EXERCISE FOR PEOPLE WITH PRADER-WILLI SYNDROME

A guide for families and individuals

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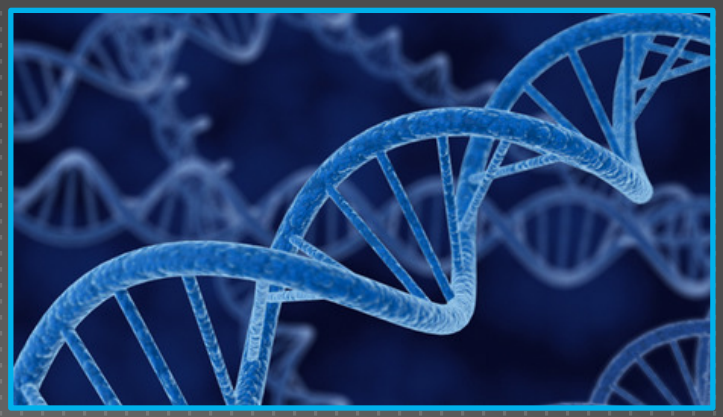
OBJECTIVES

- Gain an understanding of the anatomical and physiological processes that affect individuals' physical capabilities with Prader-Willi Syndrome (PWS)
- Understand the physiological processes that predisposed people with PWS towards obesity
- Summarize recent studies regarding exercise and its effects for those with PWS
- Provide an efficient and understandable guideline for families/individuals regarding exercise for individuals with PWS
- Empower all disciplines to provide basic information regarding exercise for those with PWS



Pathophysiology of Prader-Willi Syndrome (PWS)

THE BASICS

CAUSES, SIGNS, AND SYMPTOMS

- Genetic error on chromosome 15
  - Disruption of hypothalamus function
  - Normally controls *hunger*, *thirst*, indirectly controls *growth* and *sexual development*

- **Hypotonia** → low muscle tone
- **Distinct facial features** → almond eyes, narrowing at temples, turned-down mouth, thin upper lip
- **Hyperphagia** → food craving/lack of satiety
- **Hypogonadism** → Underdeveloped sex organs
- **Poor growth**
- **Global developmental delays**
- **Sleep disorders**
- **Behavior problems**

DIAGNOSTIC CRITERIA

- Suggested New Criteria to Prompt DNA Testing for PWS

Age Noted	Signs/symptoms
Birth to 2 years old (y.o.)	• Hypotonia w/ poor suck
2y.o. to 6 y.o.	• Hypotonia w/ history of poor suck • Global developmental delay
6y.o. to 12 y.o.	• History of or present hypotonia w/ poor suck • Global developmental delay • Hyperphagia w/ possible obesity
13y.o. through adulthood	• Cognitive disabilities (usually mild) • Hyperphagia w/ possible obesity • Hypothalamic hypogonadism and/ or behavior problems

Prader-Willi Syndrome Association website. http://www.pwsausa.org/about-pws/testing/diagnostic-criteria. Accessed March 19th, 2015

Standard of Care for PWS

MEDICAL MANAGEMENT

EVIDENCE-BASED RECOMMENDATIONS

- Early Intervention Services
  - Therapies, educational, parent support, nutrition
- Growth Hormone (GH) treatments
- Sleep studies
- Orthopedic treatment (scoliosis)
- **Obesity management**
- Sex steroids replacement (adults)
- Psychology/behavioral treatment

PT + PWS

- Early Intervention Services
  - For developmental milestones, strengthening, mobility
- But mostly → exercise for obesity management!

THE VICIOUS CYCLE
OBESITY + PWS

- What we know →
  - Children with PWS are already at high risk for obesity due to hypotonia, decreased GH, and hyperphagia
- Plus → also shown to have underlying muscle fiber distribution abnormalities
  - Makes movement even more difficult
  - Can decrease typical caloric expenditure
  - Altered resting metabolic rates
  - Increased fat mass = decrease in lean body mass = low energy expenditure
- And then → the more weight they gain, the more difficult it is for them to get adequate physical activity, furthering the weight gain

- Children with PWS are genetically predisposed to having a harder time losing and/or maintaining weight
  - Need only ~60% of typical caloric intake of age- and gender-matched peers to maintain current weight
  - Naturally have lower resting metabolic rate than either normal weight or even obese age- and gender-matched peers without PWS even when adjusted for fat-free mass.

