

Exercise and Physical Activity for children with Prader Willi Syndrome

A Guide for Parents and Carers

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The information provided in this Guide is not intended to be medical advice and is provided as general information only. If you have a particular question or concern about your child's health you should see a qualified medical practitioner.

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Foreword

Raising a child with Prader-Willi syndrome is often very challenging, frustrating, and worrying. Parents have many questions, “am I doing this right? What else can I do? Where can I find good information?” and difficulties can seem insurmountable. It’s hard enough to bring up a child under ordinary circumstances and those with PWS bring their own set of different and special needs. In the past, too often the only information parents could access was in the form of medical research, or textbook-like instructions that did not provide the information needed. Parents need a hands-on approach and in easy-to-understand language in order to know they are doing the right thing at the right time, and in the right way. This excellent handbook, which focuses on one of the major aspects in PWS, does this admirably.

Professor Peter Davies has had a long term interest in Prader-Willi syndrome, being awarded the British Nutrition Society medal in 1991 for his work in the UK, evaluating body composition in children. He has since published a number of papers in international literature relating energy metabolism and growth hormone treatment in the syndrome. Kristy Reid, an exercise scientist and researcher, has worked in Australia and Canada during the last three years and returned to the Children’s Nutrition Research Centre in January, 2010, to continue her research. Together they have

produced this book, which, I am sure, will be welcomed by every parent and care giver into their ‘must have’ library. It is refreshing to find authors who relate easily to the subjects of their topic, who can understand the genuine anxieties of children with PWS, and who offer sensible options for different levels of ability, while hammering out their one simple message – the amount of exercise you do every day must be equal to, or more than, the amount of calories you consume in order to stay fit and healthy, or lose weight.

Even with the advantages of greatly improved body composition that growth hormone treatment will give a child with PWS, it is critical that a regime of exercise and activity is started early and maintained into adulthood. No one knows better than a child with PWS how boring exercise can be, and the strategies and ideas in the booklet to make exercise fun, will help parents overcome the “I don’t want to!” syndrome that seems to appear in the older child.

We are fortunate to have researchers of such high calibre working on our behalf.

Linda Thornton

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Preface

This booklet is a guide to exercise and physical activity for parents and carers of children with Prader Willi Syndrome (PWS).

As a parent/carer, you will already understand that one of the key characteristics of children with PWS is their tendency to eat excessively. Individuals with PWS simply do not experience the capacity to feel “full” after eating.

This guide is intended to give you a better understanding of the importance of managing your child’s energy balance, exercise and/or activity levels. We hope the information will help you to ensure they are limiting their calorie intake and expending enough energy to prevent excessive weight gain over time.

In a discussion about energy balance, we understand that diet is also a critical issue of concern for parents and carers. To address this, we are currently developing a ‘Need to Know Nutrition’ booklet to accompany our exercise and activity guide.

Other members of the community are important in supporting you and your child. We encourage you to share this guide with your extended families, other carers, educators, and people in your community. To assist their understanding, we have included plain language explanations of medical terms and some general definitions of Prader Willi Syndrome and its characteristics.

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What is Prader Willi Syndrome?

Prader Willi Syndrome (PWS) is a genetic condition affecting between 1 in 10,000 to 1 in 15,000 live births. PWS occurs equally in males and females, affects all races, and is the most common genetic cause of obesity.

A list of PWS support organisations is supplied on page 31 of this guide book.

What are the characteristics of PWS?

There are two distinct phases of PWS Syndrome – the “Failure to Thrive” phase, followed by the “Hyperphagia” phase (excessive eating/insatiable appetite).

Infants with PWS are commonly reported as having “Failure to Thrive”. They are often hypotonic (floppy) and lethargic, with a poor suckling reflex and a weak cry. They are also slower to develop motor skills, taking longer to sit up, crawl, walk and talk.

As the child gets older, typically around four to five years of age, they enter a pattern of hyperphagia, and a preoccupation with food develops. This phase is often characterised by increased appetite and excessive weight gain.

Children with PWS often have a shorter stature, small hands and feet, poor muscle tone, hypogonadism (a condition where the male testes or female ovaries do not produce enough hormones), and begin to build excess weight around the abdomen, buttocks and thighs. They have a higher level of body fat and increased fat has been noted in all stages of development, along with decreased lean mass, bone mineral content, and bone density.

Individuals with PWS have distinct facial characteristics including almond shaped eyes, narrow nasal bridge, narrow forehead and thin downturned lips. Other characteristics may include altered temperature sensitivity, high pain threshold, delayed puberty and behavioural problems such as temper tantrums and obsessive compulsive behaviour.

Why are children with PWS more likely to become obese?

Children with PWS are at higher risk of developing obesity due to two key characteristics of the syndrome. The hyperphagia (excessive eating) results in an increased energy intake while hypotonia (low muscle tone), often coupled with poor co-ordination, reduces their ability and desire to engage in exercise or physical activity. Consequently, energy expenditure is limited. Children consuming more energy than they expend can store the excess energy as fat.

Excessive eating, coupled with low muscle tone, produces a chronic imbalance between energy intake (in the form of food and drink) and energy expenditure (the amount of exercise and activity undertaken). This results in excessive weight gain.

Many believe that children with PWS have a lower basal metabolic rate (amount of calories burned at rest) than other children. This belief is unfounded. Studies have shown that once a child's height, weight, body composition and age are accounted for, the basal metabolic rate of a child with PWS is similar to that found in other children.

However, it is true that children with PWS have more fat mass and less lean body mass than other children. Lean body mass consists primarily of muscle. Muscle burns a significant amount of energy at rest. This means that children with low muscle mass will burn less energy throughout the day than others with high muscle mass. Therefore, children with PWS are unlikely to expend as much energy throughout the day as other children simply because they have less muscle to burn calories.

