Q. How do we access the Maudsley Clinic?

A. Approach the local CAMHS service first either via your GP or SENCO. If the local CAMHS are in agreement a referral can be made to the Maudsley specialist clinic.

Q. We managed to get referred to the local CAMHS but they would not take our child. Is this normal?

A. CAMHS service, like many things is falling off. It might be worth approaching the Educational Psychologist, who may know about PWS – it is a concerning situation. You need to demand better service - it is difficult being a parent, as they do not carry the weight that a consultant does. The NHS should have a duty of care to ensure that everyone who plays any part in the care of a child with PWS knows what it is and how to best care for that child. Even parents who are GPs and therefore know the system still have to fight for everything. Local services should educate themselves sufficiently if they have a child with PWS. An IQ cut off of 70 cannot be given as a reason for not seeing a child – it is a violation of human rights to discriminate on the basis of IQ. The issue is about commissioners putting money into the services.

In some schools the child has its own plan and inclusion assistant. Schools should only accept a child with PWS if they are able to get it right. There seems to be a lack of consistency throughout the country. Advice varies from PCT to PCT. There needs to be standard guidance but the research is not yet at the stage where there are textbook answers.

Behaviour in children may be a form of communication. Understanding the function of behaviour helps in the understanding what they are telling you and makes the way forward easier.

Q. I am working with a six year old child with no speech yet who uses Makaton. What progression in speech should I expect?

A. Continue to use a combination of speech and Makaton and don’t stop assuming that speech will never develop. Children can acquire language at 11. Makaton enhances the ability to speak. A cognitive assessment would enable you to know what you can and can’t expect in school.

Q. What do you feel about the use of ABA in a PWS child on the autistic spectrum?

A. Applied Behaviour Analysis as sometimes described in autism is an early intervention and an extreme approach of rigorous monitoring which would need to be applied in all settings, not just school. It would not be particularly suited to children with PWS and the schools it is offered in are for Autism and do not think that any have children with PWS. The wider Applied Behaviour Analysis for children generally is an approach used by all.
Q. How do you stop skin picking, in particular rectal picking in the loo and head picking during the night?

A. Keep a diary for all types of skin picking to see what happens directly before the episode and directly afterwards. Is it socially rewarded – e.g. sent to the teacher and gets one to one attention, or avoids having to do something? Ensure that rewards come without skin picking.

It is thought that there is little pain feedback, so that the child never learns not to do it. Sometimes it is a routine – perhaps always picking a scab. It may be uncontrollable. You need to try and outcompete it, offering a reward for one hour without skin picking, then two hours etc so that there is an incentive not to do it. Try teaching techniques for avoiding the urge rather than just saying don’t do it. When looking at the diary it may highlight subtle problems that you might not ordinarily notice but are problems to people with PWS.

Rectal picking will require supervision when child is on the toilet and using rewards, or allowing use of a game which occupies the hands. Finding the right reward is hard, but absolutely never under any circumstances use food as a reward. Some schools use sweets as a reward and is it quite wrong, demonstrating a lack of understanding. Some school’s SENCOs will not modify the school’s behaviour for one child. It can become a battle between school, home and health.

Professionals can also be naïve, suggesting use of items relating to food when testing various abilities.

Pulling out hair during the night or occasionally during the day if very distressed, and sometimes tries to eat it. This is a basic natural grooming behaviour and so very hard to control. It could also be self-stimulatory behaviour, so it would be helpful to do observations on what happens before the behaviour starts in order to determine why your child pulls their hair. In case of self-stimulatory behaviour you should consider offering alternative sensory experiences based on child’s preferences, for example, squoosh balls or tactile fidgety toys. Try a hat or gloves? Rewards relating to hair decorations?

Q. If skin picking is compulsive could it be medicated like OCD?

A. It is unlikely to be helpful as often these behaviours are when very relaxed.

Chris Oliver felt that it would be very useful if the Association produced guidelines about the minimum requirements for every child with PWS. It could be the standards of care recommended by the Association, and signed off by someone such as Tony Holland and the Research Advisory Board.

