Aging in Prader- Willi Syndrome

2025 United In Hope Conference

Objectives

Recognize	Recognize key aging-related changes in individuals with PWS.
Understand	Understand the physical, emotional, and behavioral challenges associated with aging in PWS.
Apply	Apply best practices in managing aging-related care needs in the group home setting.
Foster	Foster a supportive and respectful environment for older individuals with PWS.

Introductions

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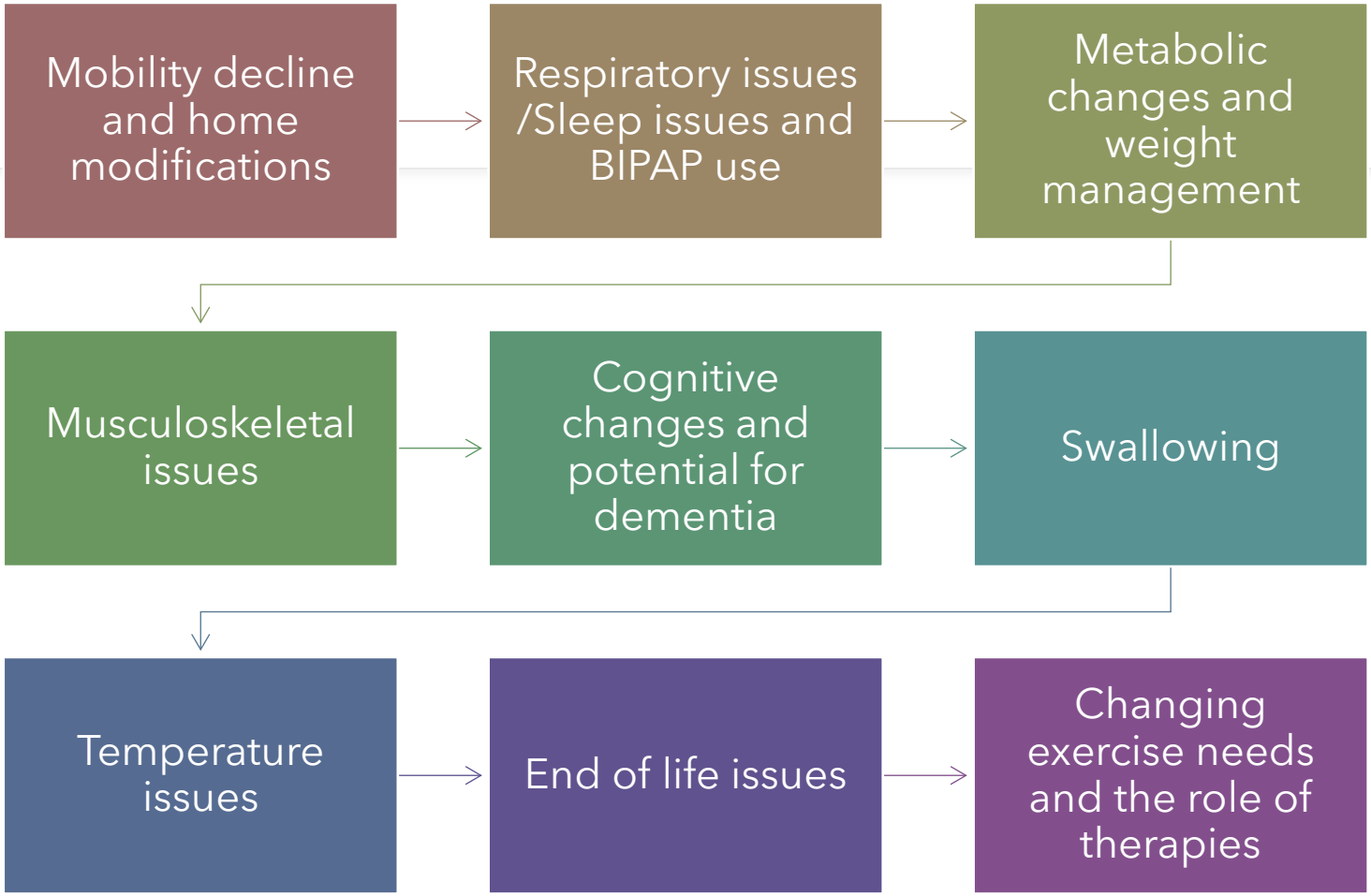
Where are you from?

