



ECCO Guideline/Consensus Paper

# ECCO-ESCP Consensus on Surgery for Crohn's Disease

**Willem A. Bemelman,<sup>a</sup> Janindra Warusavitarné,<sup>b</sup> Gianluca M. Sampietro,<sup>c</sup> Zuzana Serclova,<sup>d</sup> Oded Zmora,<sup>e</sup> Gaetano Luglio,<sup>f</sup> Anthony de Buck van Overstraeten,<sup>g</sup> John P. Burke,<sup>h</sup> Christianne J. Buskens,<sup>a</sup> Francesco Colombo,<sup>c</sup> Jorge Amil Dias,<sup>i</sup> Rami Eliakim,<sup>j</sup> Tomás Elosua,<sup>k</sup> I. Ethem Gecim,<sup>l</sup> Sanja Kolacek,<sup>m</sup> Jaroslaw Kierkus,<sup>n</sup> Kaija-Leena Kolho,<sup>o</sup> Jérémie H. Lefevre,<sup>p</sup> Monica Millan,<sup>q</sup> Yves Panis,<sup>r</sup> Thomas Pinkney,<sup>s</sup> Richard K. Russell,<sup>t</sup> Chaya Shwaartz,<sup>u</sup> Carolynne Vaizey,<sup>b</sup> Nuha Yassin,<sup>v</sup> André D'Hoore<sup>g</sup>; on behalf of the European Crohn's and Colitis Organisation [ECCO] and the European Society of Colo-Proctology [ESCP]**

<sup>a</sup>Department of Surgery, Academic Medical Center [AMC], Amsterdam, The Netherlands <sup>b</sup>Department of Surgery, St. Mark's Hospital, Harrow, UK <sup>c</sup>Department of Surgery, ASST Fatebenefratelli Sacco – Ospedale "Luigi Sacco" Polo Universitario, Milan, Italy <sup>d</sup>Department of Surgery, NH Hospital, a.s., Horovice, Czech Republic <sup>e</sup>Department of Surgery, Sheba Medical Center, Tel Hashomer, Israel <sup>f</sup>Surgical Coloproctology Unit, University of Naples Federico II, Naples, Italy <sup>g</sup>Department of Abdominal Surgery, UZ Leuven, Campus Gasthuisberg, Leuven, Belgium <sup>h</sup>Department of Colorectal Surgery, Beaumont Hospital, Dublin, Ireland <sup>i</sup>Pediatric Gastroenterology Unit, Hospital S. João [University Hospital], Porto, Portugal <sup>j</sup>Department of Gastroenterology and Hepatology, Sheba Medical Center, Tel Hashomer, Israel <sup>k</sup>Servicio de Cirugía, Complejo Asistencial Universitario de León, León, Spain <sup>l</sup>Colorectal Unit, Ankara University Medical School, Ankara, Turkey <sup>m</sup>University Department of Paediatrics and Referral Center for Paediatric Gastroenterology & Nutrition, Children's Hospital Zagreb, Zagreb, Croatia <sup>n</sup>Department of Gastroenterology, Hepatology, Feeding Disorders, and Pediatrics, Children's Memorial Health Institute, Warsaw, Poland <sup>o</sup>Paediatric Gastroenterology of the Children's Hospital, University of Helsinki, Helsinki, Finland <sup>p</sup>Department of General and Digestive Surgery, Hôpital Saint-Antoine and University Paris VI, Paris, France <sup>q</sup>Department of Surgery, Hospital Universitari Joan XXIII de Tarragona, Tarragona, Spain <sup>r</sup>Department of Colorectal Surgery, Beaujon Hospital [APHP] and University Paris VII Denis-Diderot, Clichy, France <sup>s</sup>Academic Department of Surgery, University of Birmingham, Birmingham, UK <sup>t</sup>Department of Paediatric Gastroenterology, Royal Hospital for Children, Glasgow, UK <sup>u</sup>Department of Surgery, Sheba Medical Center, Ramat Gan, Israel <sup>v</sup>IBD Unit, University of Birmingham, Birmingham, St Mark's Hospital, London, UK

Corresponding author: W. A. Bemelman, PhD, Department of Surgery, Academic Medical Center, University of Amsterdam, Meibergdreef 9, 1105 AZ The Netherlands. Email: [w.a.bemelman@amc.uva.nl](mailto:w.a.bemelman@amc.uva.nl)

