

# European (ESPEN/ESNM) Survey on Clinicians' Perspectives on the Diagnosis and Management of Severe Gastrointestinal Dysmotility in adults

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## A. You and your team

1. **Name:**  
\*optional if you would like your participation to be acknowledged at the end of the published manuscript

2. What is the name of your institution?

3. In which country is your unit based? \*

4. Which department(s) do you work in?\*

Select at least 1.

- Intestinal Failure
- Gastroenterology
- Upper GI surgery
- Colorectal surgery
- General surgery
- Intestinal Transplantation
- Neurogastroenterology and Motility
- Other, please specify

5. What is your role?\*

- Clinical Academic (Professor/Associate Professor)
- Consultant (Attending) physician/surgeon
- Clinical Trainee/ Fellow
- Other, please specify

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## A. You and your team

6. Which description best fits your hospital/unit?\*
- National referral centre for intestinal failure and/or intestinal transplantation
  - Regional referral centre for intestinal failure and/or intestinal transplantation
  - Teaching hospital, non-referral centre for intestinal failure and/or intestinal transplantation
  - General hospital, non-referral centre for intestinal failure and/or intestinal transplantation
  - Other, please specify
- 
7. What is your background?
- Physician
  - Surgeon
  - Other, please specify
- 
8. Which of the following multidisciplinary team members do you have in your unit/hospital?
- Intestinal failure clinician
  - Gastroenterologist with a sub-specialty interest in Functional/ Neurogastroenterology and Motility disorders
  - Specialist dietician
  - Clinical Psychologist
  - Chronic Pain management team
  - Histopathologist with specialist interest in GI neuromuscular disorders
  - Intestinal Transplant surgeons
  - Clinical Pharmacist with specialist interest in parenteral nutrition
  - None of the above

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**Based on findings from radiological and motility tests, severe intestinal motility disorders can be sub-classified into Chronic Intestinal Pseudo-obstruction (CIPO) and Enteric Dysmotility (ED).**

• CHRONIC INTESTINAL PSEUDO-OBSTRUCTION (CIPO) is defined as chronic/recurrent obstructive type symptoms with radiological features of dilated intestine with air/fluid levels in the absence of any lumen-occluding lesion. • ENTERIC DYSMOTILITY (ED) refers to patients with abnormal small bowel motility tests but without radiological features of a dilated intestine.

9. In your opinion, is it clinically important to recognize CIPO and ED as separate clinical entities?\*
- Yes  No
10. Approximately how many new referrals (all patients) with confirmed or suspected CIPO or ED do you see per year?\*
- <5
  - 6-10
  - 11-20
  - 21-30
  - 31-40
  - 41-50

- 
- > 50

11. Based upon your response to question 10, approximately what percentage of these patients meet the clinical and radiological criteria for CIPO?\*

Please see definitions of CIPO and ED above

- 0-25%
- 25-50%
- 50-75%
- 75-100%

12. In your opinion, has there been a recent increase in the relative proportions of ED vs. CIPO patients that you see in clinical practice?\*

-- Please Select --

13. Based upon your response to question 12 related to the proportion of referrals with ED vs. CIPO, which of the following statements is most accurate?\*

- The incidence of CIPO appears to be rising
- The incidence of ED appears to be rising
- There is no relative change in the proportion of CIPO vs. ED cases that I see in practice
- The incidence of both CIPO/ED may be increasing

14. Please estimate (in your experience) the typical time interval from onset of symptoms to diagnosing CIPO and ED?\*

|      | <b>&lt;6<br/>months</b> | <b>6-12<br/>months</b> | <b>1-5<br/>years</b>  | <b>5-10<br/>years</b> | <b>&gt; 10<br/>years</b> |
|------|-------------------------|------------------------|-----------------------|-----------------------|--------------------------|
| CIPO | <input type="radio"/>   | <input type="radio"/>  | <input type="radio"/> | <input type="radio"/> | <input type="radio"/>    |
| ED   | <input type="radio"/>   | <input type="radio"/>  | <input type="radio"/> | <input type="radio"/> | <input type="radio"/>    |

15. Of the total population of patients with CIPO or ED under your care, please estimate the proportion that you see who have a recognized **secondary** cause (i.e. a systemic disease such as a Scleroderma, Ehlers-Danlos etc.) as opposed to idiopathic (primary) dysmotility.\*

- 0-10%
- 10-25%
- 25-50%
- 50-75%
- 75-100%

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16. In your clinical practice, have you noticed a change in the frequency CIPO or ED referrals in association with any of the following list of disorders? Please select the statement that best reflects your recent experiences.\*

