

# Prader-Willi Syndrome: A Parent's Perspective

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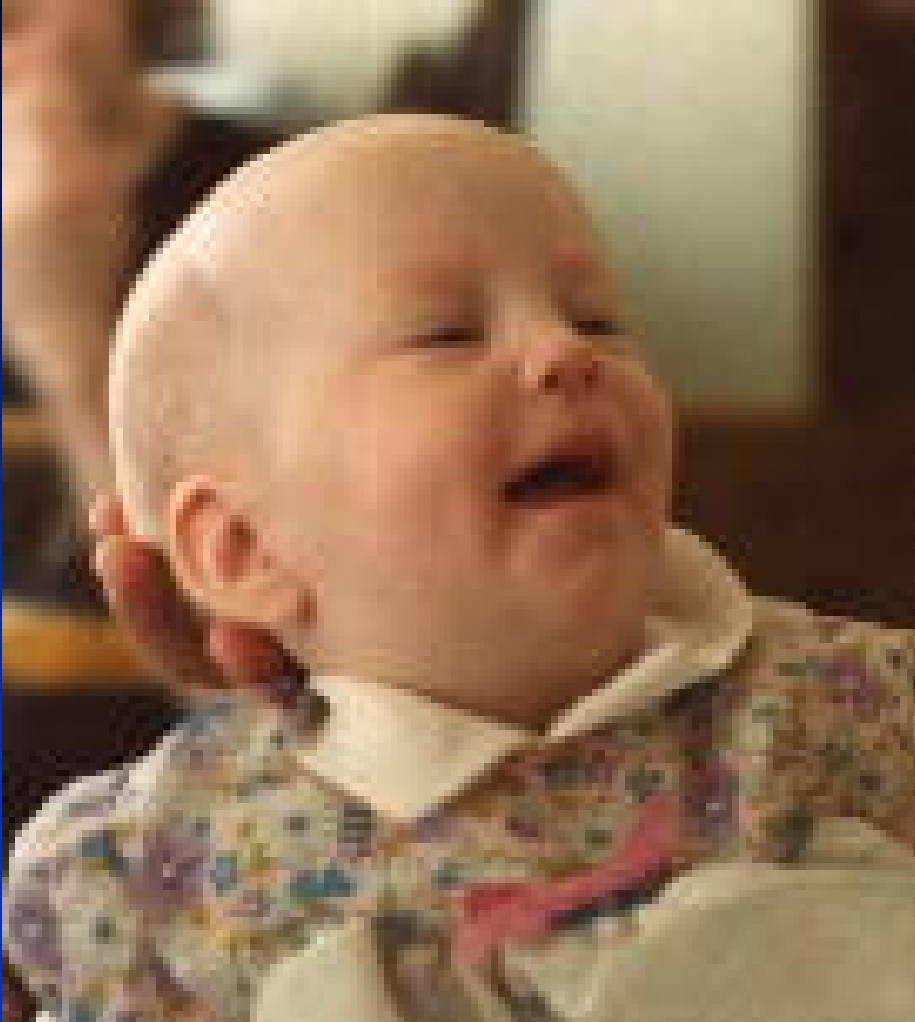
# Karie Sweet 16



# Critical Questions

- What is the most difficult challenge?
- How do we prioritize issues?
- What are realistic expectations?
- How can we maximize success?

# Symptoms during infancy



- Poor muscle tone
- Lethargy
- Poor suck
- Lack of appetite
- Developmental delays

# Developmental Stages



- Infancy (First 2 to 4 years)
  - ◆ Failure to thrive
  - ◆ Below normal weight
  - ◆ Difficulty gaining weight
- Childhood and Adult Years
  - ◆ Insatiable appetite
  - ◆ Tantrums
  - ◆ Learning disabilities
  - ◆ Mental health issues