What is your area of practice?

What's one thing you've noticed about aging in the individuals you support?

Understanding Aging in Prader-Willi Syndrome



Mobility Decline and Home Modifications

Due to improvements in medical care, early diagnosis, better management of symptoms, and supportive living environments, individuals with PWS are living longer.
What are we seeing?
Accelerated Aging.

Mobility Decline Factors

- Decreased muscle tone
- Abnormal Gait
- Small hands/feet
- Obesity

Supportive Devices

- Walkers
- Wheelchairs
- Transport wheelchairs
- Sit to stand machine
- Ceiling/freestanding lifts
- Shower chairs
- Ramps
- Vehicles

Respiratory issues

- Sleep-Disordered Breathing
- Obstructive & Central Sleep Apnea
- Hypoventilation during sleep
- Impaired Respiratory Drive
- Reduced response to low oxygen/high CO_2
- Increased risk for pneumonia

Respiratory issues: contributing factors

- Muscle Weakness & Hypotonia
- Ineffective ventilation
- Poor cough and airway clearance
- Scoliosis & Chest Wall Deformities
- Restrictive lung disease
- Reduced lung expansion

- Higher risk of pneumonia and aspiration
- Poor swallow coordination
- Sedentary lifestyle and weak cough effort
- Sedating meds (e.g., antipsychotics) worsen risks

Respiratory Management strategies

- Sleep Studies and use of CPAP/BiPAP
- Weight management & nutrition
- Vaccinations: flu & pneumococcal
- Chest physiotherapy, assisted cough devices
- Avoidance of sedatives where possible
- Consider continuation of GH therapy where appropriate

METABOLIC CHANGES AND WEIGHT MANAGEMENT

- Aging may result in Type 2 Diabetes Mellitus
- Higher risk of metabolic syndrome, especially with obesity
- Cardiovascular diseases
- Body composition changes—decrease in lean body mass, increase in fat mass
- Endocrine dysregulation
- Potential for increased age-associated diseases, especially if not treated with GH

Musculoskeletal Issues in Aging Individuals with PWS

Key Risk Factors:

- Hypotonia (low muscle tone)
- Growth hormone and sex hormone deficiencies
- Reduced mobility and physical activity
- Nutritional challenges (e.g., low calcium/vitamin D)
- Obesity

Common Issues:

- Osteopenia/Osteoporosis → Fragility fractures
- Scoliosis/Kyphosis → Pain, breathing issues
- Joint laxity → Dislocations, early arthritis
- Fatigues easily/ difficult to motivate to exercise

Musculoskeletal management strategies

Monitoring:

