

PREPLADDER

NEET-SS

PEDIA

VOLUME -2

## INDAEX

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# APPROACH TO VOMITING IN CHILDREN



## Q. What is Vomiting?

00:01:01

- Vomiting refers to a **highly** coordinated reflex process characterized by the active movement of gastric contents from the stomach into the esophagus.
- Vomiting is often preceded by **increased salivation and involuntary retching**.
- There is a violent ascent of the diaphragm, constriction of the abdominal muscles, and the relaxation of the gastric cardia, thus producing vomiting.
- The vomiting center is present in the medulla oblongata.
- Medulla oblongata is affected by various direct factors such as toxins, or it may be affected by a rising from a specific area called a chemoreceptor trigger zone (CTZ).
- Medulla is also affected by cortical sensations, including **psychic and visual sensations**.

## Vomiting Versus Regurgitation

- Regurgitation refers to the passive movement or expulsion of gastric contents into the esophagus.
- No nausea, no increased salivation, no distress, or other autonomic symptoms exist in regurgitation.
- The child remains comfortable immediately afterwards.
- In regurgitation, the child becomes hungry soon after the episode.

## Causes of Vomiting in Children

### Infancy

- Common causes include acute gastroenteritis.
- It includes overfeeding and GERD.
- Anatomic GIT obstructions include CHPS, Malrotation, Intussusception, Hernias, and Adhesions.
- It can be produced by systemic infections such as sepsis and meningitis.
- It can be seen due to pertussis and acute OM.
- Rare causes include inborn errors of metabolism, brain tumors, subdural hemorrhage, food poisoning, renal tubular acidosis, uretero pelvic junction obstruction, pseudo-obstruction, etc.

### Childhood

- The common causes include acute gastroenteritis or acute gastritis.
- Drug-induced vomiting, along with GERD.
- Anatomic GIT obstruction includes CHPS, Malrotation Intussusception, Hernias, and Adhesions.
- Systemic infections like sepsis and meningitis can be there.
- Pertussis, Acute OM and sinusitis may also be found.
- Eosinophilic esophagitis may also be there.

- Rare causes include Reye's Syndrome, Brain tumors, Pancreatitis, Hepatitis, IEM, Food Poisoning, RTA, Pseudo-obstruction, and Gastroparesis.

## Complications of Vomiting

- Metabolic
  - Metabolic complications will include loss of gastric juice which can produce hypochloremic alkalosis, and loss of potassium can cause hyponatremia and hypokalemia.
- Protein Energy Malnutrition and Failure To Thrive.
- Mallory-Weiss Tears and Hematemesis.
  - The most common site is the lesser curvature of the stomach adjacent to the gastroesophageal junction.
- Boerhaave Syndrome
  - There is a rupture of the esophagus in this syndrome.
  - It is less commonly seen in children.
- Esophagitis.
- Aspiration
  - Aspiration results in pneumonitis in the patients.
- Shock.
- Pneumothorax And Pneumomediastinum.
- Petechiae And Retinal Haemorrhage.

## Management of Vomiting

- Treat the cause, for example:
  - Relieving GIT obstruction.
  - Antibiotics will be needed for Infection.
  - PPI will be needed for reflux.
- Management Of Complications.
- Specific anti-emetics will be needed to stop the vomiting.
  - The first line drug for Infection associated vomiting and cancer chemotherapy-induced vomiting is always **Ondansetron** with a dose of about 0.1-0.3 mg/kg.

## Antiemetics in Selected Conditions

- Prokinetics, specifically Dopamine D2 receptor antagonists, should be given if a patient has reflux-associated vomiting.
- Domperidone is preferred over metoclopramide because domperidone does not cross the blood-brain barrier, and thus it is better tolerated.
- If a patient has Gastroparesis, then Dopamine D2 receptor antagonists are preferred, and if the dopamine D2 receptor is absent, erythromycin can be given, which acts as a motilin receptor.
- The drug of choice in cancer chemotherapy-induced vomiting includes
  - Serotonergic 5-HT<sub>3</sub> antagonist such as Ondansetron which is considered the first-line drug.

- Dopamine antagonist includes prochlorperazine.
- Steroids can also be used, such as Dexamethasone, and cannabinoids, specifically Tetrahydrocannabinol.
- The post-operative choice will include Ondansetron.
- In case of motion sickness, and vestibular disorders, antihistamines such as dimenhydrinate and anticholinergics such as **scopolamine** can be used.
- In case of an adrenal crisis, cortisol can be given.

#### Approach to a Vomiting Child

00:16:32

To examine a vomiting child, a detailed history is taken, which includes several points to focus on

- Whether it is vomiting or regurgitation.
- If it is vomiting, looking at a child's age is important.
- Then, the frequency of vomiting to know about the episodes of vomiting that have happened so far.
- To ask whether it is bilious or non-bilious.
- To ask whether it is projectile or not.
- To know about the contents of the vomitus.
  - If food particles consumed 6 hours back come in vomit, there is a chance of Gastroparesis or obstruction.
  - If blood in the vomiting appears, then there can be hematemesis.
- Suppose the frequency is more than two times. In that case, a patient should be enquired about any associated symptoms like oliguria, fever, CNS Symptoms, recent outside food intake, and drug history.
- In the examination, always focus on the vitals and signs of dehydration.
- If signs of dehydration are present, a patient should focus on hydration therapy and anti-emetics.
- If signs of dehydration are absent, oral hydration will be given if the child can tolerate it orally, and the child should be investigated.

#### Extra-Edge: Red-Flag Signs in Vomiting

- Blood or bile in the vomitus.
- Severe abdominal pain and distention.
- Projectile Vomiting.
- Persistent tachycardia or hypotension.
- Photophobia or neck stiffness.
- If any of these signs are present, they should be considered in hospital management and workup.

#### Functional Nausea

All of the following criteria should be fulfilled over the last 2 months

- There should be bothersome nausea as the predominant symptom, which occurs more than 2 times per week and is generally unrelated to meals.
- It should not be consistently associated with vomiting.
- After appropriate evaluation, another medical condition cannot fully explain nausea.

- Cyproheptadine is effective in some cases of functional nausea.

#### Functional Vomiting

All of the following criteria should be fulfilled

- Vomiting episodes more than 1 time per week.
- Absence of self-induced vomiting or an eating disorder, or rumination.
- After appropriate evaluation, another medical condition cannot fully explain the vomiting.

#### Cyclic Vomiting Syndrome (CVS)

00:23:00

- It is an idiopathic disorder characterized by repeated attacks or cycles of intense vomiting more than 4 times per hour with eventual resolution between attacks.
- Some have monthly attacks, while others occur every 3–4 months.
- Each attack lasts about 24–48 hours and responds to hydration.
- Clinical suspicion should happen if **5 episodes of vomiting or at least 3 episodes have occurred in the last 6 months**.
- It affects 2% of Caucasians and affects females rather than males.
- A family history of migraine or motion sickness happens in 30–40% of cases.
- The median age of onset is about 5 years.
- Most children outgrow the problem by the adolescent period but may develop **migraine** in later life.