# Many Therapies



- Speech therapy
- Occupational therapy
- Physical therapy
  
- We taught her to sign
- She learned 24 signs
- Her favorite sign: eat

# Subtle Transition – Age 3



- From sweet and placid to  
Terrible tantrums
- From not interested in food to  
Obsessed with eating
- From “easy baby” to  
Constant crisis intervention

# Weight Management



- Lock up the food
- Count the calories
- Plan meals carefully
- Control the environment

# Symptoms during childhood and adulthood



- Short stature
- Small hands and feet
- Narrow forehead
- CNS dysfunction
- Poor large muscle strength
- Behavior difficulties
- Hyperphagia

# Neurological Dysfunction

- Average IQ: 70 (40-105)
- Functional ability usually below IQ
- Generalized brain dysfunction
- Social development stunted at age 6
- Skilled in getting food
- Difficulty with abstract concepts
- Poor decision making skills, judgment
- Inability to consistently control behavior



# Karie's Special Skills

- Argues like a lawyer
- Picks locks like special agent
- Hears cellophane through walls
- Finds hidden scissors
- Counts calories with 100% accuracy
- Pushes buttons for reaction

# Physiological Concerns

- Decreased metabolism (60%)
- Increased appetite
- Low muscle tone
- Poor balance
- Vision problems
- Low bone density
- Abnormal pain/temperature sensation
- Sexual development



# Conditions and Risks

- Strabismus
- Myopia
- Scoliosis
- Diabetes
- Infertility
- Inability to vomit
- Unusual reaction to medications
- Risk of cardiac problems
- Risk of choking, abdominal rupture
- Risk of accidental suicide



# Psychosocial Concerns



- Emotional immaturity
- Impaired conscience
- Strong compulsions
- Perseveration
- Stubbornness, tantrums
- Low frustration tolerance
- Skin picking
- Risk of Depression
- Potential for Psychosis
- Increased vulnerability

# Effective Intervention



- Understanding
- Acceptance
- Expectations
- Environment
- Focus on prevention

# Common Problems

- Obesity
- Hypogonadism
- Poor motor skills
- Speech/language
- Insatiable appetite
- Poor gag reflex
- Social relationships
- School issues
- Personal stress
- Sleep disturbances
- Rigid thoughts
- Perseveration
- Emotional lability
- Peer interactions
- Poor stamina
- Skin picking
- Stubbornness
- Pain tolerance
- Running away

# What Works for PWS



- **Plans**
  - ◆ Nutrition
  - ◆ Behavior
  - ◆ School
  - ◆ Medical
- **Weight Management**
  - ◆ Balanced calorie-restricted diet
  - ◆ Control access to food
- **Supervision**
  - ◆ 24/7 for life

# Behavior Management: How Behaviors are Perceived

- Laziness, lack of motivation
- Manipulative and controlling
- Lying and stealing
- Lack of empathy, lack of conscience
- Fickle affection
- Stubborn, willfully defiant
- Illogical, unreasonable
- Temperamental, touchy

# Behaviors: What's Really Going On

- Low muscle tone
- Hypothalamus in starvation mode
- Sensory integration disorder
- Low metabolism
- Reaction to medications
- Sleep disorder
- Attachment disorder
- Perseveration, OCD
- Lack of impulse control, poor judgment

# Behaviors: Where Are They Coming From?

- Chromosome defect causes hypothalamus to dysfunction
- Neurological impairment, especially in the frontal lobes (similar to TBI)
- Neurochemical imbalance
  - ◆ GABA neurotransmitters
  - ◆ Serotonin
  - ◆ Adrenaline

# Other Factors That May Affect Behaviors

- Fatigue
- Frustration or stress
- Dehydration or hunger
- Change in routine
- Illness or discomfort
- Broken commitments, changed plans
- Excessive sensory stimulation
- Medications (missed, wrong dose)
- Artificial additives (aspartame, colors)

