Shropshire Community Health NHS
NHS Trust

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2	V2 July 2017	Discussed at peer meeting and alterations made and accepted Birmingham Women's Genetic department permission to use PIL SATH informed re updated guidelines and increased risk of VTE SSSFT CAMHS lead informed re updated guideline
3	V3 Nov 2020	General review and discussed with community paediatrics team
4	V4 Jan 2025	General review and discussed with community paediatrics team
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Prader Willi Syndrome:
Management and Health Surveillance Guidelines

1. Introduction

Prader Willi Syndrome (PWS) is a rare inherited genetic condition that has a recognisable set of physical findings and can lead to cognitive, neurological, endocrine and behavioural problems. This can lead to complex management issues for patients, carers, families and medical professionals.

2. Purpose

The purpose of this guideline is to aid medical professionals working in Shropshire Community Health NHS Trust to be able to identify patients with PWS and investigate, diagnose and treat this condition effectively within a multi-professional environment.

3: Definitions.

Meiotic non disjunction: The failure of homologous chromosomes to separate equally during meiosis cell division resulting in daughter cells with improper chromosome content
Imprinting: Phenomenon where certain genes are expressed depending on being either maternally or paternally derived.
Uniparental Disomy: Usually 1 copy of a gene is inherited from each parent. In uniparental disomy 2 copies of a gene are inherited from one parent and none from the other. This is usually due to meiotic non disjunction. In PWS both genes on chromosome 15 will be inherited from the mother with no copy from the father.
Hyperphagia: excessive hunger or appetite.

4. Duties

4.1 Chief Executive

The Chief Executive has ultimate accountability for the strategic and operational management of the Trust, including ensuring there are effective and appropriate processes in place for the medical management and health surveillance of children with Prada Willi Syndrome.

4.2 Director of Nursing & Medical Director

The Director of Nursing & Medical Director have responsibility for ensuring that children with PWS are offered appropriate medical management and health surveillance checks and to support patient safety at all times.

4.3 Service Managers

Service Managers are responsible for the day-to-day operational management and coordination of the medical management and health surveillance of children with PWS in accordance with clinical guidelines and to be aware of MDT responsibilities in managing these patients.

4.4 All Clinical Staff

Clinical staff is key essential members of the MDT in ensuring that children with PWS are managed appropriately as per national/ local guidelines and are offered regular health surveillance checks. All clinical staff are required to comply with this guideline

and to report any adverse care related issues to their line manager and to complete a Datix incident report in line with Trust Incident reporting policy.

5. Prader Willi Syndrome: Management and Health Surveillance Guidelines

5.1 Genetics

PWS arises from absent expression of the PWS 'critical region' on chromosome 15q11.2-q13, typically due to deletions from the paternal chromosome, maternal disomy, or an imprinting defect.
Approximately 70% of cases are caused by deletions of genes on chromosome 15q11-q13. 28% of cases are due to maternal uniparental disomy caused by meiotic non-disjunction.
Less than 1% of patients with PWS have deletions of this chromosome related to the imprinting centre, which carries a risk of recurrence.
Genotype-phenotype correlations show that individuals with deletions often have higher BMI and scoliosis, while those with uniparental disomy may have milder physical features but a higher likelihood of autism or psychosis.

5.2 Epidemiology

Prader-Willi Syndrome (PWS) affects 350,000–400,000 individuals worldwide. Prevalence estimates vary, ranging from 1:8,000 in rural Sweden to 1:76,500 in Flanders. In the UK, the estimated prevalence is approximately 1:45,000.

5.3 Clinical manifestations

PWS has a great range of signs and symptoms, and there are varied causes of morbidity and mortality throughout the natural history of the condition. Infants typically present with hypotonia, poor sucking, and feeding difficulties, often leading to growth hormone deficiency and failure to thrive. Motor milestones are delayed, with an average age of 27 months for walking and 39 months for speaking. Key behavioural features include temper tantrums, stubbornness, obsessive-compulsive behaviours, and, in some cases, features resembling autism. As the patient ages, a second phase of symptoms ensue, characterised by hyperphagia, obesity and behavioural problems.
Without adequate control of this hyperphagia and behavioural problems, the child will develop long-term sequelae such as hypertension, diabetes mellitus, obstructive sleep apnoea and right-sided heart failure.