**Key Words:** Crohn's disease, guideline

## 1. Introduction

The goal of this consensus initiated by the European Crohn's and Colitis Organisation [ECCO] and European Society of Colo-Proctology

[ESCP] was to establish European consensus guidelines for the surgical treatment of Crohn's disease [CD]. The strategy to reach the consensus involved several steps and follows the standard operating

procedures for consensus guidelines of ECCO. An open call for chairs and participants for this consensus was made among members of the ESCP, surgeons of ECCO, and ECCO members. Participants were selected by the Guidelines' Committee of ECCO [GuiCom] and the guideline committee of the ESCP on the basis of their publication record and personal statement. Five working groups [WGs] were formed: WG 1, perioperative management; WG 2, upper gastrointestinal tract and small bowel; WG 3, large bowel; WG 4, perianal CD; and WG 5, special conditions. Participants were asked to answer relevant questions on current practice and areas of controversy related to the surgical treatment of CD based on their experience as well as evidence from the literature [Delphi procedure].<sup>1</sup> In parallel, the WG members performed a systematic literature search of their topic with the appropriate key words using Medline/PubMed/ISI/Scopus, and the Cochrane database, as well as their own files. Provisional guideline statements [with supporting text] were then written by the WG chairs based on answers to the questionnaire and were subsequently circulated among the WG members, prompting discussion and exchange of available evidence. The proposed statements and the supporting text were submitted to an online platform for discussion and two online voting rounds were conducted. The first round involved all participants of the consensus development process and, for the second voting round, all national representatives of ECCO and ESCP were invited to participate. The consensus participants finally met in Dublin on September 22, 2015 for face-to-face discussion and voting for consensus on the statements. This was done by projecting the statements and revising them on screen until consensus was reached. Consensus was defined as agreement by more than 80% of participants; the consensus statements were numbered for convenience in the document. The level of evidence was graded according to that published by the Oxford Centre for Evidence-based Medicine 2011 [[http://www.cebm.net/mod\\_product/design/files/CEBM-Levels-of-Evidence-2.1.pdf](http://www.cebm.net/mod_product/design/files/CEBM-Levels-of-Evidence-2.1.pdf)]. The final manuscript was written by the WG chairs [OZ, GS, JW, WB, ZS] in conjunction with the WG members and revised for consistency by WB and AD. Officially, WB is representing the ECCO and AD the ESCP. It was agreed to publish the consensus guideline in *JCC* and to post links to the original publication in *JCC* on the websites of ECCO and the ESCP.

The surgical treatment of CD was covered in the Third European Evidence-based Consensus on the Diagnosis and Management of Crohn's Disease 2016. Part 1: Diagnosis and Medical Management, and Part 2: Surgical Management and Special Situations.<sup>2</sup> The following guidelines add to the surgical aspects of the multidisciplinary management of Crohn's disease. The present document is to be seen as complementary to the surgical section published in the 2016 consensus. Some consensus statements are modified or expanded and others are entirely new. In order to have a complete overview,<sup>2</sup> the surgical statements of the existing Third European Evidence-based Consensus on the Diagnosis and Management of Crohn's Disease 2016: Part 1: Diagnosis and Medical Management, and Part 2: Surgical Management and Special Situations<sup>3</sup> are added, without supporting text.

## 2. General statements

A number of statements account for the general management of CD, and are listed in this section.

### 2.1. Existing statements

#### 2.1.1. ECCO Statement 6B

If a patient has a relapse, escalation of the maintenance treatment can be considered to prevent disease progression [EL2]. Steroids

should not be used to maintain remission [EL1]. Surgery should always be considered as an option in localised disease [EL4].

#### 2.1.2. ECCO Statement 7J

Whether there is a higher rate of postoperative complications from abdominal surgery during or after anti-tumour necrosis factor [TNF] therapy remains controversial [EL3].

#### 2.1.3. ECCO Statement 7K

Prednisolone 20 mg daily or equivalent for more than 6 weeks is a risk factor for surgical complications [EL2]. Therefore, corticosteroids should be weaned if possible [EL5].

