Cyclic Vomiting Syndrome: A Paroxismal Disorder of Brain-Gut Interaction

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Cyclic Vomiting Syndrome (CVS) consists of recurrent, stereotyped episodes of intractable nausea and vomiting lasting hours or days and separated by symptom-free intervals [1,2]. It is a functional disorder in the sense that no organic disease has been shown to cause its symptoms. Few naturalistic descriptions or clinical investigations of CVS appeared in the medical literature subsequent to Samuel Gee's publication of a series of patients in 1882. It was presumed to be a rare disorder. Lack of recognition of CVS caused frustation in both patients and clinicians.

Renewed interest in CVS earlier in this decade coincided with the development of support groups in the United States and Great Britain, the production of the 1st International Symposium on CVS in 1994, and publication of its proceedings [3]. In 1995, Abu-Arafeh and Russell [4] reported a population-based study which showed the prevalence of CVS to be 1.9% in school children of Aberdeen, Scotland. If future epidemiologic studies confirm these data, a disorder previously thought to be rare will have a prevalence in children double that of epilepsy and diabetes combined.

CVS affects boys and girls in nearly equal numbers [5]. CVS can begin during infancy or mid-life, most commonly between 2 and 7 years of age [5]. The frequency of episodes in a series of 71 patients ranged from one to seventy times per year, averaging twelve times per year [1]. The term “cyclic” implies regularity and predictability of attacks. However, in about 50% of patients, attacks recurred sporadically [1]. Migraine, increased susceptibility to motion sickness and functional bowel disorders are more prevalent in patients with CVS as well as their families [1].

The features of CVS episodes are consistent relative to each patient. Most patients' attacks begin at the same times, most commonly during the night or morning [1,5]. The duration of most episodes, i.e. the interval of time between the onset of nausea and its subsidence, tends to be the same in each patient over months or years. Episodes may last 2 hours or a week or more, the commonest durations being 12 to 24 hours and 1 to 2 days [1]. Cyclic vomiting may become more severe as episodes become longer and/or more frequent.

Some patients experience recognizable prodromal symptoms that provide opportunities to administer oral medications that might abort episodes if the medications can be retained long enough to be effective. Once vomiting begins, it reaches a typical intensity of 20 or more times per hour, usually the first hours of an episode [(2)]. Episodes are tipically separated by days or weeks of no symptoms, in contradistiction on the vomiting of dyspepsia or giardiasis which tends to be low grade (1-2 times per day) and recurs almost daily [(2)]. About 80% of patients can identify circumstances or events that trigger the onset of attacks, the commonest being heightened emotional states, which may either be noxiuos (e.g. acute anxiety) or nonnoxious (e.g. pleasant excitement) [(1)].

Numerous signs and symptoms may accompany ciclic vomiting, e.g. pallor, weakness, hypersalivation, tachycardia and hypertension, heartburn, fonophobia, photophobia, headache, abdominal pain, loose stooling, fever, skin blotching, leukocytosis, and insulin-resistent hyperglicemia in diabetic cyclic vomiters. There is intense autonomic discharge and release of ACTH, vasopressin, norepinephrine and prostaglandin E2 [(6)]. The extent to which these acute phenomena are accompaniments of nausea per se, migrainous events, primary neuroendocrine pertubations, or somatic components of emotional arousal requires investigation.

Patients' behavioral states during episodes seem to be of three types: 1) subdued but responsive; 2) an immobile, unresponsive state referred to as “conscious coma”; and 3) writhing and moaning. Patients who are subdued but responsive prefer to lie down, but are able to walk about and perhaps watch television or attempt to play between bouts of vomiting. Patients in the state of “conscious coma” lie motionless with their eyes closed and are so unresponsive that it is difficult to know if they are awake or asleep. Patients who writhe and moan between bouts of vomiting have intense abdominal ache or severe retrosternal pain or both. Other unusual behaviors are observed in some patients that may be misinterpreted by the observer. Some are unwilling or unable to swallow their saliva; they may be somewhat alert, but cannot speak because of accumulated oral secretions. “Spit-out” behavior is typified by patients who lie, sit or walk about holding an emesis basin or towel into which they repeatedly expectorate saliva. Although eating is intolerable during episodes, some patients are compelled to guzzle large amounts of fluids only to vomit almost immediately after each draught. If spontaneous vomiting doesn't occur promptly, they may attempt to induce vomiting by putting their fingers down their throat. This should not be mistaken for bulimia. These behaviors have two purposes: drinking dilutes the acid and biles in gastric juice thereby lessening esophago-pharyngeal pain caused by vomiting; vomiting is induced in order to achieve a transient lessening of nausea that follows successful evacuation of the stomach. Some patients are comforted by prolonged showering or bathing. Concerned parents understandably feel the need to evoke a response from their sick, uncommunicative child and repeatedly ask, “are you OK?” Although children need to know that their parents are available and supportive, they feel burdened by such demands for responsiveness. When they are well, they often don't like to talk about or be reminded of how they felt during episodes.

The course of the illness is unpredictable. CVS may involve several or several hundred episodes, which may recur during the course of a few months or a few decades [(1)]. In some cases, patients may experience remissions lasting a year or more, only to suffer a resumption of episodes. This is especially troubling to young adults who are expected to finish school, pursue a career and become self-sufficient. Although they may be bright and talented, they live from day to day, unable to plain their futures or hold jobs because of unpredictable interruptions in their ability to work. Nevertheless, most patients eventually recover. Some go on to develop migraine headaches [(7)], a condition more easily treated than CVS in most cases.