A more comprehensive list of clinical features is listed here :

Neonatal

- Breech position
- Reduced foetal activity
- Polyhydramnios

Growth

- Short stature
- Failure to thrive in infancy
- Central obesity

Head and neck

- Narrow bitemporal diameter
- Almond-shaped eyes
- Strabismus
- Up-slanting palpebral fissures
- Myopia
- Hyperopia
- Thin upper lip

- Small-appearing mouth
- Down-turned corners of mouth
- Thick, viscous (reduced) saliva
- Enamel hypoplasia
- Early dental caries
- Dental crowding and malocclusion

Ocular

- Strabismus
- Nystagmus
- Cataracts (rare)
- Retinal hypopigmentation
- Foveal hypoplasia
- Hyperopia
- Myopia

Respiratory

- Hypoventilation
- Obstructive sleep apnoea
- Central sleep apnoea

Gastrointestinal

- Feeding problems in infancy
- Gastroesophageal reflux
- Increased vomiting

Genitourinary

- Small penis
- Scrotal hypoplasia
- Cryptorchidism
- Hypoplastic labia minora
- Hypoplastic clitoris

Skeletal

- Osteoporosis
- Osteopenia
- Scoliosis
- Kyphosis
- Small hands and feet
- Narrow hands with straight ulnar border
- Clinodactyly

Skin, nails, hair

- Hypopigmentation
- Blonde to light-brown hair
- Frontal hair upsweep
- Skin picking

Neurologic/ developmental

- Severe neonatal hypotonia that improves with age
- Poor neonatal suck and swallow reflexes
- Poor gross motor coordination
- Poor fine motor coordination
- Mild-to-moderate Learning difficulties.
- Increased risk of seizures
- Global developmental delay
- Speech-articulation problems
- Hyperphagia

Sleep

- Snoring/obstructive sleep apnoea
- Central apnoea during sleep
- Excessive daytime sleepiness
- Early-morning waking
- Night-awakening for food-seeking

Voice

- Hypernasal speech
- Weak or squeaky cry in infancy

Endocrine

- Hyperinsulinemia
- GH deficiency
- Hypogonadotropic hypogonadism
- Diabetes mellitus (type 2)

Behaviour/mental health

- Temper tantrums
- Difficulty with transitions
- Stubbornness
- Obsessive behaviours
- Perseverant speech
- Obsessive-compulsive disorder
- Psychosis
- Elopement

Miscellaneous

- Temperature instability
- High pain threshold
- Unusual skill with jigsaw puzzles

Also refer to appendix 1: unique features

5.4 Clinical Diagnosis

The diagnosis of a child with possible PWS is usually apparent from the clinical phenotype however genetic testing is becoming more freely available and is considered the gold standard of diagnosis. There are widely agreed situations when genetic testing of children for PWS is deemed appropriate. This is outlined in the following table:

Age at assessment	Features sufficient to prompt DNA testing
Neonatal period	• Neonatal hypotonia • Poor feeding
2–6 yr	• Global developmental delay • Short stature and/or growth failure associated with accelerated weight gain
6–12 yr	(hypotonia often persists) • Global developmental delay • Excessive eating (hyperphagia, obsession with food) with central obesity if uncontrolled
13 yr through adulthood	• Cognitive impairment, usually mild • Learning difficulties • Hypothalamic hypogonadism • typical behaviour problems (Including temper tantrums and Obsessive-compulsive features)

5.5 Management

Management of children with PWS is complex and involves a multi-disciplinary approach summarised in Appendix 4. There are different things to focus on depending on the age of the patient.

5.5.1 Neonatal period

- Most neonates with PWS are susceptible to faltering growth.
- The main problem in the neonatal period is hypotonia and poor suck leading. to feeding difficulties and failure to thrive with requirement of tube feeding.
- Dietetic input is mandatory.