## 2.2. ECCO-ESCP statements

### 2.2.1. ECCO-ESCP Statement 1A

Laparoscopy, when feasible, should be the preferred approach in surgery for CD. It results in reduced morbidity, shorter hospital stay, reduction in adhesions and hernia formation, and improved cosmesis [EL1]

### 2.2.2. ECCO-ESCP Statement 1B

Anti-TNF therapy is associated with higher postoperative septic complications after abdominal surgery for CD. The safest period for omission of anti-TNF therapy is unknown [EL2]

The impact of preoperative medical therapy on postoperative surgical complications has been widely studied. Prednisolone 20 mg daily or equivalent for more than 6 weeks before surgery is a significant risk factor for surgical complications. Therefore, steroids should be weaned if possible. Conversely, thiopurines can safely be continued in the perioperative period. In the past, the association of anti-TNF therapy before surgery with postoperative complications was controversial, but recent studies and meta-analysis have shown an increased risk of postoperative sepsis, intra-abdominal abscesses, anastomotic leak, wound infections, and readmission in CD patients. The combination of steroids and anti-TNF enhances the risk for complications. At present, no data are available to determine the safest period of discontinuation before surgery in order to reduce the risk related to anti-TNF therapy.<sup>4-12</sup>

## 3. Perioperative management

### 3.1. Indications for surgery

#### 3.1.1. Emergent setting

##### 3.1.1.1. ECCO-ESCP Statement 2A

Free perforation with peritonitis and massive haemorrhage unresponsive to other therapies are indications for emergency surgery in CD [EL3]

CD can present with acute complications requiring emergency surgery in approximately 6–16% of cases.<sup>13,14</sup> Patients with symptoms and signs of free perforation with peritonitis should undergo emergency surgery.<sup>13,15,16</sup> Resection of the diseased segment with or without anastomosis is the operation of choice in these cases.

Massive haemorrhage is a rare complication of CD.<sup>17,18</sup> Other causes of gastrointestinal [GI] tract bleeding should be excluded using endoscopy and interventional radiology. If these approaches are unsuccessful or unavailable, surgery is indicated.

Patients with CD who require emergency surgery should undergo fluid and electrolytes resuscitation to allow adequate tissue oxygenation before induction of anaesthesia; blood transfusions may be necessary in cases of haemorrhage. Broad-spectrum antibiotics should be used for prophylaxis or treatment of sepsis.<sup>19,20</sup> Septic patients undergoing emergency surgery are at risk for hypothermia, and aggressive corrective measures have been shown to improve survival after sepsis.<sup>20</sup> Foley urinary catheters and central venous access are recommended for monitoring optimisation. Preoperative stabilisation should not, however, delay emergency surgery.<sup>19,20</sup>

### 3.1.1.2. ECCO-ESCP Statement 2B

Patients with CD who present with acute severe colitis should be under daily and interdisciplinary surveillance by a specialised gastroenterologist and surgeon, in haemodynamically stable patients without peritonitis. [EL5]. Any clinical deterioration or failure to improve within approximately 1 week despite optimal medical treatment should prompt consideration for an emergent surgery [EL3]

Acute severe colitis in CD often presents a clinical scenario undistinguishable from severe colitis of UC.<sup>21</sup> These patients should be hospitalised for investigation and treatment, and close monitoring by a multidisciplinary team including a gastroenterologist and colorectal surgeon.<sup>22</sup> Medical treatment for acute severe colitis should be initiated<sup>21,22</sup> and clinical signs and blood tests should be monitored daily to detect signs of systemic toxicity. Any clinical deterioration or signs of toxicity should prompt consideration of emergent colectomy.<sup>23</sup> Recommendations for overall surgical treatment are similar to the management of acute severe colitis in UC.<sup>22</sup>

### 3.1.1.3. ECCO-ESCP Statement 2C

Acute intestinal obstruction caused by an inflamed or fibrotic segment should be initially treated with conservative measures. Emergency surgery is indicated in rare cases of complete bowel obstruction, or if bowel ischaemia is suspected. In cases of partial bowel obstruction nonresponsive to medical therapy, surgery can usually be scheduled after the patient is optimised [EL4]

Acute intestinal obstruction is characterised by nausea, vomiting, a distended abdomen, and absence of gas or stool passage per rectum, and usually responds to conservative management. These measures include bowel rest, intravenous hydration, and nasogastric decompression. Intravenous steroids should be considered in active inflammatory disease.<sup>14</sup> If there are signs of peritoneal irritation or suspected bowel ischaemia, emergency surgery may be indicated, and a resection of the diseased segment should be performed.<sup>6</sup>

