

Cyclic Vomiting Syndrome: An Easily Overlooked Disorder



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Dear Editor,

An article published by Haghghat et al in December 2016 issue (vol. 1, no. 2) of the *International Journal of Basic Science in Medicine* examines a new guideline for using erythromycin in the management of childhood cyclic vomiting syndrome (CVS).¹ This is an interesting and informative article that urges researchers to explore other management approaches as well as the information around the incidence, course, and etiology of CVS. CVS manifests itself with symptoms of recurrent (hours to days long) bouts of nausea and vomiting, separated by symptom-free periods (weeks to months). As an idiopathic disorder first mentioned in 1861 (French literature), CVS is no longer considered as a rare condition now.² It is known to be more common among children and females (girls > boys by a ratio of 60: 40) and has also been seen among adults.^{2,3} The reported prevalence of this disease is around 2% and it was also found to be 3.15 per 100 000 children in an Irish study in 2005.³

The research shows some connection between CVS and autonomic nervous system irregularities, gastrointestinal dysrhythmias, mitochondrial disorders, and hypothalamic-pituitary-adrenal activation.⁴ Furthermore, associations of estrogen sensitivity (among girls during menstruation), food allergies (e.g. cow's milk, soy, egg white protein), and migraines with CVS have been established.^{5,6} Moreover, CVS is usually triggered by physical and/or psychological stressors.⁵ Similar to the causes of abdominal migraine, chocolate, cheese, and monosodium glutamate can also trigger an episode of CVS; however, abdominal migraine less likely manifests itself with vomiting.⁷ Some other triggers could be sinus or respiratory infections, motion sickness, and hot weather.⁷ Even though CVS symptoms are episodic, they

can lead the children and adolescents to miss the school days, as well as causing an average annual healthcare cost of ~\$17 000 per individual, which can be a significant economic burden on the sufferer as well as on the healthcare system.⁸

The diagnostic criteria for CVS as described by the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) comprise of having at least 5 attacks in any timeframe or 3 attacks in a 6-month period in a stereotypical pattern, without any connection to another disorder. The attacks are defined as episodes of severe nausea and vomiting (≥ 4 times/hour) lasting from one hour to 10 days with an at least 1 week interval. This period is followed by an asymptomatic period with normal health parameters.⁵ It is an adoption of the Rome III criteria mentioned in the study of Haghghat et al.¹ Diagnosis of CVS can be challenging since the sufferers can display symptoms that are common to some other disorders including gastrointestinal, neurological, renal, metabolic/endocrinological, mitochondrial, and even psychological disorders.⁹ A list of some differential diagnoses based on the reviewed literature is shown in Table 1.^{4,5}

Haghghat et al describes the effect of erythromycin in the improvement in gastric motility that can be beneficial as an abortive therapy during prodromal phases of CVS.¹ In another study, we saw the role of antimigraine medications such as amitriptyline and cyproheptadine as effective prophylactic treatments in managing CVS as well.¹⁰ NASPGHAN review suggests cyproheptadine or propranolol prophylaxis for children aged 5 or younger and amitriptyline or propranolol for over 5 year-old individuals.³ In acute phase, symptomatic and supportive treatment with medications such as lorazepam,

Table 1. Some Differential Diagnoses Associated with the Cyclical Vomiting Syndrome

Gastrointestinal disorders (including autoimmune and congenital disorders)	<ul style="list-style-type: none"> • Bowel obstruction • Intermittent volvulus • Internal hernias (e.g. <i>Bochdalek hernia</i>) • Intestinal lymphangiomas • Duodenal webs and atresias • Peptic ulcer disease • Crohn's disease • Pancreatic disease (pseudocyst, inflammation) • Functional gastrointestinal Disorders (dysmotility, hypomotility) • Abdominal migraine
Brain and neurological disorders	<ul style="list-style-type: none"> • Space occupying lesions • Epilepsy • Vascular disorders/malformations • Migraine
Endocrine disorders	<ul style="list-style-type: none"> • Pheochromocytoma • Addison disease • Diabetes and related complications
Metabolic/mitochondrial disorders	<ul style="list-style-type: none"> • Inborn errors of metabolism • Mitochondrial encephalomyopathy • Amino acid, fatty acid, urea cycle defects
Urological/renal disorders	<ul style="list-style-type: none"> • Pelvi-ureteric junction obstruction • Nephrolithiasis
Psychiatric disorders	<ul style="list-style-type: none"> • Munchausen's syndrome • Bulimia

ondansetron, promazine, and diphenhydramine is indicated in both NAPGHAN and the study of Haghghat et al.^{1,5} Supportive treatment with 10% Dextrose and 0.45% Sodium Chloride solution is also recommended especially during acute attacks to avoid dehydration and metabolic irregularities coupled with the antiemetic medicines, as well as intravenous administration of nutritional supplements with minimal or no oral intake.^{1,5} In addition to the pharmaceutical interventions, lifestyle modifications are also recommended to avoid or control the CVS attacks.⁴ Some examples of lifestyle modifications include adopting good sleep hygiene; avoiding triggering food items such as chocolate, cheese, caffeine, monosodium glutamate; counseling/support to avoid excessive emotional excitement; and finally adopting balanced dietary and physical activity habits.^{5,6}

In summary, CVS is a complex disorder that could be a diagnostic challenge for the health care practitioners necessitating to rule out other potentially serious pathologies. Inaccurate or delayed diagnosis can be detrimental for the patients both physically and financially. Furthermore, ruling out other diseases through diagnostic procedures and tests can be costly for the health care system. It is also crucial to explore family and personal history of migraine while case management, as children with migraine or a family history of it are more likely to develop CVS.⁷ Finally, the prognosis of CVS varies case by case, that is, while some children recover from it, others continue to suffer throughout their adulthood, and some develop migraine.

Ethical Approval

Not applicable.

Competing Interests

Authors declare no competing interests.

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