5.5.2 Cryptorchidism

- This occurs in over 80% of boys with PWS.
- As with children with cryptochordism without PWS, orchidopexy should be performed by year 2 of life, to avoid the small chance of testicular cancer if orchidopexy was not performed. It is also a much more technically demanding procedure after the onset of scrotal hypoplasia and obesity.

5.5.3 Hypotonia

- Children with PWS syndrome display muscle hypotonia, decreased muscle mass, general developmental delay, and a reduced number of activity-dependent type 2 muscle fibres.
- Early involvement in physiotherapy is required, although there is no definitive evidence that physiotherapy improves gross motor delay or time to sit or walk.
- Continuing exercise is necessary to combat the ongoing reduction in muscle mass as hypotonia persists into adulthood.

5.5.6 Growth and Growth hormone treatment

Paediatric endocrinologist involvement is essential from as early as 3 months of age because the early start of growth hormone therapy (even < 6 months old) can significantly increase lean body mass (earlier walking) and, therefore, improve body composition, reducing the risk of obesity, metabolic syndrome and type 2 diabetes.

- Pre and postnatal Growth restriction
- Short stature due to growth hormone deficiency and lack of pubertal growth spurt.
- There is no evidence that GH status is directly linked to height velocity; therefore, GH testing before starting treatment is not essential but can be helpful.

The aims of GH treatment in children with PWS are:

- to improve childhood growth velocity
- to increase final adult height
- to improve body composition.

Early initiation of growth hormone treatment, ideally before 12 months of age, can significantly improve body composition, enhance motor development (such as earlier walking), and improve bone density. Regular monitoring of thyroid function and growth velocity is required. Growth hormone therapy should be carefully managed, especially in patients with sleep-disordered breathing.

Most studies evaluating the use of GH test the dose of 0.6-1mg/m2/day and current evidence shows this significantly improves these parameters in the first year following starting treatment.

There are positive effects on body composition and bone mineral density once maximal height velocity has been achieved.

Hypothyroidism is common in children with PWS, and measurement of TSH, free T3, and free T4 should be done pre and post-commencement of GH therapy.

Monitoring of children in preparation for, and whilst on GH therapy should consist of the following:

Before starting GH treatment in improving the developmental milestones:

1) Genetic confirmation of PWS
2) Evaluation of IGF-I status and, if possible, GH status
3) Nutritional evaluation
4) Prior control of food environment is vital, especially in obese children
5) Sleep Study and ENT evaluation
6) OGTT (in older children)
7) Family instruction on GH treatment
8) Scoliosis evaluation consider x-ray
9) Evaluation of hypothyroidism (TSH, free T4, free T3)
10) Bone age study (not in very small infants)
11) Weight, height, height velocity and BMI evaluation

On GH treatment:

1) Regular clinical assessment of height, weight, BMI, body composition
2) ENT assessment and polysomnography within the first 6 months
3) If development or worsening of sleep-disordered breathing, snoring, or enlargement of tonsils and adenoids, ENT assessment +/- polysomnography
4) Orthopaedic assessment for scoliosis
5) Regular bone age determination
6) Monitoring for Thyroid function and IGF.

Cessation of GH treatment should be considered if:

1) Uncontrolled progression of obesity
2) Continued worsening of glycaemic control
3) Continued worsening of sleep-disordered breathing
4) Attainment of final height (but because there are potential benefits in adults on
5) body composition, peak bone mass, cognition, and quality of life, reassessment of persistent GH deficiency, and replacement with adult doses may be warranted)

5.5.7 Management of puberty

Paediatric endocrinologist role is essential as there is no universally agreed treatment regimen for induction of puberty in children with PWS.

- Hypogonadism (central and peripheral) is almost a universal problem in both boys and girls with PWS.
- Most people with PWS will either have delayed, incomplete or absent puberty.
- There can be isolated premature pubarche, and there is also a small incidence of precocious puberty in males.
- At some point almost all children with PWS will require sex steroid therapy for induction, promotion or maintenance of puberty.
- Sex steroid therapy is also very important for bone mineralisation (bone density).

