

A Parent's Experience of Schooling for a Child with Prader-Willi Syndrome

Prader-Willi Syndrome (PWS) is a genetic disorder. Along with the physical characteristics of insatiable appetite, small stature and low muscle tone, children with the syndrome typically have mild to moderate general learning disabilities. This article presents a parent's perspective on raising a child with PWS and the difficult decisions involved in finding the most suitable school for him.

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INTRODUCTION

Our son, Dara, is fifteen. He is our third child and he has blue eyes and blond hair...and he has Prader-Willi Syndrome (PWS). I could write and better still, I could talk about PWS for days and I would still only be able to give you some idea of what it means for Dara and for us his family, so I will give you a brief outline of the syndrome and some detail about Dara and in particular our experience of school life over the last ten years or so.

There are approximately 100 to 150 adults and children with PWS living in Ireland. As PWS is generally not hereditary, it does not re-occur in families but there is a hereditary cause in approximately 1% of families and this can lead to other children being born with the syndrome. The characteristic which is most often used to describe the syndrome is an insatiable appetite. This means that children born with PWS crave food and many feel hungry all day long regardless of how often or how much food they have eaten. They cannot control this urge for food and will do anything to get food. They can be highly manipulative and they may do things such as raid the kitchen at night or steal school lunches from other children. Some children will resort to eating toothpaste or even soap.

A small minority of children don't have the same level of craving but all have a heightened interest in food and need constant reassurance about the next snack or mealtime and cannot cope with the possibility that they might miss a meal. They will wake thinking about breakfast and as soon as that is over they will begin to talk about snack time and after that they need reassurance about lunch and after that dinner and so on. Each day their thought pattern will be the same. Generally once reassured they will relax until it gets near to meal time again or they see food. Having food on display such as fruit bowl or cake or biscuits left out, or even a glass of water on a counter is temptation that they simply cannot resist. Their eyes will constantly be drawn to the food or drink and they will be unable to focus on any topic or conversation. People with PWS have been found not to burn calories at the same rate as the general population. This means that even if people with PWS eat only the normal recommended daily amount of calories for their height and weight, they will still become overweight and so their diet has to take this into account.

In appearance, the children are usually small in height for their age and in adulthood are often of short stature. All tend to have very small hands and feet and boys will have small genitalia. Some children have almond shaped eyes and a down-turned mouth. Many will develop weight problems at an early age which can easily lead to the child becoming very overweight and obese.

The children often have low muscle tone and can tire very easily. This can affect their ability to do physical exercise. Scoliosis (curvature of the spine) is also a common medical condition associated with the syndrome. Children with PWS have been found to produce a low level of growth hormone and in recent years many children have been prescribed treatment which involves a daily injection of growth hormone. This has a benefit in terms of gaining height, reducing weight gain and improving muscle strength. Most children have impaired intellectual ability ranging from a mild to moderate general learning disability.

DIAGNOSIS AND INFANCY

Dara was diagnosed with PWS at about two weeks of age. He was very floppy at birth, couldn't suck and had to be tube fed. He had the characteristic soft skin, small hands and feet. He also had blue eyes and blonde hair which many children with PWS tend to have. However, my two older children have blonde hair and blue eyes too and they don't have the syndrome!

Dara, like many children with PWS, reached the developmental stages with a delay of roughly twice the normal age. He was unable to hold his head up until he was over six months old. He didn't sit until he was over a year old and he didn't walk until he was nearly three years of age. Children with PWS don't cry very much as babies, if at all, and so they are very quiet children. Dara began to say a few words only when he was four, and only after much persistence by all the family and my daughter in particular. He always had a huge will to speak and simply refused to learn even simple sign language. He survived for about twelve months with "dada", "mama" and "help" and he got lots of use out of that last word.

Language

His language skills "took off" after that and language would be one of his strengths. My friends poke fun at me at times and say that Dara speaks like a civil servant – yes I am a civil servant – and his usual language includes words like "agenda" and "plans" and "focus" and even "eliminate" but as yet he isn't using that term "going forward"! He follows the pattern of other children with PWS and will ask the same question over and over again. He will try to interrupt conversations with questions, questions and more questions. It is like being stuck on the repeat button. You find yourself answering the same question three or four times before saying, "Enough", or getting cross with the persistent questioning and interruptions to your line of thinking. I often say that since Dara starting talking, I haven't been able to think through any thought or idea in my head without being interrupted. It can be very frustrating at times.

Physical and Social Development

Dara continues to have low muscle tone and tires very easily. He has difficulty, for example, opening heavy doors or walking uphill and can't sustain long periods of walking or running. Often, he seems fine at the time but will be sluggish and tired the following day or two. He has also developed scoliosis which may require surgery at some time in the future and meanwhile it also restricts his physical activity.