**The  
frequency**

**The  
frequency**

**The  
frequency**

|  | <b>of this condition in referrals with CIPO/ED is increasing</b> | <b>of this condition in referrals with CIPO/ED is the same</b> | <b>of this condition in referrals with CIPO/ED is decreasing</b> |
|--|--|--|--|
| Scleroderma  | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Neurological disorders (e.g. mitochondrial disorders)                  | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Endocrine disorders  | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Ehlers-Danlos syndrome/ joint hypermobility disorders                  | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Paraneoplastic disorders   | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Autoimmune disorders   | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Autonomic dysfunction (e.g. postural orthostatic tachycardia syndrome) | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |
| Hirschsprung's disease   | <input type="radio"/>  | <input type="radio"/>  | <input type="radio"/>  |

17. Based upon your response to question 16, which (if any) of the listed conditions associated with CIPO/ED have you seen the largest increase in referrals? Please select **one** of the following.

- Scleroderma
- Neurological disorders
- Endocrine disorders
- Ehlers-Danlos syndrome/ joint hypermobility disorders
- Paraneoplastic disorders
- Autoimmune disorders
- Autonomic dysfunction (e.g. postural orthostatic tachycardia syndrome)
- Hirschsprung's disease
- Other, please specify

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### C. Diagnosing CIPO and ED in clinical practice

18. When comparing your experiences making the diagnoses of CIPO and ED, which of the following statements best reflects your opinion?\*
- CIPO is a more difficult diagnosis to make
  - ED is a more difficult diagnosis to make
  - Both CIPO and ED are equally difficult diagnoses to make
  - CIPO and ED are not difficult diagnoses to make

19. Based on your answer to question 18, in your opinion, how often do the following factors contribute to difficulties or delays in the diagnosis of CIPO or ED?\*

|   | <b>Never<br/>(0%)</b> | <b>Sometimes<br/>(&lt;50%<br/>cases)</b> | <b>Often<br/>(50-<br/>99%<br/>cases)</b> | <b>Always<br/>(100%<br/>of<br/>cases)</b> |
|---|-----------------------|--|--|---|
| Non-specific nature of symptoms                       | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Difficulty excluding mechanical obstruction           | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Limitations of diagnostic tests                       | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Difficulty in eliminating opiates as the cause        | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Psychological Comorbidity in CIPO/ED                  | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Difficulty obtaining histopathology                   | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Lack of awareness of CIPO/ ED amongst non-specialists | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |

20. How often do you perform the following investigations when establishing the diagnosis of CIPO or ED?\*

|   | <b>Never<br/>(0%)</b> | <b>Sometimes<br/>(&lt;50%<br/>cases)</b> | <b>Often<br/>(50-<br/>99%<br/>cases)</b> | <b>Always<br/>(100%<br/>of<br/>cases)</b> |
|---|-----------------------|--|--|---|
| Oesophageal Manometry                                   | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Small Bowel Manometry                                   | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Colonic Manometry                                       | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Anorectal Manometry                                     | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Gastric emptying study (scintigraphy)                   | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Motility Small Bowel MRI (Cine MRI)                     | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| MRI Enterography (MRE)                                  | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Barium Meal and Follow through                          | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Wireless Motility Capsule (SmartPill)                   | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Lactulose/Hydrogen breath test (Orocaecal transit time) | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| X-ray colonic transit study                             | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Full thickness jejunal biopsies                         | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Plain X-ray Abdomen                                     | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |
| Breath tests for Small Intestinal Bacterial Overgrowth  | <input type="radio"/> | <input type="radio"/>                    | <input type="radio"/>                    | <input type="radio"/>                     |

Jejunal aspirates for  
Small Intestinal  
Bacterial overgrowth

21. At the time of CIPO or ED diagnosis, please estimate the percentage of patients that previously underwent surgeries that were, in your opinion, inappropriate?\*

- 0-10%
- 10-25%
- 25-50%
- 50-75%
- 75-100%

22. When do you request full thickness biopsies in ED or CIPO?\*

- Never
- Routinely
- Only if a specimen is available from previous or planned surgical procedure
- When the diagnosis is unclear
- Other, please specify