- In children with significant learning difficulties hygiene issues surrounding puberty can be an enormous problem and need to be discussed with carers. However learning difficulties should not be seen as a contraindication for usual treatment to stimulate normal puberty

5.5.8 Sleep Related Breathing Disorders

- Sleep-disordered breathing, including both obstructive and central sleep apnea, affects over 80% of individuals with PWS.
- OSA can occur due to hypotonia, obesity, upper airway dysfunction, kyphoscoliosis and adenotonsillar hypertrophy.
- OSA leads to a variety of complications such as systemic hypertension, pulmonary hypertension, cor pulmonale and cardiovascular disease.
- PWS can have problems with sleep-disordered breathing (SDB) with frequent hypoxic events, sometimes complicated by an abnormal central ventilation response.
- Growth hormone treatment can exacerbate sleep apnea, and therefore, polysomnography with capnography should be performed before starting GH therapy.
- Sleep-disordered breathing can cause cognitive deficiencies, behavioural problems, poor school performance and psychiatric disorders.

Recommended management:

- A paediatric respiratory specialist is required to manage apnoea.
- All patients with PWS should be referred for an annual inpatient overnight oximetry WITH capnography.
- PWS patients starting Growth Hormone should have pre-therapy inpatient polysomnography with a capnography study.
- A referral is to be made for an overnight oximetry or a paediatric respiratory team.
- Annual oximetry with capnography should be part of routine care to monitor for changes

5.5.9 Orthopaedic Management

- Scoliosis and kyphosis are common in children with PWS due to hypotonia and obesity (relative prevalence of between 30 and 70%).

- Weight control is also required to reduce the risk of scoliosis.
- Assessment for scoliosis is needed before starting growth hormone replacement, as scoliosis generally worsens in children who are receiving it.
- Worsening scoliosis is not an absolute indication to stop growth hormone treatment, but careful evaluation is required.
- An early orthopaedic referral is necessary for monitoring scoliosis progression
- Complications of surgical management of scoliosis are more common in children with PWS with a high risk of paraplegia (20%) and of major complications (30%, deep infections, pneumonia).

5.5.10 Management of hyperphagia/ obesity

- Nutritional management is a crucial aspect of care, as hyperphagia begins at around 2 years of age and progressively worsens, leading to severe obesity if environmental controls are not in place.
- Psychological support for children with PWS and their parents/ carers.
- Early onset of calorie-controlled diet with restriction to food access and money to buy food.

- Dietician referral to support dietary control.
- Regular exercise, with appropriate support/support groups
- No evidence supports the use of appetite inhibitors, somatostatin analogues, and bariatric surgery.
- Type 2 diabetes mellitus is common in patients with PWS, occurring in around 20% of cases with a mean time of onset of around 20 years; treatment should follow current national guidance.
- It should be noted that patients with PWS and diabetes need routine health surveillance related to their diabetes (retinopathy screening, nephropathy screening, etc) as well as health surveillance related to their PWS.

5.6 Ethical considerations

The support of vulnerable people; whether children or adults, is an essential core value to the medical profession. However this involves the careful balance of respecting an individual's autonomy and to avoid exploitation, harm and potential abuse.

Parents need to be educated early about the prospects of hyperphagia and the risks of developing obesity and obesity related problems. Once this education has occurred parents have a duty to act in the child's best interest to avoid these problems as much as is conceivably possible. It's therefore difficult to control patient's decision making, especially as they gain age and increasing autonomy.

It should be noted however that patients with PWS almost always have reduced satiety, and poor control over their eating disorder. Also metabolic complications of obesity are almost certainly of a poorer prognosis in PWS.
Therefore early death is likely without control of these problems from an external source.

There is good evidence that control of access of food and to money to buy food is essential for good long term prognosis. Therefore systems need to be in place for families and carers to put these systems into place with support from the medical professionals. These systems should have the agreement of the patient as much as is possible.

Problems occur when patients do not agree to such external controls. It can be argued that patients with PWS do not have capacity to make decisions related to food and diet and that we therefore have a duty of care to put these systems into place as much as is possible.

In reality this will involve intensive discussion and negotiation with the patient and carers over a period of time.

