

Tuesday, 3:00 – 4:30, C4

Exploring the Complexities of Supportive Living Options for Individuals with PWS


Jackie Mallow

Objectives:

Identify effective methods for the practical application of concepts related to improving the delivery of services for persons with developmental disabilities

Notes:

**Best Practices –
Exploring the Complexities of
Supportive Living Options for
Individuals Diagnosed with
Prader-Willi Syndrome**
Presented By Jackie Mallow




Prader-Willi Syndrome

▶ **Prader-Willi Syndrome (PWS)** is a complex genetic disorder that typically causes low muscle tone, short stature, incomplete sexual development, cognitive disabilities, problem behaviors, and a chronic feeling of hunger that can lead to excessive eating and life-threatening obesity."

Prader-Willi Syndrome Association (USA)

Prader-Willi Syndrome

First Described in
1956
by three Swiss doctors named:
**Andrea
Prader
Alexis
Labhart
Heinrich
Willi**

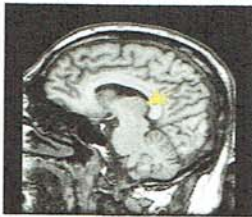


Genetic Explanations

- **Paternal Deletion:**
(about 70%) - a deletion (break in band) on the 15th chromosome contributed by father; non-inherited- the deletion occurs at time of conception.
- **Maternal Disomy:**
(about 20-25%) - two 15th chromosomes are contributed by the mother & no contribution by father.
- **Imprinting:**
(about 2-5%) - "an error in the 'imprinting process' that renders the paternal contribution nonfunctional"

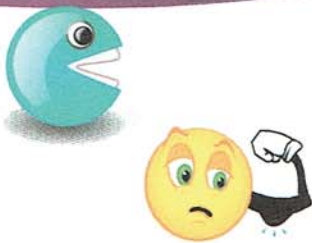
The 4th Explanation (Not Genetic)

- **Acquired:**
trauma to the hypothalamus (injury, surgery, tumor...)

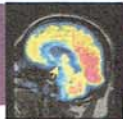


The Three Major Characteristics of PWS

- ▶ **Hyperphagia-**
insatiable appetite
- ▶ **Hypotonia-**
poor muscle tone
- ▶ **Hypogonadism-**
underdeveloped genitalia



What We Know – The Hypothalamus



The **hypothalamus** is a tiny (size of thumb nail tip, weighing ¼ oz.) brain structure located between the thalamus & brainstem.

Functions include:

- Controls Appetite
- Body Temperature Regulation
- Water Balance
- Influences Blood Pressure

The Hypothalamus



The **hypothalamus** is also involved in the regulation of:

- Sexual Behavior
- Aggression
- Fear
- Sleep (in communication with the pineal gland)

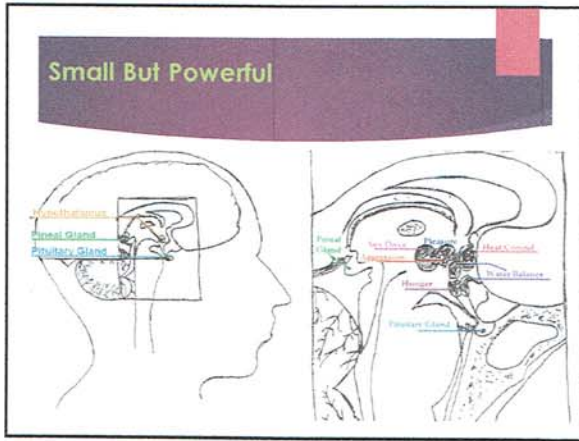


The Hypothalamus




- The **hypothalamus** controls the pituitary gland, which produces hormones that affect:
- Growth
- Sexual Development
- Metabolism





Special Health Concerns

- Restricted Diet
- High Pain Threshold
- Altered Temperature Regulation
- Stomach /GI Issues
- Choking
- Low Muscle Tone
- Low Stamina
- Skin Picking
- Bruising
- Daytime Sleepiness
- Strabismus
- Scoliosis
- Dental Issues



Behavioral Obstacles

- Sequential Processing Deficit
 - ✓ If and Then
- Impulsivity
 - ✓ Action Before Thought
- Rigid Thought Process
 - ✓ Stuck Thinking
- Version of Truth
 - ✓ Miss representation
- Sensory Issues
 - ✓ Overload
- Social Awareness
 - ✓ Poor Insight
- Medical Challenges
 - ✓ Hunger
 - ✓ Sleep

Precipitating Factors



- ▶ Environment
- ▶ Health
- ▶ Cognitive Functioning
- ▶ Relationships
- ▶ Peers
- ▶ Family
- ▶ Staff
- ▶ Weather
- ▶ Change
- ▶ Medications
- ▶ Work
- ▶ Loss/Death
- ▶ Moves
- ▶ Seasons
- ▶ Money
- ▶ Spirituality
- ▶ Sleep Patterns
- ▶ Perceptions
- ▶ Social Events
- ▶ Hunger
- ▶ Psychological
- ▶ Psychiatric
- ▶ Unknown
- ▶ Community
- ▶ Holidays


Types of Living Environments

So where do we go with what we know...

- Type of Supports
- Integrated or Specialized
- Number of Roommates
- Location
- Apartment
- Homes
- Semi Independent
- Resident Rights




Best Practices



To provide quality care for individuals diagnosed with PWS a program must have the basic guiding principles in place...

** Individual's opinion should be taken into consideration **

- Environmental Supports – 24 hours
- Highly Trained Caregivers
- Comprehensive Medical Services
- Continuity of Services & Care





Best Practices

- > Home Like Environment
- > Community Integration
- > Diet & Exercise
- > Personal Growth & Development
- > Maintaining Healthy Relationships
- > Promote a Safe & Supportive Environment




Best Practices

- > Communication & Information Systems
- > Routines & Consistency
- > Rules & Guidelines
- > Motivational Plans
- > Behavior Management Plans
- > Crisis Plans


Resources for Training – Initial & On-Going

- > PWSA-MI 517-764-2483
- > PWSA (USA) www.pwsausa.org
- > Videos – PWSA (USA) On line publications
- > Books – Best Practice Guidelines
- > PWSA(USA) Gathered View
- > PWSA Informational Brochures
- > PWS Local & National Conferences
- > Parents
- > Other Professional Providers

Training

Knowledge is the backbone of a training program.