For example:

Compare a plane with two engines, to a plane with four engines. While each engine might burn fuel at the same rate, the plane with four engines will consume more fuel, simply because it has more engines!! In this example, the engines represent lean muscle mass. A child with more muscle mass will burn more energy – like the plane with four engines, than a child with less lean muscle mass – even though their basal metabolic rate is the same.

With so many factors placing children with PWS at higher risk of obesity, it is crucial for parents to help their child get the balance right between energy expenditure and energy intake to prevent obesity (refer to Diagram 1).

Diagram 1. Preventing weight gain - energy must balance with energy out.



Why is obesity a major concern?

It has been established that children with PWS are at higher risk of obesity. Studies have shown that up to one third of individuals with PWS weigh more than 200% of their ideal body weight. Indeed, body fat percentages in those with PWS are 2-3 times higher than in the general population, with body fat accounting for 40-50% of their body weight.

This is of major concern. Obesity is one of the major causes of disease and death in people with PWS. Obesity can lead to health problems including Type 2 Diabetes Mellitus, high blood pressure, coronary artery disease, cor pulmonale (failure of the right side of the heart), sleep apnoea and premature death.

Preventing obesity in children with PWS

To reduce the risk of obesity, it is important for children with PWS to be taught the importance of adopting a healthy lifestyle early in life. The prevention and management of obesity is dependant on a combination of:

- a sensible diet (energy intake)
- the modification of eating behaviours (to ensure children adhere to prescribed diet)
- an exercise plan (energy expenditure)

Energy Intake – the importance of diet

Children with PWS develop an early obsession with food and have an urge to overeat.

It is important to ensure your child expends the same (or more) energy through exercise, than they are consuming. This will help prevent excessive weight gain. Weight control is essential to prevent obesity and the associated risks of chronic disease and premature death. The earlier your child is taught the importance of strict calorie controlled eating, the easier it will be for them to stick to their routine and incorporate it into their lifestyle.

To discuss an individualised dietary program for your child, contact a specialised dietitian. To find a dietitian in your area, contact:

1. The Dietitians Association of Australia at www.daa.asn.au or call their toll free number on 1800 812 942
2. The New Zealand Dietetic Association at www.dietitians.org.nz or call (04) 473 3061.

Modifying Eating Behaviours

Eating behaviours (such as how much your child eats and what they eat) should be established early in life to enable them to maintain a healthy weight over time. This is important because obesity and inappropriate eating behaviours are lifelong complications of PWS.

It is important to ensure your child is eating the correct types and amounts of food, and also to restrict your child's access to food to ensure they are not consuming extra energy.

When preparing food for your child, remember that not all calories are equal.

1g protein = 4kcal (17kJ)

1g carbohydrate = 4kcal (17kJ)

1g fat = 9kcal (38kJ)

1g alcohol = 7kcal (29kJ)

A kcal is commonly referred to as a calorie

Fat has the highest calorie count per gram. Fat is also very efficient at being converted to body fat. Carbohydrate is high in energy but less efficient at being converted to body fat. Protein is the most satisfying food.

Therefore one way to minimise energy intake for your child is to offer foods that are **LOW IN FAT**.

A dietitian can prepare a dietary program for your child to assist you with minimising energy intake. Refer to the previous page for information about how to contact a dietitian in your area.

Helpful Resources

Some other helpful resources to assist you in preparing healthy foods for your child are listed below:

1. Nutrition Australia's website:
www.nutritionaustralia.org/dynamic/recipes.php
2. Dietitians Association of Australia website:
www.daa.asn.au/recipes
3. Nutrition Society of New Zealand:
www.nutritionsofnewzealand.org.nz
4. New Zealand Nutrition Foundation:
www.nutritionfoundation.org.nz
5. New Zealand Dietetic Association:
www.dietitians.org.nz

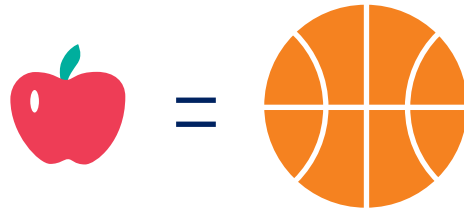


How long does it take to “burn off” some of the common foods that we eat?

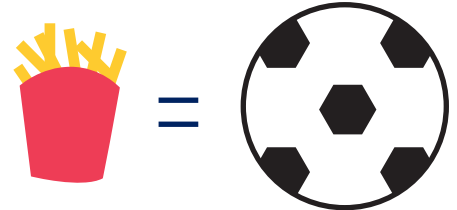


When trying to minimise your child’s energy input through diet, it’s very important to understand how much activity is needed to “burn off” the energy consumed through food.

Table 1, combined with the below pictures will assist you in understanding the energy levels of common foods and how much time we need to spend doing common activities to expend this energy.



An Apple = 11 minutes Basketball
302kJ = 72kcal



Hot Chips = 46 minutes Soccer
1260kJ = 300kcal



Ham salad sandwich = 63 minutes Tennis
1344kJ = 320kcal



Banana Muffin = 111 minutes Cycling
2255kJ = 537kcal

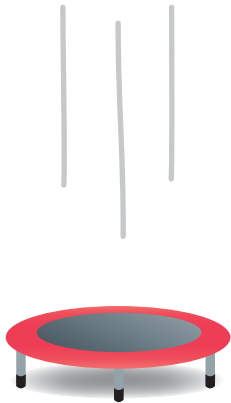


Table 1.