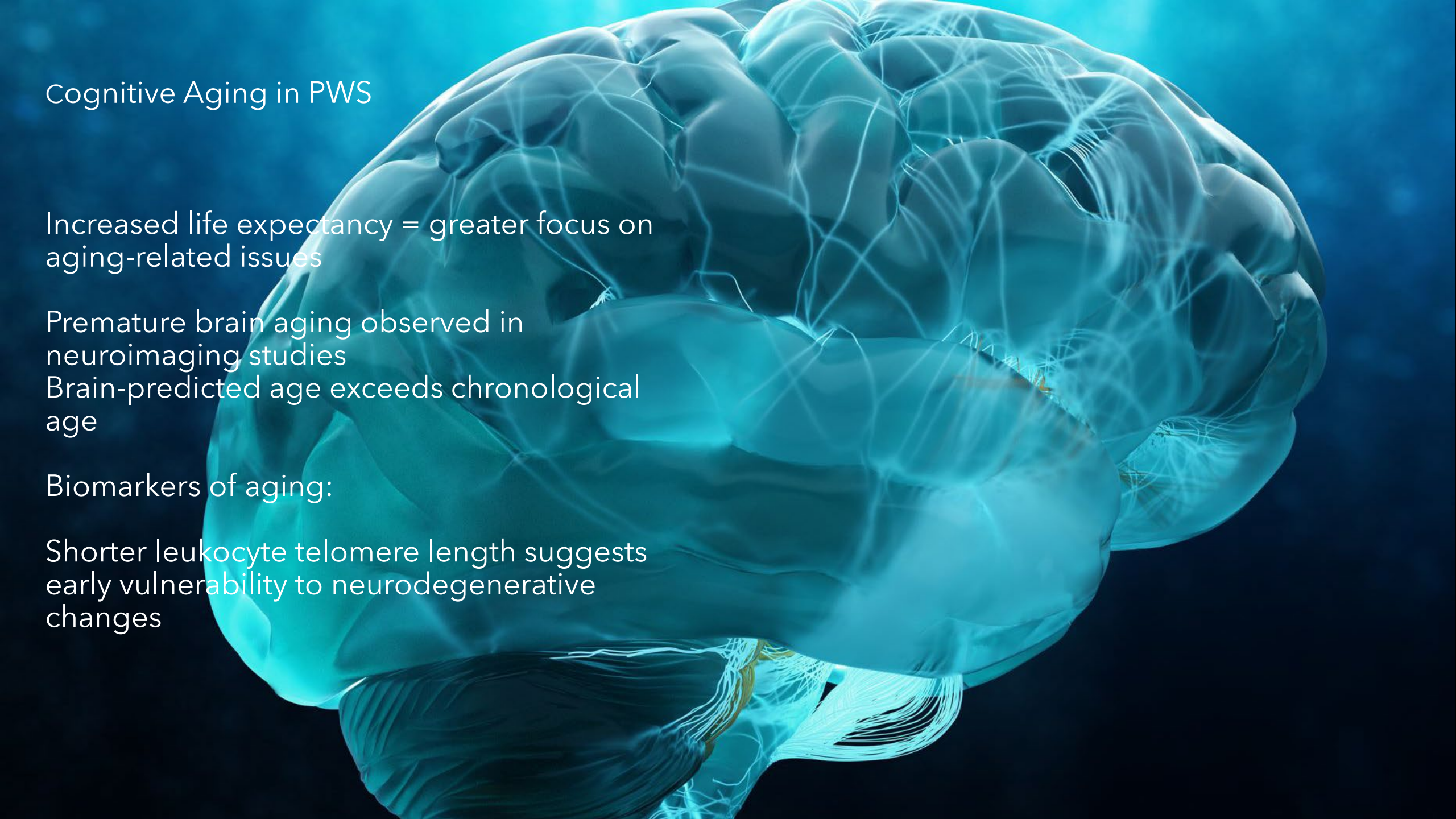
- DEXA scans for bone density/ routine labwork

Orthopedic evaluation for scoliosis and joint health

- Therapies:
- Physical therapy for strength, flexibility, and posture develop HEP
- Hormone replacement (GH, testosterone/estrogen)
- Calcium + Vitamin D supplementation

Multidisciplinary Approach:

- Endocrinologist, orthopedist, therapist, dietitian, behavioral team
- Emphasize early detection, consistent activity with focus on core strength and balance