In the majority of cases, the patient will present with several episodes of partial obstruction alleviated by reducing oral intake and/or increasing medication. When there is partial bowel obstruction nonresponsive to medical therapy, cross-sectional imaging is recommended to define disease extent and location,<sup>24</sup> and the nutritional and immunosuppression status may be optimised before surgery.<sup>14</sup>

## 3.1.2. Elective setting

### 3.1.2.1. ECCO-ESCP Statement 2D

Surgery is the preferred option in patients with localised ileocaecal CD with obstructive symptoms but no significant evidence of active inflammation [EL4]

Short ileal stenosis in a patient naive for surgery is a common indication for surgery, especially if there is no significant evidence of active inflammation.<sup>6</sup> The definition of 'short' is arbitrary and also depends on the remaining amount of small bowel. Alternatives to surgery include endoscopic treatment with dilatation<sup>25,26</sup> or stenting<sup>27,28</sup> for accessible strictures shorter than 5 cm.

### 3.1.2.2. ECCO-ESCP Statement 2E

Patients with active inflammation should generally be first treated by medical treatment. In cases of failure of conservative therapy, surgery is indicated [EL5]

Patients with symptoms and signs of active inflammatory disease are usually treated first with medications as suggested in the ECCO guidelines for medical treatment of CD.<sup>6</sup> If symptoms are not controlled by medical treatment or patients are steroid-dependent, surgery should be considered. The term 'nonresponse to medical therapy' is hard to define, as there are often alternative medical treatments that have not been exhausted. The potential benefit of any medication should be weighed against the additional surgical risk should this medication fail to achieve symptomatic relief.

The choice between medical and surgical treatment in case of a short segment can be difficult and both options are usually legitimate. Multidisciplinary team discussion for case-by-case decision making and attention to the patient's wishes are recommended. An ongoing randomised study compares surgery and medical treatment in patients with short segment ileal disease and may help in treatment selection in the future.<sup>29</sup>

### 3.1.2.3. ECCO-ESCP Statement 2F

In patients with symptomatic perforating/fistulising disease, surgery should be considered at an early stage [EL4]

In perforating CD, surgery should be considered at an early stage. In patients with significant symptoms owing to fistulas between diseased bowel loops and adjacent organs, there is a higher risk of nonresponse to medical treatment. This should be weighed against the additional risk of surgery following prolonged medical treatment. In patients with intra-abdominal abscesses who have had successful percutaneous drainage, several case series favour delayed elective resection.<sup>30</sup> Patients with a concomitant stenosis, an enterocutaneous fistula, or refractory active disease are more likely to need surgery. An ongoing French prospective study from the GETAID group [MICA study] may help guide treatment in these patients in the future [personal communication]. The finding of an interloop fistula in an asymptomatic patient should not be considered an absolute indication for surgery [incidental finding at MR or CT enterography].

### 3.1.2.4. ECCO-ESCP Statement 2G

Surgery should be considered in children with CD in pre-pubertal or pubertal age if height velocity for bone age is reduced over a period of 6–12 months in spite of an optimised medical and nutritional therapy [EL4]

In children with CD, growth retardation is a major indication for elective surgery as the window of opportunity may be narrow.<sup>31</sup> It is important to assure that conservative treatment with measures such as an elemental diet are optimised before the decision of surgery is made, since surgery is not curative and the disease may relapse after surgery.<sup>32–34</sup> Refusal, intolerance, or increased risks of maintenance medications such as immunomodulators or anti-TNF $\alpha$  agents may also be considered as relative indications for elective surgery.

## 3.2. Preparation for surgery

### 3.2.1. ECCO-ESCP Statement 2H

Intra-abdominal abscess should initially be treated with antibiotics and/or percutaneous drainage. Emergency surgery is only indicated if unresponsive to treatment or percutaneous drainage is not feasible [EL3]

Intestinal imaging plays a vital role in distinguishing between active CD and septic complications. Intra-abdominal abscesses should initially be treated conservatively with antibiotics and/or percutaneous drainage [if > 5cm], whereas for active CD, medical therapy is first-line treatment. Large abscesses may not resolve with antibiotics and early percutaneous drainage is recommended.<sup>6,30</sup>

Emergency surgery may be necessary if patients who do not respond to conservative treatment become septic.<sup>34</sup> Surgery for septic complications carries a significant risk of anastomotic leak, recurrent abscesses, and fistulas. A staged procedure includes resection of the diseased segment, drainage of abscess, and a stoma in the first stage, and anastomosis in the second stage should be considered in these cases.