23. **In your opinion**, how often do full thickness biopsy results alter the following factors in managing patients with CIPO or ED?\*

|  | <b>Never</b>          | <b>1-25%<br/>of<br/>cases</b> | <b>25-50%<br/>of<br/>cases</b> | <b>50-75%<br/>of<br/>cases</b> | <b>75-100%<br/>of<br/>cases</b> | <b>Not<br/>sure</b>   |
|--|-----------------------|-------------------------------|--------------------------------|--------------------------------|---------------------------------|-----------------------|
| Can lead to targeted medical therapies e.g. immunosuppression                    | <input type="radio"/> | <input type="radio"/>         | <input type="radio"/>          | <input type="radio"/>          | <input type="radio"/>           | <input type="radio"/> |
| Influences nutritional management decisions (i.e. oral vs enteral vs parenteral) | <input type="radio"/> | <input type="radio"/>         | <input type="radio"/>          | <input type="radio"/>          | <input type="radio"/>           | <input type="radio"/> |
| Influences surgical management decisions   | <input type="radio"/> | <input type="radio"/>         | <input type="radio"/>          | <input type="radio"/>          | <input type="radio"/>           | <input type="radio"/> |
| Influences choice of prokinetic drug   | <input type="radio"/> | <input type="radio"/>         | <input type="radio"/>          | <input type="radio"/>          | <input type="radio"/>           | <input type="radio"/> |
| Can help determine patient prognosis   | <input type="radio"/> | <input type="radio"/>         | <input type="radio"/>          | <input type="radio"/>          | <input type="radio"/>           | <input type="radio"/> |

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24. How often do you find the following treatment approaches useful in patients with CIPO or ED?\*

|             | <b>Never<br/>(0%)</b> | <b>1-25%<br/>cases</b> | <b>25-50%<br/>cases</b> | <b>50-75%<br/>cases</b> | <b>75-100%<br/>cases</b> |
|-------------|-----------------------|------------------------|-------------------------|-------------------------|--------------------------|
| Domperidone | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>    |

|   |                       |                       |                       |                       |                       |
|---|-----------------------|-----------------------|-----------------------|-----------------------|-----------------------|
| Metoclopramide  | <input type="radio"/> |
| Erythromycin  | <input type="radio"/> |
| Prucalopride  | <input type="radio"/> |
| Linaclotide   | <input type="radio"/> |
| Octreotide  | <input type="radio"/> |
| Neostigmine/<br>Pyridostigmine  | <input type="radio"/> |
| Cisapride   | <input type="radio"/> |
| Naloxegol   | <input type="radio"/> |
| Antibiotics for small<br>intestinal bacterial<br>overgrowth   | <input type="radio"/> |
| Neuropathic<br>analgesics (e.g.<br>Pregabalin,<br>Gabapentin,<br>antidepressants)                   | <input type="radio"/> |
| Opiate analgesia  | <input type="radio"/> |
| Venting<br>gastrostomy  | <input type="radio"/> |
| Venting Colostomy   | <input type="radio"/> |
| Enteral Tube<br>feeding   | <input type="radio"/> |
| Parenteral Nutrition  | <input type="radio"/> |
| Clinical Psychology<br>interventions (e.g.<br>cognitive<br>behavioural<br>therapy,<br>hypnotherapy) | <input type="radio"/> |
| Surgery   | <input type="radio"/> |

25. Have you noticed any difference in the prevalence of psychological co-morbidity between the ED and CIPO sub-types?
- Patients with CIPO have higher prevalence of psychological co-morbidity compared to ED
  - Patients with ED have a higher prevalence of psychological co-morbidity compared to CIPO
  - The prevalence of psychological co-morbidity is similar in ED and CIPO
26. Have you noticed any difference in the health care utilization between patients with ED and CIPO (select any that apply)?
- Patients with CIPO have a higher readmission rate than patients with ED
  - Patients with ED have a higher readmission rate than patients with CIPO
  - Patients with ED and CIPO have similar readmission rates
  - Patients with CIPO have a longer length of inpatient stay than patients with ED.
  - Patients with ED have a longer length of inpatient stay than patients with CIPO
  - Patients with ED and CIPO have similar lengths of inpatient stay

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### Functional/dysmotility patients who do not meet CIPO or ED criteria

27. Have you ever referred patients for long term parenteral nutrition (PN) with suspected functional/dysmotility diagnoses who **do not** meet the diagnostic criteria for CIPO or ED?  
\*

-- Please Select --

28. If you answered yes to question 27, **what is the reason** that these patients do not meet the diagnostic criteria for ED or CIPO?

If you answered NO to Question 27, then please proceed to question 29.

- Small Bowel motility studies not available  
 Small Bowel motility studies not tolerated  
 Small Bowel motility studies either normal or not done, but enteral tube feeding not tolerated  
 Clinical decision not to order Small Bowel motility studies because these are not usually useful  
 Other, please specify

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### E. Home Parenteral Nutrition (HPN) dependent patients with CIPO and ED

If you do not look after patients on HPN then please answer '0' in question 29 to complete the survey.

29. Overall, approximately how many HPN dependent patients with **any cause** of Type III intestinal failure are currently under your care?\*
- 0  
 1-20  
 20-50  
 50-100  
 100-150  
 150-200  
 200-250  
 250-300  
 >300

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30. What proportion of your total HPN dependent patients have CIPO or ED?

