

Behavioural challenges in older children and adults with PWS

FAMCARE Information Session
Managing Challenging Behaviour
Saturday 22nd April 2023

Aims

- Provide an understanding for why specific types of challenging behaviours are associated with PWS
- Consider the implications for the assessment, prevention and management of such behaviours
- Reflect on why transition and adult life can be particularly challenging

Some general principles

- The importance of knowledge and understanding
 - Prader-Willi Syndrome (PWS)
 - Your child with PWS (strengths and weaknesses etc)
 - The behaviours of concern and their circumstances
- Behaviours arise and repeat over time
 - Biological vulnerability (atypical brain development)
 - Environmental circumstances and family/support context
 - Shaping and maintaining of behaviours

PWS specific factors that impact on mental health?

Atypical brain development
Genetic sub-type differences
Cortical & subcortical structures
Hypothalamic neural and neuroendocrine pathways

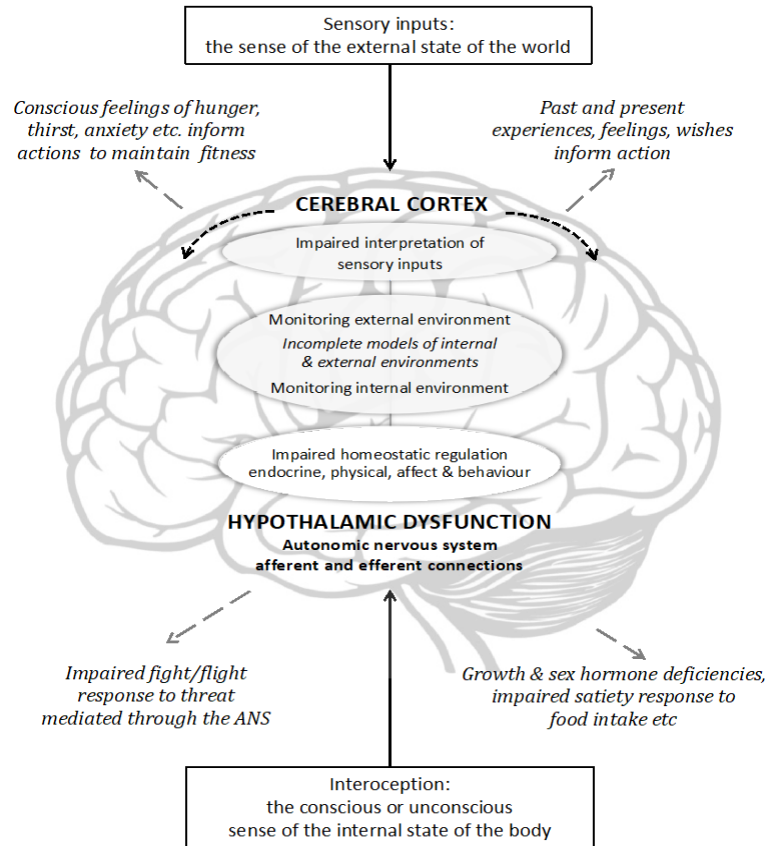
Dimensions of mental ill-health associated with PWS

Cognition, Communication & Social Functioning
Impairments in intellectual & executive functioning, social cognition & communication
Cognitive inflexibility/rigidity
Intellectual Developmental Disabilities (IDD)
Autistic Spectrum Conditions (ASC)

Behaviours of concern
Hyperphagia
Emotional (temper) outbursts
Body-focused self-harming behaviours
(e.g. skin picking etc.)
Repetitive & ritualistic behaviours
Rigidity and perseveration

Abnormalities of mental state
Mood instability
Situational anxiety
Affective disorder
Psychotic illness (mUPD>deletion)

ATYPICAL BRAIN DEVELOPMENT
& THE PWS NEUROPSYCHIATRIC PHENOTYPE
Maintaining homeostasis in a changing environment



BIOLOGICAL VULNERABILITIES - CONSEQUENCES

Impaired perception of internal mental states

- Hunger, fullness (satiety and reward systems)
- Pain

Impaired perception of the external world

- Temperature
- Threats
- Emotions of others

Impaired cognition

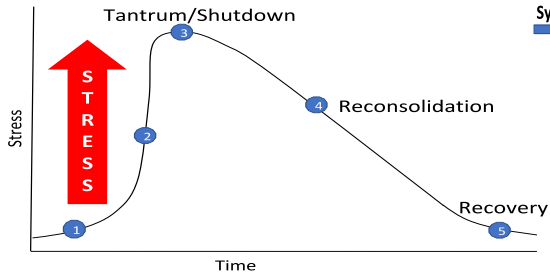
- Social cognition
- Higher order cognitive abilities
- Cognitive flexibility

Consequences of the above

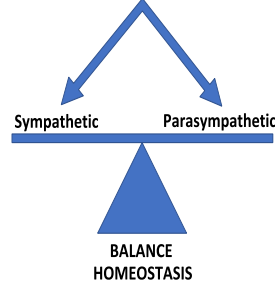
- Impaired homeostatic regulation of energy balance, mood and emotional responses, temperature
- Uncertainty
- Anxiousness
- Heightened arousal

How is this knowledge applied to an understanding of emotional outbursts?

