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NATIONAL CONVENTION

June 21 – 24, 2023 • Orlando, FL

Improving mental health and wellbeing for people with Prader-Willi Syndrome

Tony Holland, Department of Psychiatry, University of Cambridge, UK
President, International Prader-Willi Syndrome Organisation

PWSA USA Family Conference 23rd to 24th June 2023

Declarations: Advisor to pharmaceutical companies on clinical trials



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World Health Organisation (WHO) definition of mental health

(Galdeseri et 2015, Towards a definition of mental health. World Psychiatry, DOI:10.1002/wps.20231)

Good mental health is **more than** 'the absence of mental disorder'. It includes the presence of:

- A dynamic internal state or equilibrium
- The ability to maintain harmony in line with universal values
- The ability to recognise, express and modulate emotions
- The ability to cope with adverse events and function socially

This definition is analogous to that for physical health

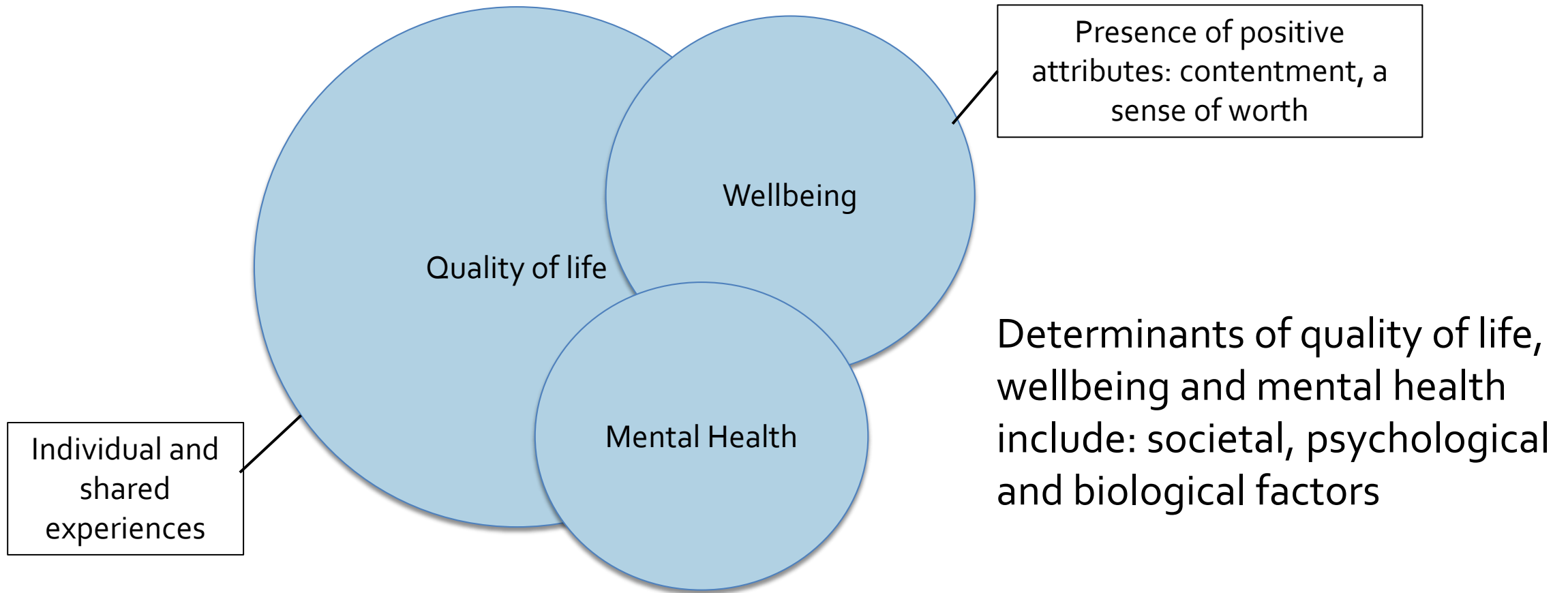


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Quality of life, wellbeing and mental health



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Improving mental health, wellbeing, quality of life

Impact of the individual, support network and society

The person with PWS

- Strengths and weaknesses
- Extent and nature of behaviours of concern
- Physical and mental health
- Their understanding & acceptance of PWS
- Access to professional expertise and treatments

Family and close support network

- Early diagnosis
- Access to information on PWS
- Family, friends and professional support
- Financial resources
- Housing

Society

- Opportunities (education, employment, leisure)
- Social inclusion
- Financial support (benefit systems)
- Civil society (PWSA, Social networks etc)
- Policies and practices that are inclusive
- Public acceptance and understanding



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Good mental health

For people with PWS having and maintaining good mental health, and a sense of positive wellbeing and quality of life are not just issues for health services.