Cognitive Aging in PWS

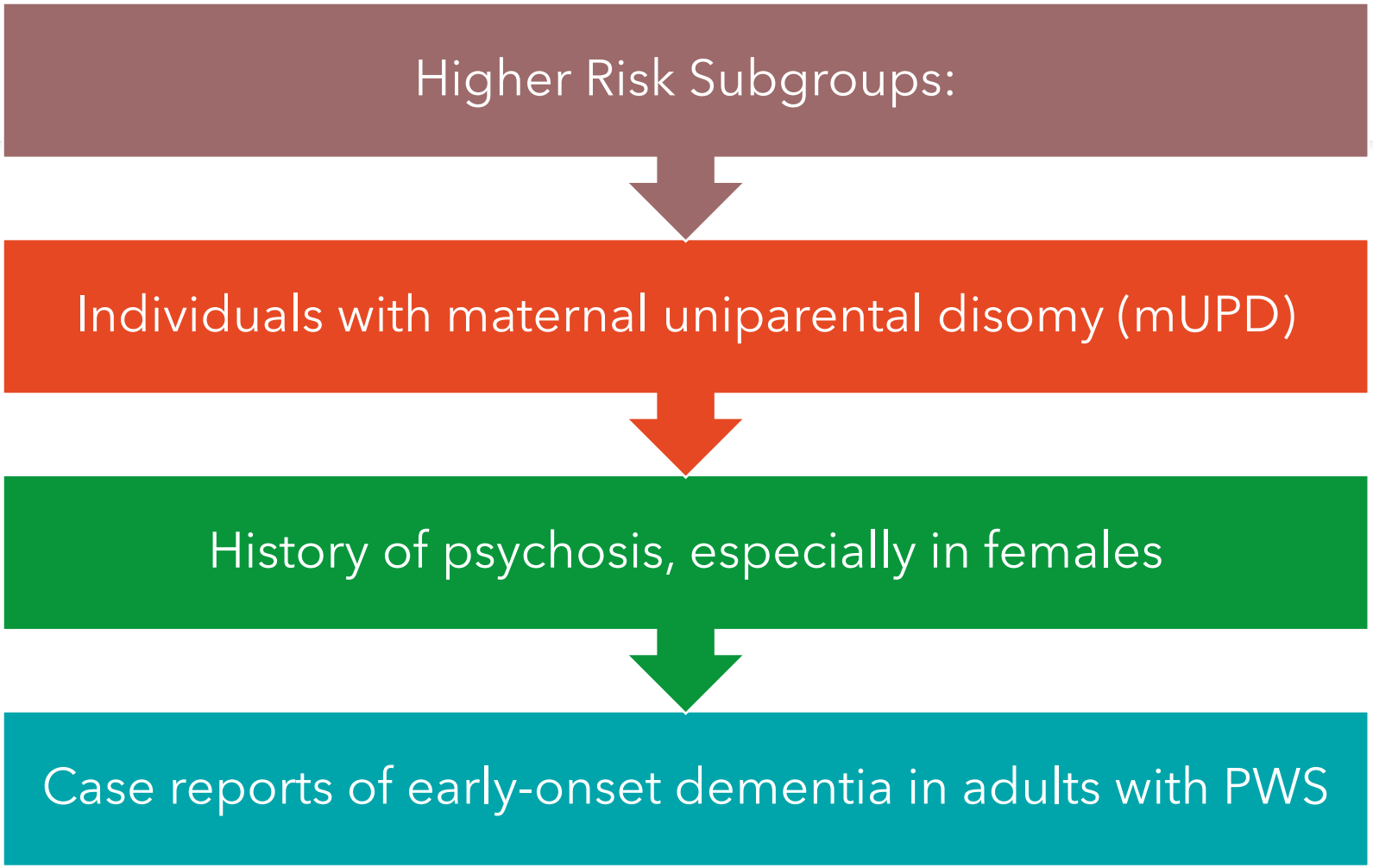
Increased life expectancy = greater focus on aging-related issues

Premature brain aging observed in neuroimaging studies
Brain-predicted age exceeds chronological age

Biomarkers of aging:

Shorter leukocyte telomere length suggests early vulnerability to neurodegenerative changes

Dementia Risk & Cognitive Testing



Cognitive Tests for Individuals with Intellectual Disability

Specialized Assessments:

- Dementia Questionnaire for People with Learning Disabilities (DLD)
- CAMDEX-DS (for those 30+ with ID)
- Vineland Adaptive Behavior Scales (Vineland-3)

Adapted Tools:

- Modified WAIS and BADS for attention, memory, executive function

Best Practices:

- Establish cognitive baseline in early adulthood
- Re-test every few years to track individual changes
- Multidisciplinary approach recommended

INCREASED ISSUES WITH TEMPERATURE DYSREGULATION

- Overall prone to overheating or becoming dangerously cold can increase with age
- Increased risk of malignant hyperthermia with age
- Absent Fever with infection
- How to manage?
- Be aware of the rituals and routines and set limits
- Be attentive to possible illness in other ways
- Know baseline temperature

End of Life Issues

- Palliative and symptom management
- Feeding and nutrition
- Emotional and psychosocial issues: Grief and Loss
- Collaboration with healthcare services
  - Hospitals
  - Skilled nursing facilities
  - VNA services

MANAGING EXERCISE NEEDS/ROLE OF THERAPIES IN AGING PERSONS WITH PWS

- Inactivity and obesity related health issues can lead to decreased mobility
- Muscle loss and weakness can impact endurance and performance
- Scoliosis can increase with age, as can complications like Type 2 diabetes
- Therefore, the role of all therapies become even more important to address…

- Mobility and strength
- Daily activities
- Muscle mass
- Home safety

Increase and maintain independence

Sustain/improve quality of life

Cognitive functioning

Improve balance

Group question

What are some of the most concerning aspects of aging in PWS that you have noticed?