CVS patients as a group have an unexpectedly high prevalence of concomitant disoders. My review of 52 consecutive CVS patients treated between 1991 and 1996 revealed that 3 had idiopathic epilepsy (10 times the expected prevalence), 3 had neurologic disease attributable to perinatal or post-natal brain damage, 6 had histories indicative of placental insufficiency, fetal distress or perinatal anoxia without obvious sequela, 6 had major congenital anomalies (e.g. gastroschisis, congenital lung cysts, congenital heart disease, craniofacial deformity) (twice the expected prevalence), and 3 had Type I diabetes (more than 300 times the expected prevalence). Sixteen (31%) had clinically significant psychiatric conditions, e.g. anxiety disorders or depression. Panic attacks may trigger cyclic vomiting episodes and it is sometimes difficult to distinguish such cyclic vomiting patients from non-CVS patients with panic or anxiety attacks who also vomit, have abdominal pain and signs of excessive autonomic discharge.

Lack of generally accepted clear distinctions between the terms “migraine”, “abdominal migraine”, and CVS has resulted in nosologic confusion. In my view, all of these disorders are migrainous. In migraine, headache is the predominant symptom. In abdominal migraine, sustained, incapacitating, non-colicky, mid- to upper abdominal ache that lasts for hours is the predominant symptom. In CVS, nausea and vomiting are the predominant symptoms. Patients with any one of these syndromes are more likely tu suffer symptoms tipycal of the other two disorders. All three disorders seem to be paroxysmal dysfunctions occuring in individuals who are costitutionally predisposed. It must be stated, however, that the above definitions are semantic in nature and will probably be revised or discarded once pathogenetic mechanisms become known.

The differential diagnosis of CVS includes many disorders that may mimic it [(1)]. Misdiagnosis of CVS is more likely in patients with intracranial space-occupying lesions, unsuspected obstructive uropathy and inborn error of metabolism.

No standard management has yet emerged for CVS. The literature contains numerous therapeutic recommendations [(3)], but all of them should be viewed as points of departure in the search for the regimen most effective for each individual patient. The general goals of management are: 1) Interruption of episodes; among the most effective anti-emetics for this purpose are ondansetron, granisitron and lorazepam. 2) To abort episodes; this may be accomplished if the patient can retain oral medications, e.g. ondansetron for impending nausea, ibuprofen for abdominal migraine-like pain, an H2 blocker or proton pump inhibitor to protect esophageal mucosa and dental enamel, and lorazepam p.o. or sublingually for its sedative, anxiolytic and antiemetic effects. 3) Search for and alleviate conditions that may predispose to episodes, e.g. anxiety disorders, chronic sinusitis. 4) Prophylaxis of episodes in patients whose attacks are frequent or prolonged; cyproheptadine, pizotifen, amitriptylene and propanolol have been used successfully in many cases. 5) Make the patient whose episode cannot be prevented or shortened comfortable until the attack runs its course; this requires effective sedation, since sleep is the only state that provides respite from intractable nausea.

The following complications of CVS episodes should be anticipated; 1) Water and electrolyte deficits in extracellular fluid; 2) peptic esophagitis - the vomitus should have a pH above 4.5 and be free of gross and occult blood; 3) deficits of intracellular potassium and magnesium that may be present even when serum levels are near or at the low normal range; 4) fluid retention due to nausea-induced inappropriate secretion of antidiuretic hormone.

Management of CVS involves more than finding the anti-emetic or prophylactic agent that works. It also requires an understanding of the patient as a human being, as a member of a family, as someone who is oppressed by a misterious, unpredictable disorder that causes fear and despair. In many cases, improvement is principally brought about by a relationship with a physician who is genuinely caring, accessible, collaborative and unwilling to be defeated by failure.

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REFERENCES

1. Fleisher Dr, Matar M. The Cyclic vomiting Syndrome: a report of 71 cases and literature review. *J Pediatr Gastroenterol Nutr* 1993; 17: 361-9.
Cited Here... | View Full Text | PubMed | CrossRef

2. Pfau BT, Li B U.K. Differentiating cyclic from chronic vomiting patterns in children: Quantitative criteria and diagnostic implications. *Pediatrics* 1996; 97: 367-8.
Cited Here...

3. Li B U.K. (guest editor). Cyclic Vomiting Syndrome. *J Pediatr Gastroenterol Nutr* 1995; 21 (Supplement 1).
Cited Here...

4. Abu-Arefeh I, Russell G. Cyclic vomiting in children: A population based study. *J Pediatr Gastroenterol Nutr* 1995; 21 (4) 454-458.
Cited Here...

5. Fleisher D. Cyclic vomiting syndrome described. *J Pediatr Gastroenterol Nutr* 1995; 21 (Supplement 1) S1-S5.
Cited Here...

6. Sato T, Igarashi N, Minami S et al. recurrent attacks of vomiting, hypertension and psychotic depression: A syndrome of periodic catecholamine and prostaglandin discharge. *Acta Endocrinologia* (Copenh) 1988; 117: 189-197.
Cited Here...

7. Hammond J. The late sequelae of recurrent vomiting of childhood. *Develop Med Child Neurol* 1974; 16: 15-22.
Cited Here... | PubMed