#### Diagnostic Criteria for CVS

- Episodic attacks are more than 2 times or more of intense nausea and paroxysmal vomiting lasting hours to days within 6 months.
- Stereotypical pattern and symptoms in the individual patient.
- Episodes are separated by weeks to months.
- There is a return to baseline between the episodes.
- It is not attributable to another disorder.

#### Possible Etiological Factors

- An over-responsive HPA axis, including the role of CRH.
- Autonomic dysfunction with sympathetic overactivity.
- Mitochondrial dysfunction.
- Role of nuclear mutations.
- It may share features with cannabis-induced hyperemesis syndrome.

#### Management of CVS

- Early aggressive hydration shortens the acute attacks with a fluid of choice, dextrose in normal saline (DNS).
- Abortive medications must be given, which is Sumatriptan of 20 mg intranasally. C/I – Basilar Artery Migraine.

- Supportive or add-on medications include
  - Ondansetron or diphenhydramine.
  - Ketorolac as analgesic.
  - Consider PPIs for reflux or acute epigastric pain.
- Manage complications.
- Resume enteral nutrition as soon as possible. but parenteral nutrition should be considered if enteral feeds can't be started in 3-4 days.

#### **Prophylaxis of CVS**

- Avoid potential triggers. For example, fasting or anxiety or exhaustion or sleep deprivation, etc.
- Avoid skipping meals or caffeine intake.

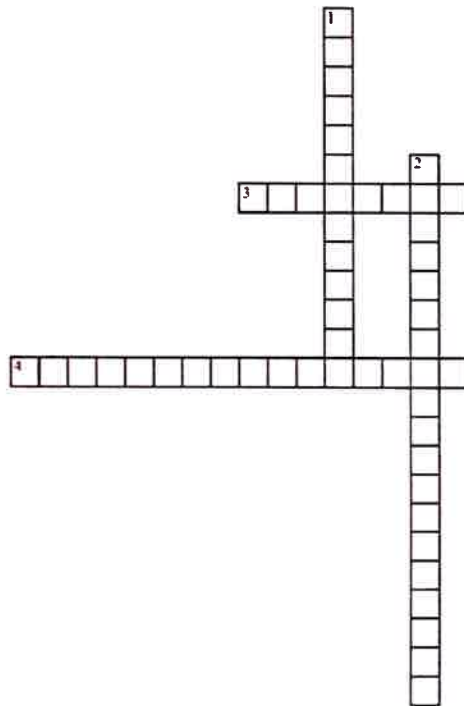
- Prophylactic medication can be used.
  - An age less than 5 can consider **Cyproheptadine** followed by **Pizotifen** or **Propranolol**.
  - Prophylaxis for those above 5 years of age is **Amitriptyline**, followed by **Propranolol** also **Phenobarbitone**.
- **Topiramate** is helpful in debilitating and refractory long-standing cases.
- Dietary supplements were found to be useful adjuncts, such as L-Carnitine and Coenzyme Q-10.
- For Catamenial CVS (menstruation-related): OCPs can be considered.



# CROSS WORD PUZZLES



Crossword Puzzle 1



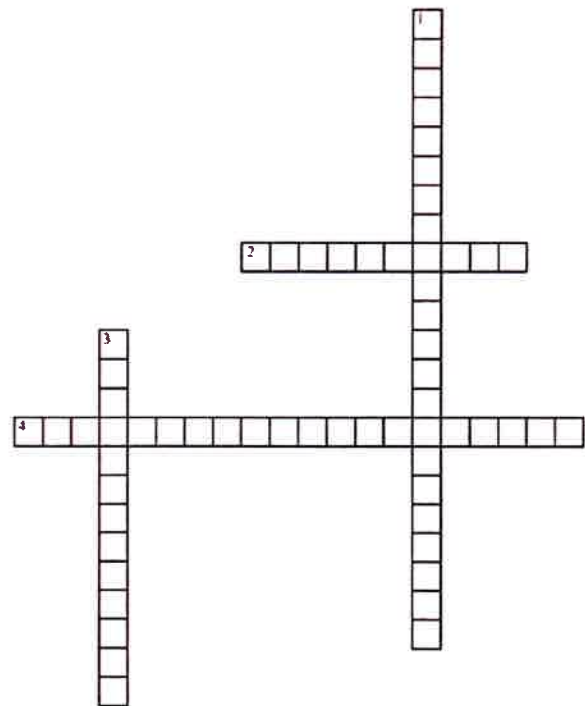
## Across

3. \_\_\_ refers to a highly coordinated reflex process characterized by the active movement of gastric contents into the esophagus.
4. The vomiting center is present in \_\_\_.

## Down

1. \_\_\_ refers to passive movement or expulsion of gastric contents into the esophagus.
2. Vomiting is often preceded by increased salivation and \_\_\_.

Crossword Puzzle 2



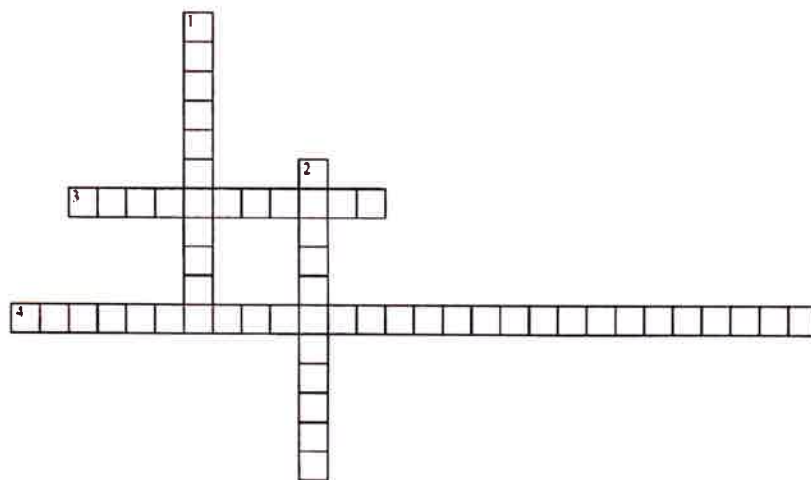
## Across

2. \_\_\_ is helpful in debilitating and refractory long-standing cases.
4. Early aggressive hydration shortens the acute attacks with a fluid of choice is \_\_\_.

## Down

1. \_\_\_ is an idiopathic disorder characterized by repeated attacks or cycles of intense vomiting.
3. If food particles consumed 6 hours back come in vomit, there is a chance of \_\_\_.

### Crossword Puzzle 3



#### Across

3. \_\_\_\_ is preferred over metoclopramide.
4. If a patient has Gastroparesis, then \_\_\_\_ is preferred.

#### Down

1. First-line drugs for Infection associated vomiting and cancer chemotherapy-induced vomiting are always \_\_\_\_.
2. In regurgitation, the child remains \_\_\_\_.