Supporting health and wellness in aging adults with PWS

Case presentations with group discussion

Case study 1: Aging Adult with Prader-Willi Syndrome and Complications from Aspiration Pneumonia

Name: Michael
Age: 54 years
Diagnosis: Prader-Willi Syndrome (PWS), confirmed deletion subtype
Living Situation: Group home for adults with PWS
Medical History: Obesity, Type 2 Diabetes, GERD, mild scoliosis, mild intellectual disability
Functional Status Prior to Hospitalization: Independent with ADLs with staff support, attended a day program, stable in weight for the past 2 years, exercised 6 days a week with staff support

Acute Event: Michael was admitted to the hospital after group home staff noticed him becoming increasingly fatigued, shortness of breath and having increased irritability. Upon arrival to the ED, Imaging confirmed **right** lower lobe pneumonia. He became increasingly hypoxic and required oxygen supplementation which progressed to needing mechanical ventilation for 7 days.

During hospitalization: Michael was noted to cough while drinking fluids. A speech-language pathologist (SLP) was consulted, and a swallow study revealed **s**ilent aspiration of thin liquids. This likely contributed to the pneumonia. He was placed on a Level 2 dysphagia diet (mechanical soft solids, nectar-thick liquids) and received antibiotics and supportive care. He was discharged to a rehabilitation facility for intensive physical and occupational therapy.

While in rehab:

-He gained 40 pounds in 6 weeks, partly due to lack of consistent food security and difficulty enforcing his PWS-specific nutritional plan.

-He showed reduced mobility and endurance, attributed to deconditioning, weight gain, and reduced participation in physical therapy.

-rehab staff noted increased behavioral issues, including food-seeking, irritability, and resistance to therapy—possibly exacerbated by change in environment, reduced activity, and unfamiliar caregivers. The group home staff attempted to visit frequently and to educate staff and dietitian relating to behavioral and food security needs of individuals with PWS.

Discharge: He was discharged back to the group home with a significantly higher need for care than before including- continuous oxygen, PT, OT exercised to complete three times daily, need for a walker for ambulation, change in diet.

- How do we train staff to recognize change in status, and when to intervene
- How do we communicate with hospital staff the needs of the PWS individual
- How can staff advocate in both hospitalization and rehab placement for behavior/safety/food security and meal planning
- What would need to be done to prepare and train staff for the increased needs of the individual upon return to the home

Case study 2 : Aging adult with complicated hospital course

Name: Lauren

Age: 55 years

Diagnosis: Congenital Malformation Syndrome (Prader-Willi Syndrome), Bi-Polar 1 Disorder, Mild Intellectual Disabilities, High cholesterol, Constipation, Flat feet-Left club foot, Unsteady Gait, Osteoporosis, Hypertension, Non-specific T wave abnormality, Astigmatism, Seborrhea Dermatitis, GERD, Vitamin D Deficiency, Allergic Rhinitis, Nightmare Disorder

New after hospital: Cardiovascular Disease, Congestive Heart Failure (Diastolic), Obstructive Sleep Apnea, Chronic Hypoxemic and Hypercarbic Respiratory Failure

Living Situation: Group home for adults with PWS

Medical History:

Functional Status Prior to Hospitalization: Independent with ADLs with staff support, used a walker as needed when outside the house, attended a day program.

