



Cyclic Vomiting Syndrome

By: Robert M. Issenman M.D., Chief, Pediatric Gastroenterology and Nutrition and Director, Ambulatory Services, McMaster Children's Hospital and Professor, Pediatrics, McMaster University, Hamilton, Ontario, Canada

Introduction

Cyclic vomiting syndrome was first described about 120 years ago by Dr. Samuel Gee, the erudite British physician. Interest in the syndrome was revived when Kathleen Adams, a parent of an affected child founded the Cyclic Vomiting Syndrome Association in 1993. She enlisted the support of pediatric gastroenterologists, Drs. David Fleisher and B.U.K. Li, who recognized that better treatment of the disorder would only occur if the syndrome could be scientifically defined for medical researchers. The result of this effort has been three scientific meetings on the Cyclic Vomiting Syndrome. The first international meeting was in 1994 and the latter two in Milwaukee in 1998 and 2000.

Definition of Cyclic Vomiting Syndrome

These meetings and some 70 publications since have defined cyclic vomiting syndrome (CVS) as a condition characterized by recurrent, stereotypical bouts of intense vomiting interspersed with "periods of completely normal health." The average child with CVS starts bouts of cyclic vomiting at 5.2 years of age and has been affected for 2.6 years before diagnosis. Females and males are equally affected (55:45).

Description of Attacks

The vomiting is 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 with an average of 25 bouts of vomiting (emesis) per episode. Affected children characteristically appear almost motionless during episodes, refusing to swallow saliva for fear of inducing vomiting. Others compulsively drink water, possibly to reduce the upper abdominal (epigastric) pain due to the continual emesis of acid content from the stomach. This is often followed by a period of exhausted sleep at which time the child wakes and is eager to eat. This has been described as a signature "on/off" pattern by Dr. Li. Similarly there is often a pattern with 34% of children awaking from sleep at 2-4 a.m. with vomiting and intense nausea. Attacks tend to be stereotypical, in that 98% of children experience the same progression and character of attack with each episode. Symptoms include abnormal drowsiness, or lethargy (91%); paleness, or pallor (87%); abdominal pain (80%); headache (40%); diarrhea (36%); and occasionally fever (29%).

Chronic Vomiting and Cyclic Vomiting

Work by Dr. Li has clarified that cyclic vomiting syndrome needs to be distinguished from a pattern of chronic vomiting in which a child vomits on a daily basis. Chronic vomiting lacks the intensity and periodicity of CVS and is much more likely to be associated with underlying disease. Chronic vomiting is a "final common symptom pathway" for disease affecting numerous body systems and processes. For example, chronic infection of the urinary or gastrointestinal tract and inflammation of the pancreas (pancreatitis) may produce chronic vomiting. Raised intra-cranial pressure typically results in early morning vomiting. Anatomic problems affecting the urinary or gastrointestinal tract can produce regular or irregular vomiting. Obstruction of the kidneys and a twisted bowel (midgut volvulus) present at birth can be particularly hard to pinpoint. In both instances, ultrasonography [an imaging technique using high-frequency sound waves] and specialized radiographic [x-ray] techniques must be done at the time of vomiting to find the underlying disease.

The Cyclic Vomiting Syndrome

In most cases, the cause of cyclic vomiting syndrome remains unexplained. Eighty-two percent of children are found to have a pattern described as "migraine associated CVS." Many have a family history of migraine headache. This pattern has a good prognosis and generally the problem either resolves between 9 and 14 years of age or matures into a more clearly recognized pattern of migraine headache (33%).

Some of the most severely affected children have a pattern that manifests symptoms typical of the body's emergency "fight or flight" response. These children often have profound nausea and vomiting episodes lasting up to six days. They are more often resistant to conventional treatment. Bouts are accompanied by elevated blood pressure and output of certain amino acids found in the urine (urinary catecholamines).

Recently, Dr. Rick Boles, a geneticist at UCLA has defined the presence of a particular pattern of cellular abnormality seen more commonly in some CVS children. These children have a large rearrangement and several mutations in a part of their DNA. This pattern may also be seen in some children with classic migraine, but is most often recognized in children with other problems such as

developmental delay, growth failure, seizures, muscle weakness, or eye problems. Laboratory tests often reveal a persistent presence of lactic acid in the blood.

Approach to Treatment

Treatment of CVS begins with identification of the condition and making the distinction 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 these patients. However, in some patients, these problems co-exist with cyclic vomiting syndrome. The child with typical migraine pattern CVS, and a normal abdominal contrast x-ray that visualizes the entire small bowel, can first be treated with anti-migraine therapy.

Not every child requires every medical test. However, certain warning signs may require urinalysis, upper GI endoscopy, abdominal ultrasound, brain CT or MRI, and metabolic blood tests. Warning signs include chronic daily symptoms or other suspicious symptoms the physician should exclude, anatomic problems, space occupying neurological lesions (hemorrhage or tumor), and known metabolic causes.

Cyclic vomiting syndrome is highly responsive to the “placebo effect.” Once the child and family learn that the condition is familiar and treatable, there is often up to a 50% improvement in episodes.

Acute Attacks

Dr. Fleisher has identified three levels of treatment; *abortive treatment*, *rescue therapy*, and *prophylaxis*. Numerous agents have been used to abort attacks including acetaminophen (e.g., Panadol, Tempra, Tylenol), non-steroidal anti-inflammatory agents (e.g., Advil, Motrin), drugs used to treat or prevent nausea and vomiting (anti-emetics), and anti-migraine preparations. Be sure to discuss the use of medications with your child’s physician.

Rescue Therapy: Keeping the Patient Comfortable

If it proves impossible to abort the attack, keeping the patient comfortable is of paramount importance. There is increasing evidence that these children have an overly reactive autonomic stress response, which may result in “anticipation stress” from fear of an attack beginning. These children may need hospitalization and intravenous fluid replacement, intravenous medication to control acid reflux, and sedation. A treatment plan should be organized in advance (either in hospital or home care) to allow expedited treatment for children experiencing recurrent attacks.

Prophylactic Treatment

Most children will respond to the measures outlined above. For the rare patient with persistence of recurrent attacks, daily preventative (prophylactic) measures are indicated. Patients should certainly avoid suspected “migraine triggers” such as chocolate, cheese, caffeine, and licorice to see if that helps. Otherwise, a number of medications have been tried with some success. Discuss this with your child’s physician.

Summary

The recognition and treatment of CVS has come a long way in just ten years. Children’s Memorial Hospital in Chicago and Northwestern University, in association with the Cyclic Vomiting Syndrome Association (CVSA), has established the Cyclic Vomiting Center. Cases that are resistant to treatment can be assessed in this center. The CVSA has established a network of knowledgeable physicians across North America and around the world who have the interest and experience necessary to vastly improve a lot of children (and families) suffering from cyclic vomiting syndrome.

For information and support contact:
Cyclic Vomiting Syndrome Association
10520 W. Bluemound Rd.
Milwaukee, WI 53226
Website: www.cvsaonline.org

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