- **"Accompanying obesity can increase energy cost of gait and daily activities and lower energy reserve for educational, social, and recreational activities".**[1]
  - And have reduced energy reserves due to GH deficiency
    - Even with long-term GH treatment, only partially helps
  - Many have some delays/intellectual disabilities, so they will need all the energy they can get for daily academic tasks

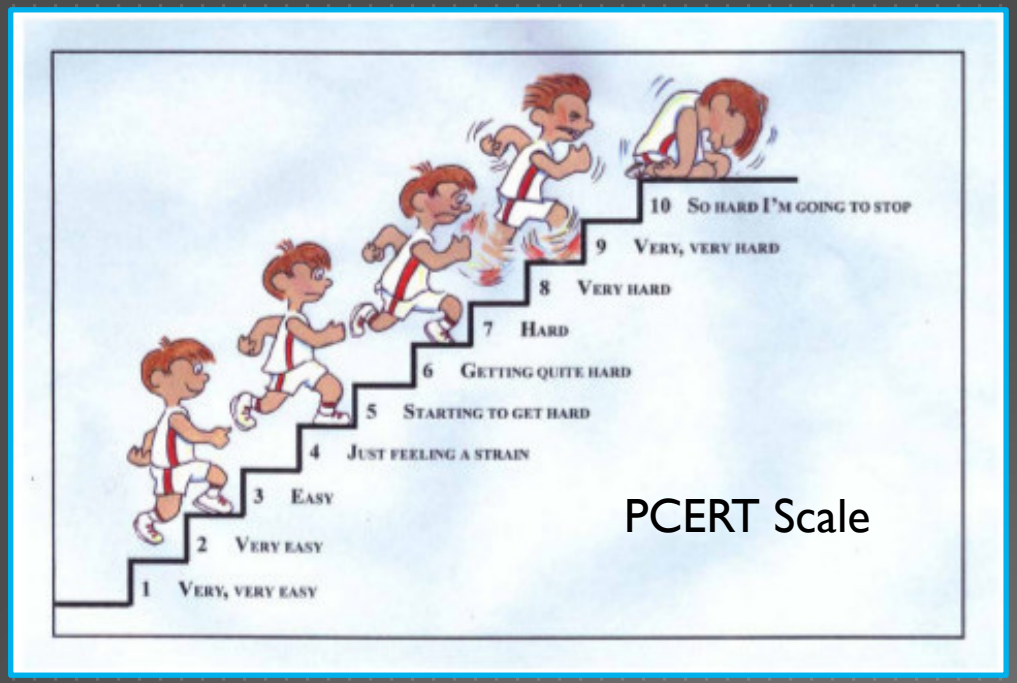
SO WHAT CAN WE DO?

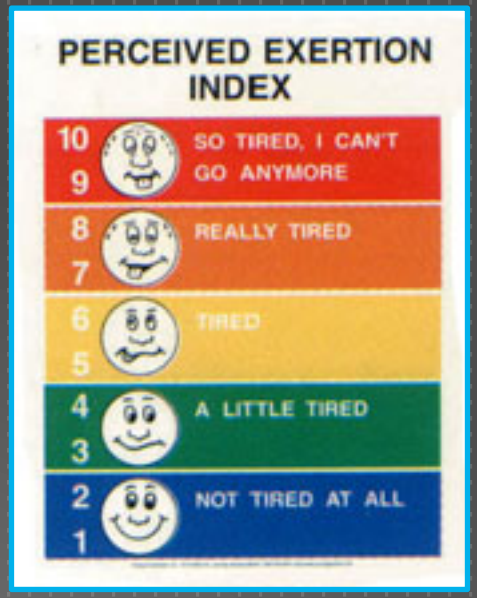
MAKE EXERCISE A DAILY ROUTINE!