Dara started growth hormone about seven years ago but stopped the treatment last Christmas as his scoliosis had deteriorated. At fifteen, Dara looks like your average twelve or thirteen-year-old in appearance and in height. He doesn't have any weight problems and indeed was slightly below his expected weight until recently. You can't tell immediately that he has

PWS. There are advantages and disadvantages to this. He is friendly, cheerful and generally very well behaved. He gets on well with people and they with him. It is clear from his speech and his conversation that he is a child with special needs. He tends to engage people working in shops and restaurants and he is always remembered and welcomed with affection.

Diet

Dara has been on a very strict diet from birth and I manage this through calorie counting and very limited access to “treats”. For instance, Dara always uses ‘Super Milk’ because it has lower calories but enhanced calcium which he needs. He loves his breakfast cereal whether it is ‘Cornflakes’, ‘Krispies’, or ‘Weetabix’. Equally, he likes porridge and no sugar muesli. Basically he doesn’t have the cereal with the sweet sugar tastes but yet he is quite happy. Dara has the same dinner as everyone else but he gets a smaller portion. I use a smaller dinner plate for Dara and give him more salad or vegetables than potato or rice or noodles. He loves Chinese food. Dara has never had sweets or chocolate, ever. He wasn’t given crisps or biscuits as a small child but as a teenager he now has low calorie crisps or packets of snacks provided the calorie count per packet is less than 100 calories. He might have one or two rich tea biscuits in a week. His treat is a scone which he loves. He wouldn’t swap a scone for any piece of cake or chocolate. He doesn’t object to these dietary restrictions and has no problem with other children around him having their “treats”, so long as he has his “treat”. So we supply low calorie snacks to school and to summer camp and friends’ Mums and seldom encounter any problems.

EARLY EDUCATION

So, I have told you a lot about Dara and PWS and as yet very little about school. Dara’s school life began when a very nice lady agreed to take him into her morning pre-school playgroup. He was about four at the time but fitted in nicely with the other two to three year olds and enjoyed his time there. At five Dara still wasn’t toilet trained and he was still very small. Although he could walk, he would topple over easily and had very little strength so we continued to use a buggy for anything but very short walks. He was very quiet, very happy to sit and watch others play but not interested in the rough and tumble. Shortly after the diagnosis of PWS he was referred to an agency providing support services for children with general learning disabilities, and had the support of a multi-disciplinary team which included a psychologist.

Dara was offered a place in a special school. The school had excellent facilities and teachers. Meanwhile, I also looked at a local Montessori school which took children up to the age of about six years. This school didn’t have a place for him the previous year but was prepared to offer him a place at this stage. We gave both options serious consideration but on balance we decided to try the local school for three main reasons. Firstly, the school was local and had a mix of children, including two children with special needs in a class of about ten pupils. Secondly, the classroom environment was calm and quiet, and finally, the children each worked at their own pace. We felt that Dara would learn by example and we liked the idea that he would have children without special needs to engage with and hopefully he would learn from both the teachers and the children. When visiting the special school, he loved the teachers but found the noise and the boisterous behaviour to be very frightening. During this time, we had the support of a liaison teacher from the special school who came once a week and worked with Dara and his teacher for an hour or two. This was a new experience for both the liaison teacher and the Montessori teacher and worked really well for all concerned. I

really appreciate the fact that both were willing to try this arrangement although it was new and different.

At six years old, we began to consider the options available to us after Montessori. Dara had an educational assessment and he was considered to be functioning in the high mild range. His language skills would have pulled up his score while his number skills were extremely low. He could recognise and sound letters but couldn't read. He found holding a pen or pencil difficult but could tell you the names of the continents. He loved speech and drama and trips to the library. He was very well behaved but would tire very easily. In appearance, he looked much younger than his age, maybe more like a three or four year old. The Montessori teacher and some parents were interested in setting up a Montessori school for six to twelve year olds and I was asked to get involved. After a huge effort, much stress and with a very small grant, we set up the school and employed a teacher to teach six or seven children in a premises loaned by a parent. The school was run by a "not for profit" company and the fees were set to cover the salary and expenses. No one made any money out of it. We offered one or two places each year to children with special needs. It was very hard work and obviously I intended that Dara would go there.

BIG DECISIONS

At seven, Dara was still very small but toilet trained and walking to school, about a fifteen-minute walk. His language was improving all the time. He was happy with other children, chatted away and was very social. He still couldn't do his shoe laces or his buttons and was only beginning to be able to use zips. He could now hold pens and colour. He was much more successful with markers than crayons because he didn't need to use so much strength to get the colour on the page. He was becoming very good with scissors. I began to question whether the small Montessori class would be best for him and also the school didn't have access to a playground or any of the extra curricular things like choir or games etc. I went back to the special school and reconsidered that and other local special schools but I had the same concerns that I had before, mainly he wouldn't improve his social skills and would be more likely to copy and learn the less desirable behaviours. The children were much stronger, more vocal and simply more boisterous in a happy and friendly way but this still was very frightening for him and he wasn't happy in that environment. He wouldn't join in and simply withdrew.