# FOOD ALLERGY AND ADVERSE REACTION TO FOODS



## What are Adverse Reactions to Foods?

00:00:26

- According to Nelson, an adverse reaction to food is an untoward reaction provoked by ingesting food particles or additives.
- There are mainly 2 types of food reactions:
  - Physiological reactions - Food intolerance, i.e., lactose intolerance, can be called food intolerant.
  - **Pathological reactions: immune-mediated reactions** - Any type of food allergy is called a pathological allergy. Allergy to fish, eggs, and others will be compiled into it.

## Classification of Adverse Reactions to Food

### A. Food Intolerance (Non-Immune)

- They can be broadly classified into 2 types

#### 1 The first one is related to host factors.

- Enzyme deficiencies: lactase deficiency, sucrase/isomaltase deficiency, hereditary fructose intolerance, and galactosemia.
- **GIT disorders are IBD, IBS, and pseudo-obstruction.**
- **The idiosyncratic reaction can also be a part of it.**
- **When certain children take caffeinated beverages can produce hyperactivity, which lasts as long as caffeine in the body.**
- Psychological deficiencies like food phobias and OCD.
- Migraine can also cause food intolerance in people.

#### 2 Some factors are related to food factors.

- Infectious organisms like E. coli, staph aureus, Cl perfringens, shigella, salmonella, botulism, yersinia, and Campylobacter.
- Toxins are like histamines (scombroid poisoning) and saxitoxin (shellfish poisoning).
- Drugs like caffeine, tyramine (cheese), theobromine (chocolate, tea), tryptamine (tomatoes), and benzoic acid in citrus fruits (perioral flare).
- Contaminants: Heavy metals, Antibiotics, Pesticides.

### B. Food allergy

- The food allergy can be either IgE-mediated or non-IgE mediated.

#### In IgE-mediated,

- Cutaneous- urticaria, angioedema, morbilliform rashes, flushing.
- They can be in the form of gastrointestinal reactions like oral allergy syndrome and GIT anaphylaxis.

- **Perioral and lip-swelling, gum-swelling, which can further develop into angioedema, can be seen in the Oral allergy syndrome.**
- **Respiratory features that include acute rhinoconjunctivitis and bronchospasm.**
- **It includes generalized food allergies, which are anaphylactic shock, exercise-induced anaphylaxis**

### Mixed Non-IgE as well as IgE-Mediated Forms

- They have cutaneous-like atopic dermatitis and contact dermatitis.
- **GIT: Allergic eosinophilic, esophagitis, and gastroenteritis.**
- **And respiratory, like asthma.**

### Non-IgE mediated

- **Are mediated by T-cells.**
- Cutaneous like contact dermatitis, dermatitis-herpetiformis.
  - GIT like celiac disease, food-protein induced enterocolitis, and proctocolitis.
- Respiratory like food-induced pulmonary hemosiderosis, Heiner syndrome.

### Epidemiology

00:11:21

- Worldwide estimates of food-allergy prevalence are 1-10%
- Up to 6% of children experience food allergy in the 1st 3 years of the life
- Cow-milk allergy is estimated to be up to 2.5%
- **Peanut allergy is estimated to be up to 2-3%**
- **Egg allergy is estimated to be up to 2%**
- Most children "outgrow" milk and egg allergies approx 50% by school-age
- In contrast, 80-90% of children with peanut or seafood allergies retain their allergy for life

### Genetic Factors in Food Allergy

- Positive family history has up to 2-10 fold higher risk
- **Association with HLA-DQ variants, especially differential methylation at the HLA-DR and -DQ regions**
- **Implicated genes are (I) filaggrin, (II) IL-10, (III) STAT6 (IV) FOXP3**
- The Filaggrin gene has been implicated in both atopic dermatitis and food allergy in children
- DNA methylation effects on interleukins 4, 5, and 10 and IFN- $\gamma$  genes and in the MAP-kinase pathways have also been implicated

## Pathogenesis of Food Allergies

- Different for IgE-mediated and non-IgE mediated
- In patients who have IgE-mediated allergies, in the 1st few years of life, there will be exposure to the allergen which will lead to the production of IgE antibodies
- These antibodies will get attached to the surface of mast-cells and basophil cells by Fcε receptors
- This initial phase is called priming/sensitization
- Whenever the allergen comes into contact, there will be a rapid release of inflammatory mediators from the mast cells
- These mediators will directly cause systemic effects as well as these mediators will also recruit the inflammatory cells they will get involved in creating food allergies
- The non-IgE mediated allergies occur by a cell-mediated immune response
- T-lymphocytes do them
- There is an undefined response of innate immune disorder which also contributes to the development of reaction

## Types of Food Allergens in IgE Mediated Food Allergies

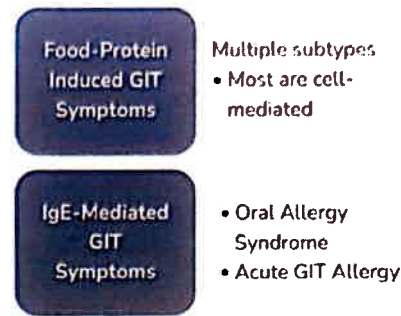
- Class 1: those penetrating GIT barrier
- Any food may serve as a class 1 food allergen, but an egg, milk, peanuts, tree nuts, fish, soya, and wheat account for 90%
- Cell-mediated sensitivity develops only in class 1 IgE-mediated food allergies
- Class 2 allergens are those that are partially homologous to plant pollen penetrating the respiratory tract
- They are typically vegetables, fruit, or nut protein that are partially homologous with pollen proteins
- Cell-mediated sensitivity does not develop to this class of allergens
- Nelson says, Intermittent ingestion of allergenic foods causes acute symptoms such as urticaria or anaphylaxis
- Prolonged exposure to allergenic foods can cause chronic disorders such as atopic dermatitis and asthma

## Clinical Manifestations

00:19:13

### A. GIT Manifestations

- Often the 1st form of food allergy in infants and young children
- Cell-mediated mechanisms are more common than IgE-based
- Manifest as vomiting, irritability, diarrhea, and FTT
- Broadly classified into 2 categories



## Food Protein-Induced GIT Symptoms

They are mainly of 4 types

1. FPIES = food protein-induced enterocolitis syndrome
  - In 1st few months, it will have vomiting/irritability/ diarrhoea/ abdominal distension
  - Cows milk protein or soya-based formula is the cause of the majority of these allergens
  - Nelson says -About 15% of these children may tend to develop hypotension
  - Most of them improved by 3-5 years of age
2. FPIAP = food protein-induced allergic proctocolitis
  - These children with features like diarrhoea/vomiting/ abdominal distension will have it in the 1st few months of life
  - 60% of them will have a breast milk allergy, and the other 40% is either due to cow's milk protein or soya-based formula
3. FPE = food protein-induced enteropathy mainly due to cow's milk protein allergy, the most severe form of FPE is celiac disease
4. Eosinophilic gastro enteropathy has two types: eosinophilic esophagitis and eosinophilic gastroenteritis

## Ig E-Mediated Symptoms

00:25:19

### 1. Oral Allergy Syndrome

- Many patients are associated with pollen-induced allergic rhinitis
- Oral pruritus, tingling, and angioedema of the lips, tongue, palate, and throat; pruritus in the ears and tightness in the throat
- Short-lived symptoms can be precipitated by raw fruits and vegetables

### 2. Acute GIT Allergy

- Acute abdominal pain, vomiting, or diarrhea when IgE-mediated allergic symptoms develop in the target organs

### B. Skin Manifestation

- Also common in infants and young children.
- Associated with atopic dermatitis. According to Nelson, at least 30% of children. With moderate to severe AD, have food allergies.

