

Important considerations for Supported Living and Prader-Willi Syndrome

Introduction

Prader-Willi syndrome (PWS) is a complex genetic condition, involving a range of medical, cognitive and behavioural characteristics, which vary in extent from one individual to another. This means the success or otherwise of interventions and approaches can differ in different situations. People with PWS are unique and require individualised person centred approaches to their social, therapeutic and medical care. However, because many adults appear relatively able, it is common for the needs of someone with PWS to be under-estimated by those providing or purchasing care for them. Carers require good training and understanding of the syndrome in order to support individuals with PWS successfully.

In almost every case, it is necessary to restrict access to food. This is because individuals with PWS have a dysfunction in the hypothalamus which means they feel hungry most of the time and will hence seek out food. Research shows that the messages which normally reach the stomach from the brain to tell someone they have had enough to eat are lacking in someone with PWS. Due to an “impairment of brain or mind”, people with PWS therefore lack the **mental capacity** to know when they have had enough to eat. Added to this is the fact that low muscle tone means they require fewer calories than other adults of the same age and height. Account also needs to be taken of:

- **challenging behaviour**
- **physical problems**
- **hidden risk factors**
- **any mental health issues**

PWS and supported living

Supported living (sometimes called “assisted living”) is the term used to describe an individual living in his or her own tenancy, either alone or with others, and receiving support from a care provider for between one and 24 hours per day.

The PWSA UK has worked with a number of people who have been in a supported living scheme over recent years. The work we have undertaken has been in an advisory capacity for the individual or the scheme itself, or in a parental support context. Placement breakdown is not uncommon, leading to a move back into residential care, home to families or, in the case of a few, their untimely death. The reasons for the breakdown in these tenancies have been varied, although there are common themes:

- There is no plan in place to restrict access to food (eg by locking cupboards or providing just enough food for each meal during the day) or staff are unwilling or unable to restrict access to food. People with PWS can be highly manipulative in their quest to obtain food, and they may also obtain food from other service users or members of the public. Food-stealing from shops is not uncommon.
- Insufficient, inconsistent or non-existent support - there is no monitoring provision made to ensure support staff arrive on time or indeed arrive at all.
- There is a gap between the support planned and that actually delivered.
- Staffing shortages leading to agency staff being used, usually untrained in PWS.

- At the commencement of a scheme, highly trained and knowledgeable support staff are put in place. However, when, for whatever reason, these individuals leave the scheme, they are not replaced by staff with similar knowledge or skills.
- There may be over-reliance upon particular support staff to deal with situations, leading to "burn out".
- The location of a supported living unit is unsuitable because due attention has not been paid to the fact that a person with PWS does not have capacity in respect of food and, therefore, temptations must be put beyond their reach. Examples include a supported living unit behind a chip shop, takeaway or supermarket; or the kitchen facilities and food being fully accessible to the person with PWS.
- People with PWS invariably, from time to time, exhibit behavioural challenges which may be unpredictable and/or become aggressive.
- There is unsuitable accommodation for support staff within the unit e.g. no "safe" space for staff to have "time out" or record and store important information without the person with PWS being able to access that information and possibly remove or destroy it.
- People with PWS are vulnerable adults and there is an inappropriate balance between the competing needs of independence and protection: the desire for food can be extremely compelling, leading to financial and sexual exploitation.
- Some people with PWS have a higher intellectual level, which may take them out of the learning disability range. However, their social and emotional immaturity means it is usually very difficult for them to function at their intellectual level. This can lead to inappropriate, under-resourced care packages, with service users being perceived to be much more able than they actually are. This has led to expectations not being met and relationship breakdown between support staff and the individual concerned. A person with PWS who does not have a measured learning disability may have an exaggerated sense of their coping skills, and can be vulnerable to the exploitation mentioned above.
- Inappropriate assessment and ongoing monitoring of a placement. In one case, a person with PWS began to feel isolated. This issue was not identified and the individual absconded on several occasions, eventually leading to placement breakdown.
- People with PWS are prone to behavioural difficulties and have limited social skills. The timing and extent of outbursts are highly unpredictable. A provider of supported living is required, therefore, to ensure 24/7 support or have in place monitoring and support at very short notice. There have been instances of screaming, shouting, and exhibiting tantrums in public places resulting sometimes in physical and verbal assaults.
- Due to insufficient support of staff, the threshold at which staff begin to feel uncomfortable is reduced, resulting in placement breakdown.