Acute Event: 55-year-old female who lives in a group home with past medical history of Prader-Willi syndrome, bipolar disorder, ADHD, sinus bradycardia. She had recent hospitalization last month with COVID pneumonia, with hospital course complicated by acute blood loss anemia secondary to hepatic artery pseudoaneurysm rupture. During that hospitalization she required 6 units PRBC transfusion, and underwent left hepatic artery embolization, CHF exacerbation, NSTEMI and was discharged on 02/24. She returned on 3/10/25 with fatigue, generalized weakness, shortness of breath, cough with yellowish sputum. Oxygen saturation found to be 83 % on room air. In the ED, CTA of the chest which was negative for PE but showed multifocal pneumonia. She was given broad spectrum antibiotics and admitted to the hospital.

On hospital day 2 in the morning she was noticed to be hypotensive with systolic blood pressures in the 70s. She was asymptomatic with this. She was given 1 L IV fluid bolus with no improvement in blood pressure. She was transferred to ICU for further care in the setting of possible shock. 3/11 CT A/P finding of incidental pseudo aneurysm noted of gastric artery and fusiform aneurysmal dilatation of intrahepatic hepatic arterial branch. 3/12 S/ P embolization of left hepatic artery fusiform aneurysm on 3/12 by IR Dr. Shah.

During hospitalization- social worker recommended a short term rehab due to Lauren needing PT, OT, SLP and oxygen.

While in rehab:

Was sent to the hospital on 3 different occasions for shortness of breath and low oxygen. Rehab unable to raise oxygen levels, unclear on BiPap availability/and use- Lauren reported that she felt unwell and shortly after remembers passing out. She woke up to the EMTs after receiving Narcan. Lauren was brought to the ICU but later transferred to general medicine floor a day or so later. She was weaned off supplemental oxygen during day time hours and was directed to use her BiPap machine nightly. Upon arrival to Tobey she was retaining fluids, she since diuresed which in turn helped with the concerns for chronic heart failure and her ability to breathe easier. She was treated for aspiration pneumonia and her diet was downgraded to pureed and nectar thick liquids. Still admitted as of 4/27/25

· She was discharged home with a significantly higher need for care than before , to a new group home. Non invasive home ventilator, PT OT SLP VNA services, Dietary guidelines changed to pureed and nectar thickened liquids.

· How do we train staff to recognize change in status, and when to intervene

· How do we communicate with hospital staff the needs of the PWS individual

· How can staff advocate in both hospitalization and rehab placement for behavior/safety/food security and meal planning

· What would need to be done to prepare and train staff for the increased needs of the individual upon return to the home

Case study 3: Aging adult with a small bowel obstruction

Name: Robert
Age: 62years
Diagnosis: Prader-Willi Syndrome
Living Situation: Group home for adults with PWS
Medical History: Obesity, Type 2 Diabetes, Obstructive sleep apnea, chronic constipation,

Functional Status Prior to Hospitalization: Independent with ADLs with staff support, attended a day program, history of weight gain during home visits

Acute event: Robert was brought to the emergency department by his group home staff after returning from an unsupervised weekend visit with extended family. According to family reports, Robert was left unattended briefly and accessed a large amount of food, including high-fat, high-fiber items such as nuts, cheese, pastries, and leftover meat. Over the next 12 hours, he began experiencing progressive abdominal distension, pain, nausea, and had not passed stool or gas. Group home staff noticed his discomfort and brought him to the hospital.

During hospitalization: Robert was admitted to the surgical service and initially managed conservatively with bowel rest, nasogastric decompression, and IV fluids. Due to worsening abdominal tenderness and increasing lactate levels, a decision was made for exploratory laparotomy. A large, dense mass of undigested food material was found in his colon and 12 cm of colon reeded to be removed.

Following surgery, Robert began picking at his surgical site which led to infection. He also became increasingly behavioral because of new dietary restrictions. He was ultimately discharged back to the group home.

- How do we help the family plan for food security during home visits?
- What would need to be done to prepare and train staff for the increased needs of the individual upon return to the home?
- How can staff help the hospital staff decrease behavioral episodes while inpatient?

