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

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

Dimensions of mental ill-health associated with PWS
Abnormalities of mental state
Mood instability
Situational anxiety
Affective disorder
Psychotic illness (mUPD>deletion)

PWS specific factors that impact on mental health?
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?

Diagrams courtesy of Janice Forster

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

Development and maintenance of behaviours

Interventions
- Environmental
- Psychological
- Biological

Environmental factors

Motivation

Triggers

Skills deficits

Behaviours of concern

Consequences

Extract from diagram courtesy of Kasey Bedard and Annette Griffith

The Autonomic Nervous System (ANS)

Sympathetic

Parasympathetic

BALANCE HOMEOSTASIS

Anatomy of a Tantrum

Stress

Time

Tantrum/Shutdown

Reconsolidation

Recovery

International Prader-Willi Syndrome Organisation
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 (pain, vomiting)
  - What about mental health – mood, voices etc.
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 outburst 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
• 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
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

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