The energy cost of some common activities for 20-49kg children

Energy Cost of Activity			
Activity	20-29 kg	30-39 kg	40-49 kg
Bowling	2.0	2.9	3.7
Car Washing	1.3	1.8	2.3
Cycling (9km/hr)	2.0	2.8	3.5
Dancing (easy-aerobically)	2.3	3.2	4.1
Shopping	1.0	1.4	1.7
Gardening - mowing	2.3	3.2	4.1
Gardening - raking	1.8	2.5	3.2
Housework - mopping	1.5	2.1	2.7
Housework - laundry	1.1	1.5	1.9
Housework - vacuuming	1.7	2.4	3.1
Jumping Rope (70 per minute)	4.2	6.0	7.7
Roller-skating (outside on the pavement)	3.3	4.7	6.0
Running - on flat surface (8.4 km/hr)	3.2	4.5	5.7
Sitting Quietly	0.6	0.8	1.0
Soccer (casual)	2.7	3.8	4.9
Swimming - laps	4.1	5.7	7.4
Swimming - treading water	1.6	2.3	3.0
Tennis (recreational)	2.9	4.1	5.2
Walking (5.5 km/hr)	1.9	2.6	3.5

* Refer to the column that is closest to your child's body weight and then multiply the number in the column by the number of minutes they spend doing the activity to determine the approximate energy cost of their participation in kcal. To convert to kJ multiply by 4.2.

These values are approximate and estimated.



Values calculated using the information supplied in Ridley, Ainsworth et al. 2008.

Table 2. The energy cost of some common activities.

* Refer to the column that is closest to your child's body weight and then multiply the number in the column by the number of minutes they spend doing the activity to determine the approximate energy cost of participation in kcal. To convert to kJ multiply by 4.2.

These values are approximate and estimated.

Energy Cost of Activity										
Activity	50-59 kg	60-69 kg	70-79 kg	80-89 kg	90-99 kg	100-109 kg	110-119 kg	120-129 kg	130-139 kg	140-149 kg
Bowling	5.3	6.3	7.2	8.2	9.2	10.2	11.2	12.2	13.2	14.2
Car Washing	3.8	4.6	5.2	5.9	6.6	7.3	8.0	8.7	9.4	10.1
Cycling (9km/hr)	3.5	4.2	4.7	5.4	6.1	6.8	7.5	8.2	8.9	9.6
Dancing (easy-aerobically)	5.4	6.4	7.2	8.3	9.4	10.3	11.2	12.1	13.0	13.9
Shopping	3.4	4.0	4.6	5.3	5.9	6.5	7.1	7.7	8.3	8.9
Gardening - mowing	6.1	7.3	8.3	9.5	10.6	11.7	12.8	13.9	15.0	16.1
Gardening - raking	2.9	3.5	4.0	4.6	5.1	5.7	6.3	6.9	7.5	8.1
Housework - mopping	3.4	4.0	4.6	5.3	6.0	6.7	7.4	8.1	8.8	9.5
Housework - laundry	3.6	4.3	4.9	5.6	6.3	7.0	7.7	8.4	9.1	9.8
Housework - vacuuming	3.5	4.2	4.8	5.5	6.2	6.9	7.6	8.3	9.0	9.7
Jumping Rope (70 per minute)	8.9	10.5	12.0	13.7	15.4	17.0	18.6	20.2	21.8	23.4
Roller-skating (outside on the pavement)	6.8	8.1	9.2	10.5	11.8	13.1	14.4	15.7	17.0	18.3
Running - on flat surface (8.4 km/hr)	7.4	8.8	10.0	11.5	12.9	14.3	15.7	17.1	18.5	19.9
Sitting Quietly	1.2	1.4	1.6	1.8	2.0	2.2	2.4	2.6	2.8	3.0
Soccer (casual)	6.7	7.9	9.1	10.4	11.6	12.8	14.0	15.2	16.4	17.6
Swimming - breast stroke	8.9	10.5	12.0	13.7	15.4	17.0	18.6	20.2	21.8	23.4
Swimming - freestyle	7.0	8.3	9.0	10.8	12.2	13.5	14.8	16.1	17.4	18.7
Swimming - treading water	3.4	4.0	4.6	5.2	5.9	6.5	7.1	7.7	8.3	8.9
Tennis (recreational)	6.0	7.1	8.1	9.2	10.4	11.5	12.6	13.7	14.8	15.9
Walking (5.6 km/hr)	4.7	5.7	6.4	7.8	8.2	9.0	9.8	10.6	11.4	12.2

Adapted from McArdle and Katch, 2001 with permission

Energy Expenditure – the importance of an exercise plan

Physical activity and exercise can help children with PWS to expend more energy than they consume through food.

How much energy a child expends is determined by four key factors:

1. The **DURATION** of the activity
2. The **INTENSITY** of the activity
3. The **TYPE** of activity
4. The **METABOLIC EFFICIENCY** of your child

Please refer to Tables 1 and 2 – “Energy Cost of Activity” on pages 9 and 10 for the energy cost of some common activities.

There are two main ways to increase the amount of energy your child expends.

1. Start them on a regular exercise program if they do not have one already.
2. Increase the amount of physical activity in your child’s routine.

Remember – any exercise is better than none, and it all adds up at the end of the day! We’ve provided some helpful hints to help you encourage your child to undertake regular exercise and physical activity.

Helpful Hints

- > Use a diary or calendar to document how much activity your child is doing and include their feelings about the activity. e.g. How they felt during the activity, whether they liked the activity and how they felt after the activity.
- > Show your child how to make exercise a priority.
- > Try to do the exercise or activity at the same time each day to develop a routine.
- > Develop a reason for exercising or performing a certain activity. e.g. We are going to feed the ducks at the lake or the horse down the road or we are going for a bike ride to see relatives or friends.
- > Perform the activity or exercise with your child to ensure they are doing it and not just telling you they are doing it (or watch them complete the activity).
- > Always keep your child updated on their progress.
- > Always give your child advance notice when you intend to increase their exercise workload and/or intensity. e.g. Explain that for 3 weeks you are going to walk for 20 minutes, 4 times per week and then you are going to increase the time to 30 minutes for another 3 weeks, etc.

The Benefits of Exercise

It was established earlier that excess energy consumed will be stored as fat if a child's energy intake is not increased through exercise and physical activity. But what are the other benefits of getting your child active?