Best practice in supported living

The following recommendations were made by members of the PWSA UK Residential Care and Supported Living Forum to facilitate successful supported living provision:

- Location – not too far, but far enough, from shops. No fast food outlets nearby.
- Staff given appropriate and ongoing training.
- Effective staffing levels
- Staff consistency (no agency staff)
- Access to OT, physio, podiatry, dietitian, etc via health service
- 24 hour support
- Weekly programme of activities
- Involvement in the local community
- PWS knowledgeable GP
- Available opportunities for exercise and leisure
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- Space to engage in activities eg large garden, pets, crafts etc
- Appropriate for the individual (person-centred)
- Good relationship with local authority
- 1:1 hours built in to care package
- Agreement about restricted food access
- Understanding landlord
- Access to public transport
- Care package which meets all the person's needs
- Communal and private spaces
- Compatibility with others sharing the provision
- Access to jobs – paid and voluntary

Lessons to be learned

Statement by Professor Tony Holland on the case of KD (widely reported in the UK press, July 2015) a young woman who died at the age of 23 years, after going into supported living provision



KD had Prader-Willi syndrome (PWS), a genetically determined disorder known to be associated with a very high risk from early childhood onwards of severe obesity due to an inability to regulate food intake. She died from the consequences of severe obesity, having moved into supported accommodation and, as reported by the Coroner, no effective action was taken to do anything about the fact that her weight was increasing.

Although a rare syndrome, there is abundant information freely available to any one concerned describing the fact that the brain mechanisms that regulate food intake, so that calories taken in as food and drink and calories used are appropriately balanced, are faulty in people with this syndrome. What experience has shown is that the consequences of this inability on the part of people with PWS to be able to regulate eating behaviour – life threatening obesity – is entirely preventable through appropriate food-security and ensuring that access to food is supervised at all times.

Efforts to control access to food may not be easy and can be initially resisted by the person with PWS, sometimes leading to quite problematic outbursts, but again as experience indicates once proper arrangements are in place and experienced and well-informed family members or staff are supporting the person, the anxiety experienced by people with PWS is reduced and behaviour improves and, importantly, life-threatening obesity is prevented.

Why then does this happen and how could it have been prevented?

First, there is a failure to appreciate that this behaviour of extreme over-eating is intrinsic to this syndrome and has its basis in a failure of the normal feedback systems in the brain. Whilst the degree of this may vary from person to person with PWS it is always present and the risk of over-eating and resultant severe obesity is ever present. This is not a matter of lack of will or weak moral fibre, this is a biological determined problem.

Secondly, whilst parents, having been informed early in life about the fact their child has PWS and the consequences of this, are expected to act in the best interests of their child and, in the case of a child with PWS, this means managing the food environment, uncertainty arises as to the duties and responsibilities of others in adult life. As was illustrated in KD's situation, severe difficulties inevitably arise with a move to independence in adult life.

Thirdly, we, as adults, take responsibility for those decisions that it is for us to make – what to do, what to wear, how to spend our money etc. We may seek the advice of

others, who may wish to encourage us one way or the other, but ultimately these are our decisions and we live with the consequences. This, as a starting point, is true for all people including adults with learning disabilities.

However, what is now well recognised and enshrined in law - the Mental Capacity Act 2005 for England and Wales and the Adults with Incapacity (Scotland) Act 2002 - there are circumstances when, because of some disorder of the brain or severe mental disability, it is not possible for the person to understand, balance the facts, and/or communicate his/her wishes when it comes to particular decisions, whether about treatment, personal matters etc. This is termed a person's decision-making or mental capacity. How this is defined, and the fact that it relates to specific decisions and particular points in time, is all described in the relevant Acts and their Code of Practices. Where an adult meets the necessary criteria and is considered to lack the capacity to make a particular decision or to undertake a specific act (e.g. consent to treatment, sign a contract) others can act on his or her behalf providing they do so in that person's best interests. In England and Wales how 'best interests' must be assessed is also described in the Mental Capacity Act and in the Code of Practice.

What happened in KD's case?

