

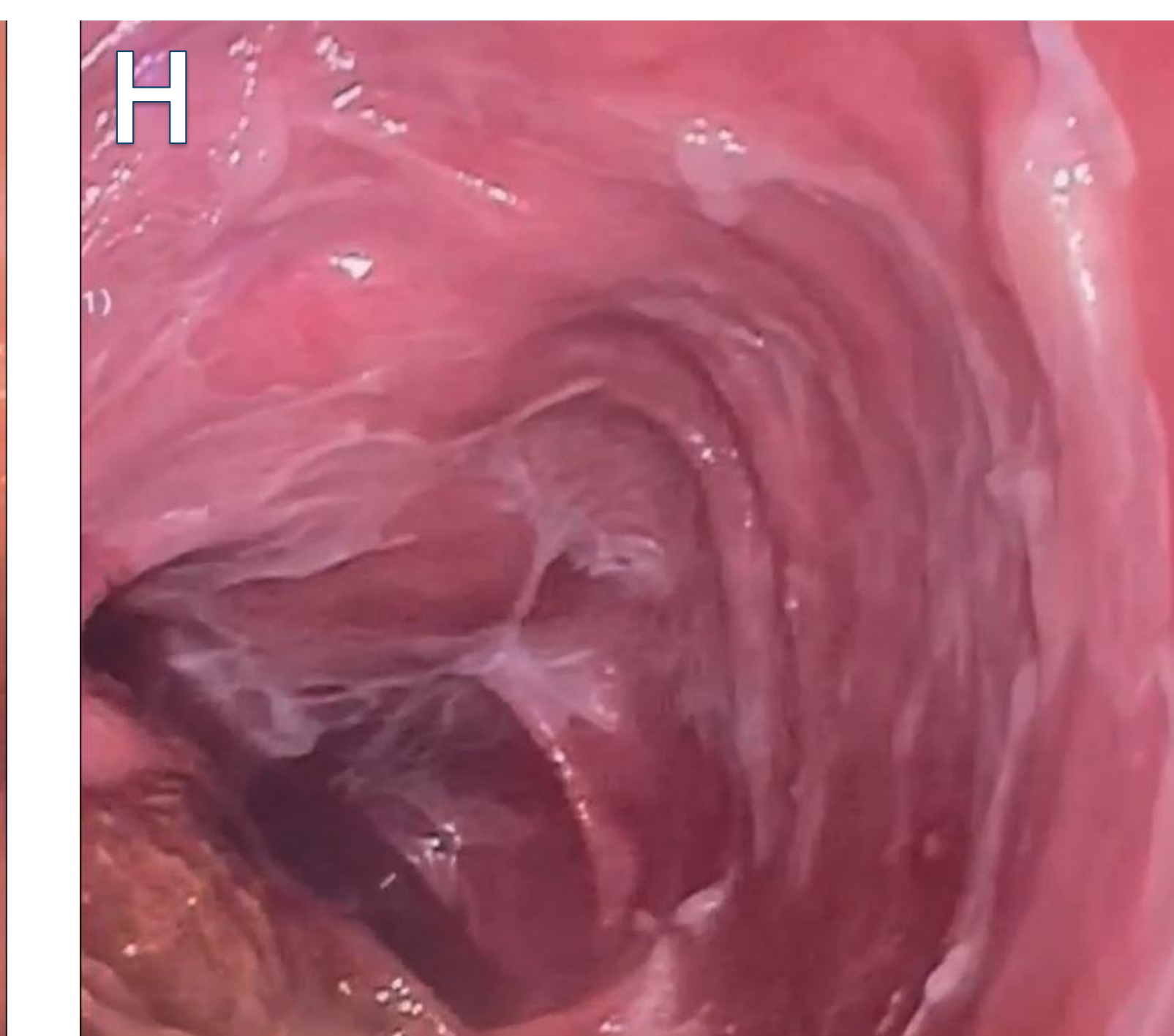
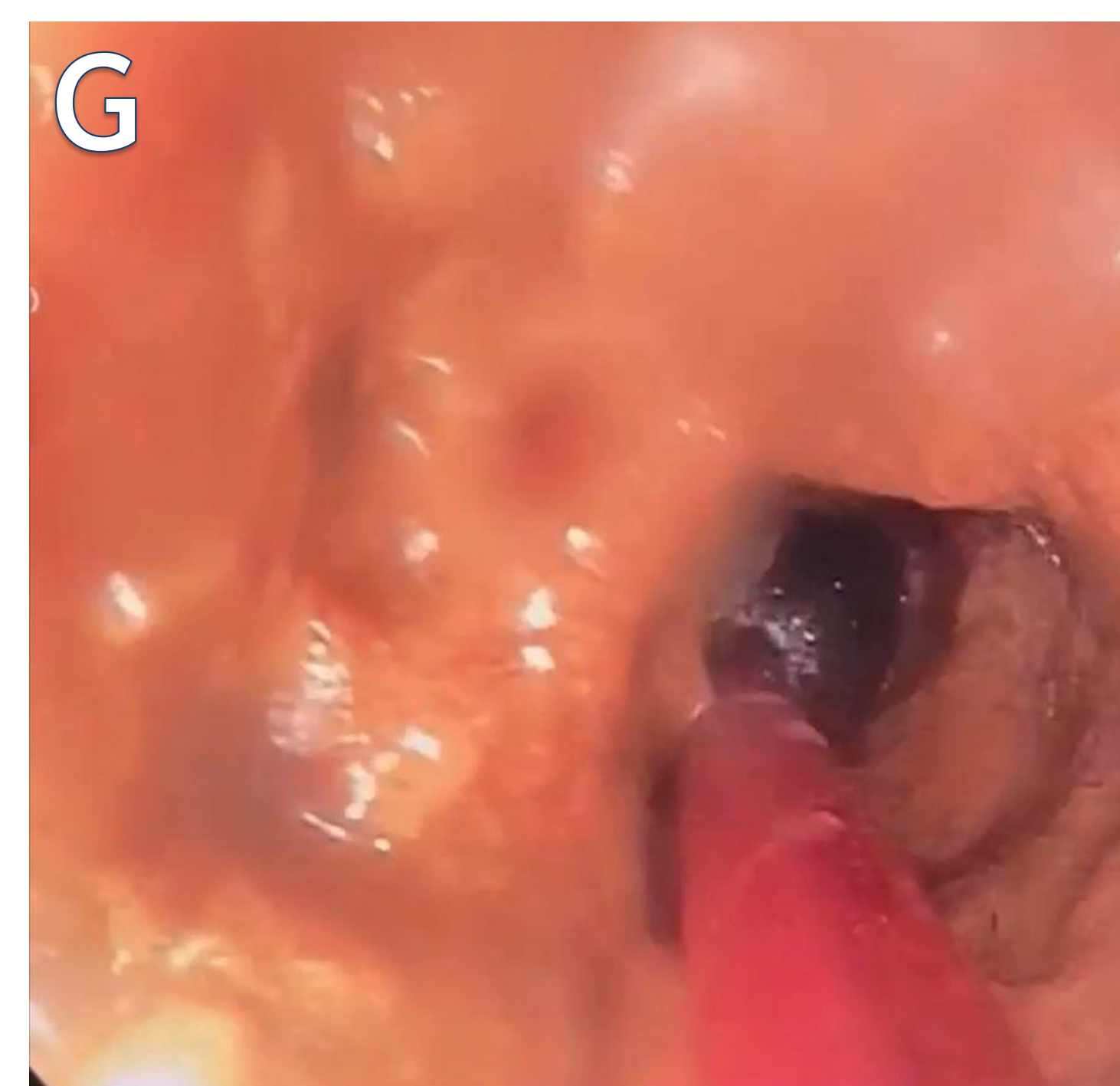
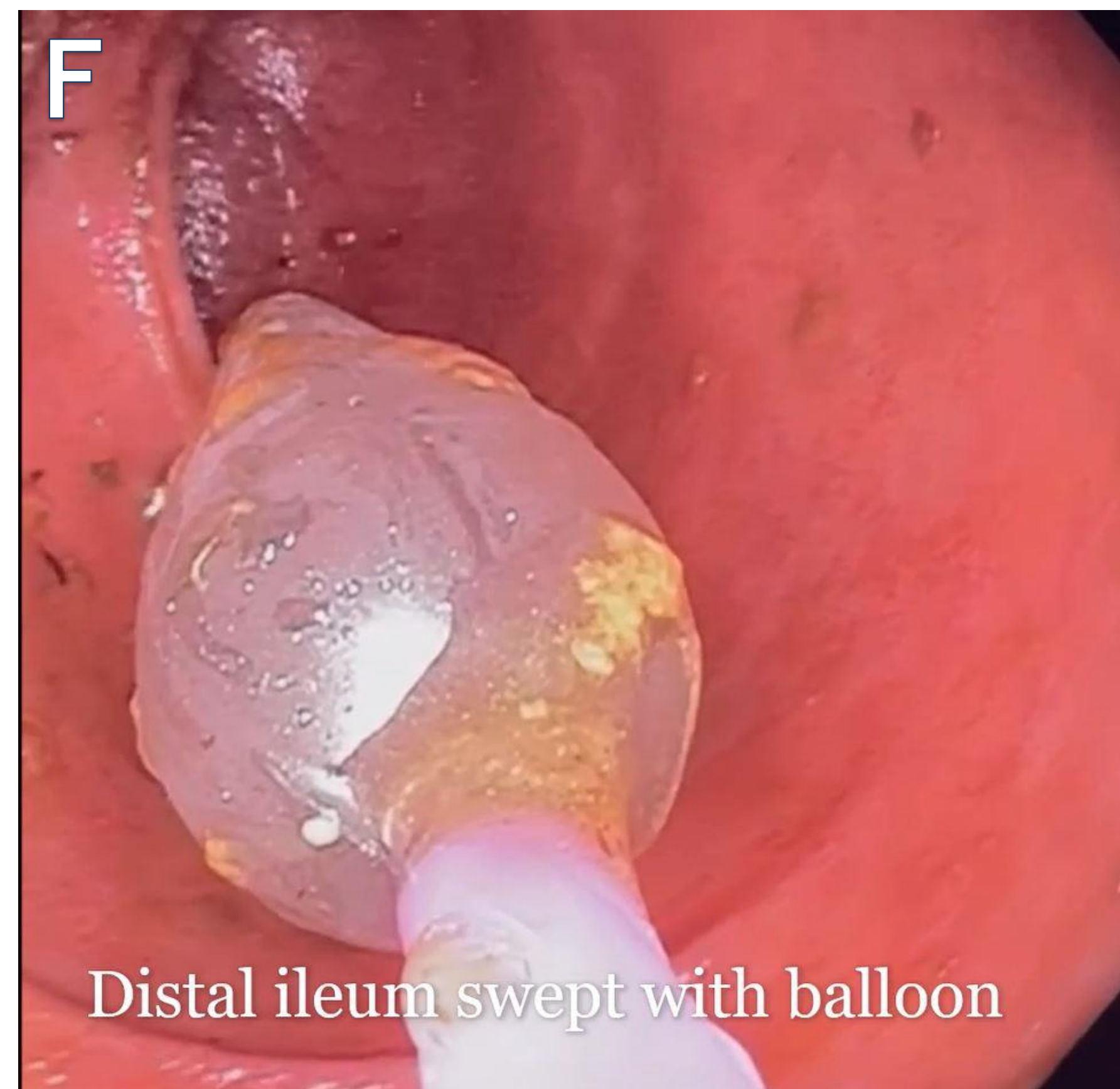
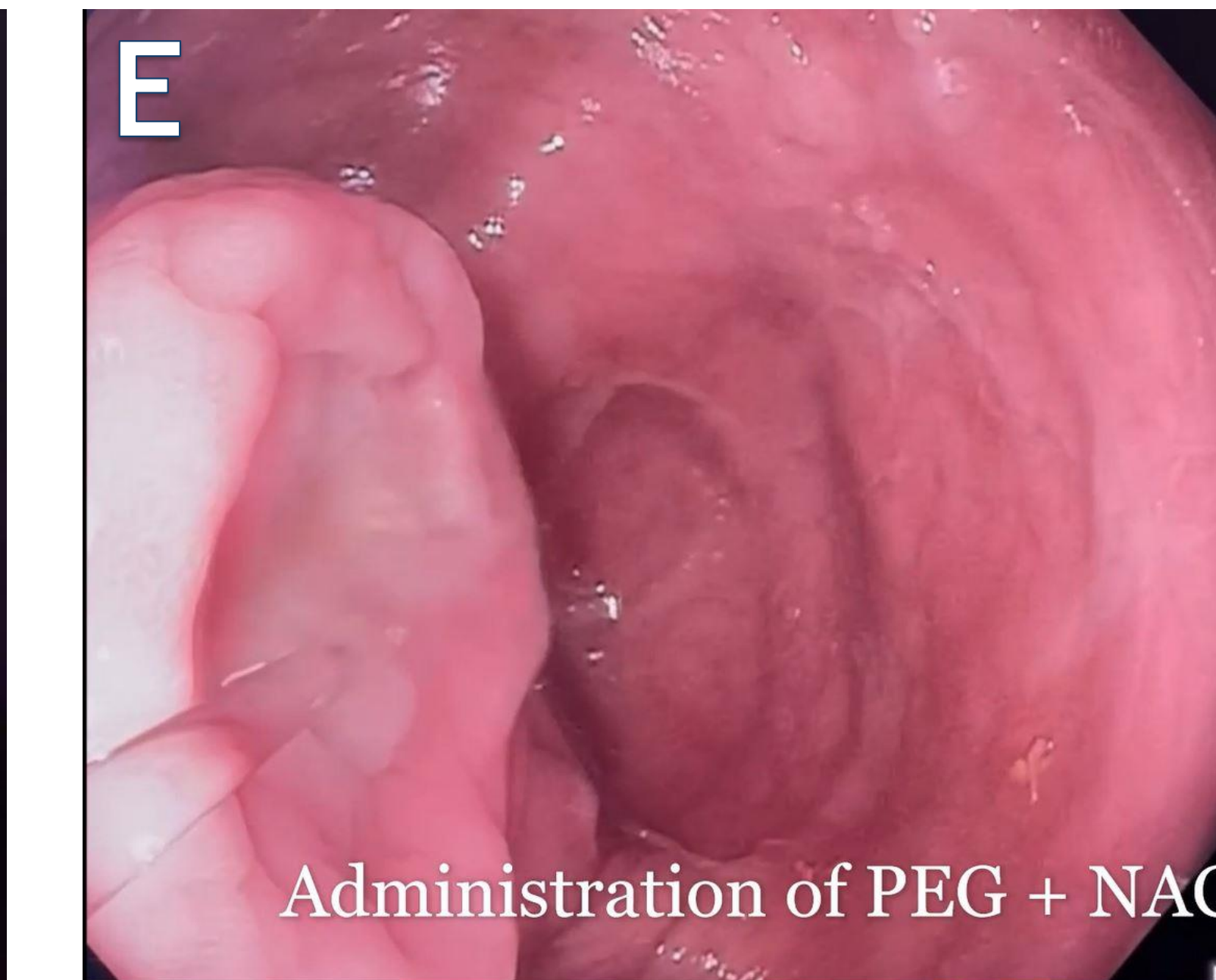
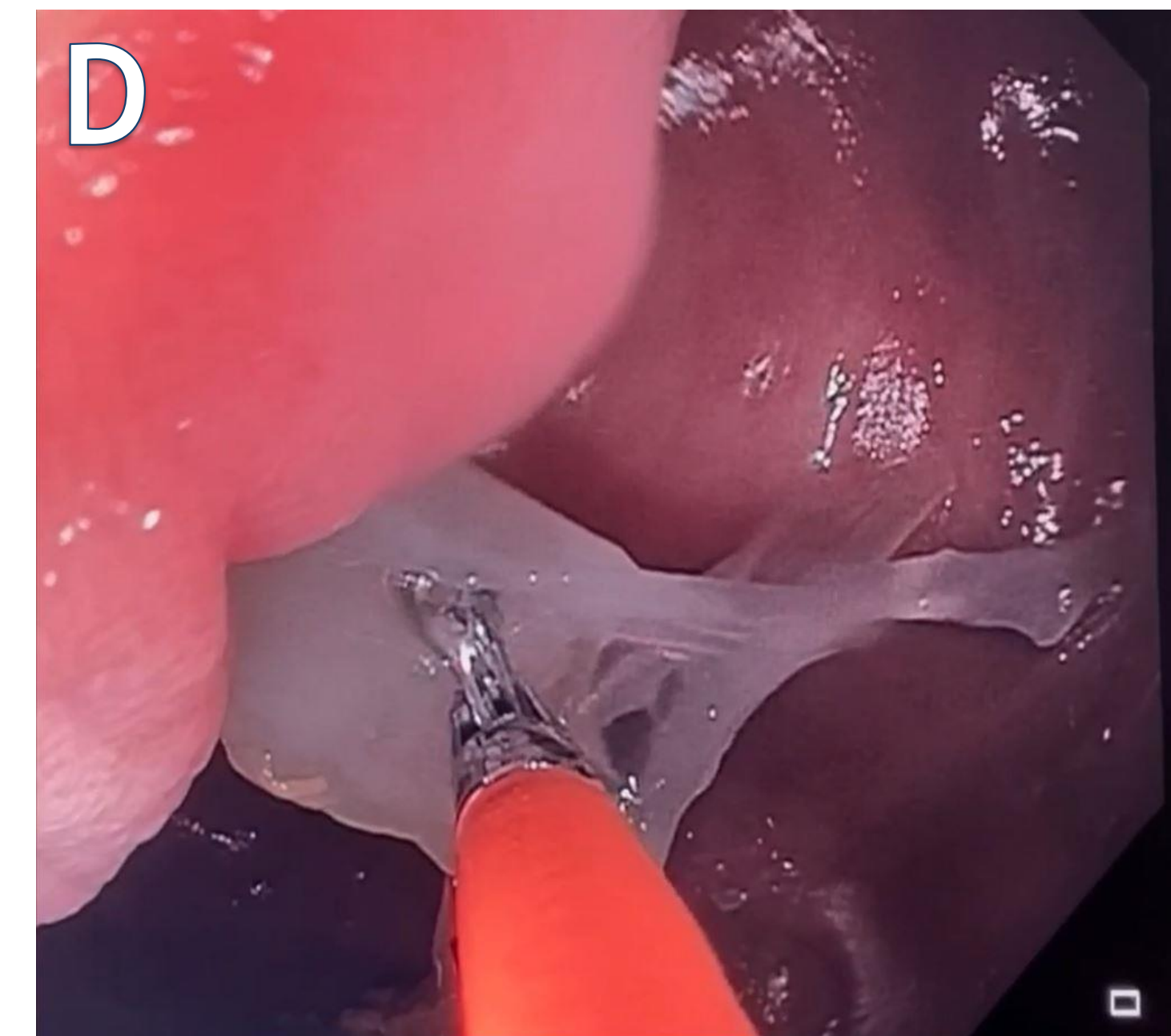
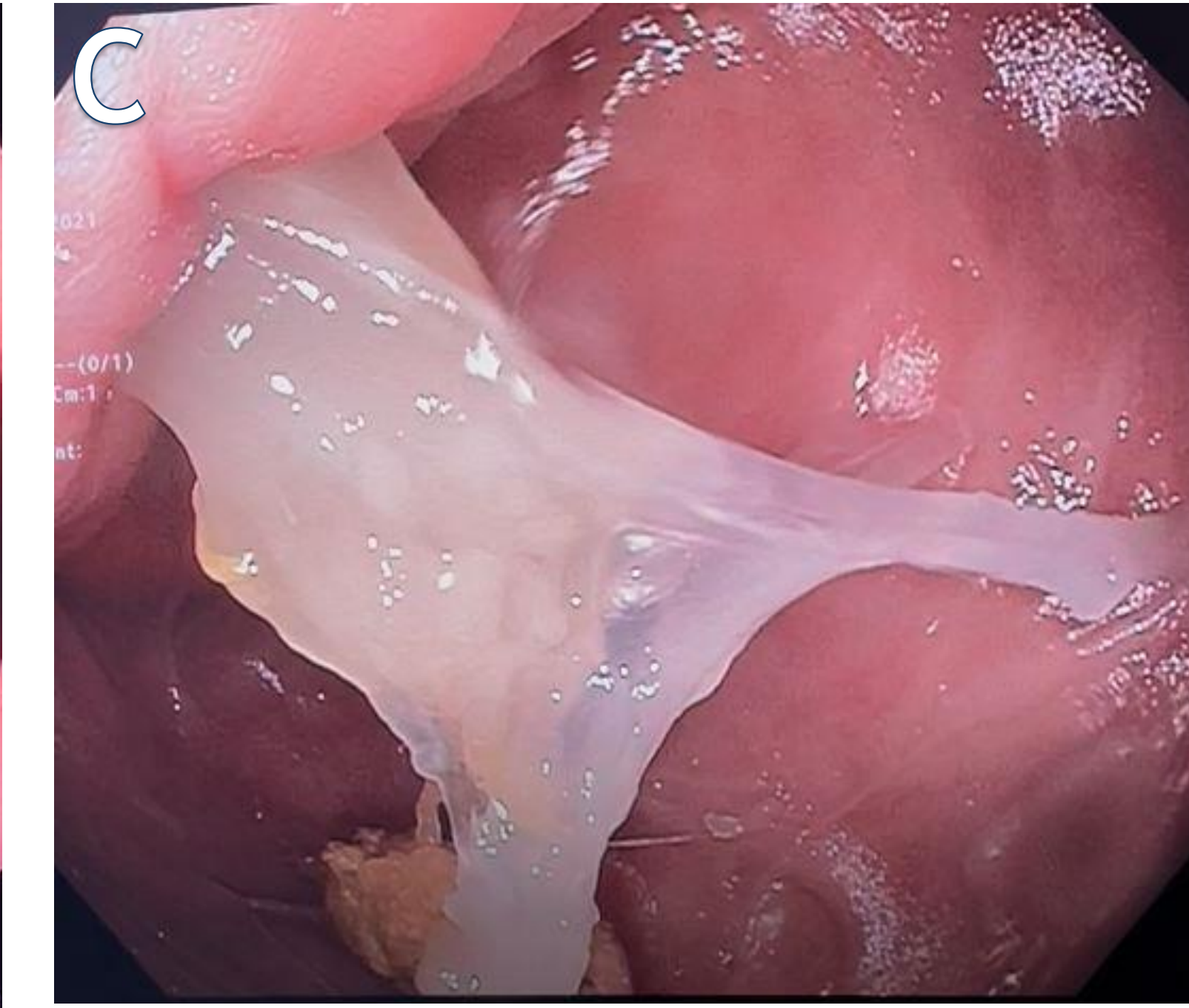
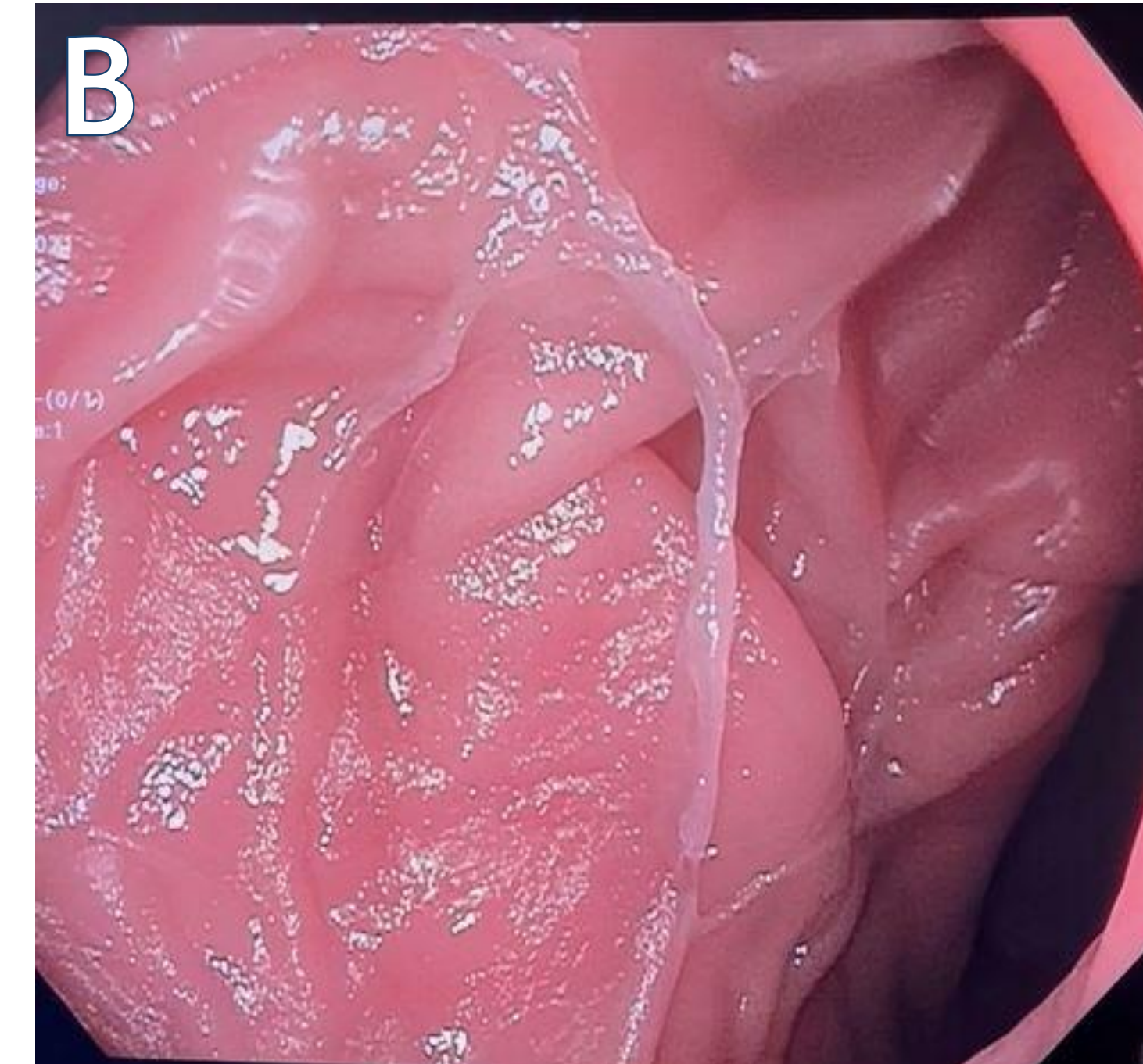


## Introduction

- Cystic fibrosis (CF) can lead to obstructive GI disease:
  - Meconium ileus in children