Mental health and emotional well-being

How aging effects emotional regulation

Strategies for managing anxiety, rigidity and new behavioral concerns

Maintaining dignity and identity in later years

HOW AGING EFFECTS EMOTIONAL DYSREGULATION IN PWS

- Some characteristic behaviors (aggression, impulsivity) may lessen in older adults; other issues like rigidity and stubbornness may persist
- Some may experience even greater difficulties recognizing and interpreting emotions in others
- Some studies suggest individuals with PWS can experience premature brain aging, contributing to behavioral and cognitive changes
- May have a decline in physical functioning, increased risk of age-associated diseases like diabetes and cardiovascular disease
- Optimal approach is to support across the life span, using behavioral therapies early, promoting adaptive scheduling and supporting the entire family.

Maintaining dignity and identity in later years

- Even if cognitive limitations exist, offer *choices* (e.g., clothes, activities within limits).
- Continue honoring their preferences, routines, and interests.
- Be conscious of language (diaper vs. brief)
- Adapting care thoughtfully
- Maintain privacy: In caregiving (e.g., bathing, dressing), explain what you're doing and give them control when possible.
- Facilitate connection with friends, family, or peer groups to reduce isolation.
- Encourage meaningful activities.

Communication and end of life planning

- Approaching aging conversations with residents and families
- Legal and ethical considerations (guardianship, advance planning)
- Supporting a program after a loss

Approaching aging conversations with residents and families

- Start early!! These are not conversations you want to have in the middle of a crisis.
- Know the individual, include them in an appropriate manner related to their cognitive level, emotional sensitivity, and behavioral patterns.
- A realistic understanding of what aging with PWS may look like.
- Support in planning for transitions: medical needs, residential changes, guardianship, end-of-life care.

Legal and ethical considerations

- Guardianship limitations
- Code status
- Medical interventions
- Clear documentation

Supporting a program after a loss

- Grief may not look “typical” – behavior is often the primary language of loss.
- Individual's responses to grief will vary.
- Consider communication within the program
- Support each person where they are at.
- Plan a celebration of life ceremony, offer space for shared grief and legacy, like planting a tree or naming a bench.
- You may utilize outside resources .

Practical tools and staff self-care

Checklists and observation tools for aging signs and plug workgroup on aging

Communication logs and team coordination

Importance of staff mental health and burnout prevention

PWS Aging Signs Checklist for Residential Care Providers

Monthly / Quarterly Observation Tool

Cognitive & Mental Health Changes

Increased confusion or forgetfulness		Neuropsych eval	
Decline in problem-solving or planning		Track cognitive tasks	
Changes in language or word finding		Speech therapy referral	
New or worsening anxiety or fears		Psychiatry consult	
Withdrawal from preferred activities		Behavioral health	
New or worsening OCD behaviors		Medication/CBT review	

Mobility & Physical Function

Decrease in walking speed or endurance		PT referral	
Increased falls or unsteadiness		Fall risk assessment	
Loss of muscle tone/strength		Review GH, PT plan	
Difficulty with stairs, rising from chairs		Functional mobility plan	
Worsening scoliosis/postural changes		Ortho review	

Cardiovascular and Metabolic Health

Fatigue or shortness of breath		Cardiology workup	
Swelling in legs or ankles		Check cardiac/kidney	
New/worsening hypertension		Medication review	
Changes in cholesterol/glucose		Lab monitoring	
Unexplained weight loss		Nutritionist referral	

Bone Health & Endocrine Signs

Frequent fractures or bone pain		DEXA scan	
Shortened height/spine changes		Endocrine consult	
Loss of GH therapy benefits		GH dosing review	
Signs of menopause/andropause		Hormone panel	
GI slowdown or constipation		GI consult/diet review	

Sleep & Breathing

Increased daytime sleepiness		Sleep study	
Snoring or apnea symptoms		BiPAP/CPAP review	
Resistance to sleep equipment		Re-education/device check	
Frequent waking during the night		Sleep hygiene adjustment	
Morning grogginess		Adjust sleep schedule	

Sensory & Neurological Health

Vision changes		Eye exam	
Hearing issues		Audiology referral	
New tremors/coordination issues		Neuro consult	
Headaches, dizziness, seizures		Urgent medical eval	

Social, Emotional, and Behavioral

Reduced social interaction		Increase engagement	
More rigid/resistant to change		Behavioral support	
Regression in self-care		OT reassessment	
Increased emotional outbursts		Medication/therapy review	
Paranoia or psychosis symptoms		Urgent psych referral	

Q & A

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