### 3.2.2. ECCO-ESCP Statement 2J

Malnutrition is a significant risk factor for postoperative complications. Nutritional status should be optimised before surgery via enteral or parenteral routes [EL 3]. If surgery is required in a malnourished patient, a staged procedure is advised [EL5]

Patients who have lost 10% or more of their body mass in a 3-month period are considered significantly malnourished and may benefit from preoperative nutritional support by either enteral or parenteral nutrition.<sup>35–37</sup> Malnutrition is a significant risk factor for postoperative complications,<sup>35</sup> including anastomotic leak, as good nutrition aids wound and tissue healing. Serum albumin level is not a proper marker of nutrition, as albumin level may be influenced by multiple factors including acute or chronic sepsis. Various other assessment tools should be used in addition to the albumin level.

There is no difference between elemental and polymeric enteral diet for preoperative optimisation in adults. Total parenteral nutrition should be considered for malnourished patients who cannot tolerate oral diet, including enteral formulas.<sup>37</sup> In patients requiring surgery while significantly malnourished, a staged procedure which includes resection of the diseased segment with formation of a stoma, followed by delayed restoration of continuity, is recommended.

### 3.2.3. ECCO-ESCP Statement 2K

Thiopurines can safely be continued in the perioperative period and beyond [EL2]

Most available literature shows no association between the use of azathioprine and an increased risk of surgical complications.<sup>6,38</sup> Discontinuation of thiopurines before surgery is not required.

### 3.2.4. ECCO-ESCP Statement 2L

When planning abdominal surgery for CD, it is advised to have a recent assessment of the disease locations, including ileocolonoscopy and cross-sectional imaging [EL3]

Ileocolonoscopy and biopsies from the terminal ileum and colon are routine procedures in establishing the diagnosis of CD.<sup>39</sup> Before scheduling surgery, a recent endoscopy should be reviewed by the multidisciplinary team. Cross-sectional imaging has evolved in recent years, and at present is widely used for the assessment of CD extension and both mural and peri-enteric involvement.<sup>24</sup> Computed tomography [CT], magnetic resonance [MR] and ultrasonography [US] have high and comparable diagnostic accuracy for the diagnosis of CD.<sup>24</sup> Recent ECCO-ESGAR guidelines<sup>40</sup> and ESPGHAN guidelines<sup>41</sup> recommend MR enterography as the modality of choice for CD evaluation, as it can identify disease site and length, assess disease activity, and detect complications such as abscesses, fistulas, and strictures. The disadvantages include high cost and limited availability. It also requires high patient compliance and a considerable amount of oral contrast for adequate distension of the lumen.

In children, imaging with MRI is the method of choice and preferred over CT as radiation should be avoided in young patients. In localised disease, transabdominal US is an additional imaging tool but requires significant operator expertise.<sup>41</sup> Both CT and MR have been shown to accurately predict intraoperative findings in CD.<sup>42,43</sup>

### 3.2.5. ECCO-ESCP Statement 2M

Patients having major abdominal surgery with recent history of chronic steroid use should receive perioperative steroid replacement therapy [EL4]

'Stress doses' of steroids doses have routinely been considered the perioperative standard of care over the past decades for patients on long-term steroid therapy. This practice is mainly based on two case reports from the 1950s of postoperative death of patients undergoing major surgery while chronically treated with high-dose steroids.<sup>44,45</sup> Patients chronically receiving steroids before surgery will commonly have a 'steroid stress dose'. There is little evidence to support this practice over simple continuation of the preoperative steroid doses. Small randomised controlled trials, several retrospective studies, and a Cochrane review did not show beneficial effect of preoperative steroid stress dose.<sup>46,47</sup> This widely used practice should be challenged, and a randomised controlled trial [RCT] should be encouraged to generate the needed evidence. Budesonide has a limited systemic effect; however, long-term use might be an indication for substitution.

### 3.2.6. ECCO-ESCP Statement 2N

Patients with CD requiring surgery are at a higher risk for venous thromboembolism and prophylaxis measures should be considered unless contraindicated [EL2]

Patients with CD requiring surgery are at a higher risk for venous thromboembolism [VTE], and the combination of mechanical and pharmacological prophylaxis should be considered unless

contraindicated.<sup>48–50</sup> Extended VTE prophylaxis for 28 days is recommended for IBD patients.

