Cyclic Vomiting Syndrome in Children

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What is Cyclic Vomiting Syndrome (CVS)

CVS is a chronic disease marked by severe, recurrent episodes of nausea and vomiting in a person who is otherwise healthy. CVS episodes often flip-flop with states of wellness. Vomiting may occur every 5-10 minutes for several hours during an episode. On average, CVS episodes last 3-7 days but vary with each person. An episode may be accompanied by stomach pain, tiredness, sensitivity to light, and/or headaches. Episodes may begin at any time of the day but often start in the middle of the night or early morning hours. They may be triggered by positive 'stress' (vacations) or negative stress (job loss).

Many patients have a long history of symptoms and frequent emergency department (ED) visits and hospitalizations. A clear diagnosis may have been difficult to obtain which can be frustrating! Therefore, we are providing you with information to help you better understand the disease, best manage your symptoms and improve overall well-being.

Symptoms

Cyclic vomiting syndrome (CVS) often begins in childhood, usually between ages 3 and 7 years. The current pediatric diagnostic criteria specify a minimum of 3 vomiting episodes over six months or total of 5 attacks. The vomiting spells start suddenly, and many children have a typical warning period of 1-2 hours of symptoms such as nausea, irritability and abdominal pain. The attacks often begin in the early morning hours and awaken the patient from sleep. During the episode, patients often like to be left alone in a dark, quiet place. The vomiting is rapid-fire and invariably accompanied by what has been described as the most intense kind of nausea a human can experience. The typical child vomits 6 times per hour at the peak and the episodes last anywhere from a few hours to 10 days.

Affected children characteristically appear almost motionless during episodes. Some children will refuse to swallow saliva for fear of inducing vomiting; others compulsively drink water. This is possibly to reduce the upper abdominal pain due to the continual vomiting of acid content from the stomach. A period of exhausted sleep often follows, with a subsequent return to baseline health.

Once awake, the child is eager to eat. Attacks tend to be stereotypical. Ninety-eight percent (98%) of children experience the same progression and character of attack with each episode.

Symptoms may include:

- Abnormal drowsiness, or lethargy (91%)
- Paleness, or pallor (87%)
- Abdominal pain (80%)
- Headache (40%)
- Diarrhea (36%)
- Sometimes fever (29%)

Testing

In general, symptoms alone will strongly suggest a diagnosis of CVS. However, because there is no specific lab or X-Ray test for CVS and many other conditions cause recurrent vomiting, other causes may need to be excluded. Testing is similar to that of adults although some additional conditions which are more common in children need to be excluded. The most important includes malrotation of the intestine, where the abnormally positioned intestines can twist on themselves. A second is uretero-pelvic junction obstruction, where urine flow is blocked from the kidneys due to a congenital malformation which can lead to episodic vomiting. In both, corrective surgery will resolve the vomiting. Metabolic and genetic diseases can also cause recurrent vomiting, particularly in infants and young children. Very rarely, brain tumors

or other lesions in the brainstem can present with recurrent vomiting.

The most important test to exclude malrotation is an upper GI series (x-ray of the stomach and duodenum). The second is an abdominal ultrasound (or dedicated ultrasound of kidneys) to exclude ureteropelvic junction obstruction. During the episode, standard blood work and urinalysis are recommended at least once to evaluate for other causes. In those who do not respond to therapy or have concerning symptoms, blood and urine testing for metabolic disorders may be needed. At times, an imaging scan of the head may be needed, especially if there are neurologic symptoms. If symptoms suggest, an endoscopy (examination of the esophagus and stomach with a scope) may be performed to exclude gastrointestinal causes of vomiting. Multiple studies show that in children with suspected CVS, the results of these tests are generally normal.

Treatment

Treatment of pediatric CVS is similar to adults and begins with identification of the condition. A distinction is made between chronic vomiting and cyclic vomiting syndrome. Underlying conditions such as gastroesophageal reflux disease (GERD) and sinus infection (sinusitis) should be sought and treated in the patient.

Three levels of treatment include:

- Abortive treatment (to try to stop an attack after it starts),
- Rescue therapy (to keep the child as comfortable as possible if unable to stop an attack), and
- Prophylactic (steps to take to try to prevent future attacks)

Numerous medicines have been used to abort attacks and most medications used in adults are also used in children. When daily, preventive medicines are required, tricyclic antidepressants are generally reserved for children over the age of 5 years. Younger children are generally treated with cyproheptadine and/or a low dose beta blocker. Rescue therapy is needed if it proves impossible to abort the attack. Most children will respond to these measures. For the rare patient with persistence of recurrent attacks, daily preventative (prophylactic) measures are indicated.

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