  - Distal intestinal obstruction syndrome (DIOS) in adults
- DIOS is characterized by:
  - accumulation of inspissated stool and viscous secretions in the terminal ileum and proximal colon
  - leads to complete or incomplete obstruction
- Currently, there are no RCTs to guide treatment of DIOS.
- We describe a case of a CF patient with DIOS, successfully treated with 4% N-acetylcysteine (NAC) and polyethylene glycol (PEG) administered via colonoscopy.

## Case Description

- A 46 yo woman with CF and prior episodes of DIOS presented with several days of abdominal pain and constipation.
- On exam, her abdomen was distended and diffusely tender to palpation.
- Abdominal CT showed extensive debris in the distal small bowel with resultant dilation proximally, thickening at the terminal ileum, and a decompressed colon (**Fig. A**).
- She was initially managed with frequent enemas and oral laxatives including Lubiprostone and PEG via nasogastric tube. Her symptoms worsened and she was taken for urgent colonoscopy.



## Case Description (continued)

- With the patient in supine position, the colonoscope was meticulously advanced to the cecum, traversing several matted, mucoid-adherent folds (**Fig. B**).
- A mucoid plug was seen extending from the ileocecal (IC) valve to the walls of the cecum (**Fig. C**). The mucoid plug was removed with standard biopsy forceps (**Fig. D**) and the terminal ileum (TI) was injected with 4% NAC and PEG (**Fig. E**).
- The scope was then advanced into the terminal ileum, approximately 20 cm to the proximal end of a large, actively dissolving fecalith with administration of additional NAC and PEG.
- A CRE 8-9-10 mm balloon was used to sweep the stool from the distal ileum (**Fig. F, G**). Following this maneuver, peristalsis was observed with inspissated stool emanating from the distal ileum towards the IC valve (**Fig. H**).
- Over the following days, the patient began to have several bowel movements daily while on oral PEG. She was discharged on daily PEG as needed in addition to her prior bowel regimen.

## Discussion

- Optimal management of DIOS in CF patients remains a challenge. Although most cases are relieved with conservative treatments, select patients will need surgery, which is associated with a higher peri- and postoperative morbidity.
- However, the administration of 4% N-acetylcysteine and polyethylene glycol via colonoscopy offers a viable remedy that may obviate the need for surgery.