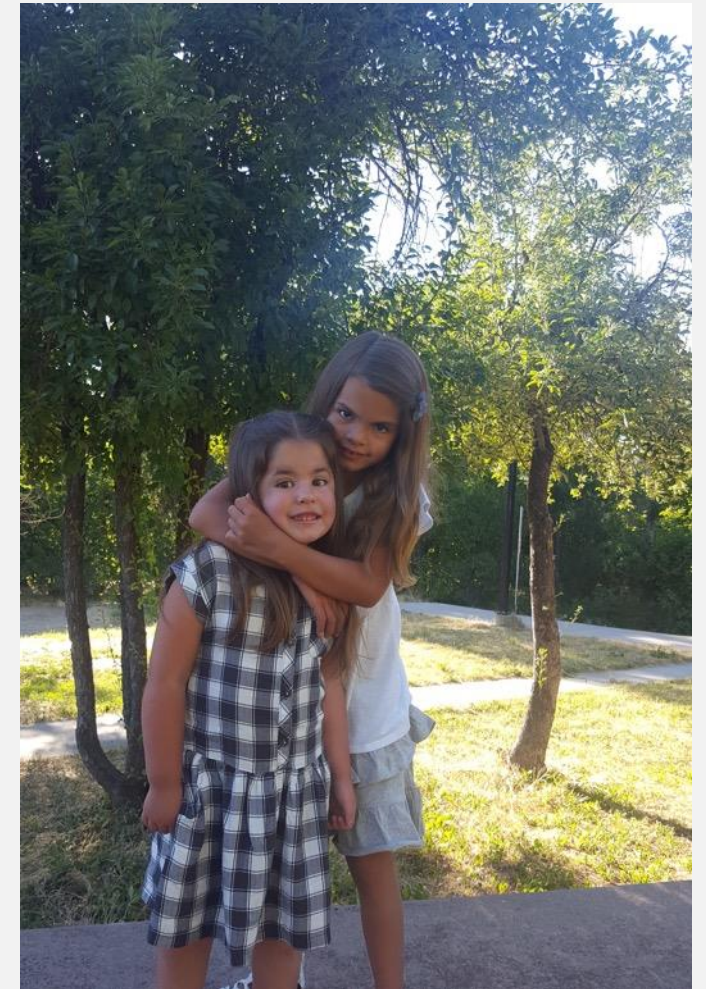
EMOTIONS



- “I am grateful for what she adds to the family.”
- “It is impossible not to love her.”
- “When he is happy, we are happy.”
- “She can always make me happier.”

Relationships With Sibling with PWS

- “She is the royalty and we are her servants.”
- “I wouldn’t want a life without him.”
- 90% of interviewees enjoy being with their sibling with PWS



Safety

“I worry about my sister’s safety. She doesn’t have common sense; she might wander off by herself.”

“I always have to be aware. Ten eyes need to be on her at all times; it is stressful and straining.”

80% of the interviewed siblings claimed they worried about their sibling’s safety often.

77% are often hurt by their sibling.

HOPE



- “It is all worth it.”
- “I don’t like the disability, but I love her.”
- “We are more mature and handle things better.”
- “I can look at different ways to solve a problem.”
- “I’m thankful because he brings blessings and hardships.”

Growth and Resilience

- Patience
- Responsibility
- Understanding
- Gratitude
- Maturity



DISCUSSION



There is no “one” family environment

- Siblings may each experience the family differently depending on their age, gender, age relative to the child with PVVS, genetics, temperament, etc.
- Families exist to support the development of all members
- Resources are limited



Build Resilience



INDIVIDUAL
RELATIONSHIPS



STRENGTHEN
POSITIVE EMOTIONS



ENCOURAGE
HUMOR

Recognize
Distress



Open channels of discussion



Listen, debrief



Allow expressions of negative emotions



Find professionals when needed




CONCLUSIONS



SIBLINGS ARE AT RISK

- Not surprisingly, there is considerable variation among families in parental well being and family functioning
- Psychological distress is high and clinically significant for the majority of the children in our sample
- 60% are reporting clinically significant levels of PTSD symptoms. In our qualitative surveys, many children report witnessing frightening events including: being attacked by sibling with PWS, seeing parents attacked, witnessing medically frightening events, and fears when sibling runs away
- As is observed in other clinical populations, having a sibling with PWS is a significant risk factor for poor adjustment

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- Growing up with a sibling who has PWS places a child at psychological risk. A number of siblings struggle with anxiety, depression, social isolation, and PTSD.
 - However, modifiable factors like family organization, parental well being, and presence of positive emotions in a child's life can foster resilience and protect against distress.

WHAT NEXT?

- How do we define a “medical home”?
 - Who is screening? Who takes ownership? (Not a typical ACE)
 - How are we educating parents? Do we need to give particular attention to subgroups (i.e. UPD)
 - If some risk factors are modifiable, how to we build these?
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- Potential benefits of family centered and wrap around care
 - Need for more local and national supports for siblings
 - Utah’s effort to develop more systematic support and intervention for siblings of children with PWS