

## **My sister or brother has Prader-Willi Syndrome: Information for siblings**

### **Someone special**

People with Prader-Willi Syndrome (PWS) are special, but so are you.

PWS is quite rare. It is unlikely, but not impossible, that there is anyone you know living near you who also has a brother or sister with PWS, so it may be hard for your friends and neighbours to understand what it is like to have someone with PWS in the family. However, there are many more people with all sorts of disabilities, and it is quite likely that someone you know has a brother or sister with a different disability who will share some of your concerns and feelings.

Another word for “brother or sister” is “sibling”.

### **Why did it happen?**

The first thing to say is that no-one is to blame. PWS just happens - it is a fluke of nature. And in the vast majority of cases it does not happen more than once in a family. There are about 5% of families who may have hereditary cases of PWS, but nowadays geneticists can test you to tell you whether or not you are one of these “at risk” families.

### **What is Prader-Willi Syndrome?**

A syndrome is a collection of symptoms or signs, which when they occur together, show that a person has a particular disease or condition. There are thousands of different syndromes known to medical science, some of which you are born with, and others which develop later in life. The symptoms that your brother or sister has are called Prader-Willi syndrome. There is a simple reason why these symptoms have this very strange name. If a scientist discovers a new law, a new disease, a new invention or a new syndrome, it is very often called after him or her. The set of symptoms your sibling has were first “described” (published in medical journals) by Dr A Prader and Dr H Willi, who come from Switzerland,

and the syndrome was then named after these two doctors. This was in 1956, but we are certain that the syndrome existed before then, even if it did not have a “name”.

If you need to know how to pronounce Prader-Willi, it is *Prahder (to rhyme with “larder), and Willi (as in the boy’s name, to rhyme with “Billy”)*.

PWS is something you are born with. If you are older than your brother or sister with PWS, you may remember how difficult it was for your parents to feed him or her, and perhaps how it took your sibling much longer than other children to learn to walk, talk, and do other things. This is because people with PWS have poor muscle tone - their muscles are not as strong as yours are. No one knows why this is, but muscle strength can be improved with lots of exercise.

Scientists think that the part of the brain that is affected in PWS is the hypothalamus. This is the “control centre” of the body, and regulates things like: appetite, growth, sexual development, and emotional stability. All of these things do not work properly in PWS.

The message that tells you that you have had enough to eat does not “switch on” for people with PWS, and so they go on feeling hungry for much longer than most people, even when they have already had a lot to eat. Sometimes they feel so hungry they will steal food from anywhere.

Most people with PWS are naturally shorter than average. This, together with the weak muscles and the need to eat, means that it is VERY difficult for your brother or sister to keep their weight down, and they need all the help and support they can get. Before much was known about PWS, many people died early because they got very fat, but with better management, they are living well into their 40s and 50s and beyond. Nowadays, too, many children with PWS are receiving growth hormone treatment, which helps them to grow taller and make their muscles stronger.

If you have a brother with PWS, you may have noticed that he has a smaller penis than other boys of his age. This is because the hormones which help sexual development do not work properly in PWS. There are no records of men with PWS fathering a child, but there are a couple of rare cases on record of a woman with PWS having a baby. Your brother or sister may find it difficult to come to terms with the fact that they are unlikely to have children, so you need to be very careful when talking about this. They may be too young to know at the moment anyway, so it is best to leave it to your parents to discuss this with them.

People with PWS can become easily confused, anxious and upset. They find it very difficult to change direction when they are set on a particular path of thought or action, and they may show their confusion, frustration and anger by having “temper tantrums”. These are very similar to the type of tantrums that

toddlers and young children have - your brother or sister may find it very difficult to grow out of them. Many people with PWS have learning difficulties, which means they may need to be in a special class or a special school, and may not be able to read, write, or do maths as well as other children. Some have more problems than others.

Your brother or sister also has many shared characteristics with you and the rest of your family. Although people with PWS are similar to one another, they are also as individual as the rest of us, so don't blame everything they do wrong on PWS!

## **Life with your brother or sister with PWS**

Here are some things which some brothers and sisters have said are good about living with a sibling with PWS:

- seeing them achieve goals which had not been thought possible
- having a balanced diet and healthy food
- the loving, funny, caring and kind personality of the person with PWS
- learning to be patient and tolerant
- their help around the house
- helps to face other challenges in life
- teaching them to do things
- better understanding of and meeting others with disabilities
- she always gives me something to laugh about
- taking part in special activities
- she has never felt sorry for herself, and this has made me less selfish

And here are some of the things that are not so good:

- can't eat food in front of him and guilty feelings about food
- coming to terms with the fact that the person may die young
- unable to do things on the spur of the moment
- sometimes she is grumpy
- sometimes he is annoying
- listening to constant repetitions of questions or topics
- temper tantrums
- slowness of person with PWS - always having to wait for them
- parents worn out with looking after the person
- worrying about taking responsibility for the person with PWS in the future
- having to "babysit" for someone who should be old enough to stay on their own

- constant arguments
- obsessiveness
- lying on the part of the person with PWS
- embarrassing behaviour in public
- person with PWS butting into conversations

## How to cope

None of us are perfect, and we can't be expected to be in a good mood all of the time, but there are some things which might help you to cope when the going gets tough.

## Food

Our whole society revolves around food. You only have to switch on the TV, open a magazine or go in a shop, and it is not too long before you are confronted with images of food. How many times have you seen an advert on TV, smelled cooking as you walk past a restaurant, and thought, "I really fancy eating that!" This is what life is like for people with PWS all the time, because they are hungry most of the time.



