

## Masqueraders of GERD

### Scope of the Problem

Gastroesophageal reflux (GER) and gastroesophageal reflux disease (GERD) are common in children and may be incompletely responsive to standard medical therapy. When this clinical scenario develops, other diagnoses must be considered. Clinical experience and the advent of endoscopy have identified a number of different diseases presenting with complaints formerly thought to fall under the umbrella of gastroesophageal reflux disease (GERD). These "masqueraders of GERD" include:

- eosinophilic esophagitis
- food allergic diseases
- achalasia
- cyclic vomiting syndrome
- rumination syndrome

The importance of recognizing these conditions lies in the fact that they require therapeutic measures different than those used for GERD.

### Eosinophilic Esophagitis (EoE)

#### DEFINITION:

Eosinophilic esophagitis (EoE) is characterized by upper gastrointestinal symptoms and dense esophageal eosinophilia, both of which are unresponsive to acid blockade.

#### CLINICAL FEATURES:

In the young child, symptoms include feeding refusal, vomiting and abdominal pain. Whereas in the teenager, dysphagia and food impaction are common. A personal or family history of allergic diseases or peripheral eosinophilia can be absent but is seen in 75% of patients. Upper gastrointestinal series may reveal isolated strictures or longitudinal narrowing as a manifestation of longstanding inflammation. Proximal strictures are seen more often than distal strictures. Abnormal features seen at upper endoscopy include whitish exudates, circular rings, longitudinal furrows, and splitting of the mucosa.

#### DIAGNOSIS:

Mucosal biopsy is required to make the diagnosis of EoE. The histological hallmark of the disease is a markedly increased number of eosinophils (>20/HPF) found in the squamous epithelium despite at least two months of proton pump inhibition. The gastric and duodenal mucosa are normal. Other causes for esophageal eosinophilia include GERD, hypereosinophilic syndrome, inflammatory bowel disease, parasitic infection, eosinophilic gastroenteritis, food/drug allergy and collagen vascular diseases.

#### TREATMENT:

Dietary elimination of specific food allergens or the use of an elemental diet (amino acid based formula) is very effective and can be used for both



induction and maintenance treatment. Potential problems are related to non-compliance.

Other off label treatment is directed at reducing allergic inflammation with corticosteroids (systemic or topically - *swallowing* rather than inhaling the dose delivered by a metered actuation).

When food/foreign body impactions are encountered endoscopic disimpaction with or without esophageal dilation may need to be performed.

#### PROGNOSIS:

The natural history of EoE is unknown. Reports to date document the presence of esophageal strictures in children and adults with EoE. No cases of carcinoma have been reported to date.

### Food Allergies

#### DEFINITION:

Mechanisms of food allergies include both non-IgE mediated inflammation (gluten intolerance), and IgE mediated reactions (peanut anaphylaxis). In some instances, mixed reactions take place (eosinophilic gastrointestinal diseases). Symptoms are relieved by the removal of the triggering food and reappear with the ingestion of the antigen.

#### CLINICAL MANIFESTATIONS:

Children with food allergies masquerading as GERD may present with vomiting and abdominal pain often associated with other complaints such as hives, rash, wheezing, edema, bleeding or diarrhea. When esophageal inflammation is present,



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manifestations may be related to esophageal dysmotility with dysphagia, food impaction or feeding refusal (see EoE above). In patients with small intestinal and/or colonic inflammation, diarrhea, bleeding or protein losing enteropathy can occur.

#### DIAGNOSIS:

The diagnosis for food allergies lies in an excellent history and physical examination. Radioimmunosorbent testing (RAST) or skin prick can be abnormal for suspected foods. An elimination trial with or without re-challenge should also be considered. If vomiting is a key symptom on presentation an upper gastrointestinal series may reveal mucosal thickening. Endoscopic examination can detect ulcers, polyps and inflammation.

#### TREATMENT:

Food elimination is the treatment of choice for food allergic diseases.

#### PROGNOSIS:

The prognosis of food allergic diseases depends on the allergen. Cow's milk allergy in infancy is usually outgrown by one year of age however, peanut allergy can be lifelong.

## Achalasia

#### DEFINITION:

Achalasia is a motor disease that leads to abnormal esophageal peristalsis and a functional obstruction of the distal esophagus from failure of the lower sphincter to relax.

#### CLINICAL MANIFESTATIONS:

Typically, achalasia is a condition seen in adults but can present during childhood (mean age around 9 years). Symptoms include vomiting, dysphagia, food impaction, weight loss/failure to thrive, nocturnal regurgitation, chest pain, cough and recurrent pneumonia.