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Questions and answers from the workshop about Teenager and Adults with PWS

(Note: this workshop had a number of questions to answer which had been sent prior to the meeting and also took questions from the floor. Some of the prior questions were not addressed at the meeting, but were later commented on by panel members in writing)
Q. My daughter always used to occupy herself with making things out of junk and colouring and wordsearches. Then around 16-19yrs odd things she started doing things such as staring at her watch and she would start to shake which at first people thought she was fitting but she wasn't. She still "does" colouring and wordsearches but actually only stares at the words and pushes the pen into the paper or repeatedly colours the same bit, it is so frustrating as she never used to be like that. If I tell her to stop she just says she’s bored.

She is on Escitlopram which is supposed to help these behaviours but she still does them, the thing it seems to help is the worrying about things such as transport being late or breaking down (all "what if" questions).

People said she had just grown up and lost interest in the activities she used to do, I am not so sure. I feel it is something more physical - some sort of change in chemicals in the brain. Does anyone on the panel have any insights into this seeming lack of motivation?

A. 16-19 years is a time of many changes in any person's life. Various questions could be asked. Is this a bi-polar disorder or depression? What are the girl's sleep patterns like? What is she doing in the daytime? However, sometimes people do just grow up and move on, and new interests could be introduced.

Q. Does growth hormone/sex replacement hormone, in the panel's opinion, have an effect on the behaviour with those with PWS?

A. Growth hormone can help to improve motivation and physical health if continued into adulthood. Growth hormone given in childhood beneficially affects the classic PWS appearance (eg small hands and feet). Both of these could help with self-esteem and indirectly affect behaviour, but there is no close correlation.

Testosterone should only be used when necessary and the dose built up slowly. Behaviour may alter but affects vary between individuals.

Q. We access positive behavioural support training with our local CTPLD. In that training it is stated that when people are in a highly agitated state that to calm them down it is prudent to give a cup of tea/biscuit/Mars bar. On our staff questioning this due to PWS the response was that even people with PWS would forget that they had been given food items when in a state of heightened agitation. What does the panel think of this statement?

A. Panel members felt that a cup of tea would be OK, but not a Mars Bar. If food and drinks are given to calm the person, would it then have the effect that the person would create a situation in order to get the "reward".

Q. What can be done about people with PWS who are refusing support, even though they clearly need it? (ie non-compliance)

A. Don't give up straightaway. Eventually the person may come around to accepting support in their own time and in their own way. Take positive, small steps. Praising what they can do and building up a consistent approach to inspire trust. Look for environmental triggers which
may be affecting this. If nothing works, it may be that the Mental Capacity/Court of Protection route should be taken.

Q. What can be done to stop/lessen or prevent skin-picking? What can be done about skin-picking on the top of the head, where it cannot be covered up? What can be done to help prevent rectal picking while the adult is on the toilet?

A. Reward charts have worked well for some - ie going through a day/week/month without picking to earn a reward. Keep finger nails short. Occupy hands with other activities, eg crochet, knitting, "worry balls", Nintendo DS etc. Skin-picking can be a symptom of anxiety and if it increases, look for reasons for the anxiety. Keep records for occurrences of skin-picking so that increases can immediately be recognised. (The panel were unable to offer any advice on rectal picking).

Q. My son is living in residential care. When he is upset he will not get up and refuses to eat.

A. He is trying to control the situation. If it is only every three months or so that he does it, don't worry. If more frequent, It might be helpful to have in place an agreement with him as to what will happen when he gets upset, or construct a reward for getting up.

Q. What is Citalopram?

A. It is a Selective Serotonin Reuptake Inhibitor (SSRI) and is generally beneficial to use in cases of depression coupled with anxiety. Dose may need to be lower for people with PWS.

Q. I have a teenager with PWS whose outbursts are getting aggressive, breaking furniture and hurting himself - what can be done?

A. Try counselling and anger management. Have something available to hit (punch-bag, cushion etc) or kick a football around. Drumming might help.

Questions not answered on the day (teenagers and adults)

Q. What can be done about people with PWS who live at home, but have little or no support when they are out and about.

A. If people are going out but have little or no support, then misdemeanours inevitably will occur. It is then down to ensuring that the relevant persons/agencies are notified so is that the correct level of support may then be identified. It is a matter of gaining the agreement of the person with PWS to accept a particular level of support. When seeking for an individual budget, the level of support should be such that there is someone available to go out with him/her.