- The most common symptoms are acute urticaria and angioedema.
- Most are IgE-mediated allergies.
- Food most commonly incriminated includes eggs, milk, and peanuts.

### C. Other Systemic Manifestation

- Respiratory and food-induced rhinoconjunctivitis symptoms typically accompany allergic symptoms in other target organs, such as skin.
- Wheezing occurs in approximately 25% of IgE-mediated food allergic reactions
- Anaphylaxis is a serious, multisystem allergic reaction that is rapid in onset and potentially fatal.

### Diagnosis

00:29:50

- A thorough history and examination are critical to elucidate the diagnosis and precipitating food component.
- Enquire for food consumed and quantity, the time interval between ingestion and onset of symptoms.
- Types of symptoms elicited, whether the history of similar symptoms on ingestion
- Any other inciting factors like exercise and any family history of atopy

### Investigation Performed

- Skin prick tests to detect sensitization that is food-specific IgE antibodies
- A negative skin prick test excludes IgE-mediated food allergy
- A positive test needs further evaluation
- Total IgE levels
- No tests for cell-mediated food allergens
- Food eliminations and food challenge tests performed in such patients



### Important Information

- Serum food-specific IgE levels
  - $\geq 15 \text{ Ku}_A/\text{L}$  for milk ( $\geq 5 \text{ Ku}_A/\text{L}$  for children  $\geq 1$  year)
  - $\geq 7 \text{ Ku}_A/\text{L}$  for egg ( $\geq 2 \text{ Ku}_A/\text{L}$  for children  $< 2$  years)
  - $\geq 14 \text{ Ku}_A/\text{L}$  for peanuts
  - Elimination time to perform food challenge test: 10-14 days for IgE-mediated food allergies and up to 8 weeks for cell-mediated
- ### Treatment
- Ideal treatment is to identify and eliminate the offending food agents.

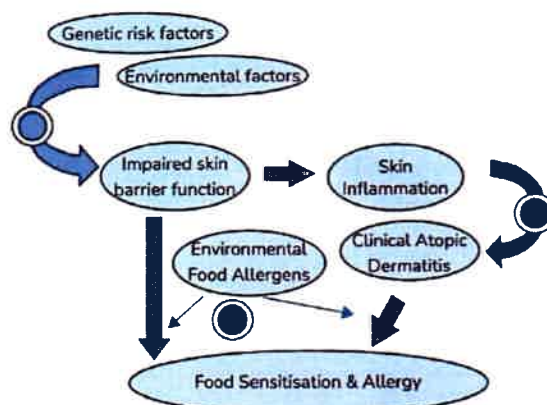
- Re-evaluate periodically if still allergic or losing reactivity with age.
- Trials like immunotherapy with omalizumab (Anti Ig E MONOCLONAL Ab) can help.
- Many milk or egg-allergic children better tolerate baked products with extensively-heated milk and egg products.
- Teach emergency management for accidental ingestion.

### Prevention

- Exclusive breastfeeding for 6 months.
- Introduce complementary feeds after 6 months. Start with low-risk complementary foods, 1 at a time.
- Do not over-delay the introduction of potentially allergenic foods.
- Do not avoid allergenic foods during pregnancy or nursing.
- Soy-based formulas do not prevent allergic disease.

### Hot: Role of Skin Barrier in Food Allergies

00:38:47



### Potential Strategies for Preventing Food Allergies

- The key potential strategy which has been proposed now is to ensure that in infancy, the skin barrier remains intact
- For that the child must receive emollients as much as possible
- In addition, there is growing research on role in probiotics to ensure gut mucosal integrity as a way to prevent food allergies

### MCQ One-Liners

- Vaccines to be avoided in children with egg allergy
- The two vaccine are Absolute CI- yellow fever vaccine and relatively CI- rabies vaccine
- Age of resolutions of food allergies are
- Hen's egg white: 75% resolve by 7 years of age
- Cow milk: 76% resolve by 5 years of age
- Peanuts: only 20% resolves, rest are life-long allergic
- Fish and shellfish: persistent allergy throughout life
- Wheat allergy: (unrelated to celiac disease) 80% resolve by 5 years of age
- Cross-reactivity in Ig E-mediated food allergy

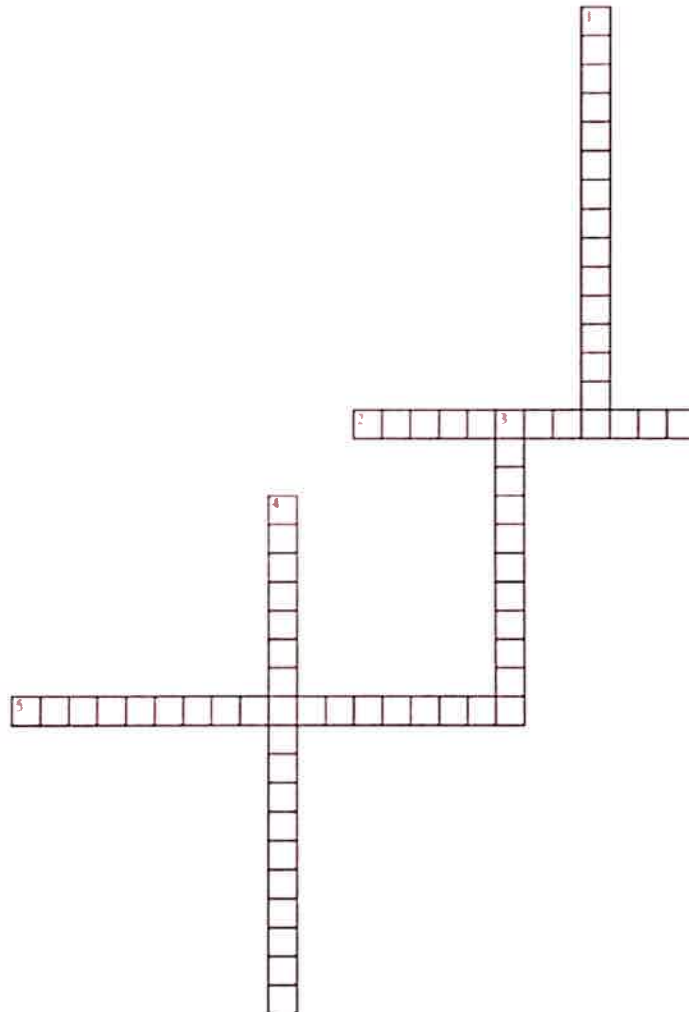
| Food Family                                               | Risk of Allergy to 1 member (%; approximate) |
|-----------------------------------------------------------|----------------------------------------------|
| Legumes                                                   | 5                                            |
| Tree nuts (e.g. almond, cashew, hazelnut, walnut, brazil) | 35                                           |
| Fish                                                      | 50                                           |
| Shellfish                                                 | 75                                           |
| Grains                                                    | 20                                           |
| Mammalian milks                                           | 90                                           |
| Rosaceae (pitted fruits)                                  | 55                                           |
| Latex-food                                                | 35                                           |
| Food-latex                                                | 11                                           |