A multi-disciplinary team is involved with patients with PWS:
Shropshire Community Health NHS trust:

- Community Paediatrician-Lead clinician
- Dietician
- Physiotherapist
- Psychologist
- Speech and language Therapy

Shrewsbury and Telford NHS Trust:

- Neonatologist
- Paediatric endocrinologist

- Gynaecologist / urologist
- Paediatric Cardiologist
- ENT specialist
- General Surgeon (for orchidopexy)
- Respiratory paediatrician
- Dentist
- Ophthalmologist
- Paediatric Gastroenterologist

Birmingham Women's Hospital:
- Clinical genetics department,

SSSFT:
- CAMHS

Robert Jones and Ages Hunt Hospital:
- Orthopaedic surgeon

Local Authority:
- Social worker
-

6. Consultation

The community Paediatric Team has discussed the guideline.
Both the neighbouring trusts involved in the care of children are informed about the update of this guideline.

7. Dissemination and Implementation

Guideline will be available in hard copy at the Community doctor office and on the trust internet.

8. Monitoring Compliance

Guideline will be audited but as incidence is infrequent cohort will be small and data will be collected over a few years from initiation of this guideline.

9. References

Recommendations for the Diagnosis and Management of Prader Willi Syndrome. A.P. Goldstone et al J Clin Endocrinol Metab November 2008 93(11) 4183-4197
Up To Date, epidemiology and genetics of Prader Willi syndrome, https://www.uptodate.com/contents/epidemiology-and-genetics-of-prader-willi-syndrome#H3
https://www.nhs.uk/conditions/prader-willi-syndrome/symptome
Prader-Willi syndrome: Clinical features and diagnosis. UpToDate, 2024.
Prader-Willi syndrome: Management. UpToDate, 2024.

10. Associated Documents

Guidelines for the Public Health Nurse Team for the Management of Growth Measurements in School Age Children 1947-40658

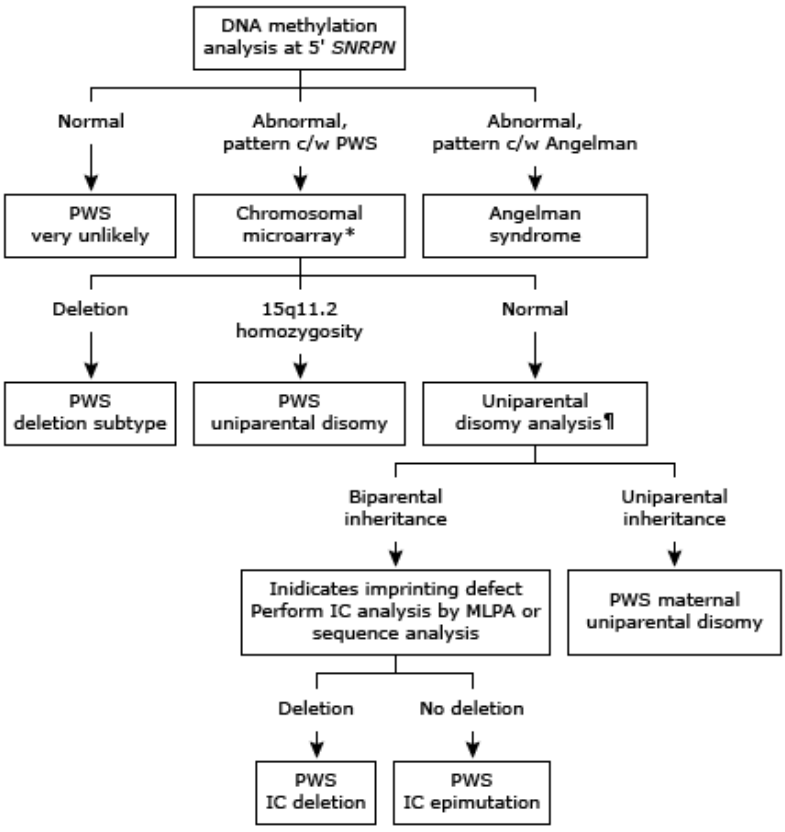
11. Appendices:

