Understanding Behaviour: behavioural diagnostic methods for caregivers in the everyday care of people with PWS

Mental illness and PWS: recognition and differentiation

Tony Holland
Mental ill-health, mental disorder, mental illness: definitions

**Mental ill-health** is a broad term covering mental disorders, psychosocial disabilities, and other mental states associated with significant distress etc.

A **mental disorder** is characterized by a clinically significant disturbance in an individual’s cognition, emotional regulation, or behaviour – developmental or acquired (DSM-V, ICD-10).

**Mental illnesses** are health conditions involving **change** in emotion, thinking, behaviour (or combination) associated with significant distress and/or problems in social, work, or family activities (DSM-V, ICD-10).
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What should alert you to the possibility of mental illness in a person with PWS?

- Be aware of the possibility – age, sub-groups at risk (mUPD), +ve family history
- Persistent or cyclical change in behaviour and/or mood
- Onset of unusual and/or abnormal mental beliefs and/or experiences (hallucinations, delusions)
- Disordered thinking
- Appears bewildered/confused,
- Change in sleep, concentration, activity levels, appetite, general interests
- Other (e.g. abnormal motor behaviours, fixed postures etc)
- Lasts over days or weeks, may fluctuate over time

‘They are different – not their normal self’
Ms X, aged 18 years, has PWS - maternal UPD
On daily GH since early childhood
Lives with her family in a food managed environment – not overweight

Two week ago – sudden onset:
• Increased irritability, over-activity, talking excessively, markedly disturbed sleep, concerned school friends might harm her and are talking about her

Mr Y, aged 17 years, has PWS - maternal UPD
On daily GH since childhood
Lives at home with family, occasional respite, mildly over-weight

Very sudden onset;
• Bizarre behaviour
• Appeared confused, talking incoherently,
• Grabbing ‘butterflies’ from the air
• Odd postures
• Not eating or drinking
What to do if you suspect the person you support with PWS may have developed a mental illness

- Re-evaluate the person’s care plan
- Manage risk to self and others (e.g. suicide)
- Monitor physical health (eating, drinking, presence of physical symptoms)
- Daily observations of mental state (phrases used, behaviour, mood)
- Seek urgent health advice – psychiatric advice
Some key messages

• Significant and persistent change in mood and/ or behaviour observed – ask why?
• Keep records of mood etc and make a note of experiences the person with PWS reports
• Emotional outbursts or skin picking characteristic of PWS are NOT a mental illness but their frequency and severity may change in the context of the onset of a mental illness
• Anxiety or anxiousness in someone with PWS is generally not the same as a generalised anxiety disorder – may be more due to uncertainty and the environment
• Where the onset of a ‘co-morbid’ mental illness is suspected
  • Review care plan
  • Refer for advice
• Psychiatric medication justified and effective providing correct diagnosis
  • Monitor treatment outcomes
  • Be prepared to question and challenge
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Physical illness and relationship to behaviour in people with PWS

Tony Holland
Mr R aged 56 with PWS

Very sleepy and not eating following immunisation against flu the day before

Increasingly withdrawn, sleepy, agitated and distressed

See by GP and in A & E - post immunisation reaction - discharged home

One day later c/o abdominal pain for the first time

Blood observed at the corner of his mouth - vomited

Admitted to hospital

Died that night from aspiration after vomiting large amounts of fresh blood.

Duodenal ulcer identified at post-mortem.
Physical illness may present atypically in people with PWS

- Inability to describe symptoms or recognise their significance
- High pain threshold
- Abnormal temperature regulation

Certain physical illnesses are common in people with PWS

- Obesity related conditions: diabetes mellitus, sleep apnoea etc
- Gastrointestinal conditions: constipation, slow gastric emptying, gastric rupture
- Respiratory conditions: aspiration and its consequences
- People with PWS also get illnesses that may present later because of the above
  - Appendicitis

A change in behaviour or the onset of a new behaviour may be an indication of a ‘comorbid’ and as yet unrecognised physical illness
Some physical illnesses may be directly cause a change in mental state or behaviour.

- Diabetes mellitus – high or low blood sugar – acute confusional state
- Sleep apnoea – excessive daytime sleepiness and morning irritability
- Respiratory infection – confusional state
- Post-operative confusion/mental illness
When an established behaviour changes in characteristics, severity or frequency, or a new behaviour develops, ask why?
Always consider the possibility of a physical illness

If the following are present take it very seriously
• Vomiting
• Complaints of pain
• Loss of appetite and not eating
• Confusion, not knowing time or place
• Excessive and unusual levels of drowsiness
• ‘Just doesn’t look well and not themselves’
Be informed, be prepared to explain to doctors and others, advocate for those you support, have information available.

- IPWSO Website
- IPWSO ECHO programme
- National PWS Associations
- Published papers, practice guidance etc
Thank you
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