Most children with Prader-Willi Syndrome (PWS) have some physiological and behavioural differences that make it difficult to assess how sick they actually are

Poor temp control
May not develop a fever or may be hyper- or hypothermic instead. Baseline temp below 37˚C/98.6˚F

Lack of vomit
Vomiting rarely occurs in PWS and should be taken seriously. Emetics may be ineffective.

Anaesthesia
Both general and regional anaesthesia are challenging. More likely to have cortisol deficiency. https://www.orpha.net/data/patho/Pro/en/Prader_Willi_EN.pdf

Abnormal body composition
affecting medication
Reduced lean tissue and increased adiposity. May affect dosages of medication: less required

Poor response to pain
May under report pain masking fractures, acute abdominal conditions, or internal injuries

Skin lesions – picking and bruises
Common and may complicate healing at IV sites etc

Top tips for triaging & treating children with PWS

Gastroparesis
Can become dangerous if overeating occurs. Severe gastric distension with ischaemia may result in stomach rupture. Child may present with abdominal distension, pain and/or vomiting but may only complain of mild abdominal discomfort.

Optimise communication strategies
Speech and language often lags behind understanding. Keep instructions clear and simple. Use visual aids to help with both comprehension and communication. Give plenty of time to process information and respond.

Explain & reassure
Children with PWS may struggle with new experiences outside of their usual routine. Take time to explain and reassure.

"What is normal for this child?"
Assessment can be difficult if you don't know the individual child at baseline. Ask the parent: they know the child best!

Don't panic! Just don't forget the hypothalamic dysfunction

More info - pwsa.co.uk Helpline: 01332 365676
https://www.pwsa.co.uk/information-for-professionals/health

PWSA UK
PRADER-WILLI SYNDROME ASSOCIATION UK