MANAGEMENT OF GASTROINTESTINAL ASPECTS IN SSC

A. MANAGEMENT OF INCONTINENCE

1. Establish diagnosis

Assess frequency, enquire dietary and fluid intake

Determine pattern if any or precipitating factors eg access to toilets, mobility issues related to SSc

Assess urge faecal incontinence, passive leakage, sensation of incomplete evacuation, post-defaecation soiling

Assess consequences eg perianal pruritus/soreness

Assess impact of symptoms on lifestyle/quality of life

Physical examination to include PR for anal tone, ability to squeeze anal sphincter voluntarily

Core investigations may include rigid sigmoidoscopy and direct visualisation with colonoscopy if red flag features are present

2. Identify and underlying contributory factors

- A history of anal sphincter injury including obstetric history (parity, difficult delivery, large birth weight and associated complications) and other perianal surgical procedures should be evaluated
- Rectal prolapse, third-degree haemorrhoids or urinary incontinence
- SSc-GI issues eg diarrhoea (**See link**)
- Consider drugs that may exacerbate faecal incontinence and loose stools including calcium channel blockers, SSRIs, sildenafil, magnesium-containing antacids, tricyclics

3. Based on baseline assessment, initial management may include

Patient education with advice on dietary modification, address access to toilet at work, review medications that may contribute to FI and prescribe anti-diarrhoeal drugs for those with loose stools and faecal incontinence

Generic advice is provided.

- Prescribe anti-diarrhoeal drugs, Loperamide hydrochloride should be first drug of choice, introduced at very low dose and to escalate as tolerated until desired stool consistency is reached. Syrup preparation may be used for doses less than 2 mg. Codeine phosphate may be used as alternative.

4.If patients remain symptomatic, consider referral for specialist investigations

- Anorectal physiology/manometry (Thoua 2011)
- Endoanal US especially those with obstetric history. Referral to specialist GI centre may be necessary
- If these investigations are not available, MRI, endovaginal US and perineal US should be considered. Defaecating proctogram may be considered as indicated

5. Specialist management

- Patient-focus and practical management
  - Address toilet access issues
  - Modify dietary and fluid intake
  - pelvic floor muscle training, bowel retraining, biofeedback, specialist dietary assessment and management, anal plugs (for people who can tolerate them) , Rectal irrigation
  - Coping strategies with advice on emotional and psychological support are important
  - Continence products with choice of disposable body-worn pads and disposable bed pads may be necessary
- Surgical approaches may be considered
  - Consider a trial of temporary sacral nerve stimulation and proceed to implantation if successful. These patients should be assessed and managed at a specialist centre that has experience of performing this procedure. Sphincter augmentation with bioprothestic device may be considered in specialist units.

NICE has published guidelines on management of faecal incontinence in adults. Further details are available on http://www.nice.org.uk/nicemedia/live/11012/30548/30548.pdf

B. MANAGEMENT OF ABDOMINAL PAIN AND DISTENSION

1. Ascertain contributing factors including warning signs of true obstruction and other acute abdominal events such as sigmoid volvulus, diabetes
2. Identify dominant symptom: Small bowel bacterial overgrowth, Gastroparesis
3. Based on initial clinical assessment, appropriate investigations should be instituted
    - Plain Xray to assess features of bowel obstruction
    - Further investigations are undertaken, if indicated, to evaluate both upper and lower GI symptoms; for example, endoscopy for upper GI symptoms, US for biliary causes, CT to exclude other intraabdominal pathologies

4. Clinical assessment and investigations will guide management and this may include a trial of empiral antibiotics for presumed bacterial overgrowth. The choice of antibiotics is governed by local guidelines, and these may include Doxycycline and Ciprofloxacin.
5. If these initial steps fail to control the symptoms , these patients should be referred for specialist assessment.
    - Specific investigations are undertaken as indicated
        - Hydrogen breath test
        - Gastric emptying test (Marie 2012)
        - Oesophageal/ Antro-duodenal manometry??

5. Treatment
    - Practical patient-focussed approaches
        - These should address anxiety and depression with professional counselling.
        - Dietary modification includes formal dietitian review – dietary balance, meal size and timing of meals. Probiotics may be helpful in selected patients (Frech 2011)
        - Lifestyle factors such as smoking and alcohol may delay gastric emptying
    - Pharmacological agents
        - Prokinetics eg Domperidone, Metoclopramide and low-dose Erythromycin, somatostatin analogues such as Octreotide (Nikou 2007)
        - A trial of rotating antibiotics for SBO without breath test is justifiable. These may include Ciprofloxacin, ……..
    - Enteral support may be required
        - Trial of nasogastric or nasojejunal feeding leading to PEG/PEJ feeding. If enteral nutrition is poorly tolerated because of pain, bloating or recurrent abdominal injection, longterm parenteral nutrition may be necessary (Brown 2008).
    - Colonoscopic decompression may be considered in selected patients ??

C. MANAGEMENT OF GASTRO-OESOPHAGEAL REFLUX

1. Establish diagnosis and determine dominant symptom (Ntoumazios 2006)

- Assess severity of symptoms (volume regurgitation, nocturnal symptoms)
- Assess comorbidities: myositis, sicca symptoms and lung fibrosis

2. Investigations should be undertaken

- Ba swallow/OGD for persistent dysphagia or strictures
- Consider OGD for odynophagia, persistent iron deficiency anaemia to evaluate candidiasis and gastric antral vascular ectasia

3. Treatment

- Pharmacological
  - High dose PPI ± H2 receptor blockade (Pakozdi 2009)
  - A majority of patients require a maintenance dose of PPI which may be increased as necessary when symptoms flare.
  - Adjunct prokinetic may be considered if there are features of oesophageal dysmotility
- Practical patient-focussed management
  - Elevation of head bed
  - Avoid meals for at least three hours prior to bed time
  - Avoid fluid overload with meal
  - Small frequent meals
  - Dietary consideration with avoidance of cold fluid may be helpful
- Specialist referral
  - Oesophageal manometry and pH studies including impedance for non-acid reflux may be required
  - Surgical intervention such as fundoplication should be avoided as this procedure is unlikely to benefit patients with SSc and may aggravate oesophageal symptoms
  - Candida infection should be considered if symptoms especially odynophagia fail to improve and empirical antifungal treatment may be indicated.