Appendix 1: Prader Willi Syndrome- Unique features

Temperature instability (Lack of fever does not rule out serious infection)	Cortisol Deficiency (May have underlying cortisol deficiency and need hydrocortisone. check with the Endocrine team)
High pain threshold (May not complain of pain despite of serious injures including fractures)	Hypothyroidism
Lack of vomiting (Vomiting should be taken up seriously it may be a pre terminal sign)	Vit D deficiency
Gastroparesis	Anaesthesia (Thick secretions, prolonged drowsiness)
Skin Pricking (may lead to bleeding or infection)	Risk of choking
Easy Bruising (NAI consideration)	Risk of Pneumonia
Risk of VTE	Transition care

Appendix 2: Diagnostic Workup for PWS

Diagnostic workup for suspected Prader-Willi syndrome



DNA: deoxyribonucleic acid; *SNRPN*: small nuclear ribonucleoprotein polypeptide N gene; PWS: Prader-Willi syndrome; c/w: consistent with this diagnosis; IC: imprinting center; MLPA: multiplex ligation-dependent probe amplification; FISH: fluorescence in situ hybridization; SNPs: single-nucleotide polymorphisms.

* FISH can also be used but is not preferred; FISH can diagnose deletion without detail of size.

¶ Uniparental disomy analysis is ideally performed with parental karyotype but can also be done with microsatellite probes or SNPs of parents and child.

Modified from: Driscoll DJ, Miller JL, Schwartz S, et al. Prader-Willi Syndrome, 1998 [Updated 2017 Dec 14]. In: Adam MP, Everman DB, Mirzaa GM, et al. (Eds), GeneReviews™ [Internet], University of Washington, Seattle 1993-2022. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1330/ (Accessed on October 27, 2022).

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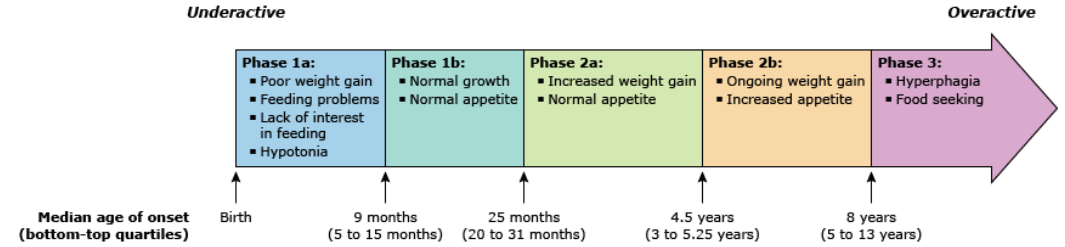
Appendix 3:

Phases of feeding and weight gain in Prader-Willi syndrome



Adapted from: Miller JL, Lynn CH, Driscoll DC, et al. Nutritional phases in Prader-Willi syndrome. Am J Med Genet A 2011; 155A:1040.