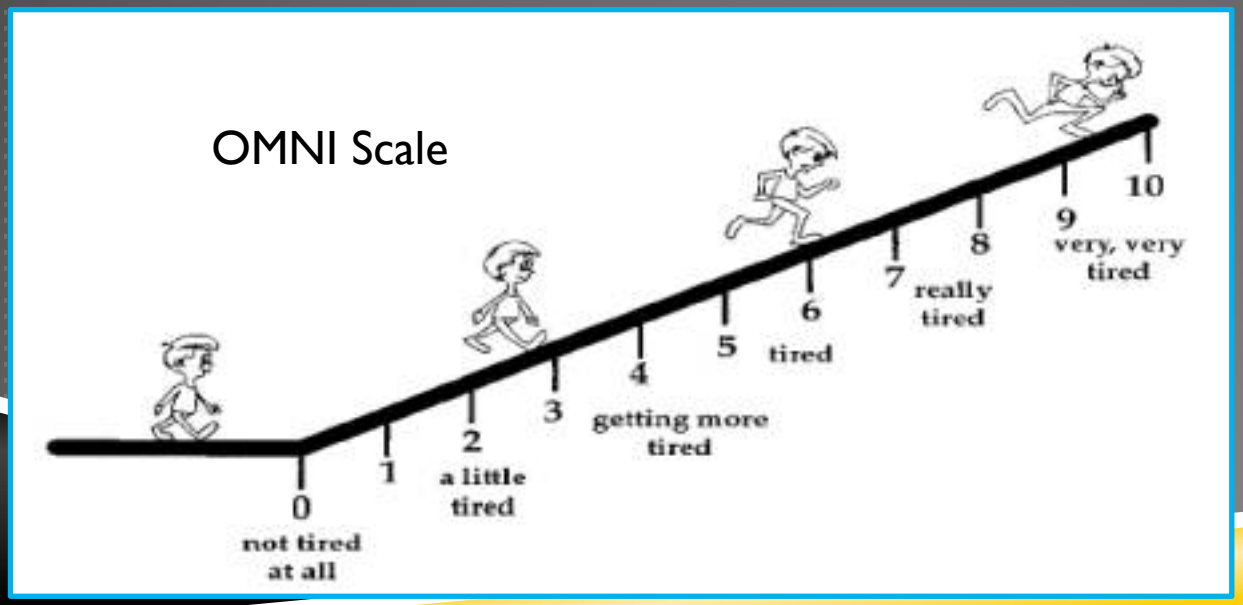
- Children with PWS need **daily physical activity/exercise** throughout their lifetime.
  - The earlier they can get used to exercising daily, the better.
  - If they start regular vigorous exercise before weight gain, it will be much easier to maintain or even lose weight as they age
- Activity Guidelines for Americans →
  - Minimum of 60 minutes of moderate-vigorous combo or vigorous only physical activity (PA) daily
    - Can be done in 10-15 minute bursts throughout day
  - Need 3 types of activity →
    1. Aerobic
    2. Muscle Strengthening
    3. Bone Strengthening

FITTE PRINCIPLES FOR EXERCISE

- Frequency → Daily
  - At the very least 3 days/week
- Intensity → Combo of moderate-vigorous or all vigorous physical activity
  - Can use scales to determine intensity
    - OMNI, PCERT, or PEIF scales (see pictures)
- Type → Whatever they enjoy!
- Time → 60 cumulative minutes
  - If low activity tolerance, start at 20 minutes and work way up
- Enjoyment → add in anything to make it fun!
  - Props, games, music, movement, silliness, etc.







- Need to be at 6-9 out of 10 to gain health benefits

SUGGESTED ACTIVITIES

Suggested Activities for Strength Training at Various Ages

Ages	Activities			
	Upper Limbs	Lower limbs	Trunk	Respiration
Younger children				
Wheelbarrow walks		Squats	Sit-ups	Blowing bubbles
Push/pull a wagon		Vertical jumping	Bridges	Straw sucking
Throwing		Stair climbing	Log rolling	Blowing balloons
Vertical drawing		On tiptoes	Trunk rotations	Singing
Lifting objects		Ball kicking	Stand from floor lying	Chair push-ups
Scooter board		Walking sideways	Swing a weighted bat	
Older children/ younger adolescents	Use of dynabands, therabands, and hand weights while performing upper limb, lower limb, and trunk resistive range of motion exercises to music or in games or dance.			
Older adolescents/ young adults	Strength training with use of weight machines and free weights requires a spotter. Specific exercises for upper and lower limbs and trunk include biceps and hamstring curls, triceps and quad extensions, latissimus pulls, abdominal crunches, squats, and toe rises. Monitoring blood pressure during weight training programs in those who are obese is important because of risk of developing hypertension.			

For strength training → can use body weight first, then add resistance/weight

SUGGESTED ACTIVITIES

Suggested Activities for Aerobic Conditioning at Various Ages

Ages	Activities
Younger children	Bunny hopping, running long jump, running up and down steps or incline, running up and down hills, riding a tricycle, sitting on a scooter board and propelling with the feet
Older children/younger adolescents	Bike riding, stationary bike riding, brisk walking, water aerobics Other Activities: As appropriate for skill level & geographical location—rolling skating, roller blading, ice skating, cross-crossing country skiing, downhill skiing
Older adolescents/young adults	Same as older-children age group, plus dancing, low-impact step aerobics, aerobic circuit training

BASICALLY…

- Doing anything that gets them moving, breathing heavier, and with a faster heart rate counts!
- Get family activities going → studies show that kids are more active and healthier if their parents/families are as well