I approached a local mainstream school, although not the same school that my older children had attended. I understood from other parents and friends, that this particular school had a special quality and that the principal and teachers were very committed to creating and maintaining a very positive culture and school environment. The school also was understanding of the needs of children with learning difficulties and/or disabilities and catered well for those needs. I met the principal and the teachers myself and I was very impressed with the school and particularly with the open, positive and encouraging atmosphere. This was evident throughout the school from the art work on the corridors, the photographs and of course the manner of the children as they went about their day. Dara would need a special needs assistant and the school had not previously had a child with an assistant but again this was never an issue but another new experience to be embraced by all, principal, teacher and class. So after much consideration of all the options, Dara went to mainstream school with a special needs assistant. I wanted Dara "to fit in" in a normal environment where he would be seen as different but accepted for all that difference. That is exactly what happened. Everyone did just that, and it was really great!

SCHOOL

Dara started primary school at seven years of age in senior infants and went all the way through to sixth class. We were very fortunate to have the facility of a special class in the school. Initially he spent most of his time with the mainstream class but as he got older and the gap between his ability and that of the rest of his class widened, Dara spent more time in the special class. He continued to do the general things like religion, art and crafts and some projects for geography and history with the main class. He took part in school plays and tours. He particularly liked choir and joined his main class in the church for singing at communions, confirmations etc. He loved being in both classes and never saw a problem moving from one to the other, in fact he thought that he did “harder work” in the special class than the other children did in the main class. He regards himself as “special” but doesn’t associate that with any negative issues. The other children were totally accepting of the two children with special needs in the class. They always engaged with them and never excluded or ignored them. I sometimes brought Dara to school but on most mornings he went to school with a friend and neighbour who also worked in the school. On the occasions when I did bring him to school, I was delighted as everyone greeted Dara on our way in and chatted as we waited for the school to open.

Of course there were challenges for Dara, for the school and for me. I remember one day shortly after he started and the teacher came out of school with him. Dara had put his head down on the desk and started crying and the teacher was worried that he had upset him and couldn’t understand what had happened. I explained that when he just couldn’t cope or was too tired, mentally and/or physically, he would get upset. He just needed a little bit of reassurance and a rest. The rest could be a distraction, like going for a little walk down the corridor or a different piece of work. He would then be able to go back later to the original work. His special class teacher was always marvellous at knowing whether to take the stern approach, the distraction approach or the patient and empathising approach and so generally there were not many serious behaviour issues.

Moving On

Dara is now coming to the end of his first year in secondary school. He is attending a special school for children with moderate general learning disabilities. We made the decision after almost two years of considering various options and visiting many schools. He found the change to a new school difficult and initially he was concerned about the behaviour of the other children. He has settled in now but still feels insecure and depends greatly on his teacher and the classroom assistants. Again his teacher is excellent and manages him really well. She has a real understanding of him and the issues that will cause him to be anxious or worried.

UNDERSTANDING CHALLENGING BEHAVIOUR

Dara’s behaviour has become more challenging as he gets older. All the research will tell you that children with PWS don’t want to be bold on purpose, but he can be stubborn and difficult without being “bold”. It certainly can be challenging for all concerned. Mostly his difficult behaviour is triggered by worry or anxiety over something and in particular over changes. When he is being difficult, he won’t do what he is told and will simply ignore you, do the opposite to what was asked or start crying. Initially he could easily be distracted by simple things, like asking him about something that he is interested in, “Did you hear about ...did

you see that...” and he would forget whatever had caused his worry or anxiety and so his behaviour would improve immediately.

There are a few occasions when things are not resolved during the school day. Then Dara will not want to go to school the next day because he feels that people were cross with him and he really needs to know that the people he relies on, like his parents, teacher and special needs assistant, are happy with him and like him. When very stressed, he will keep saying, “Mam, fix it” and even then he will often not be able to tell me the problem. I am used to trying to look back over a day’s events or even earlier and figure out what might have triggered the anxiety or worry. Dara can’t move physically or mentally until his problem is fixed. As he is unable to explain all this behaviour simply appears to be unreasonable. The answer generally lies with having lots and lots of patience and trying to work out what has upset him or what is bothering him.

CONCLUSION

Life isn’t easy for any of us but it can be very difficult for a child to be understood sometimes, particularly when they can’t always explain what they are worried or anxious about. I wish I had understood this better when I first became a parent nearly thirty years ago. I am so glad that I have found principals, teachers, assistants, escorts and minders for Dara who are willing to learn and understand Dara and PWS and who have been such willing companions for us along this journey through school life. Thank you so much for your courage, your commitment and your support.

Information on the Prader-Willi Association of Ireland is available at www.pwsai.ie