# CROSS WORD PUZZLES



## Crossword Puzzle 1



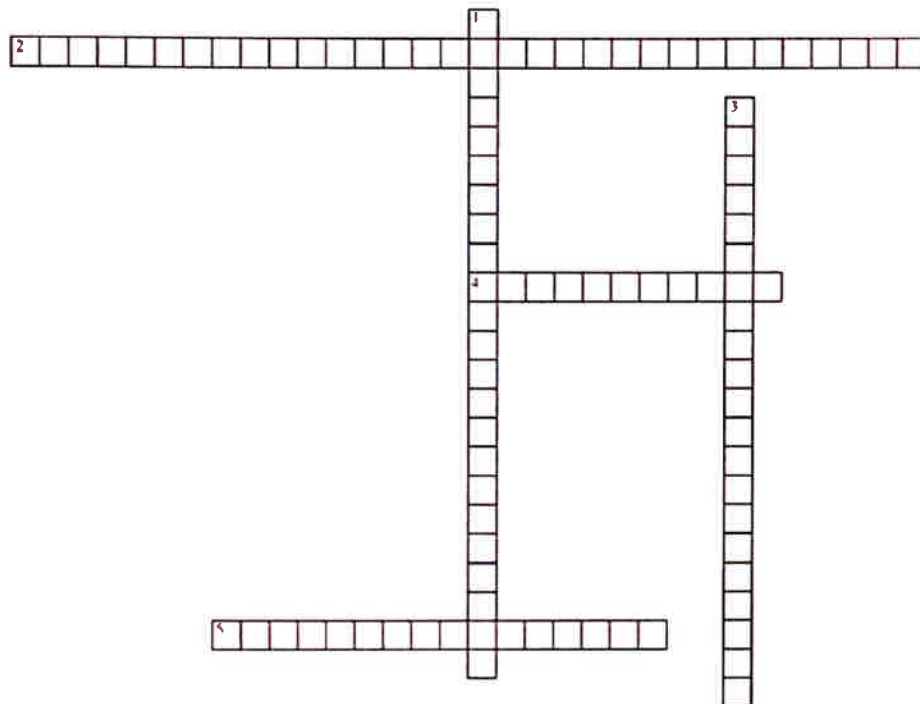
### Across

2. \_\_\_\_\_ (unrelated to celiac disease) 80% resolve by 5 years of age
5. \_\_\_\_\_ do not prevent allergic disease

### Down

1. \_\_\_\_\_ food-induced rhinoconjunctivitis symptoms typically accompany allergic symptoms in other target organs such as skin
3. \_\_\_\_\_ is defined as a serious, multisystem allergic reaction that is rapid in onset and potentially fatal
4. \_\_\_\_\_ vomiting, or diarrhoea when IgE-mediated allergic symptoms develop in the target organs

## Crossword Puzzle 2



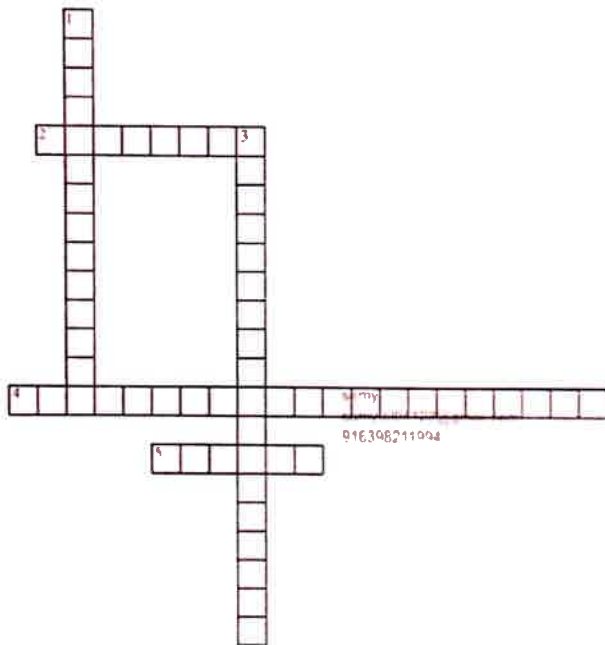
### Across

2. \_\_\_\_\_ are two types: eosinophilic esophagitis and eosinophilic gastroenteritis
4. \_\_\_\_\_ is defined as a serious, multisystem allergic reaction that is rapid in onset and potentially fatal
5. \_\_\_\_\_ According to Nelson, at least 30% of children with moderate to severe AD have food allergies

### Down

1. \_\_\_\_\_ is more common than IgE-based
3. \_\_\_\_\_ of allergenic foods causes acute symptoms such as urticaria or anaphylaxis

### Crossword Puzzle 3



#### Across

2. \_\_\_\_\_ can also cause food intolerance in people
4. \_\_\_\_\_ are also known as immune-mediated reactions
5. \_\_\_\_\_ are like histamines ( scombroid poisoning), and saxitoxin (shellfish poisoning)

#### Down

1. \_\_\_\_\_ that is lactose intolerance can be called food intolerant
3. \_\_\_\_\_ lactose deficiency, sucrase/isomaltase deficiency, hereditary fructose intolerance, and galactosemia



### Fact Based Questions

00:00:58

- The normal length of the esophagus at birth is 8-10cm.
- The normal epithelial lining of the esophagus is covered by **squamous epithelium**.
- Esophagus has two types of sphincters, the upper esophageal sphincter (UES) and the lower esophageal sphincter (LES).
- The UES is at the level of the **cricopharyngeus muscle**.
- The LES is at the level of the **gastroesophageal junction**.

### Tracheoesophageal Fistula (TEF)

00:02:54

- A **fistula** is an abnormal communication between two hollow structures.
- Here, there is communication between the trachea and the esophagus.
- Around 4-6 weeks of intrauterine life, the esophagus and stomach can be differentiated.
- Around this time, a tracheo-laryngeal groove separates the anteriorly placed trachea from the posteriorly placed esophagus.
- If there is any abnormality in the formation of this groove, there will be persistent or abnormal communication between the trachea and esophagus.
- Most fistulas develop between **4-8 weeks** of intrauterine life.
- Rarely, some fistulas tend to develop between **8-12 weeks**.