# Keys to Prevention of Serious Behavior Issues

- Know the warning signals
- Avoid fatigue, dehydration, hunger
- Provide healthy role models
- Resolve “unfair” situations
- Honor commitments
- Give time countdowns and warnings
- Negotiate, use contracts and rewards
- Reasonable expectations,
- Close supervision

# Family Support

- Respect the parents' expertise
- Respect the uniqueness of each child
- Research the web site [www.pwsausa.org](http://www.pwsausa.org)
- Search Google.com "Prader Willi Arizona"
- Read the files, reports, assessments
- Work together as a team
- Become certified for respite care
- Participate in chapter events
- Become a member of PWSAA

# Effective Solutions for Social Relationships

- Counseling - PWS trained professionals
- Minimize outings in public
- Realistic expectations
- Medications (SSRI's are helpful)
- Minimize stress
- Avoid frustration
- Positive role models
- Encourage healthy expression
- Educate all others about PWS

# Effective Solutions for School Issues

- Detailed IEP
- Support of PWS trained advocate
- Training for all staff
- One-on-one aide
- Frequent assessments
- In-house suspension
- Mentor/Buddy
- Reasonable expectations

# Effective Solutions for Personal Stress

- Process grief
- Acknowledge fears
- Avoid isolation
- Detach when necessary
- Don't take it personally
- Recognize burnout
- Seek counseling
- Keep a sense of humor

# Effective Solutions for Sleep Disturbances

- Control weight for apnea
- Encourage physical activity
- Provide interesting diversions
- Allow short nap times
- Provide pillow
- Increase arousal levels

# Effective Solutions for Rigid Thought Process

- Allow discussion of issues
- Set time limits
- Give advance warning
- Encourage journaling
- Provide interesting diversions
- Flexible schedule and routine
- Be careful with promises
- Allow control through easy choices

# Effective Solutions for Verbal Perseveration

- Work toward mutual resolution
- Gentle redirection to another topic
- Repeat what is being repeated
- Allow reasonable time for verbalizing
- Encourage written expression

# Effective Solutions for Emotional Lability

- Acknowledge your feelings
- Verbalize their feelings, self-talk
- Quiet time for self calming
- Request to accompany from room
- Everyone else leave the room
- Keep calm, use soothing approach
- Deep breathing
- Offer reassurance
- Bio-feedback
- Medications

# Effective Solutions for Peer Interactions

- Small groups
- Peer education
- Role playing “What if?”
- Limit social visits
- Acknowledge personality clashes
- Buddy/mentor
- Healthy role models
- Close supervision

# Effective Solutions for Poor Stamina

- Schedule therapy for early in day
- Academic lessons in morning hours
- Schedule after-meal activities
- Elevate arousal levels
- Allow for cyclical down times
- Take frequent breaks
- Allow for rest time

# Effective Solutions for Skin Picking

- Avoid boredom
- Reduce stressful situations
- Minimize alone time
- Redirect to healthy activity
- Encourage bandages and wraps
- Sensory Integration Therapy
- Lotions and skin creams
- Short term rewards
- Medications

# Effective Solutions for Stubbornness

- Pick your issues
- Avoid physical restraints
- Set them up to be winners
- Allow them to refuse sometimes
- Negotiation
- “Let’s Make a Deal”
- Contracts (all parties)

# Effective Solutions for Abnormal Pain Tolerance

- Watch out for “minor” symptoms
- Pay attention to complaints
- Monitor temperature daily
- Increase sensory stimulation
- Adequate fluid intake
- Moderate water temperature
- Close supervision

# Effective Solutions for Running Away

- Avoid physical restraints
- Follow them
- Talk them back home
- Block their way if necessary
- Watch out for the warning signs
- Prevent situations that set up for AWOL
- Close supervision
- Locks and alarms when necessary

# Resources

- National: [www.pwsausa.org](http://www.pwsausa.org)
- Local: [www.members.cox.net/pwsaa](http://www.members.cox.net/pwsaa)
- Youtube channel: SweetKarie
- Google Prader Willi Arizona