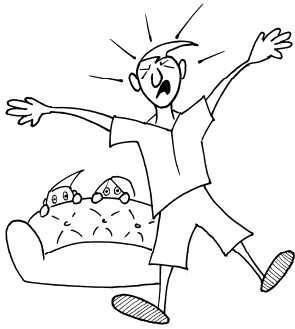
We also tend to eat when we are bored, or feeling a bit down. It is no different for someone with PWS - if there's nothing to do, the first thing they will think of is food.

It is not easy for you, as the sibling of someone with PWS, to control your eating all the time, but it can be a great help if you do not talk about food or eat sweets in front of the person. Your mum or dad can help by giving you sweets or other food treats when your brother or sister is not around.

If you find out that your brother or sister has been taking food without telling your parents, what should you do? Perhaps the best course is to discuss this with other family members first, to decide on a course of action. It is not nice to "tell" on your brother or sister, but they need to be aware of what the consequences will be if they are found out.

On the other hand, it can be very depressing for the person with PWS to be followed around all the time "just in case".

## Temper tantrums



These can be embarrassing, very noisy, and very stressful, especially if they happen in public. It can be useful to remember that your sibling with PWS does have very real difficulties in controlling their emotions, especially anger, and things can quickly get “out of control”. There are some things you can do to stop them happening in the first place, although sometimes they will happen no matter what you try to do. But try these first:

- Ask yourself if they have a point. Is someone (maybe you?) being unfair to them? If so, try to see their point of view. It sometimes doesn't cost much to agree or to give in if they are in the right.
- Are they confused or anxious? Can you help by explaining things simply?
- Turn the situation into a joke
- Don't “wind up” the situation by doing or saying things you know will annoy the person.
- Distract your brother or sister by asking a question about something they are interested in, or suggesting they do something else.
- Don't lose your temper. Try to keep cool.
- Don't shout or yell as well.
- Walk away (if this is possible, and does not leave the person on their own in a potentially dangerous situation).
- If you know they are in the wrong, and it is not a minor issue, or one you don't feel they should get away with, don't give in to their demands. If you do, they are more likely to throw a tantrum next time they want something.
- If tantrums keep recurring about the same subject, try ignoring your brother or sister every time this happens.
- Ask your brother or sister to take “time out” (if possible) eg going into the garden, into their own room etc, until they have had chance to calm down.

## Repetitive questions and conversations

If you are lucky, your brother or sister will not have this trait, but the majority of people with PWS do - and it can be very wearing, having to repeat yourself over and over again. Here are a couple of things you can do to help them “switch off”.

- Remind your sibling that they have already said something several times, and ask them what your answer was.
- Tell them that you will discuss the topic for another 5 minutes, and then you have to go and do something else. If necessary, point out when the 5 minutes will be up on the clock.

## **Arguments**

All brothers and sisters argue from time to time - it would be strange if they didn't. In some cases it may be only occasionally, and in others much more frequently, but there seems to be a difference when it comes to arguing with your brother or sister with PWS. For one thing, they just won't listen to reason - or so it seems! Researchers have found that people with PWS actually have a great deal of difficulty in understanding things which are just said to them: they need showing as well. So if you know you are really in the right, try to prove your point by showing the person what you mean, rather than just yelling at them. If your sibling is a persistent arguer you may find that your only course of action is to walk away, after warning them that this is what you intend to do in so many minutes time.

## **Bringing your friends home or being out with your sibling with PWS**

People are more likely to be understanding of your sibling if they know something about them. Showing them this leaflet may help.

Being out with your brother or sister can sometimes be very embarrassing. Nowadays, people are getting more used to seeing people with disabilities out and about, so hopefully they will be a little more understanding. The thing about embarrassment is that only you can do anything about it. Try not to let it get to you. If someone is staring at you or making remarks, it is their problem not yours - they are the ones who should be embarrassed!

## **Your responsibilities**

Every family will have different viewpoints on how much relatives should be involved in caring for a disabled member of the family, either in the present, or in the future when your parents are no longer able to care. Much will depend on your religion, culture, or family situation.

It is important that you and your family discuss what your responsibilities might be, now and in the future, so that you can iron out any difficulties before they come to crisis point. Although many people are reluctant to involve Social Services, they can be very helpful in providing practical support to families with a disabled member.

## **Helping yourself**

Life with a brother or sister can be very stressful, and everyone has different ways of coping: going out for a walk, staying overnight at a friend's house, listening to music or watching TV in your bedroom (if you have one to yourself) etc.

If you need to talk to your parents about something important, don't try to do it when your brother or sister is around. If that is difficult, let your parents know you want to discuss something with them, so that you can arrange a time when your brother or sister is either out of the house or in bed.

If your sister or brother never goes away from home, either to stay with relatives or for respite care with your local social services, ask your mum or dad if they can arrange for some time for you to be on your own with them.

### **And finally ...**

Yes, life with a brother or sister with PWS is not fun all the way, but with better understanding and managements things can get better. Here are some of the positive comments made by people who have brothers or sisters with PWS:

- *Although my sister is different she's extremely special and has brought to our family a sense of joy and binding.*
- *I didn't know there was a syndrome called PWS but now I read more about it I feel I can help my sister in different ways.*
- *Until very recently, we argued constantly with each other, and I just kept on ignoring her, and I am now beginning to understand more about her.*
- *We love him!*

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### **Thank you**

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