Q. Is there any data for PWS being on Prozac (Fluxetine) long term ie PWS patients displaying adverse side effects? My daughter was on 20mg which was deemed to be ineffective so the dose was increased fairly quickly to 60mg. Now a few months later she has had some extreme behaviours and mental reactions which are totally out of character for her. These events have put both her and others at risk so now she is to be 'weaned' off of it.
A. Prozac and particular SSRI antidepressants that help anxiety can be very helpful but they should be used at low dose and with care. In this case it sounds as if the use of a high dose led to the person with PWS becoming high or manic. This is something to be guarded against and medication should be increased with care.

A residential care manager adds: In my experience I have noted that for some it appears to work well. In others it appears to work well for a period of time, sometimes several months. In others it seems to have an adverse effect.

Q. A young lady with Prader-Willi Syndrome wants to move into supportive living. She shows challenging behaviour within her home situation, but not within the day services which she attends. The young lady has attended a residential college, where she has started to increase her independent skills. Is it suitable for a person with Prader-Willi Syndrome to move into supportive living? What would be needed to reduce this challenging behaviour within a supportive living situation?

A. Supported living models can be excellent as long as the amount and quality of support is okay. (From a residential care manager) My work is with adults with PWS in residential care situations. These work very well for most, and what is not broke don't need to be fixed. However, we are looking at the supported living model but this would need to be set up best interests of the individual and in my opinion 24/7 available support. I think that any scheme that is set up for the individual and works it doesn't matter what they call it, it works. I have seen many supported living environments which are more like a residential care than the ones we run.

Q. Have many people with PWS actually been diagnosed with schizophrenia?

A. Adults with PWS (particularly those with UPD) are prone to psychotic illness. However, whilst schizophrenia is a psychotic illness the type of illness people with PWS get is rather different.

Q. (from a PWSA staff member) I am supporting a family with at the moment re a question of capacity. The family has made a complaint against the local authority and I have been to stage one of the complaints meeting with them. The person with PW has supposedly said she does not want the family to know anything about her health issues. She is a diabetic and has also put on lots of weight. The authority has stated she has capacity to make choices around her health. She has recently been sectioned and is being detained under the mental health act. The family are asking if guardianship has ever been used in people with PWS and what was the outcome?

A. There is a case of Guardianship in Scotland, but it is likely to be different as the laws are different. In England and Wales Guardianship comes under the Mental Health Act. It does allow you to determine where someone should live but its powers are limited. I have not come across its use but in principle it could be used.

Q. How can anxiety and obsessiveness be best managed?

A. It is difficult to address this easily – for anxiety it is about the environment but also medication can be helpful if severe (see above). Obsessiveness requires a careful
constructed behavioural approach – seek advice from local community team for adults with LD.

(From a residential care manager) If I knew the answers to those I wouldn’t work the long hours I do. With anxiety getting to know the person, dealing in a very calm way, listening reading body language, learning from experiences and indeed referring to professionals for other types of intervention.

With obsessiveness this in my opinion can be more problematic to deal with. Certainly if the obsession is of a person. Again a multi-disciplinary approach needs to be adopted. In my experience, when an approach is tried it can be successful for period of time and then the obsession returns. Then a different approach needs to be adopted. Strategies can be agreed and we have tried various options overtime, many successful some not successful.

Q. My question relates to our 23 year old daughter who up to about 18 months ago was self controlled with her eating. She now is taking food surreptitiously and has gone from 8 stone to 11 stone over this period. She knows this is wrong and tries to counter by exercise. We go to the gym twice a week and swim weekly as well as walking and weight is stabilised but not going down.

A. It would be interesting to know where she is taking food from/how she is obtaining additional foodstuffs. The increase in exercise has halted the weight increase. For weight to start to reduce then the amount of calories that are being consumed it would appear also need to reduce. It may be that her body mass index is the benchmark for her ideal weight. This perhaps should be between 22-28. It would be interesting to see this as an ongoing case study.

Q. Where can the help/guidance on the issues being discussed be accessed by carers /parents etc. on an ongoing basis? How can I find psychologists with knowledge of PWS and whether they can treat people with OCD?

A. Every area should have a community team for adults with LD – providing it is accepted that your son/daughter with PWS has an LD then they should access that team. There will be psychologists etc. Your GP should know about it. Perhaps the PWSA, could have an e-mail advisory board for all sorts of issues, like it already does for dietary advice. There are many people out there that can give advice, and there are volunteers that the PWSA use the perhaps a specific advisory board for certain issues could be drawn up.