These Recommendations are based on SLHD: Royal Prince Alfred Hospital Policy for People Living with PWS

Prader-Willi Syndrome Admission	
TRIM Document No	
Policy Reference	RPAH_PD2020_007
Related MOH/SLHD Policy	N/A
Keywords	Behavioural outbursts; food seeking behaviour
Applies to	Patient Admissions to ward of Emergency
Clinical Stream(s)	All Clinical Streams
Date approved GM, RPA	*29/06/2020*
Date approved by RPA Policy Committee	*16/04/2020*
Author	PWS Clinic Staff, Metabolism & Obesity Service
Status	Active
Date	30/06/2020

Prader-Willi Syndrome Admission

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SLHD - RPA Prader-Willi Syndrome Admission Policy

1. Introduction

Prader-Willi Syndrome (PWS) is an uncommon disorder with specific needs which are often misunderstood. RPAH has the only clinic for adolescents and adults with Prader-Willi Syndrome in NSW. This policy will outline the measures that should be taken to ensure appropriate care for these patients when they are admitted to RPAH.

2. The Aims / Expected Outcome of this Policy

- To provide Emergency and Ward staff with information on the specific needs of patients with Prader-Willi Syndrome, to ensure they receive appropriate supervision and management while at RPAH.

3. Risk Statement

SLHD Enterprise Risk Management System (ERMS) Risk # 105 Minimise adverse events

1) Patients with PWS are at risk of misdiagnosis, adverse medication reactions, behavioural outbursts, absconding, weight gain
2) Hospital staff are at risk of verbal and physical aggression, damage to equipment, incorrect patient history, food & monetary theft, incorrect patient follow-up

4. Policy Statement

Prader-Willi syndrome (PWS) is a rare complex, multi-system genetic disorder requiring high care support that extends beyond usual medical and social standards. Lack of understanding of this syndrome can contribute to patient mortality and death.

5. Key Performance Indicators and Service Measures

- 1) Individual Indicators - weight changes, diabetes management and behaviour: such as food seeking, temper tantrums, stealing and absconding while in RPAH,
- 2) Clinical Indicators - length of admission, staff support strategies based on understanding of PWS.

6. Guidelines

6.1 Admission via Emergency Department

Prader-Willi Syndrome (PWS) is an uncommon disorder with a prevalence of one in approximately 15,000 live births. PWS is the most common genetic cause of obesity, resulting from a malfunctioning of the hypothalamus. Most people with PWS have a mild or moderate intellectual disability with specific cognitive problems; hypotonia and reduced muscle; short stature (in the absence of growth hormone replacement treatment); low growth hormone and low gonadotrophin levels; a high pain threshold; thermodysregulation, lack of satiety and severe obesity, when poorly managed. They are always hungry, suffer from hyperphagia and readily gain weight. They often display behavioural problems that include food seeking and emotional outbursts.

People with PWS demonstrate a cluster of specific medical, physical and behavioural features which require informed care and increased supervision. They may present with

high fevers with no obvious infection, or conversely, sepsis in the absence of fever. Hypothermia and hyperthermia can occur in people with PWS. They may react to certain medications and may require lower than prescribed doses of recommended medications. Constipation and reduced rate of gastric emptying are common in people with PWS. They can be at risk of gastric necrosis and gastric rupture after ingesting a large amount of food, especially if their energy intake is usually restricted to meet their needs.

People with PWS may demonstrate anti-social behaviour if not managed appropriately. This behaviour is generally a result of their syndrome but is often misdiagnosed as psychotic behaviour. They do, however, suffer from heightened anxiety and are at risk of psychosis. The behaviour of people with PWS can impact negatively on staff, other patients and their own health. Patients with PWS have a constant desire to eat and may consume food from sources that are unanticipated, such as food scraps from waste bins. Therefore constant supervision is required to prevent extra food consumption and weight gain while hospitalised.