- Barrett’s oesophagus will require endoscopic surveillance as per local/national guidelines.
- Recognition that treating reflux aggressively may prevent further progression of interstitial lung disease in selected patients.

D.MANAGEMENT OF DIARRHOEA

1. Establish diagnosis and assess severity.

Nutritional status should be evaluated using the Malnutrition Universal Screening Tool (MUST) to identify those who are risk of malnourishment and therefore those who may benefit from appropriate nutritional intervention (Elia M 2003, Baron 2009). These include five steps:

Step 1 and 2: Gather nutritional measurements (height, weight, BMI, unplanned weight loss over preceding months)

Step 3: Consider the effect of scleroderma to the nutritional intake

Step 4: Determine the overall risk score of malnutrition.

Step 5: Develop care plan using local management guidelines

(http://www.bapen.org.uk/must_tool.html)

Contributory factors including infection in particular C. difficile related to antibiotic use, coeliac disease and small bowel bacterial overgrowth should be addressed(Forbess 2011)

2. The following investigations should be considered as indicated

- Screen for infection with stool microscopy and culture including C. difficile
- Screen for celiac screen, thyroid function, B12 and folate

Specialist referral for further investigations may include

- Hydrogen Breath test (Marie 2009)
- Investigate for malabsorption including faecal elastase, SeHCAT for bile malabsorption

4. Treatment

- Nutrition and dietary consideration as indicated with formal dietitian review
- Medical therapy
    - Antibiotics for small bowel bacterial overgrowth
    - Symptomatic approaches should only be considered once the contributory causes to diarrhoea are identified. These may include Loperamide, ?Trial of Fybogel if overflow. Specific therapies may be required: antibiotics or Rifaximin if available and Cholestyramine for bile-salt diarrhoea (Parodi 2008).
    - Gluten-free diet for coexisting celiac disease

- Trial of pancreatic enzyme supplementation if pancreatic insufficiency
- Role of fermentable diet in selected cases

E. MANAGEMENT OF NUTRITIONAL ISSUES AND WEIGHT LOSS

1. Establish contributing causes (active SSc, major organ involvement, underlying neoplasia, depressive illness)

2. Assess severity and risk of malnutrition with MUST score..
(http://www.bapen.org.uk/must_tool.html)

3. Investigations

- Blood tests – TFT, FBC, Haematinics, Electrolytes imbalance
- Imaging – CT Abdomen
- Screening for malnutrition with change in BMI score, weight loss during preceding months and risk stratification

Specialist Referral for further management of nutritional assessment

3.Treatments

- Specific management of this aspect of the disease is governed by the primary cause or contributing factor to weight loss. These are discussed in the other sections. (See link on Diarrhoea, Abdominal pain/Distension)
- Dietician review
    - Oral nutrition support to ensure overall nutrient intake contains a balanced mixture of protein, energy and fibre, electrolytes, vitamins and minerals.

- Management of associated dysphagia is critical to evaluate the risks and benefits of modified oral nutrition support and/or enteral tube feeding
  Parenteral feeding (http://www.nice.org.uk/nicemedia/live/10978/29981/29981.pdf)

    - In those malnourished with inadequate oral or unsafe oral intake with functional and accessible gastrointestinal tract, enteral tube feeding is considered.

    - If there is associated gastric emptying abnormalities, post-pyloric (duodenal or jejunal) feeding is considered.

    - If there is severe non-functional gastrointestinal tract or failure with enteral tube feeding, continuous or cyclical parenteral nutrition is considered

    - These require coordinated multidisciplinary team, which includes input from specialist nutrition nurses, dietitians, GPs, pharmacists and district and/or homecare company nurses

- Venting gastrostomy may be considered to address abdominal distension

F.MANAGEMENT OF CONSTIPATION

1. Assessment of each symptom component (urge frequency and difficulty in evacuation). Use of patient diary should be considered.

2. Evaluation of contributory causes such as metabolic, endocrine causes and drugs (anti-depressants) Red flags such as features of malignancy must be considered.

3. Basic investigations such as calcium and TFT should be undertaken.

Based on initial assessment, appropriate generic advice may be sufficient. Specialist referral for further management may be required

4. Specialist investigations

- Colonic imaging or direct visualisation with colonoscopy is necessary if there is a change in bowel habit
- Related issues such as slow colonic transit/evacuation with radiopaque markers need to be addressed
- Defaecating proctogram to evaluate rectocoele

5. Treatment

- Dietary manipulation should be considered
- Adequate fluid intake
- Adequate fibre. Recommend foods with high fibre content such as fruits, vegetables, high-fibre bread, baked beans and wholegrain breakfast cereals. Do not recommend unprocessed bran which may aggravate bloating and flatulence.
- Oral/Suppository laxatives such as stimulants in high doses may be required.
- Prucolapride may be used in difficult cases (NICE guideline Dec 2010).
- Surgical intervention such as defunctioning stoma should only be undertaken after careful consideration and this should be undertaken at a specialist centre

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