They require a range of actions and activities including from wider society and support services, access to funding and to informed health support from the relevant disciplines, and the availability of guidance and advice as and when it is needed



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Legislation matters: changes in England and Wales

How Government policies determine attitudes and the agenda

The Mental Deficiency Act 1913

Better Services for the Mentally Handicapped, 1971

Valuing People: a new strategy for Learning Disability for the 21st Century, 2001

UN Convention on the Rights of Persons with Disabilities, (UNCRPD) 2006

Education Acts, 1971, 1981

NHS and Community Care Act 1995

Equality Act 2010

Making 'reasonable adjustments' for people with disabilities. Adapting work places, provision of aids etc



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Interrelationships between those supporting and those being supported



ORIGINAL ARTICLE

Parenting stress in families of children with Prader-Willi syndrome

Shi-Bing Wong, Tzong-Shi Wang, Wen-Hsin Tsai, I-Sheng Tzeng, Li-Ping Tsai ✉

First published: 12 October 2020 | <https://doi.org/10.1002/ajmg.a.61915> | Citations: 7

> *J Autism Dev Disord.* 1997 Feb;27(1):11-24. doi: 10.1023/a:1025865004299.

Families of children with Prader-Willi syndrome: stress-support and relations to child characteristics

R M Hodapp ¹, E M Dykens, L L Masino

Affiliations + expand

PMID: 9018579 DOI: [10.1023/a:1025865004299](https://doi.org/10.1023/a:1025865004299)

- Families of children with PWS experience higher stress levels
- Families turn to family and friends for support
- Behaviour problems associated with high parental stress

Behaviours of concern and mental ill-health arise in the context of an inter-relationship between biological vulnerability and environmental factors - helping families maintain their resilience will help the mental health of those they support

Stress and Coping in Parents of Children with Prader-Willi Syndrome: Assessment of the Impact of a Structured Plan of Care

March 2015 · *American Journal of Medical Genetics Part A* 167(5)

DOI: [10.1002/ajmg.a.36971](https://doi.org/10.1002/ajmg.a.36971)

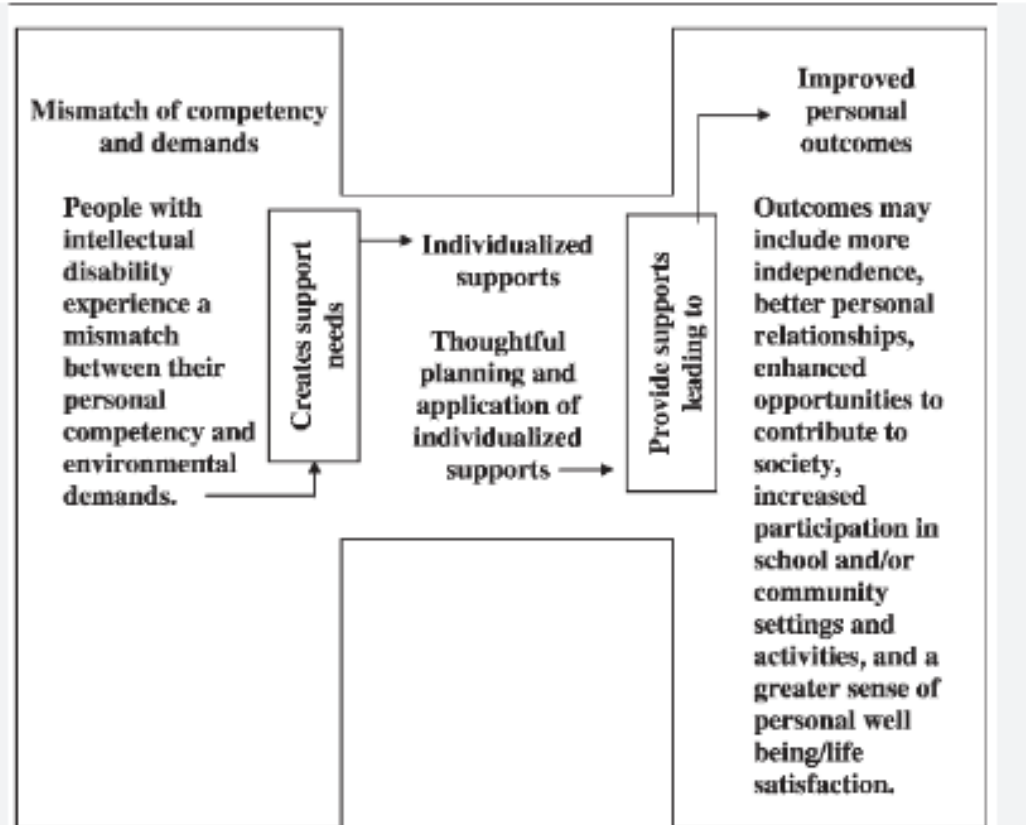
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👤 Tatiana Tvrdik · Debbie Mason · Karin M Dent · [Show all 7 authors](#) · David A Stevenson