### Epidemiology

- It is the most common congenital anomaly of the esophagus.
- The risk factors are advanced maternal age, Europeans, obesity, and smoking.
- **Advanced maternal age increases the risk of Down's syndrome and TEF.**
- The East European states (Hungary, Poland, eastern Russian states, etc.) are the areas where there is the maximum incidence of TEF.
- In India, TEF occurs sporadically.
- It is related to obesity in the mother.
- Smoking is also implicated as one of the risk factors, particularly in the first trimester.
- 50% of the cases are found to be **syndromic**, and 50% of cases are isolated (non syndromic).
- When TEF is part of a particular syndrome, it is called syndromic.
- When there is TEF with no other genetic abnormalities is called nonsyndromic.
- Rarely, syndromic TEF is associated with mutations in N-myc, SOX-2, and CHD-7 genes.

- **CHD-7 gene is involved in CHARGE syndrome.**
- The **N-myc gene** is involved in **Feingold syndrome**.
- The most common syndrome associated with TEF is **VACTERL syndrome**.
- Individually, cardiac defects are seen in 38% of cases, followed by skeletal defects in 19%.

### Vacterl Syndrome

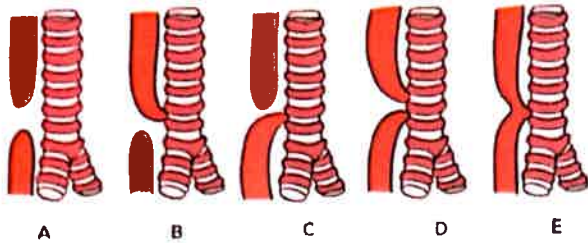
00:07:32

- V stands for Vertebral defects.
- A Stands for anorectal malformations.
- C stands for cardiac defects.
- T stands for TEF.
- E stands for esophageal atresia.
- R stands for renal anomalies.
- L stands for limb defects (radial limb defects).
- Atresia refers to the absence of lumen formation, the esophagus ending in a blind tube.
- Intelligence is always normal in VACTERL syndrome.

### Types of TEF

00:09:45

- It is classified based on gross appearance.
- It is classified as Gross Type A, B, C, D, and E.
- There is no visible fistula in Type A, but we find a blind-ending esophageal tube.
- Type A is pure esophageal atresia and is a rare form.
- In Type B, the upper end of the esophagus is attached to the trachea.
- In Type C, the upper end ends in a blind tube, and the lower end is attached to the trachea.
- Type C is proximal esophageal atresia with distal **TEF (TEF-EA)**.
- **Type C is the most common variety seen in more than 90% of the cases.**
- The upper and lower ends are attached to the trachea in Type D.
- In Type E, both ends of the esophagus join before attaching to the trachea.
- The difference between Type D and Type E is that Type D separately joins the trachea, whereas Type E forms a continuous tube and then joins the trachea.
- Type E TEF is also called **H Type TEF** due to the H-shaped structure being formed.
- H type is seen in **5-7%** of the cases; this variety has the late presentation.



This image depicts the types of TEF based on Gross Classification, where E represents the esophagus and T represents the Trachea.

#### Clinical Presentation of TEF

00:13:28

##### Features in Type C

- Type C is the most common TEF seen.
- When this TEF develops in-utero, the child cannot swallow the amniotic fluid, and normal cycling becomes disrupted.
- Antenatally- History of maternal polyhydramnios.
- Postnatally, it will be present in the neonatal period.
- The newborn cannot swallow his salivary secretion since the esophagus ends in a blind tube.
- So the saliva will come out and form frothy secretions drooling from the angle of the mouth.
- The hallmark clinical finding is frothy secretions drooling from the mouth.
- The newborn may aspirate the saliva into the airway, which may cause features of pneumonitis, or respiratory distress may be present.
- The onset of distress may either be at birth or delayed for some time.
- The lower part of the esophagus is attached to the airway, so whenever the child is breathing, some air is also going into the stomach.
- There is progressive abdominal distension found in these patients due to the air going into the stomach.
- In conclusion, the clinical features of TEF are frothy secretion, drooling from the mouth, Respiratory distress (mild to severe), and Progressive abdominal distension.
- These children may have aspirations from two parts.
- In the first part, it may be due to the pooling of secretions in the oral cavity, which may aspirate into the airway during the prone position.
- In the second part, when the child is breathing, all the air moves into the airway, and some also move into the stomach. The stomach becomes distended, and any undue pressure or manipulation of the child causes reflux. Reflux will move back into the airway, where the gastric contents aspirate into the airway.

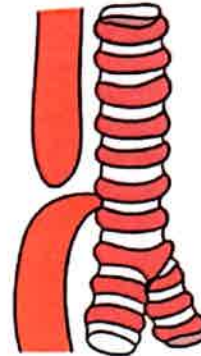


Image showing Type C fistula.

- Most of these children, if left unattended, develop features of pneumonitis.
- Type C, D, and E will have abdominal distension because the distal esophagus is attached to the trachea.
- Type A and B will not have abdominal distension as the distal esophagus is not attached to the trachea.

#### Features in Type H TEF/ Type E Gross

00:18:37

- These children will not have frothy secretion from the mouth because both ends of the esophagus are joined (no esophageal atresia).
- They present in late infancy or early childhood.
- They present as recurrent pneumonia, failure to thrive, and episodes of bronchospasm.

#### Diagnosis of TEF

00:19:46

- The neonate has frothy secretion, with or without abdominal distension and with or without respiratory distress.
- History of worsening on the attempt to feed the child.
- The milk will not go into the esophagus because the esophagus ends in a blind tube.
- It will come out or regurgitate and may be aspirated into the airway, producing aspiration.
- Polyhydramnios will also be present in the history of antenatal.
- These features raise suspicion of TEF.
- The screening test will be performed.
- A soft feeding tube is taken and is tried to pass slowly from the mouth to the esophagus.
- The tube will go up to a certain level and get coiled up.
- A chest X-ray is taken after passing the feeding tube from the mouth to get a positive screening test.
- The most accurate investigation is Tracheobronchoscopy.
- The investigation of choice is a Contrast esophagogram.
- In the past, a contrast CT scan used to be performed, but a CT scan has a radiation risk and does not give accurate results.

- A contrast CT scan is only done in a specific setting if coexisting malformations are suspected or urgent surgery is planned.
- Endoscopy and Contrast CT scans are not used nowadays.
- There is **no coiling of the tube in H Type TEF**. Thus, a contrast CT scan or endoscopy must be performed.

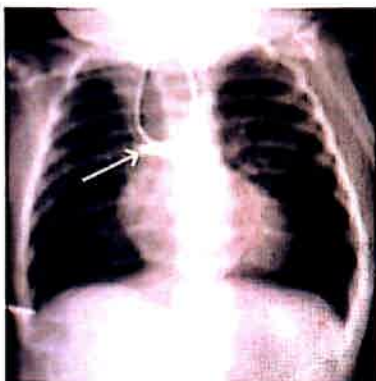


Image showing chest X-ray with the coiled-up feeding tube.