Anatomy of a Tantrum



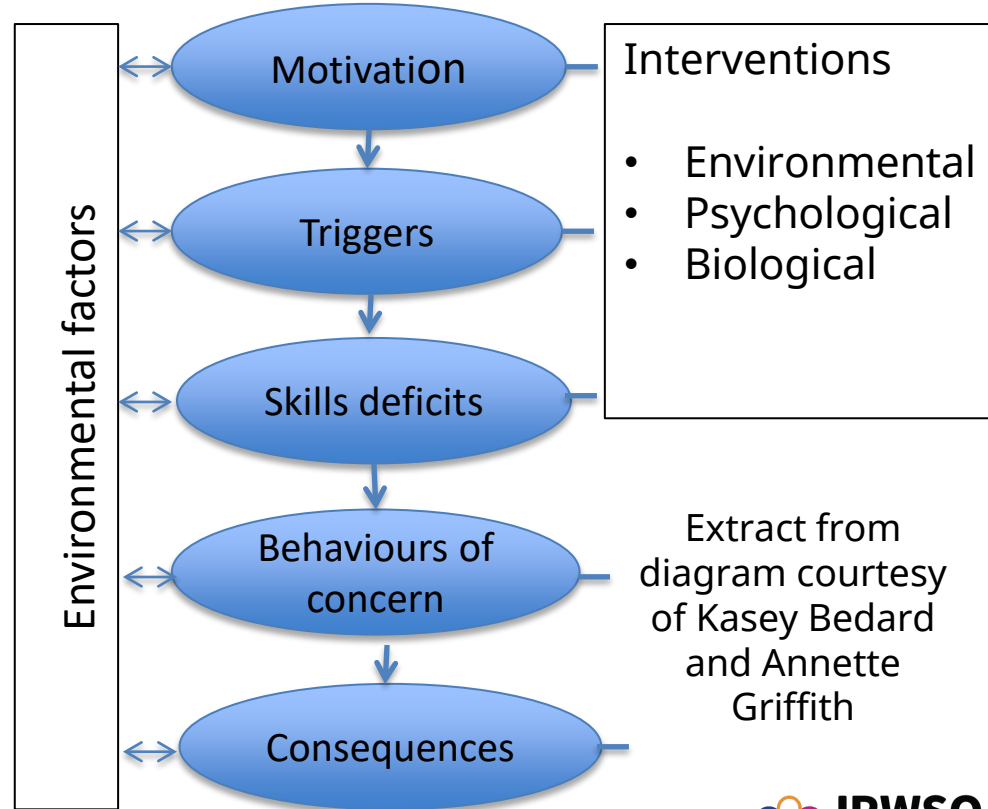
The Autonomic Nervous System (ANS)



Diagrams courtesy of Janice Forster

- Impaired perception and cognitive functioning
- Environmental change
- Increased cognitive load

Development and maintenance of behaviours



Questions to ask

- What is the nature of the problem?
 - Outbursts
 - Mood
 - Skin picking
 - All of the above
- Is this new or not, has it got worse?
- Is there a pattern to its occurrence?
- Is it what might be expected in a person with PWS or is it different?

What information to collect?

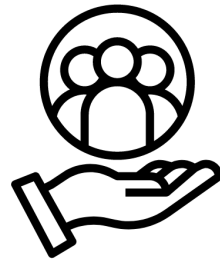
- History of the 'behaviour'
 - Change
 - Onset
 - Life event
- Record keeping (ABC charts)
 - Pattern
 - Triggers
- Physical and mental health
 - Any indication of physical illness (pa
 - What about mental health – mood, v



Interventions



With the individual



In your support



In the environment

Prevention and management of outbursts

- Optimise sensory inputs and reduce uncertainty (anxiousness)
 - food security,
 - environmental support,
 - visual timetables, calendars etc.
 - allow time for cognitive processing
 - use of language, written/visual material
- Improve emotional regulation thereby improving perception and reasoning
 - Training for people with PWS to manage change etc.
 - Consistent and agreed approach to potential triggers and for when an outbursts is beginning
 - Vagus nerve stimulation

Skin picking

Best understood as an interaction between a biological vulnerability, physical and psychological factors and environmental circumstances

- Functional analysis - behavioural interventions
- Treatment of co-morbidity (e.g. mood disorder)
- Environmental changes
- Topical treatments to skin
- Medication to modify glutaminergic pathways in the brain??