#### DIAGNOSIS:

Esophageal manometry remains the gold standard test for the diagnosis of achalasia. Suggestive abnormalities include a widened mediastinum and air-fluid level on plain chest radiograph, proximal esophageal dilation and "beaking" on the barium swallow study;

#### TREATMENT:

Mechanical dilation is accomplished by esophageal balloons or bougienage. The injection of the LES with *Botulinum* toxin temporarily relieves tonic contraction. Esophageal myotomy is a permanent solution but carries GERD as a potential complication so fundoplication may be performed prophylactically.

#### PROGNOSIS:

Patients with achalasia can redevelop obstructive symptoms even after effective treatment. This can be on the basis of recurrent achalasia or acid-related stricture.

## Cyclic Vomiting Syndrome

#### DEFINITION:

Cyclic Vomiting Syndrome (CVS) is characterized by recurring bouts of intense vomiting, with or with-

out abdominal pain, interspersed with episodes of returning to a normal state of health.

#### CLINICAL FEATURES:

CVS can develop in young children, typically around age 5 or 6 years. The vomiting is intense and protracted, often 6 times per hour. Vomiting can be associated with abdominal pain, preceding and/or accompanying the episodes of vomiting. The abdominal pain may also be associated with nausea. However, the abdominal pain and/or nausea are not always associated with the vomiting episodes in all patients. These "stereotypical" episodes can last for hours or days. The same pattern often repeats with each attack. The attacks usually stop abruptly and the child quickly resumes normal diet and activities. The bouts can occur during intercurrent infections, menstruation or at times of excitement or stress. The impact on family life and the child's daily functioning and quality of life can be significant. While both GER and CVS exhibit vomiting, the vomiting in the former is milder and more constant, not as debilitating and disruptive as in CVS.

#### DIAGNOSIS:

The causes of CVS are not known. There is often a positive family history for migraines.

When doing a differential diagnosis it is crucial to exclude raised intracranial pressure from any cause. Chronic sinusitis is sometimes implicated. Rare conditions such as midgut volvulus, ureteral pelvic obstruction and metabolic causes such as mitochondrial disease or urea cycle abnormalities can present with bouts of intermittent vomiting.

#### TREATMENT:

The mainstay of CVS management is prevention of suffering, dehydration, and ketosis once the attack begins. Benzodiazepines and ondansetron may be useful adjuncts. Prophylaxis with cyproheptadine, amitriptyline or propranolol may be effective in some patients. Recognizing the pattern of the vomiting and confirming the diagnosis of CVS is extremely important and provides reassurance, even though the attacks might not disappear. Management can be more focused and measures can be taken to prevent, anticipate, or effectively treat breakthrough attacks.

#### PROGNOSIS:

Cyclic Vomiting Syndrome resolves in most children and many children will go on to develop more typical migraines by adolescence.

## Rumination Syndrome

#### DEFINITION:

Rumination Syndrome consists of chronic regurgitation of partially digested food either ejected or re-swallowed.

#### CLINICAL FEATURES:

Rumination syndrome has long been observed in infants and young children with developmental delays or in those institutionalized or understimulated. In this setting, it has been considered a form of self-stimulation. However, more recently, otherwise normal adolescents and adults have

been described who exhibit the same clinical pattern. This occurs more often in females, but it is not considered an eating disorder. Most individuals with regurgitation syndrome, regurgitate with every meal. Typically, the regurgitation is effortless and within 10-20 minutes. The patient may exhibit halitosis or complain of a sour taste rather than discomfort or typical peptic symptoms.

#### DIAGNOSIS:

A careful history and identification of the recurrent, effortless, painless regurgitations is often sufficient to make the diagnosis. Observation of preceding air swallowing and contraction of the abdominal wall (while the lower esophagus and glottis relax) can clinch the diagnosis without need for involved and often unnecessary investigations. If available, antro-duodenal manometry will document the presence of the diagnostic pressure pattern confirming the creation of a "common cavity" between the stomach and the mouth.

#### TREATMENT:

Behavioral treatments are usually employed. In infants, this involves developmental stimulation. In adolescents and adults, teaching patients diaphragmatic breathing and other behavioral techniques seems effective in some, while the rumination can be stubborn and difficult to dissipate in others. Determination of any underlying psychiatric disorders and directing appropriate medical and psychological therapy is important in achieving successful treatment.

#### PROGNOSIS:

The recently reported Mayo Clinic experience suggests a favorable prognosis in most.



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