Cyclic Vomiting Syndrome

What is cyclic vomiting syndrome?

Cyclic vomiting syndrome (CVS) is a condition with a specific pattern of vomiting with three main features: paroxysmal (sudden onset), stereotypical (similar episodes), and intervening periods of wellness. There is not one single test that confirms CVS; correct diagnosis is made by having a doctor take a careful history, performing a careful physical examination, and conducting tests to exclude other diseases.

What are the symptoms and signs of CVS?

Patients present with vomiting episodes that tend to recur in a cyclical pattern, such as every 2 weeks, or every 2 months. The vomiting is **paroxysmal**, or with sudden onset. Most patients with CVS feel well, until they get a sudden attack of nausea, which usually progresses to vomiting a little later. The nausea and vomiting often start in the evening, and many times can even wake the patient from sleep.

Second, the vomiting episodes are **stereotypical**. Each vomiting "attack" resembles similar episodes they have had previously. Very often, the attacks last between 8 and 24 hours. However, for some patients, attacks can be as brief as 1-2 hours, and for others they can last several days. Episodes often begin with nausea, and progress to vomiting, with some people vomiting several times an hour. During the vomiting episodes, patients often like to be left alone or be in a quiet place. Other symptoms can also occur during the episode, including severe stomach pain, diarrhea, and headache. Patients can become disoriented, irritable and turn pale and clammy during an attack. Some patients vomit to the point of dry heaves or become dehydrated. The episodes often resolve by themselves without any obvious intervention or explanation.

Third, most patients feel completely well in between episodes (**intervening wellness**). After the episode resolves, the affected patient often returns to feeling "normal" within a few hours, and starts drinking and eating. The period of wellness in between episodes is between 1 and 3 months for most patients. However, some patients will have more frequent episodes (every 1-3 weeks), and others will have episodes that occur rarely (every 6-12 months).

How common is CVS?

About 1 in 33,000 children are thought to have CVS. It can occur in adulthood as well, but is more common during childhood.

What causes CVS?

The definite cause of CVS is unknown. A number of medical studies suggest that in most patients, CVS is related to migraine. The sudden onset of attacks with spontaneous resolution is also seen in patients with migraine headaches. Most (but not all) children with CVS have a family history of migraine. In addition, many of the treatments used to treat migraine headaches are also effective in treating CVS.

How is CVS diagnosed?

Diagnosing cyclical vomiting is difficult for most doctors, because CVS is a rare condition. There is a long list of diseases and problems that cause vomiting. These include: gastroesophageal reflux (acid reflux), stomach inflammation, pancreas inflammation, urinary infections, food allergies, and stomach infections. Most of these problems have a beginning and end, or are recurrent – symptoms occur every day or on most days. However, bouts of severe vomiting separated by well periods is very unusual, and should make a physician or family suspect the diagnosis of CVS.

Because there is no definitive test to prove CVS is triggered by a migraine, a number of other conditions may need to be ruled out. These include anatomic abnormalities of the bowel such as malrotation. In children with **malrotation**, the intestines are abnormally positioned in the body from birth, and can twist on themselves. A second condition to be considered is **ureteropelvic junction obstruction**, in which urinary flow out of the kidneys is blocked, leading to backup of urine into the kidney, which in turn leads to vomiting. In young adults, cyclical vomiting syndrome has been described as a complication of cannabis (marijuana) abuse. Very rarely, brain tumors or other lesions in the head can present with recurrent vomiting. Lastly, **metabolic disease (hereditary enzyme deficiencies)** can cause recurrent vomiting (particularly in infants and young children), because there is a missing enzyme in the patient leading to buildup of toxins in the blood and urine.

What tests are used in children to diagnose CVS?

In general, the history will strongly suggest CVS. However, in many cases, a physician may need to perform further tests to exclude other conditions. These tests may include: an upper GI series (x-ray with contrast to exclude malrotation), an abdominal ultrasound (ultrasound of kidneys and gallbladder to rule out pathology), and a CT scan or MRI of the head. In addition, during a CVS episode, blood tests and urine tests may need to be obtained to evaluate for other causes of the episode (including infection, inflammation of the pancreas, and metabolic enzyme problems). In some patients, endoscopy (examination of the esophagus and stomach with a scope/video camera that passes by the mouth and goes into the stomach) may be needed.

What is the treatment for CVS?

Treatment for CVS is divided into two major types: **abortive** therapy and **prophylactic** therapy. **Abortive therapy** means giving treatments to stop the episode once it starts, and only giving that treatment during the episode. In contrast, prophylactic therapy means giving a medication every day, whether the child is well or sick, in order to prevent episodes from coming on.

Once a CVS episode starts, it can be very hard to stop. For many patients, the best treatment is supportive, and can, in severe cases, include intravenous fluids and a quiet room in a hospital. Anti-nausea medicines, including ondansetron (Zofran), promethazine (Phenergan), and chlorpromazine (Thorazine) are sometimes used to reduce the feelings of nausea. Because patients may be anxious and just feel lousy during an attack, they may benefit from an antianxiety medication such as lorazepam (Ativan). Other patients may benefit from antimigraine treatments like sumatriptan (Imitrex). After enough time passes (usually hours to days), most patients come out of the episode.

Prophylactic treatments are medications given on a daily basis to try to prevent episodes from coming on. Studies suggest that in patients with frequent episodes (every 1-2 months), prophylactic treatment can lessen the frequency and severity of episodes. Therefore, in patients having frequent episodes (i.e. every 1-2 months,) prophylactic treatment should be considered. However, if episodes are infrequent (i.e. once a year), prophylactic therapy is probably unnecessary. Prophylactic medications include

cyproheptadine, propanolol, and amitriptyline. In some patients with resistant disease, anticonvulsants (i.e. medicines usually used to treat seizures) are often used. These medications include topiramate, valproate, and levetiracetam. While all these prophylactic medicines are generally safe, each has a different side effect profile, and so the benefits and risks of prophylactic therapy need to be reviewed with your physician.

What about stress and diet?

In some patients, CVS may be triggered by either physical or psychological stress. Physical stresses that can trigger episodes include infections such as colds and viruses. Some women may develop CVS or migraines around their menstrual periods. Psychological factors also play a role. Some patients will have episodes triggered by negative (unhappy) stressors, such as tests or term papers. Other patients will have episodes triggered by positive stressors (such as holidays and visits with relatives). However, a large group of patients cannot identify a specific stressful event as a trigger for CVS. While the illness is not caused by stress, stress can make things worse, and CVS is a stressful illness. Therefore, in many patients, treatments to promote relaxation (counseling, yoga, acupuncture) may help.

We don't know about the role of diet in CVS. However, some patients with migraine headaches do benefit from avoiding certain foods such as caffeine, smoked cheeses, chocolate, and legumes. Since there is no proof diet plays a role, we don't routinely recommend dietary modifications, but this is something to be discussed with your physician.

What can I expect if my child has CVS?

The disorder can affect a person for months, years, or decades. There can be complications of repetitive vomiting such as dehydration, small tears in the lining of the esophagus, inflammation (esophagitis), and tooth enamel decay. Fortunately, once properly diagnosed and treated, most patients improve. In addition, some small studies suggest that many children may "outgrow" CVS in a few years. Some patients go on to have more typical migraine headaches.

Where can I find support for my child and family?

Talk to your physician, to other patients, and also explore the website **www.cvsaonline.org**. If you are interested, join the Cyclic Vomiting Syndrome Association, which offers educational materials, conferences, and support groups.

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