Exercise and physical activity have many benefits – they help to:

- keep bones strong
- keep the heart healthy
- strengthen muscles
- increase endurance
- increase flexibility
- improve mood through the release of endorphins
- decrease feelings of depression and anxiety

Daily exercise enhances aerobic fitness and energy expenditure while minimising the loss of lean body mass and muscle tone. Aerobic exercise helps to burn body fat, while a lack of physical activity can decrease the tone and strength of muscles.

How do I introduce exercise to my child?

When your child is first starting out, sporting activities may be limited. Children with PWS usually have poor muscle strength, poor coordination and decreased muscle mass. It is not uncommon for high impact sports such as running and jumping to cause joint injuries in these children.

Low impact activities are recommended when introducing exercise to your child. Swimming, walking and stationary exercise equipment such as exercise bikes and rowing machines are all possible alternatives. They can help to build your child's strength until they are able to participate in other activities.

To preserve muscle tone, resistance activities can be effective. Try training with very light weights or use your child's own body weight as the resistance in activities such as push ups, pull ups, crunches, dips, lunges, squats and calf raises.

When starting any new exercise routine, it is always beneficial to enlist the help of a specialised trainer or physiotherapist to outline and supervise an appropriate training program for your child.

What type of exercise is right for my child?

Considerations that influence the choice of activities for your child include:

- degree of obesity
- strength
- individual interests
- level of ability to understand instructions
- stamina
- co-ordination
- medical considerations

Children with PWS often experience weakness in muscle tone and strength and decreased motor planning skills. This makes it difficult for them to gain the co-ordination and speed for regular childhood activities and competitive sport.

Specific sports can often be modified to better accommodate children with PWS. This can be achieved by modifying court size, field size, game rules and by using softer equipment. Some examples of sports that use these principles to include a wider range of children and abilities include:

- a. Auskick (AFL)
- b. NetSetGO! netball
- c. Kanga cricket
- d. Little Athletics
- e. Go-go golf
- f. Tee ball

These sports have rules that emphasise and encourage participation and play. Having said this, it is always a good idea to provide information about PWS to your child's coach or activity organisers to ensure they understand your child's abilities.

Helpful Contacts for Modified Sporting Programs

1. Australian Sports Commission – Active After School Communities Program, find out about sports available at schools and child care facilities in your area. Download fun games and activities. www.ausport.gov.au/participating/schools_and_juniors/aasc/get_involved/children
2. Auskick - www2.aflauskick.com.au
3. Net-Set-Go - Netball Australia's Junior Netball program - www.netsetgo.netball.asn.au/kids.asp
4. Kanga Cricket - www.milo.com.au/milo-and-sport/cricket/milo-kanga-cricket.html
5. Little Athletics - www.littleathletics.com.au
6. Go-Go Golf - www.getintogolf.org.au/default.aspx?s=gogogolf
7. Kiwi Cricket New Zealand - www.blackcaps.co.nz/content/grassroots/milo-kiwi-cricket.aspx
8. New Zealand Children's Athletic Association - www.athletics.org.nz/Article.aspx?ID=1115
9. Sport and Recreation New Zealand - www.sparc.org.nz

What age should I involve my child in exercise and physical activity?

It is important to teach your child sport specific skills and to get them involved in activity at a young age.

Improvement in basic skills at an early age will help to build your child's self confidence and will enable greater participation in recreational activities as they get older. Participation also fosters social skills, promotes peer acceptance and improves co-ordination, strength and endurance.

What activities will help my child lose weight?

Activities targeting both fitness (Aerobic) and strength (Anaerobic) will help your child to lose or maintain their weight through an increase in energy expenditure.

Humans use their **aerobic** energy system when completing exercise or physical activity at low or moderate intensity for an extended period of time. It results in an increased breathing rate and an elevated heart rate. Some examples of aerobic activity include:

- Walking briskly
- Cycling
- Jogging

Aerobic exercise uses oxygen to generate energy for the working muscles and uses glycogen and fat as the primary fuel sources. It is very important for burning fat stores. Aerobic exercise assists in weight control in the following ways:

- by burning excess energy (primarily fat stores)
- improving heart function and the body's ability to circulate oxygen
- preventing osteoporosis
- strengthening muscles, tendons, joints and ligaments
- improving temperature regulation at rest and during exercise in different environments
- Improving blood pressure control

DID YOU KNOW?

Most sporting activities use a combination of the aerobic and the anaerobic energy systems.

Humans use their **anaerobic** energy system when undertaking activities that are short in duration (less than 2 minutes) and require speed and power. Anaerobic fitness complements aerobic exercise to build stronger muscles. Some examples of anaerobic activities include:

- Sprinting
- Jumping
- Ball Throwing
- Gymnastics

Anaerobic exercise uses creatine phosphate (a compound stored in muscle cells providing a quick energy source for high-intensity muscle contractions) and glycogen as the main energy sources for the working muscles without the use of oxygen. Anaerobic exercises use muscles at a higher intensity and higher work rate for a shorter period of time. It helps to develop:

- Stronger muscles
- Increased muscular endurance
- Increased ability to withstand fatigue
- An improved cardio-respiratory system



Some tips to set your child up for exercise success

- > Time exercise opportunities to coincide with your child's high energy times.
- > Let your child choose the daily or weekly exercises or activities.
- > Restrict opportunities for sedentary activities such as watching television.
- > Teach your child activities in small steps that can be accomplished easily. This will help to build their confidence and encourage greater desire to participate.
- > Start introducing your child to exercise and physical activity early in life – they will carry the habits throughout their life.
- > Promote their social skill development - allow them to participate in activities with their peers to build their social confidence, peer acceptance and skills.
- > Individualise the activity or exercise for your child, based on their interests, abilities and goals.
- > Remember that variety is the spice of life. Vary activities to hold your child's interest and to provide a full range of health benefits across all body systems.