- None (0%)  
 1-10%  
 10-25%  
 25-50%  
 50-75%  
 75-100%

31. What proportion of your total HPN dependent patients have a suspected **Functional/dysmotility** diagnosis that does **not** fulfil CIPO or ED criteria?

- None (0%)  
 1-10%  
 10-25%  
 25-50%  
 50-75%  
 75-100%

32. In your experience, how often does HPN have a role in reducing any of the following CIPO or ED related complications?

|                           | <b>Never<br/>(0%)</b> | <b>1-25%<br/>cases</b> | <b>25-50%<br/>cases</b> | <b>50-75%<br/>cases</b> | <b>75-<br/>100%<br/>cases</b> |
|---------------------------|-----------------------|------------------------|-------------------------|-------------------------|-------------------------------|
| Dehydration               | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Metabolic impairments     | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Quality-of-life           | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Bacterial Translocation   | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Gastrointestinal symptoms | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Aspiration pneumonia      | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Hospital admissions       | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |

33. In your experience, once HPN is commenced in patients with CIPO or and ED, approximately what proportion of patients will be HPN dependent at 5 years?

- 0-10%  
 10-25%  
 25-50%  
 50-75%  
 75-100%

34. In your experience, how often do the following factors determine long-term dependence on HPN in patients with CIPO or ED?

|                                  | <b>Never<br/>(0%)</b> | <b>1-25%<br/>cases</b> | <b>25-50%<br/>cases</b> | <b>50-75%<br/>cases</b> | <b>75-<br/>100%<br/>cases</b> |
|----------------------------------|-----------------------|------------------------|-------------------------|-------------------------|-------------------------------|
| Primary vs. secondary CIPO or ED | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |
| Histopathological diagnoses      | <input type="radio"/> | <input type="radio"/>  | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>         |

(Myopathy, neuropathy or normal)

|                                     |                       |                       |                       |                       |                       |
|-------------------------------------|-----------------------|-----------------------|-----------------------|-----------------------|-----------------------|
| Tolerance of oral or enteral intake | <input type="radio"/> |
| Manometry findings                  | <input type="radio"/> |
| Age at diagnosis                    | <input type="radio"/> |
| Long-term opiate use                | <input type="radio"/> |
| Psychological factors               | <input type="radio"/> |

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35. In your experience, how do catheter complication rates in HPN dependent patients with CIPO or ED compare to other causes of Chronic Intestinal Failure?

- Catheter complication rates are better than other causes of Chronic Intestinal Failure
- Complication rates are similar to those observed in other causes of Chronic Intestinal Failure
- Complication rates are worse when compared to other causes of Chronic Intestinal Failure

36. In your experience, does the incidence of **Intestinal Failure Associated Liver Disease (IFALD)** differ between patients with CIPO or ED when compared to other causes of Chronic Intestinal Failure?

- Rates of IFALD are better than other causes of Chronic Intestinal Failure
- Rates of IFALD are similar to that observed in other causes of Chronic Intestinal Failure
- Rates of IFALD are worse than in other causes of Chronic Intestinal Failure

37. How does the prevalence of **psychological co-morbidity** compare between patients diagnosed with ED and CIPO and other causes of Chronic Intestinal Failure?\*

|      | <b>Higher prevalence of psychological co-morbidity than others causes of Chronic Intestinal Failure</b> | <b>Similar prevalence of psychological co-morbidity to other causes of Chronic Intestinal Failure</b> | <b>Lower prevalence of psychological co-morbidity compared to other causes of Chronic Intestinal Failure</b> |
|------|---|---|--|
| CIPO | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>  |
| ED   | <input type="radio"/>   | <input type="radio"/>   | <input type="radio"/>  |

38. In your experience, how do **five-year survival outcomes** compare in HPN dependent patients with CIPO or ED when compared to Crohn's disease related Chronic Intestinal Failure?

- 5 year survival outcomes are better than Crohn's disease related Chronic Intestinal Failure
- 5 year survival outcomes are similar to those observed in Crohn's disease related Chronic Intestinal Failure
- 5 year survival outcomes are poorer when compared to Crohn's disease related Chronic Intestinal Failure

39. What is your experience of intestinal transplantation in patients with CIPO or ED who require HPN (select any that apply)?

- I would never consider referring a patient with CIPO or ED for intestinal transplantation
- I would never consider referring a patient with ED for intestinal transplantation
- I would never consider referring a patient with CIPO for intestinal transplantation
- I have no experience in referring any patients with Chronic Intestinal Failure requiring HPN for intestinal transplantation
- I would refer patients with CIPO or ED for intestinal transplantation if necessary.