Gut involvement in PoTS – an overview

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Case Hx

- 28 year old lady presents with a long hx of constipation alternating with diarrhoea – Δ IBS
- Sports injury to the knee – arthroscopy under GA
- Post surgical flare up of GI symptoms
  - Nausea – present all day and worse in the mornings
  - Abdominal pain
  - Post prandial flushing, palpitations and diarrhoea
  - Post prandial fatigue, somnolence; dizziness esp. with CIP
  - Progresses to fainting with CIP - worse after meals
Examination and investigations

- Hypermobile with Beighton score of 7/9
- Elastic skin in phase of taking up slack
- Increase in HR of 30 beats / min between lying and standing
- Meets criteria for hEDS
- Blood profile, stool tests, gastroscopy, US abdomen all normal
- AFTs
  - Positive tilt table test
  - Post prandial Increase in HR with decrease in BP
- Δ hEDS with PoTS worsened postprandially
Symptoms-1

- Prevalence of GI symptoms: 70% - 90%.
- Most common symptoms:
  - Heartburn
  - Nausea
  - Vomiting
  - Dyspepsia
  - Bloating
  - Diarrhoea
  - Constipation
  - Abdominal pain
- Symptoms are worse after large carbohydrate meals

- Wang LB – 2015
- Huang RJ – 2103
- Park KJ – 2013
- Moak JP - 2016
Symptoms - 2

- Nausea
- Constipation
- Abdominal Pain
- Bloating

Graph showing the percentage of cases for each symptom by different studies (Wang et al., Hang et al., Moak et al.).
POTS and gut symptoms

- After eating Increased blood flow in abdominal blood vessels causes decrease in circulating volume

- Feeling of:
  - Light headedness
  - Fatigue
  - Drowsiness
  - Fainting
  - Nausea
  - Bloating
## Symptoms of dumping syndrome

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Score</th>
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<tbody>
<tr>
<td>Pre-shock or shock</td>
<td>+5</td>
</tr>
<tr>
<td>Loss of consciousness, fainting</td>
<td>+4</td>
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<tr>
<td>Will lie down or sit</td>
<td>+4</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>+3</td>
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<tr>
<td>Physical fatigue, exhaustion</td>
<td>+3</td>
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<tr>
<td>Sleep, listlessness, blurred vision</td>
<td>+3</td>
</tr>
<tr>
<td>Palpitation</td>
<td>+3</td>
</tr>
<tr>
<td>Restlessness, agitation</td>
<td>+2</td>
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<tr>
<td>Dizziness, vertigo</td>
<td>+2</td>
</tr>
<tr>
<td>Headache</td>
<td>+1</td>
</tr>
<tr>
<td>Feeling hot, sweating, paleness, clammy skin</td>
<td>+1</td>
</tr>
<tr>
<td>Nausea</td>
<td>+1</td>
</tr>
<tr>
<td>Abdominal distension, meteorism</td>
<td>+1</td>
</tr>
<tr>
<td>Borborygm</td>
<td>+1</td>
</tr>
<tr>
<td>Eructation</td>
<td>-1</td>
</tr>
<tr>
<td>Vomiting</td>
<td>-4</td>
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**FIGURE 1 - Sigstad score: value applied to the signs and symptoms of dumping syndrome**

[ABCD Arq Bras Cir Dig 2016;29(Supl.1):116-119]
Early dumping vs late dumping

- The Arts score—assesses the severity of symptoms after ingestion of glucose for diagnosis of early dumping, and one to two hours to late dumping.
- Likert scale: intensity on a scale of 0-3, where 0 represents the absence of certain symptoms, 1 mild, 2 moderate and 3 severe intensity.

**Figure 2** – Arts score: signs and symptoms to be evaluated according to their intensity

ABCD Arq Bras Cir Dig 2016;29(Supl.1):116-119
Pathophysiology of dumping syndrome

- The sudden presence of gastric contents in the proximal small intestine has the physiological response:
  - To release of bradykinin, serotonin and enteroglucagon,
  - Fluid shift
  - Leading to early symptoms in less than 30 min.
- Late symptoms: Within 90 min to 3 h, appear due to high insulin secretion causing hypoglycemia

ABCD Arq Bras Cir Dig 2016;29(Supl.1):116-119
GI physiological investigations

Gastric emptying is abnormal in two thirds of patients:
- Rapid emptying almost three times as common as delayed emptying
- Rapid emptying can cause dumping syndrome leading to postprandial symptoms seen in PoTS patients

Gastric myoelectrical activity - abnormal in PoTS patients, particularly in those with postprandial symptoms:
- variability in the gastric slow wave frequency compared to healthy controls.
- PoTS patients with postprandial symptoms have increased postprandial variability of the gastric slow wave frequency compared to PoTS patients without postprandial symptoms
Gastric Emptying in hEDS – MRI study

Menys A 2017
Gastric motility in EDS vs Healthy subjects – MRI study

All subjects motility pre/post water (A) and relative change in motility (B)