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Developing an understanding of skin picking in people with Prader-Willi syndrome: A structured literature review and re-analysis of existing data

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- What developmental, biological, psychological, social factors might be:
 - Predisposing to
 - Precipitating
 - or maintaining these behaviours
- Diverse examples:
 - Change in level of food security or its inconsistent application
 - Changing circumstances, new staff, inadvertent changes in support strategies
 - Onset of mental ill-health (mood disorder, psychotic illness)
 - Change in routine such as at Christmas or holiday time

Formulation

Transition to adult life

Maintaining homeostasis in a changing world
In adult life increasing freedoms, choices, and risks
The right to make decisions for yourself



C Poitou *et al.*

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REVIEW

The transition from pediatric to adult care in individuals with Prader-Willi syndrome

Christine Poitou¹, Anthony Holland², Charlotte Höybye³, Laura C G de Graaff⁴, Sandrine Bottius¹, Berit Otterlei³ and Maïthé Tauber⁵

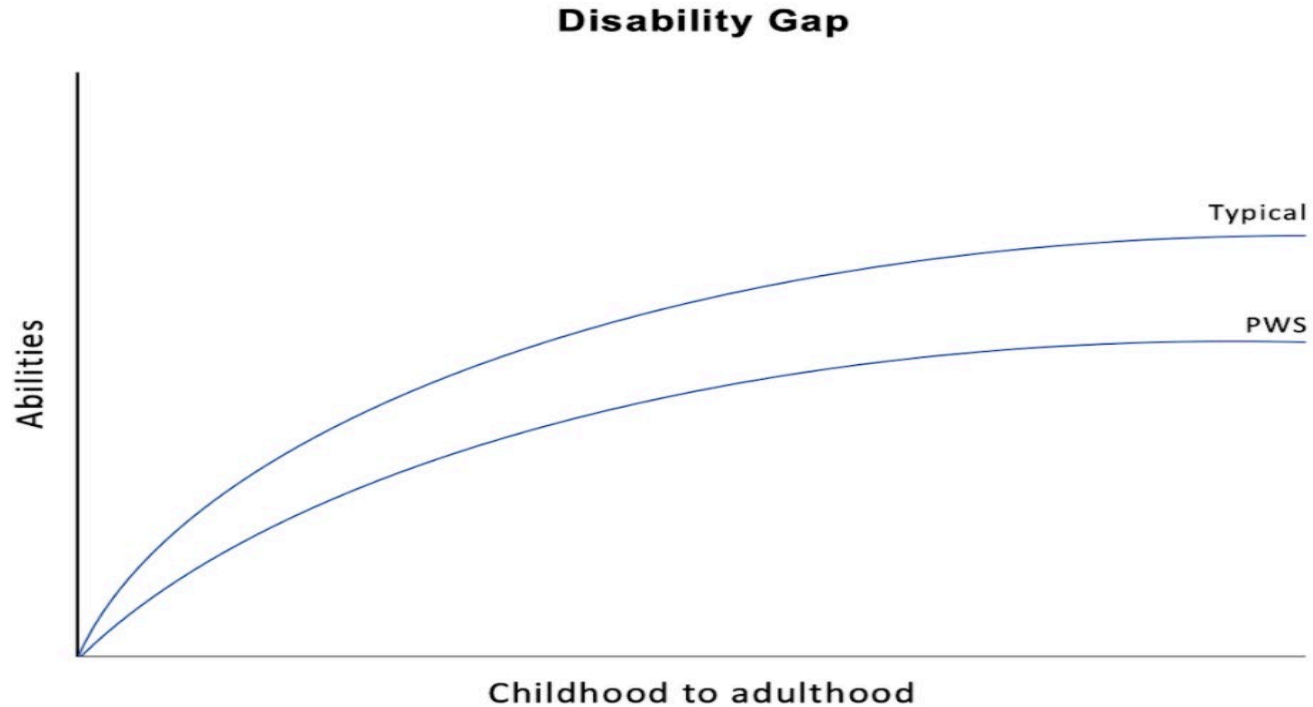
Individual characteristics of people with PWS that may impact transition

- Cognition (higher cognitive functioning, including social cognition)
- Hyperphagia
- Emotional regulation and risk of outbursts
- Risk of mental ill-health

Changes in the environment that may impact transition to adult life

- Structured to unstructured
- Access to food
- Certainty (familiarity) to uncertainty
- Increasing choice and independence
- Level of support

Disability gap



Challenges and rewards

- Engaging with the person with PWS
- Developing a formulation
- Implementing and maintaining the interventions

- Reduce frequency and severity of behaviours
- Positive impact on family life and quality of life
- Improve wellbeing and quality of life for person with PWS