On presentation to the Emergency Department or Admissions:

- Please contact Endocrinologist on call.
- Please access the Medical Alerts (Australia) attached to this policy Prader-Willi Syndrome Australia | MedAlert (pws.org.au)
- Ensure that a high level of supervision is provided
    - People with PWS need to be given a higher level of supervision due to behavioural issues. They should receive 1 on 1 nursing if possible, particularly if their behaviour is uncontrolled, they are ambulant, or are self-harming.

    - All patients for admission are to be designated to a single room located near the nurses' station, unless they require the highly specialised nursing care of another unit, eg. ICU for non-invasive ventilation or a cardiac ward for cardiac monitoring. If possible the same allocated ward should be used for all patients with PWS, so ward staff can receive information about PWS and PWS management training to improve their knowledge and understanding of this complex syndrome. A PWS patient should be started on a low energy PLUS 20gm fat diet with the following amendments: - NO juice; NO bread and jam with lunch and/or dinner. Diet cordial, tea and coffee (with sweetener NOT sugar) are to be chosen off the trolley (NO biscuits, cheese or crackers should be given or offered). Dessert should only include diet jelly. People with PWS require 7-8 kcal/cm height per day, for weight loss and 9-11 kcal/cm height per day, for weight maintenance.

    - PWS patients may take either food or money from any available source due to their uncontrollable hyperphagia and cognitive impairment. This is syndromal behaviour not a reflection of the PWS patient's character. The staff tea-room is to remain locked, when not in use, and off limits to the patient with PWS, when open for staff use.

    - The ward Dietitian should be contacted and asked to consult with the PWS Clinic regarding any changes that may be required for the patient. The patient is not to be given standard menus to choose from.

- For stays longer than 24 hours, contact the occupational therapist for consultation with the patient to provide activity options. People with PWS need to be occupied to reduce their food focus, anxiety and boredom.

- A responsible carer/parent is required to be present:
  - To obtain an accurate patient medical history
  - For the explanation of any new care plans and scheduling of future appointments for the PWS patient
  - If guardianship is in place to consent for any necessary procedures

6.2 Managing PWS Patients

- People with PWS have a high pain threshold but often exhibit a child-like fear of pain – such as for injections. Anaesthetic cream is recommended.
- Patients receiving treatment for constipation should be closely monitored for abdominal distention, faecal retention, vomiting and decreased appetite. Imaging and GI consultation may be indicated to avoid colonic necrosis, perforation and resultant sepsis.
- Sequential processing deficits preclude step-wise problem solving in people with PWS. To avoid behaviour problems all care providers are to need to slowly explain what is required of the patient, allowing time for the patient to comprehend what is being done to, or asked of, them before proceeding.
- As instructions/ procedures/ processes differ (even if slightly) between care providers, all care processes should be explained by each care provider. The patient with PWS will expect processes to remain exactly the same between care providers and behaviour problems may develop if care is even minimally varied.
- Uncooperative behaviour may escalate into major temper tantrums if they are not managed appropriately. This can be distressing for all involved, and may take some time to diffuse.
- Patients with PWS respond well to quiet, calm, confident speech that endears their cooperation rather than objects to their apprehension or oppositional attitude.
- Patients with PWS respond well to limited, controlled choices as well as distraction and humour.
- All patients should be told that they will continue to be followed up by the staff at the PWS Clinic for ongoing care once they are discharged from hospital. A follow up appointment at the PWS Clinic should be made for the patient before discharge.

7. Definitions

8. Links and tools

Medical Alerts Booklet for PWS Australia - Prader-Willi Syndrome Australia | MedAlert (pws.org.au)

9. References

- Adult Medical Care for Prader-Willi Syndrome An Overview of Medical Problems for Physicians Approved by the Clinical and Scientific Advisory Board of the International Prader-Willi Syndrome Organisation (IPWSO) January 2018

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9.1 National Safety and Quality Health Service (NSQHS) Standards, 2nd Edition


Clinical Governance Standard


Partnering with Consumers Standard


Comprehensive Care Standard


Communicating for Safety Standard