Pancreatic Exocrine Insufficiency (PEI) In Primary Care

This brochure provides a summary of the recommendations outlined in the ‘Diagnosis and management of pancreatic exocrine insufficiency (PEI) in primary care: consensus guidance of a Canadian expert panel’ document. This critical review of the literature on PEI and Pancreatic Enzyme Replacement Therapy (PERT), conducted by a panel of Canadian gastroenterologists, has developed practical diagnostic and treatment recommendations for Primary Care Physicians (PCPs). These recommendations provide guidance on identifying patients at risk of PEI, the triggers for PEI testing and referral, and best practices for co-managing patients with confirmed PEI. To review the document visit: https://www.ncbi.nlm.nih.gov/pubmed/28985688

The prevalence of PEI has been reported between 11.5% and 21.7% in individuals without pre-existing gastrointestinal disease (Campbell et al. (JGLD 2016; 25: 303-9). Depending on the underlying etiology, PEI may present at any time in a patient’s life, from infancy to advanced old age.
As PEI is a clinical diagnosis, patients should never be assumed to have PEI based solely on:

- isolated findings (e.g. low fecal elastase-1) or
- the presence of a predisposing condition such as chronic pancreatitis (CP) or cystic fibrosis (CF).

### Signs & symptoms suggestive of PEI:

- Large volume malodorous stools (stools that float in the toilet bowl, and are difficult to flush away)
- Steatorrhea; fat droplets like cooking oil in toilet bowl
- Unexplained weight loss
- Clinical sequelae of micronutrient deficiencies including iron, vitamin B12 and fat-soluble vitamins (A, D, E, K)
  - See table 3 in paper for full clinical signs/symptoms of these categories

### Tests that may be abnormal in PEI:

- Stool microscopy (Sudan stain) – fat droplets or meat fibres present
- 72-hour fecal fat excretion test – increased
- Blood levels of fat-soluble vitamins (A,D,E,K) – decreased

Symptoms of diabetes mellitus in the presence of chronic pancreatitis
When should PCP investigate the possibility of PEI?

Patients with PEI may report no symptoms or very few symptoms; equally commonly, they may have become accustomed to their symptoms. The most common symptoms relate to insufficient lipase and colipase secretion, with consequent lipid malabsorption.

In the primary care setting, a possible diagnosis of PEI should be pursued on the basis of the combination of patient history & presenting symptoms.

PEI should always be considered in individuals with a new diagnosis of pancreatic disease or a new diagnosis of a condition that may predispose to PEI or malabsorption, including:

- Chronic Pancreatitis (CP)
- Type 1 Diabetes
- Unresponsive celiac disease
- HIV/AIDS
- If they are current or former smokers* 
- Longstanding history of alcohol abuse (defined as >5 drinks/day)*

* The last two risk categories interact, such that risk of PEI with alcohol abuse is heightened among current smokers.

**PEI should be considered in patients with unexplained suggestive symptoms, even in the absence of any known predisposing conditions.**
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For patients with signs or symptoms suggestive of PEI, the PCP should make particular note of the following aspects of the patient’s history:

Patients With Signs Or Symptoms Suggestive Of PEI

- Clinical history
- Weight loss
- Prior pancreatitis
- Alcohol history
- Smoking history
- GI surgery
- Diabetes mellitus
- Family history of pancreatic disease
- GI complaints suggestive of pancreatic insufficiency

Screening & Tests for Patients with a Predisposing Condition

- Weight and body mass index
- Fat soluble vitamins and other nutritional markers (e.g. albumin)
- HbA1c
- Bone mineral density scan

In addition to the recommended screening tests, the following should be carried out regularly:

- Stool tests, including:
  - Fecal Sudan staining or
  - Fecal elastase (FE-1)

* For patients with CP = asymptomatic or minimally symptomatic PEI is common, so regular (yearly) screening is appropriate

Of the above – blood tests for deficiencies in fat-soluble vitamins (A, D, E, K) are particularly important because, although they can occur for reasons unrelated to malabsorption, deficiencies associated with an underlying condition (ex. CP) are highly suggestive of PEI. **PEI is unlikely if all the above measures are normal.**
If wait time for specialist care is long, other investigations might be appropriate such as:

- Investigating alternative diagnosis such as celiac disease or giardiasis
- Consider computed tomography (CT) imaging to assess for possible CP or pancreatic cancer
- A 2 week trial of PERT may also be considered
When and how should the PCP refer to a specialist?  
Referral letter essential elements.

Make the referral if the patient:

• shows evidence of abnormal tests following screening (Ex. FE-1, plasma fat soluble-vitamin deficiencies)
• and/or when the patient experiences ongoing symptoms (steatorrhea or weight loss)

If PEI is diagnosed in the absence of any previously established predisposing conditions, it is important for patients to be referred to a specialist for further diagnostic workup, as PEI can often be the first clinical manifestation of pancreatic cancer or CP.

Therefore, obtaining a CT scan to exclude the above is strongly considered.

A Thoroughly Documented Referral Letter Should be Made

Duration and severity of symptoms (if any)
• Chronic diarrhea ☐
• Features of steatorrhea ☐
• Abdominal Pain ☐

Quantified weight loss or failure to thrive in infants and children ☐

Summary of investigations performed:
• Biochemical markers of nutritional status ☐
• Stool tests for fecal fat ☐
• FE-1 (if done) ☐

Imaging results (if done) ☐

Response to PERT (if tried) ☐

Alcohol and smoking history ☐
History of recurrent acute pancreatitis, CP or diabetes ☐
Family history of CP, pancreatic cancer or CF ☐
List of medical conditions ☐
List of medications ☐

Given specialist wait times, a documented referral letter will improve the patient’s prospects of being seen promptly by the specialist.
How should PCP manage PERT treatment and assess response?

Patients with PEI who have an inadequate response to PERT should be referred or re-referred to a specialist. If there is a suspicion of pancreatic cancer or another serious diagnosis, referral should be urgent.

If specialist access is limited or wait time is long, the PCP may

- Assess current dosing schedule and reinforce importance of daily dosing with all meals and snacks
- Assess the patient’s adherence to PERT
- Adherence, order fFecal chymotrypsin test if available
- Investigate alternative diagnoses, such as:
  - Giardia (stool test);
  - Pancreatic Cancer (pancreatic imaging);
  - Gluten sensitivity / celiac disease (serology / EGD & biopsy)
- Initiate trial of concomitant PPIs
- Initiate trial of higher-dose PERT within accepted guidelines

What other measures should PCPs recommend for their patients with PEI?
Lifestyle/diet, etc.

Should not recommend:
Low-fat diets as patients with PEI are at risk of malnourishment although obese patients constitute a likely exception to this

Should recommend:
Quit smoking
Avoid alcohol
Psychological and other support may be needed for patients attempting to make these behavioral changes
Registered dietitian
References:

For more information, visit CDHF.ca.
CDHF is the official Foundation of the Canadian Association of Gastroenterology, we are directly connected to Canada’s leading digestive health experts, physicians, scientists and other health care professionals. You can trust us to provide you with practical, science-based information that is up to date and unbiased.