### Management of TEF

00:24:22

- Management of TEF management is primarily surgical but not done directly.
- Pre-surgical management is performed.
- The pre-surgical events are:
  - The head end of the child is elevated by 15-30 degrees.
  - Frequent oral suction is performed every 15-30 minutes.
  - IV fluids are started.
  - The first dose of prophylactic antibiotics is given (IV antibiotics).
  - Prone-position nursing can be done only when awake.
  - However, the prone position should be avoided during sleep as it is involved an increased risk of sudden infant death syndrome.
- After pre-surgical management, surgical correction is the therapy of choice.
- The surgery should be done as soon as possible.
- Surgery can be laparoscopic, open, or robotic.

### Surgical Approaches

00:26:41

- If the gap is small and the gap between the two ends of the esophagus is less than 3cm, then the two ends are pulled together and joined, and the fistula is closed.
- In other words, primary repair of the fistula and end-to-end anastomosis of the esophagus is done.
- If there is a large defect or the gap between the two ends of the esophagus is 3cm or more, the primary repair is not done, but gastrostomy is done.

- Later on, perform repair using grafts or conduits derived from the colon, stomach, or jejunum of the child.
- The surgery needs to be performed around **2-3 months of age**.



### Important Information

00:27:51

- The aspiration of gastric contents via a distal fistula causes more damaging pneumonitis than pharyngeal secretions from the blind upper pouch.
- Endotracheal intubation with mechanical ventilation should be avoided because it can worsen stomach distension.
- As it can cause severe aspiration from that area.

### Clinical Question

00:29:23

**Q.** A preterm child at 28 weeks had TEF-EA and developed HMD soon after birth. What is the approach?

**Ans.**

- First, manage HMD (hyaline membrane disease), also called respiratory distress syndrome.
- Most will require high ventilation setting or HFOV (high-frequency oscillatory ventilation).
- Frequent suctioning is done, and the patient is taken for gastrostomy as soon as he's stable.
- Primary end-to-end repair in a preterm child is usually not performed.
- Definite repair is done later, usually around 2 months of age.

### Postoperative Complications

00:30:41

- Early complications include
  - Anastomotic leak, which can cause esophagitis and mediastinitis.
- Late complications include
  - Gastroesophageal reflux is the reflux of gastric contents into the esophagus.
  - Stricture formation can occur due to surgery.
  - Tracheomalacia is the over-softening of the trachea.
- Tracheomalacia is observed when the child has hoarseness of voice, stridor, or wheezing sound.
- It can manifest a few weeks or as late as many years after the surgery.
- It is a post-surgical late complication.
- Tracheomalacia, again, requires surgery.

### Prognosis

00:32:49

- Syndromic children have a **poor prognosis**.
- Children who are **non-syndromic** (isolated TEF) have a **better prognosis**.
- Preterm children who require **ventilation** and have a gap in the esophagus of more than **5 cm** have a **poor prognosis**.

- With improved surgical features, a more than 5 cm gap is now considered a minor prognostic feature.

### Other Congenital Anomalies of the Esophagus

00:34:43

| Anomaly                           | Features                                                                                                                                                                                                                                                                                                                                                                                                                                                                                        |
|-----------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Esophageal Stenosis               | <ul style="list-style-type: none"> <li>• <b>Dysphagia</b> with food impaction is seen.</li> <li>• It may manifest from infancy to late adulthood.</li> <li>• The investigation of choice is <b>Esophagogram</b>.</li> <li>• The most accurate investigation is <b>Endoscopy</b>.</li> <li>• Therapy is Dilatation, followed by Surgery in refractory cases.</li> </ul>                                                                                                                          |
| Vascular Ring (Dysphagia Lusoria) | <ul style="list-style-type: none"> <li>• There is a <b>constricting ring</b> that compresses the esophagus and the airway.</li> <li>• So, there will be <b>dyspnea</b> with stridor or cough.</li> <li>• It may manifest from infancy to adulthood.</li> <li>• The investigation of choice is an <b>esophagogram</b>.</li> <li>• The most accurate investigation is <b>angiography</b>, the <b>gold standard</b> investigation.</li> <li>• Therapy is diet modification and surgery.</li> </ul> |

### Gastroesophageal Reflux (GER)

00:36:24

- It refers to the retrograde movement of gastric content across the lower esophageal sphincter (LES) back into the esophagus.
- LES is the thickening of the muscle at the gastroesophageal junction.
- Its function is to prevent the backward movement of the content.
- LES always has a resting tone with a pressure of more than or equal to **20 mmHg**.
- If the pressure falls **below 10 mmHg**, there is a chance of reflux occurring in the child.
- The crural part of the diaphragm acts as a supporting mechanism.
- In case when the diaphragm support is absent, or there is a hiatal hernia, the chances of GER will increase.
- So, any abnormality in the tone can lead to GER.

### Naspgan-Espghan 2018 Definitions

00:34:13

- **NASPGHAN** stands for North American Society of Paediatrics, Gastroenterology, Hepatology, and Nutrition.
- **ESPGHAN** stands for European Society of Paediatrics, Gastroenterology, Hepatology, and Nutrition.
- GER is the passage of gastric contents into the esophagus with or without regurgitation and vomiting (it may be symptomatic or asymptomatic).
- GERD occurs when GER leads to troublesome symptoms and complications.
- Refractory GERD occurs when GERD does not respond to optimal treatment after 8 weeks.

### Why reflux happens?

00:39:11

- It occurs due to a phenomenon known as **Transient LES relaxation (TLESR)**.
- They tend to occur very normally in children.
- Sometimes, they become exaggerated and prolonged, increasing the risk of reflux and causing GERD.
- Gastric distension is the main stimulant for TLESR.
- Increases in respiratory efforts can also sometimes cause relaxation.
- Other features responsible for reflux include increased movement or straining, large volume or hyperosmolar meals, anatomical factors (crura of diaphragm not being formed), etc.
- TLESR is the primary mechanism allowing reflux to occur and is defined as the simultaneous relaxation of both LES and the surrounding crura.
- TLESRs occur independent of swallowing, reduce LES pressure to 0-2 mm Hg (above gastric), and last **10-60 sec**.
- They appear by 26 weeks of gestation.
- It is a vasovagal reflex.
- Certain children have this relaxation happening more often for more prolonged periods and abnormally at all times. These are the children who tend to develop reflux.
- Transient relaxation can occur in any normal child that causes occasional reflexes called **happy spitters**.
- When fed normally and suddenly laid down, children will have transient relaxation due to abdominal distension, causing the milk to come out.
- This may not always be present, will be self-limited, and the child will not have any symptoms.
- If it becomes troublesome or persistent, then it causes reflux disease.



## Important Information

00:42:40

- A pediatric autosomal dominant form with otolaryngologic and respiratory manifestations has been located on chromosome 13q14, and the locus is termed **GERD1**.
- **AD-GERD** in children is the other name for this disease.