How do I maintain my child's interest in exercise?

Children with PWS tend to lose interest quickly so there is an increased need to keep them stimulated. Here's a list of some different activities that may assist you to keep your child active and interested:

- Visit different playgrounds for different experiences.
- Use the swings to help with their sense of balance.
- Use climbing frames to assist with concentration, co-ordination, balance and problem-solving skills.
- Use slides and water slides as they are not only fun, but require a lot of stair climbing to reach the top!
- Use monkey bars to help build upper body strength.
- Join a play gym or Gymboree club to increase opportunities for social interaction.

How do I encourage exercise with my infant or toddler?

Infants and toddlers with PWS experience muscle hypotonia (low muscle tone) and need to be encouraged to use their muscles. Frequent changes in position can stimulate them mentally and promote better motor development. Here are some ways to improve your PWS infant/toddler's muscle tone:

1. Lay your child on their side – this provides complete support and positions the arms together to promote hand use.
2. Lay your child on their stomach to encourage the development of head extension and to strengthen their trunk muscles.
3. Encourage your child to sit upright (allow them to lean on something if necessary). Allow them to freely kick their legs to encourage movement or leg strength.
4. Use a walker or jolly jumper to encourage movement and build leg strength.
5. Use ribbons, utensils, bells, toys and other common objects to suspend in front of your child to stimulate reaching and hand manipulation.
6. Encourage movement stimulation by slowly rolling, bouncing or rocking your child.
7. Place toys or objects a little beyond your child's reach to encourage them to move toward an object.



Remember

Anything that can get your child moving and expending extra energy will be beneficial to them!



How do I encourage my preschool child to exercise?

Children need to be encouraged to move as movement builds strength. Children with poor muscle tone need some extra incentive and attention to begin spontaneous activity. Here's some ways to encourage your preschooler to get moving:

1. Encourage your child to swim and play in water to stimulate movement.
2. Try rolling them across the floor or do somersaults to encourage flexibility.
3. Walking along low retaining walls can be a great balancing activity.
4. Easy ball games are great and you can vary the ball size to build different skills – using a large ball promotes the use of two hands, while a smaller ball is harder to catch but easier to throw.
5. Try walking interspersed with short bursts of running to help build strength and endurance – having a goal to run to can help – ask them to find a clue, or run to a tree, street light, or power pole!
6. Encourage your child to chase bubbles to promote running and jumping.
7. Get your child to kneel when they are playing near a low table to strengthen hip muscles and assist their balance.
8. Try heel raises to help with balance and leg strength.
9. Dancing is fun – try musical statues or get them to balance something on their head for added interest.
10. Resurrect old play favourites such as Simon Says (run on the spot, hop, jump, bend and stretch, etc), hopscotch, follow the leader (as the leader encourage them to copy you - run, skip, hop, alternate between big and small steps, etc).

11. Include music or educational elements to spice up a game and to stimulate learning e.g. Heads, shoulders, knees and toes singing game, counting flowers, or finding flowers of different colours around the garden.
12. Encourage your child to develop their own games as this is a great way to stimulate the mind!

How do I encourage my school age child to exercise?

For school aged children with PWS, it is necessary to highlight the importance of participation, rather than winning or losing. Encourage their participation in physical education classes and slowly introduce them to the importance of activity and exercise. Here are a few ideas:

1. Try entering your child in the Special Olympics.
2. Take your child bike riding through different parks and try to vary the location for a change of scenery.
3. Use different ball games and teach them basic skills.
4. Try dancing or moving around to their favourite music.
5. Encourage activities that may not be viewed as exercise by your child – try Gymnastics (tumbling), hopscotch, playing Frisbee, jumping/skipping rope, swimming, running games such as relays, egg & spoon races, or dancing.
6. Have a talk to your child's teachers and enlist their help in keeping your child active during the day at school – ask them to seek opportunities to get your child to walk at regular intervals, take breaks to stretch, and to help with errands that require walking.



Walking Tips!!

- > Take your baby for a regular walk in the pram to build the habit of getting out and moving from an early age.
- > When your toddler is old enough, encourage them to get out of the pram and walk part of the way.
- > Join a walking group.
- > On hot days, walk around an air-conditioned shopping centre or museum.
- > Take them for a walk along the beach to collect shells.
- > Schedule regular times for family walks.
- > Try bushwalking or nature walks to vary the scenery and to add interest by spotting flora and fauna.
- > Try building in some running or jogging as your child ages and muscle tone increases.

Some common activities and their benefits

The following pages list some common activities and their benefits as well as some tips for encouraging your child to get active.

Walking

Walking has the following benefits:

- Walking is a simple way to improve health and well-being.
- Walking is simple to do and can be done at any time.
- Walking can be undertaken indoors and out.
- Walking causes less shock to the lower back, hips, knees, ankles and feet compared with running and jogging.



Swimming and Water Activities

Swimming and water activities have the following benefits:

Swimming is a low impact exercise – the water supports the body and reduces stress on bones and joints.

The buoyancy of the water means decreased stress on bones, joints and connective tissues – in turn this means a decreased risk of injury for your child.

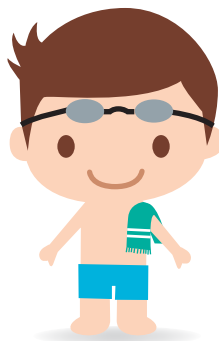
It's a great way to increase cardiovascular endurance and stamina.

Swimming is a good resistance activity – your child's own body weight helps to provide resistance when doing activities in the water.

Water helps your child to feel weightless and move freely – helping to burn more energy and increase muscle expenditure.

Swimming engages the whole body, improving cardiovascular conditioning, muscular strength, endurance, posture and flexibility.

Swimming tones the upper and lower body as most of the major muscle groups are used.