Appendix 4

Management of Prader-Willi syndrome

	Characteristics and monitoring	Interventions
Feeding and obesity		
Infants	▪ Feeding problems, poor weight gain, lack of interest in feeding	▪ Oromotor therapy ▪ Concentrated feeds (consult dietitian); commonly, nasogastric supplemental feeds
Children and adults	▪ Hyperphagia, obesity when there is a lack of environmental control	▪ Food restriction (with physical barriers) at home and school ▪ Calorie-restricted diet/low-carbohydrate diet; consult dietitian ▪ Exercise/physical therapy ▪ Consider pharmacotherapy if above actions are not effective ▪ Refer/discuss management of challenging cases with a PWS center
Behavior and neurodevelopment		
Motor skills		▪ Early intervention, occupational therapy, rhGH ▪ Carnitine supplements if deficient
Behavioral rigidity, obsessive-compulsive behaviors		▪ Behavioral specialist ▪ Pharmacotherapy
Skin picking		▪ Possible options: • N-acetylcysteine • Guanfacine • Topiramate
Anxiety		▪ Behavioral therapy ▪ Pharmacotherapy (use with caution due to risk of activation)
Cognitive and learning deficits	▪ Mild or moderate intellectual disability (mean IQ 60)	▪ Early intervention therapies (occupational therapy, physical therapy)
Endocrine issues		
Growth hormone deficiency	▪ rhGH improves linear growth, body composition, bone density, physical function, and motor development; in young children, it may promote cognitive development	▪ Initiate shortly after diagnosis of PWS (if nutrition is adequate) ▪ Benefits children and adults, with or without growth hormone deficiency ▪ Monitor for respiratory complications and hyperglycemia before and during therapy ▪ Monitor for development or worsening of scoliosis (children)
Hypogonadism	▪ Cryptorchidism ▪ Pubertal delay ▪ Infertility	▪ hCG (3 to 18 months old) ▪ Orchiopexy if needed ▪ Treatment for hypogonadism as needed* ▪ Birth control pills for fertile women
Low bone mineral density and osteoporosis	▪ DXA every 2 to 3 years beginning in late childhood	▪ Dietary calcium and vitamin D ▪ Sex steroid replacement* ▪ rhGH treatment ▪ For osteoporosis, consider treatment with bone resorption inhibitors in adults
Hypothyroidism	▪ Measure fT4 (to detect central hypothyroidism) and TSH (to diagnose primary hypothyroidism) ▪ Monitor every few months during the first year of life, then annually thereafter	▪ Management is similar to other patients with central hypothyroidism¶
Central adrenal insufficiency	▪ Association with PWS is not established ▪ Test if symptoms develop (unexplained nausea, vomiting, or hypotension, especially during physical stress)	▪ Management is similar to other patients with hypocortisolism¶
Type 2 diabetes mellitus	▪ Test with fasting blood glucose, hemoglobin A1c ▪ Test annually in patients with obesity	▪ Food security and physical activity are essential to management ▪ Management is otherwise similar to other patients with type 2 diabetes¶Δ
Sleep disorders		
Sleep apnea	▪ Mostly obstructive but may have central component ▪ Monitor for sleep-related symptoms ▪ PSG is indicated after start of growth hormone, at age 3 years, and if there is significant weight gain or change in behavior	▪ Adenotonsillectomy ▪ Residual symptoms are common ▪ CPAP may help but may be a challenge to implement, requiring behavioral intervention ▪ Tracheostomy in refractory cases
Excessive daytime sleepiness	▪ Primary disorder of vigilance	▪ Possible options: • Modafinil • Pitolisant
Orthopedic problems		
Scoliosis	▪ Clinical evaluation throughout growth ▪ Spine radiograph every 1 to 2 years, beginning at approximately 6 years	▪ Evaluation by an orthopedic specialist if scoliosis is detected
Hip dysplasia		▪ Hip ultrasound (infants); frequency is not established
Lower limb alignment abnormalities		▪ Footwear technician ▪ Orthopedic specialist

PWS: Prader-Willi syndrome; rhGH: recombinant human growth hormone; IQ: intelligence quotient; hCG: human chorionic gonadotropin; DXA: dual-energy x-ray absorptiometry; fT4: free thyroxine; TSH: thyroid-stimulating hormone; PSG: polysomnography (sleep study); CPAP: continuous positive airway pressure; GLP-1 RA: glucagon-like peptide 1 receptor agonist.

* Initiation and dosing of sex hormone therapy should be made in consultation with an endocrinologist. Considerations include benefits on bone density and potential adverse effects of sex hormones on mood, behavior, and epiphyseal closure.

¶ Refer to appropriate UpToDate content.

Δ For individuals with PWS, food security is essential. The efficacy and safety of GLP-1 RA in this population has not been established.

References:

1. McCandless SE, Committee on Genetics. Clinical report – health supervision for children with Prader-Willi syndrome. Pediatrics 2011; 127:195.
2. Duis J, van Wattum PJ, Scheimann A, et al. A multidisciplinary approach to the clinical management of Prader-Willi syndrome. Mol Genet Genomic Med 2019; 7:e514.
3. Butler MG, Miller JL, Forster JL, et al. Prader-Willi Syndrome – Clinical Genetics, Diagnosis and Treatment Approaches: An Update. Curr Pediatr Rev 2019; 15:207.
4. Duis J, Pullen LC, Picone M, et al. Diagnosis and management of sleep disorders in Prader-Willi syndrome. J Clin Sleep Med 2022; 18:1687.