Menys A 2017
Gastric accommodation – hEDS VS Healthy subjects: MRI study

A) Boxplot showing accommodation (%)

B) Scatter plot showing emptying time (min) vs accommodation (%)
Exclusion of other causes

- A thorough medical history, systems review, detailed drug history and physical examination are essential to rule out important differentials:
  - Diabetes mellitus
  - Hypothyroidism
  - Connective tissue disorders
  - Coeliac disease
  - Inflammatory bowel disease
  - Infections
  - Neurological disorders
  - Drug effects e.g. opiates can produce bowel dysfunction
Investigations to exclude other causes

- Blood testing for FBC, LFTs, ESR, CRP, thyroid function, albumin, coeliac serology and autoimmune screen.
- Suspicion of obstruction (e.g. vomiting, absolute constipation, abdominal pain and distention)
  - Abdominal ultrasound, upper endoscopy, colonoscopy or CT/MRI enterography, according to the symptoms.
- Upper and/or lower GI physiology studies
- Neurological signs esp. morning nausea:
  - CT or MRI of the head.
- Oral glucose challenge in pts. with postprandial hypoglycemia.
Management: Dietary and lifestyle modifications

- Ingestion of food is a major trigger for GI symptoms in patients with PoTS.
- Lack of strong available evidence to support specific dietary modifications
  - our experience suggests that dietary alteration can improve symptoms.
- Proper dietary history:
  - Food intake diary - identify specific triggers and avoid unnecessary dietary restrictions.
- In patients with rapid gastric emptying and postprandial hypoglycemia we recommend the following:
  - Eat small and frequent meals
  - Eat slowly and chew food thoroughly
  - Opt for low-glycemic-index foods
  - Increase fat and protein intake to balance energy requirements
  - Separate intake of liquids from solids, avoiding liquids for half an hour before and after meals.
  - Lie down for 30 minutes after meals - this can reduce postprandial symptoms e.g. palpitations, flushing or dizziness
  - Increasing intake of salt and water appears to improve symptoms of nausea
Management: Dietary and lifestyle modifications

In patients with gastroparesis, we recommend:

- Adequate chewing to reduce the size of the food
- Avoid intake of insoluble fiber
- ‘Graze’ – eat regular small meals
Reduce fat intake.
When to refer to the gastro clinic?

- When patients with PoTS present with significant postprandial symptoms in particular worsening of usual PoTS symptoms.
- Symptoms suggestive of post prandial reactive hypoglycemia.
- A proportion of PoTS patients can have delayed gastric emptying therefore symptoms such as early satiety, nausea and/or vomiting, fullness and bloating may also be referred for further specialist opinion.
What treatment is instituted in the specialist clinic?

- An empathic doctor-patient relationship-crucial.
- We need to allow some time to reassure the patient
  - Legitimacy of their symptoms
  - To explain possible causes and reassure them of the absence of a life-threatening illness.
  - Equally, it is important to explain that the management will require several steps and strategies:
- Lifestyle modification – fluid and salt intake; exercise
- Dietary modification with adequate fluid and salt intake
- Pharmacological therapy – this is symptom-directed.
  - Anecdotal experience that in some patients with PoTS - GI symptoms improve following treatment of their main non GI PoTS symptoms with;
    - Mineralocorticoids such as fludrocortisone
    - Sympathomimetics such as midodrine
    - Hormonal treatment: Octreotide
- Psychological support when the patient has difficulty with coping
Symptomatic pharmacological treatment

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Therapy Type</th>
<th>Medications</th>
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<tr>
<td>Gastroparesis</td>
<td>Motility stimulants</td>
<td>Erythromycin, Prucalopride,</td>
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<tr>
<td>Diarrhoea</td>
<td>Antimotility drugs</td>
<td>Loperamide, Ondansetron</td>
</tr>
<tr>
<td>Constipation</td>
<td></td>
<td>Prucalopride, Linaclotide, laxatives</td>
</tr>
<tr>
<td>Pain</td>
<td>Antispasmodics</td>
<td>Dicycloverine, Hyoscine, Mebeverine, Peppermint oil</td>
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<tr>
<td></td>
<td>Antidepressants</td>
<td>TCAs, SSRIs, SNRIs</td>
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<tr>
<td></td>
<td>Pain Modulators</td>
<td>Gabapentin, Pregabalin</td>
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<tr>
<td>Nausea</td>
<td>Antihistamines</td>
<td>Cyclizine</td>
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Conclusions

• PoTS - a range of gastrointestinal (GI) symptoms
• GI dysmotility in PoTS has not been proven.
• Organic GI conditions need to be ruled out
• GI physiology testing could help to define the GI phenotype and guide management strategies.
• No established guidelines for the management of GI symptoms in PoTS and patients are therefore treated symptomatically.
• Management of PoTS with conservative measures and drug treatment can improve GI symptoms especially nausea and post prandial somnolence and dizziness
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