Swimming Tips!!

- > Involve your child in water activities at an early age to build their awareness and confidence.
- > As their confidence increases, you can try swimming lessons.
- > If your child can swim, try swimming laps with them and alternate between the swimming strokes (e.g. Freestyle and then breaststroke).
- > Use a kickboard – why not try some family kickboard races.
- > Try aqua aerobics or water running.
- > Play games with your child that will get them moving in and around the pool (e.g. Throw objects in the pool and have your child dive to the bottom to retrieve them; play Marco Polo; swim in a circle; race to different areas of the pool; tread water, etc).

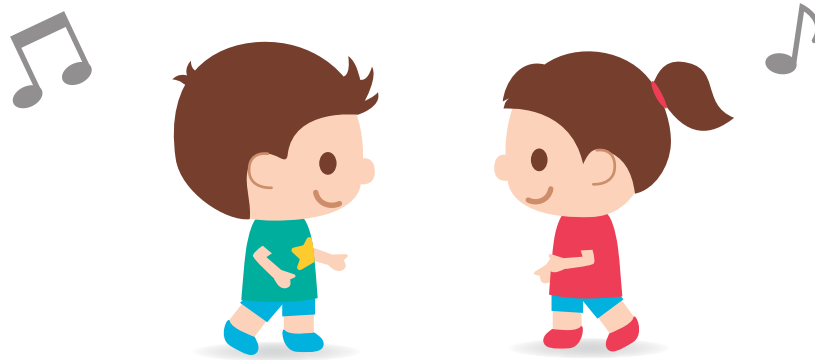
Dancing Tips!!

- > Play music from different parts of the world and encourage your child to role play.
- > Play musical games such as musical statues to make dancing more fun.
- > Enrol your child in dance lessons as their coordination and confidence grows.

Dancing and Music-based Activities

Dancing and music-based activities have the following benefits:

- Dancing encourages movement, balance and coordination.
- Dancing helps your child to be imaginative.
- Dancing benefits the heart, cardiovascular system and increases lung capacity and circulation.
- Dancing burns a lot of energy quickly, depending on the duration and speed of the dance.
- Dancing can burn as much energy as walking, swimming or riding a bike but also builds muscle strength.
- Dancing may also help to stimulate your child's brain as it requires them to remember dance steps, routines and patterns.



Playing Games and Activities

The benefits of games and activities include:

- Different games and activities are enjoyable for children and keep them interested and motivated to participate.
- Games can encourage your child to exercise without them thinking of it as exercise.
- Games that involve running and jumping get children moving while still enhancing the cardiovascular and musculoskeletal systems of the body.

REMEMBER

Clearly explain the rules of any new game to your child from the beginning and **DON'T ALTER THEM**. It may be a good idea to test out the rules on a child without PWS to make sure they are clear and to prevent problems arising.

Games & Activities Tips!!

- > Teach your child to catch and throw a ball, and other sport specific skills from an early age to build their coordination and confidence.
- > Try some old favourites – ball games, hopscotch, jumping rope and relays.
- > For ball games – try using different shaped and sized balls and change the throw to include rolling or bouncing the ball.
- > Alter the distance between you when throwing a ball to vary the length of the throw.
- > Try shifting your games to the beach – the sand makes movement harder and builds leg strength!
- > Reward your child with activities instead of food. e.g. Go ten pin bowling, skating, to the beach, for a ride in the park, etc.

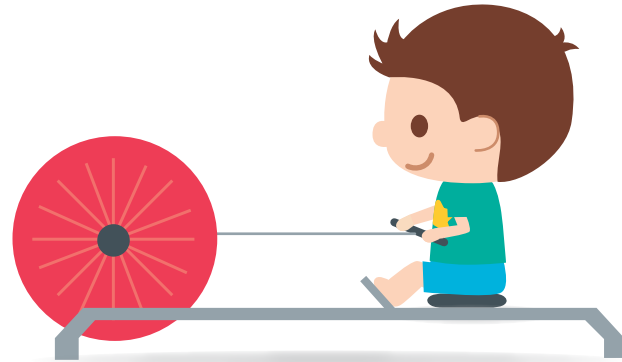
Exercise Equipment Tips!!

- > Try to vary the types of machines your child uses to target different muscle groups. e.g. Stationary cycling can provide a steady workout for hips, thighs, legs and gluteals as well as a cardiovascular workout, while rowing machines target legs and gluteals, upper and lower back and the abdominal muscles.
- > Stationary exercise bikes can be useful if your child is having difficulty learning the correct cycling technique. Pedalling on a stationary bike will help them to learn until they are ready to progress to a real bike. Training wheels may also help when they are first starting out!
- > Don't forget the trampoline – it's fun and a great way to burn energy!

Stationary Exercise Equipment

The benefits of using stationary exercise equipment include:

- Stationary exercise equipment such as cycling and rowing machines allow your child to burn energy without moving around a great deal.
- Resistance levels can be altered with equipment such as stationary cycles.
- Many types of equipment, such as stationary cycles and rowing machines help to increase cardiovascular and muscular endurance.
- Depending on the type of machine used, your child can achieve an effective workout targeting many muscle groups.
- Many exercise machines may also decrease the risk of injury to your child helping to spread the load over a number of joints. Rowing machines are a great example.



Why is incidental exercise so important?

Incidental exercise is the incorporation of exercise into daily tasks and is considered to be any movement performed during the day as a part of everyday life (e.g. hanging out the washing, vacuuming the house, washing the car, etc). Planned and structured exercise is very important but keeping active throughout the day will help your child to reduce body fat and expend extra energy.

People who keep active through the day by engaging in a high level of incidental exercise experience the following benefits:

- increased muscle mass
- improved heart and lung strength
- improved joint mobility
- improved blood flow
- injury prevention
- improved brain function
- better self esteem

Opportunities to undertake incidental exercise are becoming reduced as modern technologies remove our need to expend energy. The convenience of elevators, remote controls, drive through automatic car washers and cordless phones, causes us to become more sedentary. Many people are now not expending an adequate amount of energy in daily activity.

