The Power of Exercise for People with PWS

Georgina Loughnan
Prader-Willi Syndrome Clinic
Royal Prince Alfred Hospital
Sydney, Australia
Physical Features seen in People with Prader-Willi Syndrome (PWS)

Clinical Features
- Lower muscle tone
- Decreased % muscle
- Reduced muscle strength

Functional Impact
- ↓ resistance to movement
- Loose muscles
- Less stable joints
- Poor balance
- Reduced total daily energy output
- Reduced fat burning potential
- Poor posture
- Hypoventilation
- Reduced respiratory fitness
- Reduced physical capacity
- Prone to daytime sleepiness
- Reduced motivation

PWS Clinic RPAH Camperdown NSW Australia
Physical Features in PWS

Clinical Features

- Increased % body fat

Functional Impact

- Increased fat storage ability
- Gain fat readily
- Increased cardiac risk
- Sleep apnoea risk
- Reduced energy intake requirement
  - dieting ➔ loss of muscle loss
  - (if not exercising)
Clinical Features

- Hypothalamic $\uparrow$ hunger / $\downarrow$ satiety
- Reduced hormonal maturation
- Desire sameness

Functional Impact

- Constant food focus
- Hyperphagia
- $\uparrow$ risk of osteoporosis
- Dislike new interventions
- *Like* consistency
- *Regularity increases* compliance
The Benefits of Exercise

- Musculo-skeletal improvement
  - Muscle strength
  - Joint stability
  - Bone density
- Achieving motor milestones
- Weight management
- Cardio-respiratory fitness
- Vascular health
- Mood - endorphins / BDNF / cannabinoids
- Distraction from food focus
The Improvement of Obesity-related Co-morbidities

- Diabetes - ↑ insulin sensitivity
- Hypertension
- Circulation
- Hypoventilation / OSA
- Oedema / lymphodema
- Hypercholesterolaemia - ↑ HDL
  ↓ LDL
  ↓ Triglycerides
Exercise Picture

- Regular
- Aerobic & strengthening
- Low to moderate intensity
- Incorporated into daily routine
- One to one / group / fun / incidental
- Not an optional extra
Where to Start

- **Childhood**
  - Sensorimotor stimulation
  - Joint alignment
  - Supported weight bearing
  - Gross motor skills / coordination
  - Achievement of motor milestones

- Physical Therapy
- Occupational Therapy
- Speech Therapy
Physical Fitness & Fun

Family and carer involvement
Part of Life

- Adolescence / Adulthood
  - Muscle strength & fitness
  - Physical capacity
  - Prevent or manage co-morbidities
  - Weight management
  - Bone density
  - Mood
  - Reduction of food focus and boredom
Effective Exercise

- **Aerobic**
  - 30-60 minutes
  - 5-6 days per week
  - Before a meal or snack
  - Supported or supervised
  - Examples
    - walking / treadmill
    - cycling / air walker
    - swimming / water exercise
    - dancing / video games
    - sport / play / outings

**Include warmup & cool down**
Effective Exercise 2

- **Strengthening**
  - Specific muscle groups
  - Weighted/resisted arm exercises
  - Upper body & trunk exercises
  - Bush / sand / rock walking
  - Ball play - throw/catch/bounce
  - Up & down stairs
  - Incidental activities - tidying room, cleaning, hanging out washing

- **Encourage good posture**

URT EIHOLZER, MD, YVES NORDMANN, MD, DAGMAR L’ALLEMAND, MD, MICHAEL SCHLUMPF, SILVIA SCHMID, PHD, AND KATRIN KROMEYER-HAUSCHILD, MD – IMPROVING BODY COMPOSITION AND PHYSICAL ACTIVITY IN PRADER-WILLI SYNDROME. J. Pediatrics 2003 142:73-78
Case Examples

SB (-79.5kg) & DB (-86.3kg)

“The measure of individual freedom (in Prader-Willi syndrome) correlates with body weight”

- Prof Andrea Prader

PWS Clinic RPAH Camperdown NSW Australia
# Maintained Fitness

<table>
<thead>
<tr>
<th>Day</th>
<th>Activity</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monday</td>
<td>Maui Thai or Special Olympics swimming or Dog walking</td>
<td>60 mins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>60 mins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>60 mins</td>
</tr>
<tr>
<td>Tuesday</td>
<td>Gym- X trainer &amp; weights</td>
<td>60 mins</td>
</tr>
<tr>
<td>Wednesday</td>
<td>Walk on treadmill at home</td>
<td>40 mins</td>
</tr>
<tr>
<td>Thursday</td>
<td>Gym or treadmill at home</td>
<td>45-60 mins</td>
</tr>
<tr>
<td>Friday</td>
<td>Dog walking, washing and feeding</td>
<td>90 mins</td>
</tr>
<tr>
<td>Saturday</td>
<td>Walks on treadmill at home</td>
<td>40 mins</td>
</tr>
<tr>
<td>Sunday</td>
<td>Gym - Tough Class</td>
<td>45 mins</td>
</tr>
</tbody>
</table>

August 2021

Age: 28 yrs
Weight: 72 kg
BMI: 23.5 kg/m²
Waist: 88 cm
References


- Kristy Reid, Peter Davies. Exercise and Physical Activity for Children with Prader-Willi Syndrome. Children’s Nutrition Research Centre - The University of Queensland (IPWSO Website)