### Types of Reflux

00:43:22

#### Physiological Reflux

- It is a **transient** and occasional reflux.
- It occurs post-feed but not daily.
- It does not cause nutritional problems or respiratory problems.
- They usually begin in the late neonatal period.
- They peak around 4 months of age.
- In **88%** of patients, this stops at around 12 months.
- In almost **99-100%** of patients, this stops happening by 18 to 24 months.
- Children with **physiological reflux** are also called happy **spitters**.

#### Pathological Reflux (GERD)

- GERD is known to occur when the child fails to thrive or has respiratory or esophagitis-related symptoms.
- Sandifer Syndrome refers to reflex arching and dystonia affecting the child's neck, back, and limbs.
- Sandifer syndrome may be mistaken for seizures.
- It tends to improve with age.
- It is more of a manifestation and not a disease by itself.

#### Red Flag Signs or Alarm Signs in GER

00:48:46

- Symptoms like weight loss, fever, lethargy, and dysuria are red flag signs.
- Abdominal distension in GER is a **danger sign**.
- If there is a late onset of the disease (12 months, 14 months, 2 years of age, etc.) or persistence of more than 18 months is to be taken seriously.
- Vomiting, which is forceful, bilious, nocturnal, or hematemesis, is a dangerous sign.
- If the child has any neurological abnormality, it is a danger sign.
- If the child has **microcephaly** or **macrocephaly**, it is a danger sign.
- If the child has a **bleeding rectum**, it is a danger sign.

#### Diagnosis

00:50:27

- **Esophageal manometry** does not help demonstrate gastroesophageal reflux but might be used to evaluate transient LES relaxations and pressures.

- Contrast studies- Barium and water contrast studies can be performed but have low sensitivity and specificity. They are usually indicated only if an anatomic abnormality is suspected.
- Extended or 24-hour esophageal pH monitoring can also be done to diagnose reflux. If the pH is less than four, it often indicates significant reflux.
- Endoscopy and Scintigraphy using radionuclide scans can also be performed, but they play a limited role in diagnosis.
- Multichannel Intraluminal Impedance Monitoring is the most sensitive and specific test to diagnose gastroesophageal reflux.

### Management: NASPGHAN-ESPGHAN 2018 Guidelines

00:52:39

- The flowchart says that if you have any infant suspected of GERD, you take history and examination for the presence of alarm signs.
- If yes, you need to refer appropriately to a **pediatric gastroenterologist**.
- If there are no alarm signs, you will start with conservative management yourself.
- You should avoid overfeeding, thicken feeds, and continue breastfeeding the child.
- If not improved, consider 2-4 weeks of a protein hydrolysate or amino acid-based formula and avoid cow's milk.
- If the child improves, continue management.
- If not improved, refer to a pediatric gastroenterologist.
- If the referral is not possible, we can consider a trial of acid suppression therapy, where PPIs are the therapy of choice.
- According to current guidelines, pharmacological therapy should always come after conservative management.
- PPI should be used only for 4-8 weeks.
- You need to refer the patient if the patient has not improved.
- No further treatment is required if the patient is successfully weaned off.
- If referred to a pediatric gastroenterologist, further evaluation, work-up, etc., will be done.

Refer Image 3.1

This image shows the flowchart for the management of GER.

#### Summary

00:55:15

- When a patient is diagnosed with GERD, the first line of treatment in conservative management is the thickening of the feeds, head end elevation around 15-30 degrees, and positioning of the child (the child is in a prone position when awake and supine position while it is asleep) and small volume frequent feeds.
- Along with conservative management, we can rule out CMPA (cows milk protein allergy) and consider protein hydrolysate or amino acid-based formulas.

- If conservative management fails, the second line of treatment is drugs, and the drugs of choice are proton pump inhibitors for 4 to 8 weeks.
- If this also fails, surgery may be performed. The surgery performed is **Fundoplication**.

#### Role of Radiofrequency Therapy in GERD 00:38:07

- Endoscopic application of radiofrequency therapy to a 2-3cm area of the LES and cardia to create a high-pressure zone to reduce reflux is **Stretta Procedure**.
- It has a limited role in children and is currently not recommended.

#### Clinical Question 00:59:10

Q. What do AAP, NSPGHAN, and Nelson say regarding positioning?

Ans.

- Lying in the flat prone position and semi-seated positions in the postprandial period is considered provocative for GER and, therefore, should be avoided.
- After feeding the child, bring the child to your chest, slowly pat the back, and make the child burp.
- Burping calms the child helps improve peristalsis, and decreases the chances of GER.
- The head should be elevated by elevating the head end of the bed, rather than using excess pillows, to avoid abdominal flexion and compression that might worsen the reflex.

Q. Do prokinetic drugs have any role?

Ans.

- Prokinetic drugs include drugs like Metoclopramide (Dopamine-2 and 5-HT<sub>3</sub> antagonist), Bethanechol (Cholinergic agonist), and erythromycin (Motilin receptor agonist).
- None of the drugs affect TLESRs.
- RCTs have not demonstrated any useful effect in GER.
- **Metoclopramide can cause tardive dyskinesia.**
- However, Baclofen is a centrally-acting GABA agonist that decreases reflux by decreasing TLESRs. It can be useful in children with neurological disorders (like spastic cerebral palsy) prone to GERD.

#### Side Note 01:03:40

- **Barret's esophagus**
  - Recurrent refluxes can cause a metaplastic change of the normal squamous epithelium of the esophagus into the intestinal columnar epithelium.
  - It occurs due to esophagitis.
  - It is a precursor for **esophageal adenocarcinoma**.

#### GERD and Stridor

- Studies have shown that children with GERD have a higher risk of developing respiratory involvements like stridor and sometimes wheezing, which can manifest later as asthma.
- Patients with GERD can have reflux laryngitis. This reflex is episodic and resembles croup-like illness, stridor, and hoarseness of voice.

#### Santmyer Syndrome

- Facial stimulation by a puff of air can induce swallowing and esophageal peristalsis in healthy young infants.

#### Achalasia 01:06:37

- Achalasia is a primary **esophageal motor disorder** of unknown etiology characterized by loss of LES relaxation and esophageal peristalsis, both contributing to a functional obstruction of the distal esophagus.
- It can be caused due to degenerative, autoimmune (antibodies against components of LES), or infectious (Chagas disease caused by *Trypanosoma cruzi*) causes.
- It can also be a part of Allgrove syndrome, also known as Triple A syndrome. This syndrome is caused due to Achalasia, Alacrima, and adrenal insufficiency.
- Achalasia manifests with regurgitation and dysphagia for solids and liquids and may be accompanied by undernutrition or chronic cough.

#### Investigations

- Barium fluoroscopy will show **Bird's beak appearance**.
- **Manometry is the most sensitive test.**

#### Treatment

- Pneumatic dilation is done.
- Heller's myotomy is the surgery of choice.
- BOTOX injection into the LES endoscopically is the new line of treatment. It has a 50-65 % success rate, is costly, and requires repeated endoscopic procedures.