Exercise Benefits for People with PWS

CURRENT EVIDENCE

EVIDENCE

- "Prader-Willi Syndrome: A Review for Pediatric Physical Therapists"
  - Cynthia L. Lewis, PT, PhD at Elon College
- In studies reviewed →
  - Children with PWS have significantly ↓ energy expenditures and resting metabolic rate (RMR) than children without PWS
  - "Vigorous exercise can raise RMR by approximately 10%, but mild to moderate exercise has minimal effect on RMR."
    - ↑ levels of activity increased total energy expenditure and aided in controlling weight
  - Children with PWS who exercised regularly = ↓ resting HR, ↑ aerobic capacity, ↓ percentage body fat, and weight loss
  - May reduce risk for scoliosis and childhood-onset osteoporosis

EVIDENCE

- "Improving Body Composition and Physical Activity in Prader-Willi Syndrome"
  - Foundation Growth Puberty Adolescence and Bethanien Private Clinic, Zurich, Switzerland and Institute of Human Genetics and Anthropology, Jena, Germany
- PWS is the most common syndromal cause of obesity
- Even long-term GH treatments only partially compensate for defect of lean muscle mass.
- Study completed →
  - Kids with PWS wore pedometers for 3 consecutive days, compared against control
  - Filled out activity protocols to record/rate each half-hour (scale 1-4)
  - Muscle strength training program for 3 months with heel raises on stairs
    - 1x/day; 3sets of 12 (36 reps total)

EVIDENCE

- "Improving Body Composition and Physical Activity in Prader-Willi Syndrome"
- Results →
  - With heel lift training program, calf circumference significantly ↑ and calf skinfold significantly ↓ in PWS group
    - Indicates gain of muscle mass and loss of fat
  - Physical capacity significantly ↑ in both groups (PWS and control)
  - Mean walking distances over 3 consecutive days:
    - PWS = 11.1 km (6.89 miles)
    - Control = 24.6 km (15.28 miles)
    - Controls walk over 2x as much as PWS on a typical day

EVIDENCE

- "Improving Body Composition and Physical Activity in Prader-Willi Syndrome"
- Discussion →
  - "Considerable spontaneous hypoactivity in PWS despite long-term growth hormone treatment and in the absence of severe obesity".
  - Muscle mass increased similar amount as controls with strength exercise
    - Spontaneous hypoactivity likely large part of decreased muscle mass rather than muscle disturbance at cellular level
  - Significant ↑ in spontaneous PA and physical capacity at end of program
    - Daily walking distance ↑ 45.1% to 70.7% for PWS group
    - Accomplished with only 3-minutes a day training program (heel raises)
  - Retested 3 months after end of program →
    - ↓ calf circumference and ↑ calf skinfold in both groups
    - Significantly ↓ physical capacity but still higher than baseline (long-term effect?)

EVIDENCE

- "Participation, Preferences, Perceived Barriers and Perceived Benefits of Physical Activity in Individuals with Prader-Willi Syndrome: A Parent's Perspective"
  - California State University
- "Physical activity or exercise is part of the recommended standard of care."
- Survey of parental perceptions of enrolling child with PWS in physical activity program →
  - **Benefits**:
    - Overall program participation (77%)
    - Improvement of motor skills/balance (78.2%)
    - Improvement of stamina/strength (74.4%)
  - **Barriers:**
    - Time commitment (<40% agreed/strongly agreed)
    - Travel to program site (<40% agreed/strongly agreed)
    - Problems with program facilitators (15.1% agreed/strongly agreed)

EVIDENCE

- "Participation, Preferences, Perceived Barriers and Perceived Benefits of Physical Activity in Individuals with Prader-Willi Syndrome: A Parent's Perspective"
  - **Facilitators:**
    - Financial support (>70.1% agreed/strongly agreed)
    - Needing more time in personal schedule (54.7% agreed/strongly agreed)
    - Needing more equipment (51.2% agreed/strongly agreed)

- Conclusions → parent's/caregiver's time and financial support are key barriers to physical activity participation

For individuals with PWS and their families

HANDOUT OF EXERCISE GUIDELINES

EXERCISE HANDOUT

- Created a handout for families and individuals at Prader-Willi Clinic at the CIDD
- Includes basic information about why exercise is so important for PWS, what is recommended, and ideas for activities
- Benefits of having a handout:
  - Can be a reminder about exercise recommendations between clinic visits
  - Gives basic education about physiology behind obesity with PWS
  - Provides concrete reasons why obesity with PWS can be harmful/limiting
  - Allows other healthcare providers to provide information to families regarding exercise for PWS without substantial knowledge base needed

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