Boston — Cyclic vomiting syndrome in adults often goes unrecognized for years after onset, despite its severe and disabling consequences.

The disorder may be the cause of repeated visits to the emergency department, unnecessary surgeries and diagnostic tests, and substance abuse, according to participants at the first-ever symposium devoted to cyclic vomiting syndrome (CVS) in adults. The gathering was held as a satellite meeting following a meeting on neurogastroenterology and motility.

Diagnosis of CVS in adults is complicated by the variability in age of onset and pattern of symptoms. The result is that the average delay in making the diagnosis is 8 years from the time symptoms first appear, noted Dr. B. U. Li, director of the Center for CVS at the Medical College of Wisconsin, Milwaukee, in a presentation at the meeting. “Where were we? How did we miss these folks?” Dr. Richard McCallum of the Kansas University Medical Center, Kansas City, rhetorically asked the audience, composed of physicians, patients, and family members.

As the director of the first center for CVS in adults, Dr. McCallum said, “We’re getting a continued trickle of patients coming to us, and an avalanche of phone calls from patients and physicians from around the world.”

According to the Rome 3 criteria H1b, the definition of CVS is two or more periods of intense nausea or unremitting vomiting or retching lasting hours to days, with a return to the usual state of health lasting weeks to months (J. Gastro. Liver Dis. 2006;15:237-41).

In its mildest form, the symptoms of CVS do not interfere with a patient’s ability to work and perform usual activities; however, the patient may have fewer of these episodes in a year than the number of hours they spend sleeping. The symptoms of CVS can range from mild to severe, with an average delay in making the diagnosis of 8 years from the time symptoms first appear. In adults with CVS, 70% had migraines both prior to and during CVS episodes, and 57% had first and second degree relatives with migraines. For these patients, migraine prophylaxis is essential to prevent CVS symptoms. In a group of 41 adults with CVS, 70% had migraines during or between episodes, and 57% had first and second degree relatives with migraines. For these patients, migraine prophylaxis is essential to prevent CVS symptoms.

The effects of CVS on patients can be severe and disabling, leading to a decreased quality of life and a decreased ability to work and perform daily activities. CVS can also lead to complications such as dehydration, malnutrition, and psychiatric conditions. The diagnosis of CVS is often delayed, with an average delay of 8 years from the time symptoms first appear. The average delay in making the diagnosis is 8 years from the time symptoms first appear. In adults with CVS, 70% had migraines both prior to and during CVS episodes, and 57% had first and second degree relatives with migraines. For these patients, migraine prophylaxis is essential to prevent CVS symptoms. In a group of 41 adults with CVS, 70% had migraines during or between episodes, and 57% had first and second degree relatives with migraines. For these patients, migraine prophylaxis is essential to prevent CVS symptoms.

Several patients experienced eight or more vomiting episodes per hour for months on end, leading to some to more than 100 visits to the emergency department or hospitalizations. As CVS progresses, episodes may become more frequent with less time for recovery, a process Dr. Fleisher terms “coalescence.”

When rushed to the emergency department during the emetic phase, CVS patients can present with blood in the vomitus due to pylorospasm or Mallory-Weiss tears resulting from forceful heaves, erosive esophagitis, and aspiration. Dehydration and electrolyte imbalance, especially hypokalemia, often accompany the GI symptoms. Patients often have intense abdominal pain, and may demand narcotics; they may show signs of narcotic dependence. Tooth decay can be evident, and patients may describe chronic weight loss, Dr. Fleisher said.

Patients also may show signs of a hyperadrenergic state, including low-grade fever, rapid pulse, and hyperreactivity. Neurophysiologically, without bandemia, accompanied by vomiting and abdominal pain, may be misdiagnosed as pancreatitis, peptic ulcer, appendicitis, or pylonephritis. The vomiting attacks, with or without hypotension, may lead to confusion and give the impression of porphyria, phaeochromocytoma, abdominal distension, intermittent small bowel obstruction, or endometriosis.

