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Romanian Prader-Willi Association

ASOCIACIÓN MADRILEÑA  
PARA EL SÍNDROME DE  
PRADER-WILLI



## GASTRIC NECROSIS IS ASSOCIATED WITH NORMAL BMI IN PRADER-WILLI SYNDROME

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**INTRODUCTION:** Gastric necrosis and rupture is a rare and life threatening complication of a number of predisposing conditions. Gastric necrosis is caused by vascular insufficiency secondary to gastric dilatation and increased intra-gastric pressure. Gastric dilatation is associated with emptying delay and includes symptoms such as abdominal distension, tenderness and vomiting. This requires immediate attention including decompression. Where gastric necrosis or rupture is suspected, surgical intervention is unavoidable. In PWS, Wharton (1997) reported 6 individuals with gastric distension, 3 developed gastric necrosis. Schrandner-Stumpel (2004) reported two adults with PWS with similar gastric findings. The natural history of gastric dilatation and necrosis in PWS is still poorly understood. High pain threshold and lack of vomiting in PWS are contributing factors, but probably not the only predisposing factors in PWS.

**OBSERVATION:** We report three adults with clinical PWS, all with documented gastric necrosis and rupture. In two of them, a 26 year old female and a 32 year old male, the diagnosis of PWS was confirmed by post mortem molecular testing. The third individual, a 36 year old female, had a normal post-mortem molecular test result - this information came as a surprise. The PWS-like condition and behaviour of this woman is most likely due to a childhood-acquired hypothalamic dysfunction following meningitis and subsequent hydrocephaly requiring shunting. None of the individuals had binge eating behaviour during the days prior to death. All three individuals had been grossly overweight until started on a nutritional regime for PWS individuals. Their BMIs had since been within the normal range. None of them had received growth hormone treatment.

**DISCUSSION:** People with PWS and PWS like hypothalamic dysfunction seem to be at risk of developing gastric emptying problems, gastric necrosis and rupture. This risk appears at least partly to be due to weight loss. The risk seems to increase in those with BMI below 25.

To prevent this health hazard from occurring, individuals with PWS and PWS-like conditions should be allowed a BMI of not less than 25. Slimming below this margin should be avoided until the predisposing mechanisms are clarified in more detail and tools are available that would help to optimise and monitor the nutritional state and body composition in individuals with PWS or PWS-like hypothalamic dysregulation.

### References

Wharton RH et al. Acute idiopathic gastric dilatation with gastric necrosis in individuals with Prader-Willi syndrome. *Am J Med Genet* 1997; 73, 437-41  
Schrandner Stumpel CT et al. Prader-Willi syndrome: causes of death in an international series of 27 cases. *Am J M Genet A.* 2004 .124, 333-8