

# 6th International Prader-Willi Syndrome Caregivers' Conference

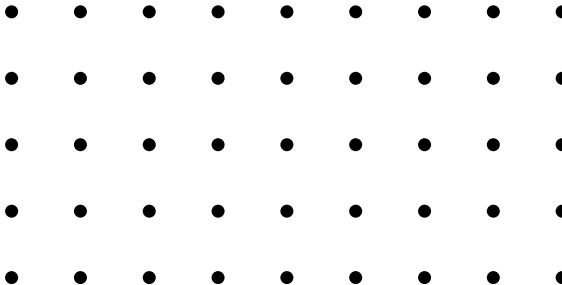
21-23 May 2024, Berlin, Germany

Understanding Behaviour: behavioural diagnostic methods for caregivers in the everyday care of people with PWS  
**Mental illness and PWS: recognition and differentiation**

Tony Holland



#PWSCARE24



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## Mental ill-health, mental disorder, mental illness: definitions

**Mental ill-health** 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'*

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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

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## 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

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## 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

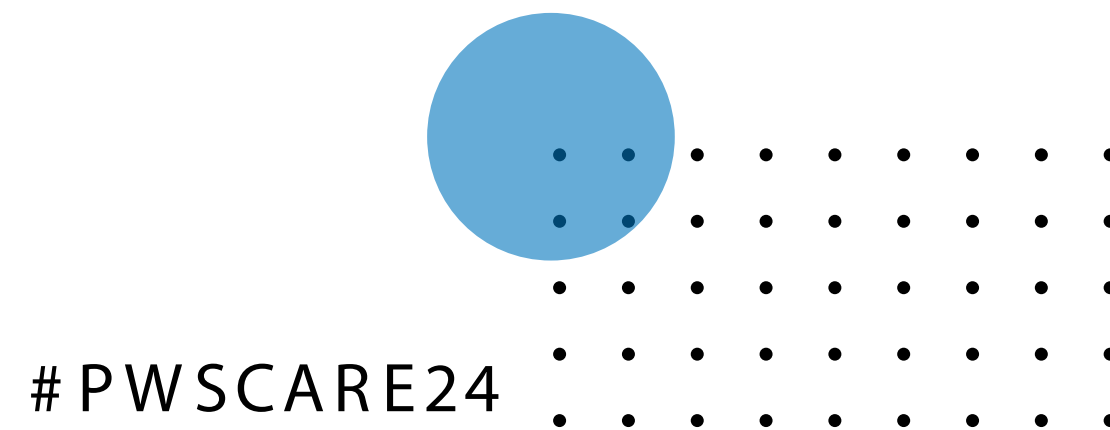


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Physical illness and relationship to behaviour in people with PWS

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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

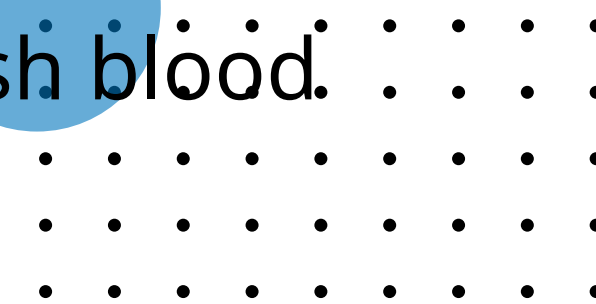
Blood observed at the corner of his mouth - ?vomited

Admitted to hospital

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

Duodenal ulcer identified at post-mortem

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Physical illness may present atypically in people with PWS

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

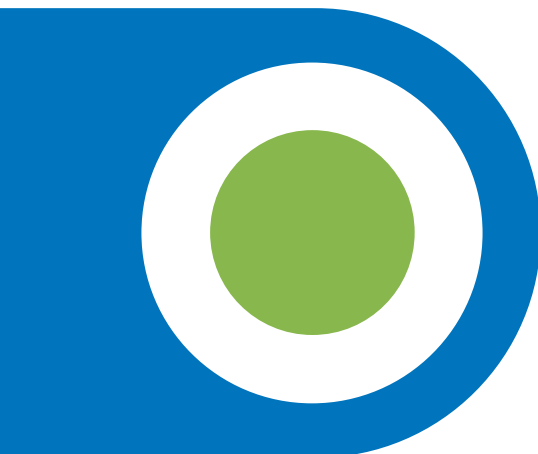
Certain physical illnesses are common in people with PWS

- 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**

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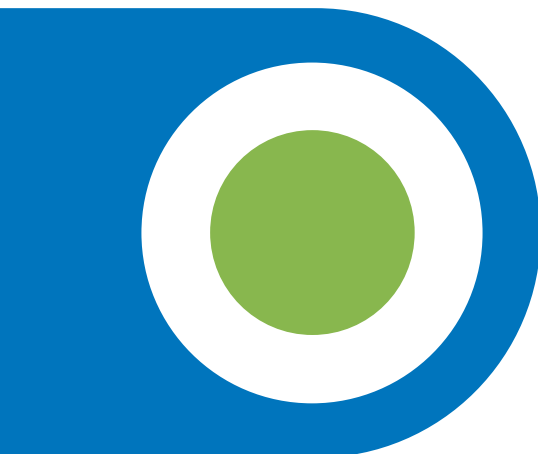


Some physical illnesses may be directly cause a change in mental state or behaviour

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- 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

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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

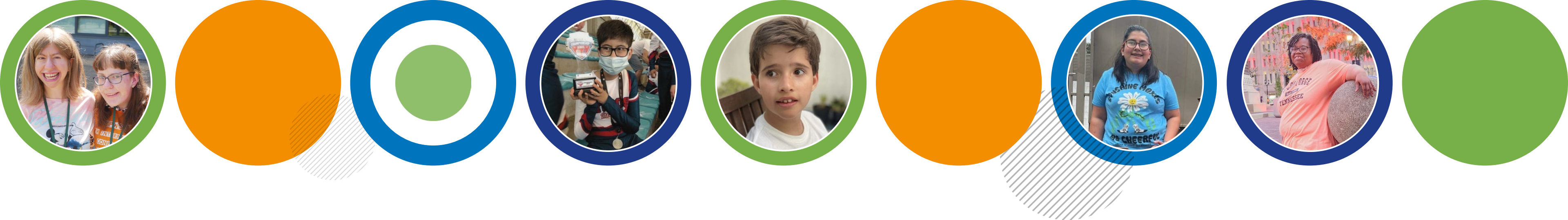
- Vomiting
- Complaints of pain
- Loss of appetite and not eating
- Confusion, not knowing time or place
- Excessive and unusual levels of drowsiness
- 'Just doesn't look well and not themselves'

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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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