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Supporting families and paid care staff to meet the mental health and behaviour needs of people with PWS



AAIDD support model

What helps?

- Being prepared for different phases (diagnosis, onset of hyperphagia, the nature of PWS, planning for the future)
- Access to informal and professional support
- Structured care plans
- Strategies to prevent and manage behaviours of concern, including crisis planning

> [PLoS One](https://doi.org/10.1371/journal.pone.0273295). 2022 Sep 1;17(9):e0273295. doi: 10.1371/journal.pone.0273295. eCollection 2022.

Experiencing illness as a crisis by the caregivers of individuals with Prader-Willi Syndrome

Katarzyna Kowal ¹, Michał Skrzypek ², Janusz Kocki ³

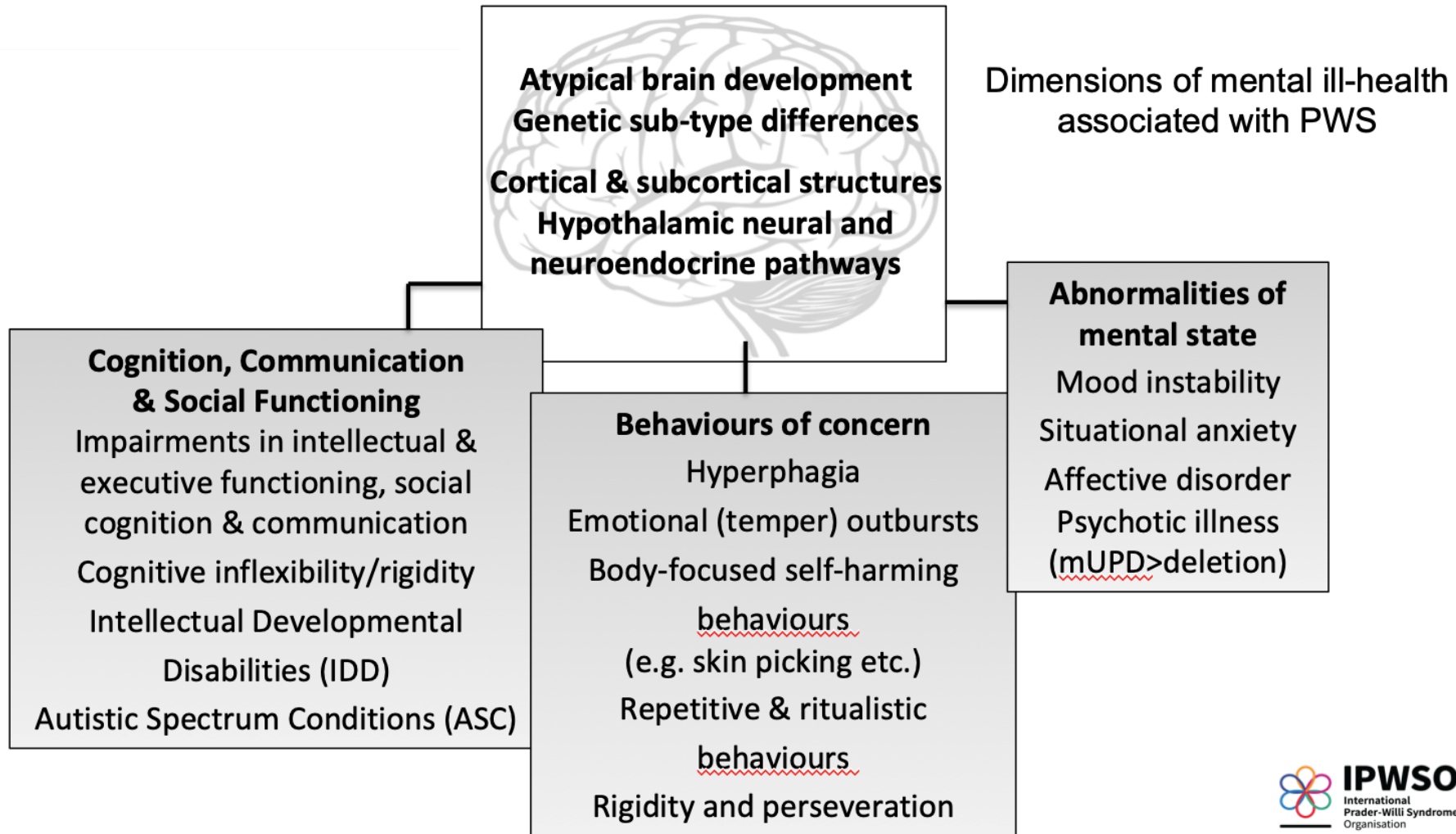
Affiliations + expand

PMID: 36048794 PMCID: PMC9436047 DOI: 10.1371/journal.pone.0273295



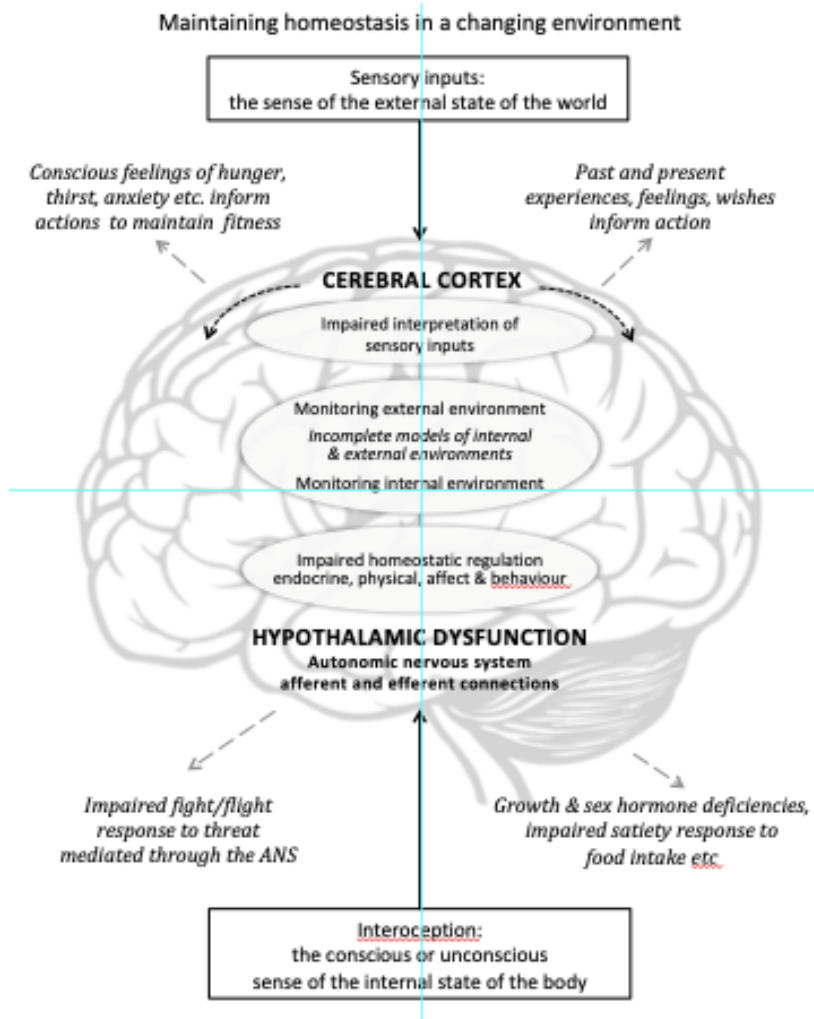
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The individual with PWS: behaviour and mental health



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Maintaining homeostasis in a changing environment



Mental ill-health, including behaviours of concern, arise as a consequence of:

a) Dysfunction of the hypothalamus and its networks

- Hyperphagia
- Mood regulation
- Emotional control

b) impaired brain development affecting cortical and sub-cortical regions and networks:

- impaired cognition (incl. social)
- reduced ability to understand and to effectively respond to change

Interaction between biological vulnerability and environmental



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Mental health at time of transition to adult life



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**Maintaining
'homeostasis' in a
changing world**

**In adult life
increasing freedoms,
choices, and risks**

**Importance of
acceptance, support
and opportunity**

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



C Poitou et al.

