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Organisation

Blind Rage

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PWS is known for hyperphagia and temper outbursts. Researchers have discovered that temper tantrums precede the typical food related behaviors in early childhood. For all children, including those who have PWS, tantrums occur when the reward centers of the brain begin to mature. The reward center enables all children to learn from experience and to form expectations about things that happen in their world.

When a child wants something and does not get it, a tantrum occurs. Or, when something occurs that is different from what the child is expecting, a tantrum occurs. Among children who have PWS, this tantrum behavior differs from typical children in 2 important ways:

1. It is more intense
2. The biochemical braking system to stop the tantrum is broken *

Each tantrum is unique to the child, but in PWS it is usually very quick in onset and may appear to occur out of the blue. If one looks carefully at the antecedents of the situation from the perspective of the child, often the etiology can be determined. Once a tantrum begins, there is nothing that can be done to stop it, other than to keep everyone and everything in the environment safe. The tantrum will run its course. After the tantrum is over, some children will be exhausted and require a nap. Other children will get back on track fairly quickly and resume their daily activities; they may or may not acknowledge their behavior.

The cortex of the brain not only governs reward, but it is responsible for thinking and reasoning behavior. Another part of the brain is involved in behaviors such as tantrums and shut downs. The amygdala directs brain mechanisms associated with survival (fight, flight or freeze); it is responsible for emotional expression, aggression, and impulsivity. For this reason, the cortex has been referred to as the 'baby sitter' of the amygdala. Although neural connections to the reward center of the cortex begin around age 2, the thinking and reasoning behavior is slower to mature in typical children and takes even longer to develop in children with PWS. Using the analogy of a computer, when a child has a full-blown tantrum, their thinking and reasoning ability in the cortex goes "off-line". This is why children with PWS appear to be in a 'blind rage" during the tantrum; they are literally behaving in survival mode, acting without thinking. In PWS it takes a while for the brain to "reboot." After they recover, they may act as if nothing has happened (brain reboot), and they may or may not remember what has occurred.

A tantrum is a learning experience for the caregiver. The keys to successful behavioral management are determining what the trigger is, redesigning the daily plan to avoid it, and developing coping strategies to deal more effectively with it.

[1] GABA (gamma-aminobutyric acid) is the major inhibitory neurotransmitter in the brain; it regulates muscle tone and provides the braking mechanism for behavior, allowing a person to "stop and think" before acting. GABA receptors are reduced in PWS, resulting in decreased GABA action in the brain that may also contribute to epilepsy, anxiety disorders and depression.