Patients also may present with unusual behaviors and mental states that compound the difficulty of identifying CVS. Normally pleasant and affable patients may become irritable and verbally abusive, demanding medications. Ant and affable patients may become irritable and verbally abusive, demanding medications. Physicians may describe thirst so intense that they drink surreptitiously from toilet bowls. The patient may demonstrate guggle-and-vomit sequence that can be mistaken for bulimia, and images may also be explained by the transient relief provided by the sudden emptying of the stomach.

Other patients may ask for repeated hot showers or baths. Some patients may appear so immobile that it is impossible to establish whether they are asleep or awake; they may also withdraw from social contact, a frightening condition Dr. Fleisher describes as “conscious coma.”

CVS patients often undergo batteries of tests, including upper GI series, abdominal ultrasound and CT scan, colonoscopy, barium enema, endoscopy, MRI of the head, sinuses, EEG, and lab work, usually with negative results.

In Dr. Fleisher’s series, the patients underwent almost 300 diagnostic studies—none of which were indicative of an organic etiology for CVS.

Recent findings suggest that about 75% of patients with CVS show rapid gastric emptying on electrogastrograms, and this may help distinguish CVS from other vomiting disorders (Neurogastroenterol Motil. 2006;18:728 [abstract 200]).

Unnecessary surgeries are common as well. Of the 41 patients in Dr. Fleisher’s series, 38 had undergone cholecystectomies, 2 had appendectomies, 5 had laparoscopies, 1 had a hysterectomy, and others had undergone other GI procedures. None of these procedures relieved the CVS symptoms.

Part of the problem is the lack of continuity of care for these patients, especially those who present repeatedly to hospital emergency departments. Dr. Fleisher suggests that more CVS centers should be created and staffed by two to three physicians available 24/7, as well as nurses and mental health professionals. A patient can be managed effectively by his primary care physician, and then referred when necessary to a center based on the patient’s centrognostic of his history, he added.

The Cyclic Vomiting Syndrome Association (www.cvsaonline.org) is establishing a referral network and provides resources for physicians who want to learn more about CVS.

Treatment Options for Adult CVS

A physician must be knowledgeable, patient, non-judgmental, and quick to respond when treating adults with cyclic vomiting syndrome, Dr. Fleisher said.

The physician should follow a rational treatment plan, tailored to the phase of the disease, that includes sedation when symptoms range uncontrolably, be advised.

The CVS cycle has four phases, said Dr. Fleisher, who has treated more than 350 pediatric and adult CVS patients at the University of Missouri Hospital and Clinics, Columbia.

The time between episodes, when the patient feels well, can be considered the first phase. The goal in this phase is to prevent a CVS episode by recognizing and controlling triggers, such as menstruation, increased stress, pleasant excitement, or fatigue, or infection. Some patients have interepisodic dyspeptic nausea and abdominal discomfort, especially in the mornings, and they may respond to proton-pump inhibitors. In some cases, IV opiate administration is indicated.

Patients should be checked for electrolyte depletion, tetany, hematemesis, and secretion of inappetant antidiuretic hormone. If the vomiting is not controlled, Dr. Fleisher recommends sedating the patient in a dimly lit and quiet room until the episode passes.

“A CVS patient needs to know there is an escape hatch that gets them out of their misery. Without that, the more they will suffer and the more they will coalesce,” Dr. Fleisher said, referring to the process in which CVS episodes become more and more frequent. He recommends chlorpromazine (0.5-1.0 mg/kg) plus diphenhydramine (0.5-1.0 mg/kg) in normal saline over 15 minutes, which can be repeated as often as every 3-4 hours if needed.

The length of the recovery period reflects the adequacy of management of the emetic phase. Patients with severe fluid or electrolyte deficits will have a more difficult and prolonged recovery. Some patients can tolerate a normal diet soon after the emetic phase passes, while others will tolerate only clear liquids.

Long waits in emergency rooms, encounters with caregivers who are unfamiliar with CVS, receiving implausible diagnoses, the repetition of unwarranted diagnostic procedures, and stopgap intravenous hydration followed by being sent home still sick are common experiences that reinforce patients’ feelings of being out of control of an illness that no one understands or can treat,” Dr. Fleisher wrote in his 2005 report on the 41 adult patients (www.cvsaonline.org). “Dr. Fleisher noted that, the more they will suffer and the more they will coalesce.”

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