Exploring the Complexities of Supportive Living Options for Individuals with PWS

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

Notes:
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)
**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"

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**The 4th Explanation (Not Genetic)**

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

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**The Three Major Characteristics of PWS**

- **Hyperphagia:**
  instable appetite

- **Hypotonia:**
  poor muscle tone

- **Hypogonadism:**
  underdeveloped genitalia
What We Know – The Hypothalamus

The hypothalamus is a tiny (size of thumb nail tip, weighing 1/4 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
Small But Powerful

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
  - Misrepresentation
- Sensory Issues
  - Overload
- Social Awareness
  - Poor insight
- Medical Challenges
  - Hunger
  - Sleep

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Precipitating Factors
- Environment
- Health
- Cognitive Functioning
- Relationships
- Peers
- Family
- Staff
- Weather
- Change
- Medications
- Work
- Loss/Death
- Moves

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) [Website URL]
- Videos – PWSA (USA) Online publications
- Books – Best Practice Guidelines
- PWSA (USA) Gathered View
- PWSA Informational Brochures
- PWSA Local & National Conferences
- Parents
- Other Professional Providers