The Power of Exercise for People with PWS

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Fit for Life, through Exercise
**Physical Features seen in People with Prader-Willi Syndrome (PWS)**

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>Functional Impact</th>
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<tbody>
<tr>
<td>Lower muscle tone</td>
<td>↓ resistance to movement</td>
</tr>
<tr>
<td>Decreased % muscle</td>
<td>Loose muscles</td>
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<tr>
<td>Reduced muscle strength</td>
<td>Less stable joints</td>
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<tr>
<td></td>
<td>Poor balance</td>
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<tr>
<td></td>
<td>Reduced total daily energy output</td>
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<td></td>
<td>Reduced fat burning potential</td>
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<td></td>
<td>Poor posture</td>
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<tr>
<td></td>
<td>Hypoventilation</td>
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<tr>
<td></td>
<td>Reduced respiratory fitness</td>
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<td></td>
<td>Reduced physical capacity</td>
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<tr>
<td></td>
<td>Prone to daytime sleepiness</td>
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<td></td>
<td>Reduced motivation</td>
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</table>
### Clinical Features

- Increased % body fat

### Functional Impact

- Increased fat storage ability
- Gain fat readily
- Increased cardiac risk
- Obstructive sleep apnoea risk
- Reduced energy intake requirement
  - *dieting*  →  loss of muscle loss
  - (*if not exercising*)
Physiological Features in PWS

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 Importance of Exercise

- Musculo-skeletal improvement
  - Muscle strength
  - Joint stability
  - Bone density

- Achievement of 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 - endorphins, BDNF
  - Reduction of boredom and food focus
Getting Started

- 10 -20 mins
- 1-2 per day (x 6 days per week)
- Add to “work out” time
- Physical therapist / personal trainer
- Extra incidental activity
  - park the car further away from destination
  - walk/cycle rather than travel by car
  - use stairs rather than lift or travelator
  - walk the long way around to school / work
  - physically active outings
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
## Intensity of Exercise

<table>
<thead>
<tr>
<th>PER</th>
<th>Hot</th>
<th>Puffed</th>
<th>Sweaty</th>
<th>Exhausted</th>
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<tbody>
<tr>
<td></td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>1</td>
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<tr>
<td></td>
<td>2</td>
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<td>4</td>
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Effective Exercise

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

- Slow rhythmic movement + stretches
- 3-5 mins slow start/finish of exercise

- To prevent soft tissue injury
- To prevent sudden change in BP
Encourage Good Posture

Remind person to:

- Lift head up when talking/walking
- Keep a straight back / “walk tall”
- Sit up straight in the chair
- Elbows at table height
Case Examples

SB (79.5kg) & DB (86.3kg)

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

- Prof Andrea Prader

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# Maintained Fitness

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

August 2021

Age: 28 yrs

Weight: 72 kg

BMI: 23.5 kg/m²

Waist: 88 cm
Motivation Starts with You

- R(ed) respect
- C challenge
- A acquired agreement
- R responsibility
- R rewards
- P praise
- S security
exercise works...
References


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