Incidental exercise is a great way to increase your child's energy expenditure.

Incidental Exercise Tips!!

- > Try walking the dog with your child for a few minutes extra each day.
- > Encourage your child to take the stairs instead of the elevator as this will increase their heart rate and strengthen leg muscles.
- > Park the car further away from the supermarket/school entrance to increase the distance you need to walk.
- > Encourage your child to help you wash the car by hand.
- > Involve your child in the garden – get them to weed the garden, mow the lawn and rake up leaves.
- > Don't use the clothes dryer. Encourage your child to help you hang out the clothes on the washing line.
- > Walk while you talk on the telephone instead of sitting down – encourage your family to do the same!
- > Put limits on activities such as watching TV, playing on the computer, playing video games and encourage physical activities instead.
- > Encourage your child to get up to change the television channel instead of using the remote control – why not try hiding it for a bit of fun!
- > Try walking or cycling with your family instead of driving wherever possible.

Key Point to Remember

The recommended amounts of activity and exercise can be achieved by:

- > Doing all the activity in one session.

OR

- > Doing several shorter bouts of activity of 10 minutes or more (i.e. Three 10 minute sessions per day will make up the recommended daily amount of 30 minutes of activity).

How much exercise and physical activity should my child be doing?

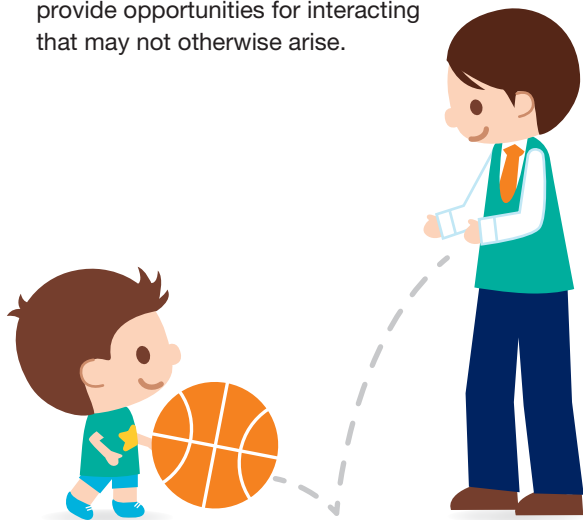
The amount and type of exercise for children should be determined individually. Exercise choices will be influenced by your child's maturity level, medical status, skill levels and prior exercise experiences.

Some guidelines for activity are based around the F.I.T.T. Principle.

- F = frequency or regularity of activity: children should participate in activity on most, or preferably all, days of the week.
- I = intensity of the activity: the intensity of exercise should be moderate. This is where the child's heart rate becomes slightly elevated and they feel an increase in their breathing rate. You can monitor this with your child based on their exercise tolerance. They should begin exercise at a lower intensity and gradually increase to a comfortable level as they continue.
- T = time or duration of the activity: children should aim to accumulate at least 30 minutes of planned and structured exercise or activity every day to achieve the benefits associated with activity and exercise.
- T = type of activity: children should be encouraged to participate in a variety of activities that exercise all the major muscle groups (e.g. those in their arms and legs). They should also undertake weight bearing activities to optimise basic skill development, weight management, aerobic fitness and bone mineral content.

How do I set a good example for my child?

Children learn from the behaviour of their parents and other older people around them. It is very important to set a good example for your child by practising good physical activity and dietary habits. The benefits to your child and the rest of the family will be immense. Undertaking activity and exercise together will improve your family's health and sense of well-being and provide opportunities for interacting that may not otherwise arise.



Some Tips to set a good example for your child

- > Get involved yourself – play catch with your children, run relay races, show them you can participate and have fun!
- > Start the chore challenge – set up some friendly competition between your children and yourself by counting the number of chores completed. e.g. Number of weeds pulled out, most clothes pulled off the washing line, etc.
- > Make exercise the feature of your parties or special occasions – have your child's birthday at a local bowling alley, take the kids to a climbing wall, or have a backyard Olympics at your next play date!
- > Undertake exercise as a family - Walk places instead of taking the car, take your child for regular walks around the neighbourhood, go for a family bike ride in the park.
- > Take your child to the park – supervise them while playing and try to join in where you can.
- > Demonstrate sport related skills to your child – help them to improve their throwing and catching, kicking skills, etc.
- > Involve family friends in activities where you can – go for a group cycle, or join a swimming club!
- > Make exercise part of the whole family's daily routine – just vary the activities or scenery to keep your child interested. Try fitting in 10-15 minutes of exercise several times a day, if this is an easier way to build up to the daily total of 30 minutes.

Growth Hormone Treatment (GHT)

More and more children with Prader-Willi Syndrome are being given GHT. Growth hormone has a significant impact on many tissues in the human body. An increase in linear growth i.e. getting taller, is only one effect of this treatment. Indeed it has been suggested that increasing linear growth should be considered a “side effect” of treatment with other effects being more important to the child with Prader-Willi Syndrome.

Studies show those with PWS have different body composition to those who are simply obese.

An Italian research team (Brambilla & colleagues) undertook studies relating to the body composition of individuals with PWS. The work has confirmed what has been anecdotally suspected for some time. The Body composition (amount of fat and fat free mass) of children and young adults with Prader-Willi syndrome is different to the body composition of individuals with simple obesity.

Simple obesity means that an individual is overweight or obese but **does not** have PWS. When you consider that the effort of moving a larger mass builds muscle size and strength, it’s not surprising that people who have simple obesity have increased fat mass **and** fat-free mass. For example, a person of larger body weight will expend more effort when walking, usually causing the leg muscles to

increase in size and strength. This “companionship of lean and fat”, as it has been described, was not apparent in those with Prader-Willi syndrome. Instead, patients were found to have an increased amount of fat in their limbs compared with their trunk, thus reducing their total fat-free mass.