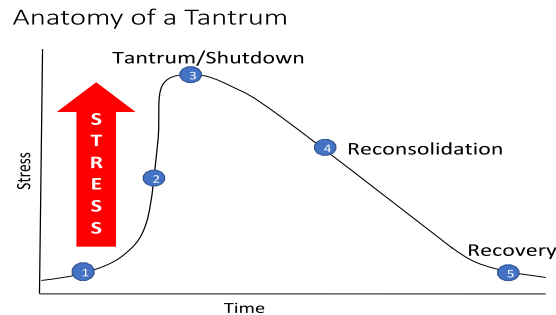
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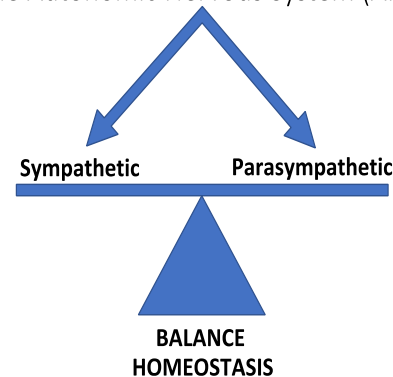
**The transition from pediatric to adult care in
individuals with Prader-Willi syndrome**

Christine Poitou¹, Anthony Holland², Charlotte Höybye³, Laura C G de Graaff⁴, Sandrine Bottius¹, Berit Otterlei⁵
and Maïthé Tauber⁶

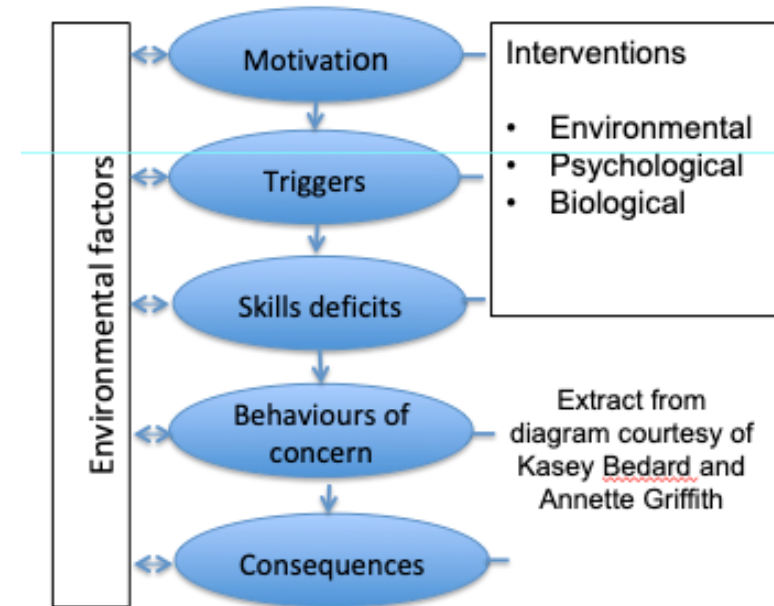
Preventing and managing emotional outbursts



The Autonomic Nervous System (ANS)



Development and maintenance of behaviours



Diagrams courtesy of Janice Forster

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

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 agreed approach to potential triggers and when an outbursts is beginning
- ?vagus nerve stimulation
- ?medication



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Mental illness in people with PWS



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- Risk of mental illness developing is increased in teens in people with PWS due to mUPD
- Mental illness can be treated and is an indication for the careful prescribing of psychiatric medication
- Where changes in behaviour and mental state are out of the ordinary and bizarre – asked why – importance of an accurate diagnosis and formulation

Presents with a deterioration in behaviour and/or the onset of new bizarre behaviour

Onset usually acute but can also be gradual

Associated with abnormal mood state and the development of abnormal mental experiences (confusion, hallucinations, delusions)

Interventions

Medication based on accurate diagnosis

Reduce demands

Consistent informed support

Prevention of harm

Maintaining good mental health, wellbeing and quality of life for the person with PWS



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Society

- Policies, practices and attitudes that are understanding and facilitate social inclusion
- Availability of services to meet needs

The person with PWS

- Early intervention
- Informed support
- Skills development
- Access to informed expertise and interventions

Family and professional support

Information and understanding
Network of support
Finance and housing



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 Prader-Willi Syndrome Association | USA

THANK YOU



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