The facts are not fully available to us and we comment with caution but what we can say is what we believe should have happened.

Our understanding is that KD was considered to have the capacity to make decisions about her care and what she ate - if that was the case, then to override her wishes would have been unlawful. There is much to indicate that, particularly when living in conditions where food is readily available, people with PWS do not have the capacity to make such decisions - whilst they may appear to understand the basic issues, they do not have the ability to balance the relevant facts and to act in a manner consistent with their usually-expressed wish not to put on weight.

Frequently people with PWS will actively deny there is a problem when all the evidence is quite clearly to the contrary - his/her weight is going up and severe physical problems such as swelling of the legs and resultant leg ulcers, diabetes mellitus, and increasing physical limitation are developing, all indicative of the seriousness of the rapid weight gain. Whilst it is accepted that people can and do make unwise decisions, in this particular situation of someone with PWS, continuing excessive eating and life-threatening weight gain taken together with knowledge about PWS should at the very least have called into question the initial assessment that she had capacity. In the event that her capacity to make decisions about her eating was in doubt it should then have raised concerns that by not acting to prevent obesity her right to life was not protected.

The Mental Capacity Act and the Court of Protection (England and Wales)

We do not force other obese people to diet. However, usually those who are obese for other reasons do recognise that they are obese and would wish to do more to prevent it becoming worse. In the case of people with PWS, the neuroscience tells us that there are fundamental and severe problems for people with PWS when it comes to the regulation of eating behaviour, such that, if not managed, life threatening obesity is inevitable. In such a context the Mental Capacity Act for England and Wales makes it clear that, for decisions, the outcome of which might be death, those questioning a person's capacity must be very sure that the person making that decision really does have capacity and that he/she fully understands that he/she will develop serious health problems and he/she is at a very high risk of death.

Again, we do not know what happened in this case, but where decisions are complex and the outcome potentially so serious, for England and Wales it is the Court of Protection that is the Court that arbitrates when it comes to issues relating to the Mental Capacity Act. This is particularly important where a person's capacity is in doubt, or where what is in the best interests of a person who may lack the capacity to make a particular decision is disputed. We would argue that in such difficult situations, as in this

case, an application to the Court of Protection is a very sound and sensible course of action.

Recommendations

In the light of this sad event and knowing that it has happened before and could easily happen again, the Prader-Willi Syndrome Association UK make the following recommendations:

1. Social care, Local Authority, and health personnel responsible for, or involved in the support of either a child or adult with PWS, have a duty to fully familiarise themselves with the basic facts pertinent to PWS and to appreciate their responsibilities towards that person with PWS, his/her family and to relevant others.
2. Social care, Local Authority and health personnel responsible for, or involved in the support of an adult with PWS, must be fully aware of the risk of severe and life-threatening obesity that will arise with greater independence and act in accordance with the law and their duty of care towards that person.
3. Those responsible for the commissioning of, and providing support to, adults with PWS have a very specific duty to ensure that food security is in place, ideally with the full agreement of the person with PWS, but where that agreement is doubtful or absent to then fully and comprehensively discharge their duty to determine the capacity of the person with PWS to make the relevant decision as set out in England and Wales in the Mental Capacity Act and the Code of Practice and in Scotland in the Adults with Incapacity (Scotland) Act 2000. We anticipate that consideration will need to be given as to the extent of restrictions that are necessary where the person is found to lack the capacity to consent to his/her care plan, and importantly whether this amounts to a deprivation of liberty and, if this is likely, to seek authorisation according to the relevant legislation across the countries of the UK.
4. Where there is doubt, we would advise that the responsible Local Authority should seek leave to apply to the appropriate Court in that jurisdiction (e.g. Court of Protection in England and Wales and Court of Sessions in Scotland) for the necessary ruling. We also ask that all Local Authorities recognise that, in the case of people with PWS, there is the very real potential for their being rights that may conflict, such as the right to life and the right to a private and family life. Given this, we call upon Local Authorities to recognise the need for services skilled in the support of people with PWS that are fully informed about PWS and are aware of questions of decision-making capacity and are able to provide support locally in such a manner that can ensure that people with PWS can have good, safe and healthy lives close to where they want to live and are supported to make those many decisions in their lives that they are well able to do.

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