



**Pittsburgh  
Partnership**

*Specialists in Prader-Willi Syndrome*

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## Behavioral Considerations During an Emergency

*It is characteristic of Prader-Willi syndrome to have increased behavior problems when the individual is under stress. Stress may be reduced and behavior problems may be averted or managed during a medical or behavioral emergency with the following considerations:*

- The presence of a family member or familiar caregiver is generally helpful; this may include during the ambulance ride, if permissible.
- At all times interpret disruptive behavior as a stress reaction and respond to the person in the most calming, reassuring way possible. This is true whether or not psychiatric illness is involved.
- If the Emergency is behavioral, as with other unruly persons, simply your arrival may be enough to end the outburst. Showing up “in force” (2 persons), in uniform can be very effective.
- Take your cues from the family member or caregivers and as much as possible concerning how to manage the behavior.
  - Ask caregivers how they want you to become involved
  - If considering physical restraint, always ask caregivers if they see this as necessary or advisable
  - Destructive or aggressive behavior can often be disrupted by stepping between the individual and the target; this is more likely to be successful than speaking to the individual, giving warnings or making threats or physically restraining the individual. Caregivers can advise you on whether or not this procedure is likely to be *effective* or *unsafe*.
- Never offer, promise, or make threats about food to a person with Prader-Willi syndrome.
- If he has stolen food, make as little issue about the theft as possible and focus on helping him to calm. Allow him to hold onto the food if he refuses to give it up. Reassure him that you will not take it away, however, if he starts to stuff large amounts into his mouth, there is a risk of choking and you will have to judge the safest course to take regardless of what you have promised him.
- Doses of sedation safe enough to give outside the hospital are unlikely to be effective in ending a disruptive behavior. If higher doses of sedation are to be used in the hospital setting, one must assume that there is a significant danger of respiratory suppression. This is especially true in obese patients.
- Physical restraint of an obese person with PWS is especially dangerous. They can hypoventilate in the prone or supine position and may not appear to be in respiratory distress even when they are retaining CO<sub>2</sub> and are in danger of arrest.