Body composition in those with PWS is similar to that found in those with Growth Hormone Deficiency.

Brambilla’s study shows that body composition in those with Prader-Willi syndrome is unlike that found in people who have simple obesity. It is, however, similar to that found in children with growth hormone deficiency (GHD). Indeed, a large proportion of children with Prader-Willi syndrome are unable to regulate growth hormone due to impairment of the hypothalamus. The hypothalamus is a part of the brain that influences hormone regulation, body temperature, hunger, thirst and fatigue. Therefore, many of the features associated with PWS, namely short stature, hypotonia (poor muscle tone) and obesity, might be affected by administering Growth Hormone injections. The effects of Growth Hormone (GH) on body composition are likely to be of more benefit to children with Prader-Willi syndrome than an increase in linear growth or height.

How can Growth Hormone help those with PWS?

Throughout this guide, it has been discussed that a disturbance in normal body composition and the development of obesity are issues of major concern in Prader-Willi syndrome. However, latest studies are showing that GH injections can have beneficial effects on body composition for children with PWS. One recent study noted an increase in fat-free mass and a decrease in fat mass resulting in a reduced body fat percentage. The study also noted secondary benefits from GH injections such as improved exercise capacity, strength and agility. These benefits were likely to be caused by changes to body composition, rather than direct impacts from the GH itself. However, the changes are important if we are trying to increase the exercise capacity of a child with PWS.

The effect of GH administration on muscle capacity and endurance

GH administration can change the anatomy (structure) of the body. These changes to body composition might also have important physiological (functional) consequences for individuals with PWS related to exercise.

A study by Eiholzer and colleagues found considerable improvement of 'physical capability' (i.e. an estimate of

physical fitness) in those with PWS following 12 months of GH administration. This improvement was probably due in part to the effect of the administered GH, which acted to increase fat-free mass over the study period. The Wingate Anaerobic test is a well documented and validated test for measuring muscle endurance and peak power. It is a 30 second cycling test, requiring maximal effort from the subject. This test was used in the Eiholzer study and data obtained from four of the 12 subjects showed dramatic improvements. Mean peak power, when expressed as a percentage of normal, changed from 57.5% to 100.4% over the 12 months of GH administration. Previously mean power had been 58.9% to 82.5% of normal.

A separate Scandinavian study used computerised axial tomography (CT scan) to assess changes in muscle and fat mass. Subjects had their thighs scanned, with measurements taken at the middle of the femur, before and after GH administration. The thigh **muscle area increased** by 33% over the first year of GH administration, while thigh **fat area decreased** by 25%. Such changes in body composition certainly help to explain why GH administration might increase physical performance.

Extensive testing of physical strength and agility were carried out in a more recent US-based study. Agility was tested using a modified Bruinites-Oseretsky test, in which

the time taken to run and pick up a block and return was recorded. Lower extremity strength was assessed using a standing broad jump, trunk strength was assessed by measuring the number of sit-ups achieved in 20 seconds, and upper extremity strength was measured using dumbbell weights. All subjects recorded significant improvements across all of these measures. For example, upper strength in Prader-Willi Syndrome measured by the mean number of repetitions of dumbbell lifts changed from 13.1 to 15.6 in the treatment group, but showed no such change in the control group (13.2 changing to 12.1). Study authors concluded that **improvement in physical abilities and exercise tolerance were the most important results of GH therapy** for children with PWS and their families. This improvement was related to what was termed 'real life' function including activities such as carrying large milk containers and independently climbing steps and stairs. It is noteworthy that the increase in linear growth (height) also found in the study was not seen as the major improvement in children's lives.

The last study infers that spontaneous physical activity may increase with GH administration. If this is true, then inevitably total energy expenditure will also increase, thus improving the energy balance of the individual. This, coupled with the increased resting metabolic rate associated with increased fat free mass, could begin to impact upon body composition in a beneficial way – tipping the scales in favour of a brighter future for kids with PWS.

Conclusion

We hope this guide has assisted you in understanding the importance of good diet and an active lifestyle in managing weight gain in children with PWS. Children with PWS are at higher risk of developing obesity. Therefore it is crucial for parents/carers to implement a healthy diet and regular exercise routine in the household.

There are many organisations and specialists in the community who can assist you to develop a diet and exercise plan that is right for your child and your family. Many of these organisations were featured throughout this guide book, and we would encourage you to contact them for individual advice relating to your child.

The most important message for a parent of a PWS is to remember the energy equations:

Energy In = Energy Out = no net weight gain/loss

Energy In > Energy Out = weight gain

Energy in < Energy Out = weight loss

If you can get the energy balance right, then you are on the way to preventing obesity and ensuring your child can look forward to a healthier future.

Prader Willi Support Organisations and other useful links

Prader Willi Syndrome Association of Australia www.pws.org.au

Prader Willi Syndrome Association of New South Wales www.pws.org.au/nsw

Prader Willi Syndrome Association of Victoria www.pws.asn.au

Prader Willi Syndrome Association of Queensland www.pwsaqld.com.au

Prader Willi Syndrome Association (N.Z.) www.pws.org.nz

International Prader Willi Syndrome Organisation www.ipwso.org

Prader Willi Syndrome Association (UK) www.pwsa.co.uk

Prader Willi Syndrome Association (USA) www.pwsausa.org

The Halberg Trust (N.Z) www.halberg.co.nz

The Halberg Trust is committed to ensuring people with a disability can participate in inclusive sport and active leisure within their community.

The Children's Nutrition Research Centre, The University of Queensland www.uq.edu.au/cnrc

This booklet is available on the Children's Nutrition Research Centre website.

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