In children, the absolute risk of VTE is low<sup>14</sup> but, similar to adults, additional risk factors including central venous catheter, parenteral nutrition, an identified hypercoagulable condition,<sup>51</sup> steroid use,<sup>50</sup> and an active disease<sup>52</sup> may increase this risk. There are no published trials for the efficacy and safety of thromboprophylaxis related to CD surgery in children. Therefore, the risks and benefits need to be considered individually.

### 3.3. Early postoperative care

#### 3.3.1. ECCO-ESCP Statement 20

Application of the principles of enhanced recovery after elective intestinal surgery [ERAS] is recommended for CD, and conveys advantage in postoperative recovery [EL 2]

Numerous studies have demonstrated the advantages of enhanced recovery protocols after colorectal surgery, with a shorter hospital stay and lower overall complication rates.<sup>53,54</sup> For this reason, the principles of fast track rehabilitation after colorectal surgery should be applied whenever possible to the surgical management of CD patients.

One study has specifically addressed the application of early recovery after surgery [ERAS] protocol in surgery of CD. Spinelli *et al.* compared 20 consecutive patients undergoing elective laparoscopic ileocaecal resection for CD with enhanced recovery pathway with 70 patients with conventional perioperative care.<sup>55</sup> Hospital stay, time to first flatus, and first bowel movement were significantly shorter in the enhanced recovery group.

Recommendations of ERAS protocols may include: preoperative counselling; preoperative cessation of smoking and alcohol; no mechanical bowel preparation; clear fluids allowed up to 2 h and solids up to 6 h before induction of anaesthesia; use of well-fitting compression stockings, intraoperative intermittent pneumatic compression, and pharmacological prophylaxis with low-molecular-weight heparin; antibiotic prophylaxis; and early ambulation. Extended VTE prophylaxis is also recommended for 28 days in patients with IBD.<sup>56</sup>

## 4. Upper gastrointestinal tract and small bowel

### 4.1. Mouth, oesophagus, stomach and duodenum

#### 4.1.1. General considerations

##### 4.1.1.1. ECCO-ESCP Statement 3A

CD affects the upper GI tract more commonly in children than in adults, but need for surgery does not seem to be increased [EL2]

Upper gastrointestinal [GI] Crohn's disease is a well-known phenotypic feature of childhood onset of CD. This finding has been recently confirmed in an audit of diagnostic work-up of children with newly diagnosed inflammatory bowel diseases [IBD] recruited within the European Paediatric IBD Registry [EUROKIDS registry].<sup>57</sup> In this cohort, 585 of 714 patients [82%] completed the whole diagnostic work-up known as 'Porto Criteria' which included

endoscopic visualisation and biopsies of the upper GI, colon, and terminal ileum [IBD Working Group 2005].<sup>58,59</sup> Isolated upper GI involvement [L4] was diagnosed in 4% [ $n = 21$ ] of patients, and 30% of children had macroscopic upper GI disease [ $n = 172$ ] either isolated or together with the involvement of other intestinal segments.<sup>57</sup> In paediatric patients with CD without overt upper GI symptoms, isolated granulomas in the upper GI tract were detected in 2% to 21% of the cases.<sup>60</sup> Despite the high frequency of the upper GI involvement in children with CD, there are almost no paediatric data on the necessity of surgery for this disease location.<sup>61</sup> Therefore, it seems that more frequent upper GI CD involvement does not require more frequent surgical treatment.

#### 4.1.2. Mouth

Oral lesions in CD have an overall prevalence of 0.5–20%, with a large number of different manifestations. Lip swelling with or without fissures, oral mucosal swelling or cobblestoning, deep linear ulcerations, mucosal tags, mucogingivitis, glossitis, and palatal ulcerations are all typical presentations. The main differential diagnosis is orofacial granulomatosis, where the lesions may be found in the absence of gastrointestinal disease. Patients with oral manifestations of CD tend to be male, present at a younger age [paediatric onset], have upper GI involvement, and perianal involvement. Gastroenterologist and colorectal surgeons working in an IBD multidisciplinary team need to be familiar with oropharyngeal lesions.<sup>62–64</sup>

#### 4.1.3. Oesophagus

##### 4.1.3.1. ECCO-ESCP Statement 3B

The need for surgical treatment of oesophageal lesions is very limited. The lesions may require endoscopic dilatation or segmental resections. Specific cases should be discussed during multidisciplinary meeting with an upper-GI surgeon [EL5]

Oesophageal CD is a rare condition and almost never occurs in the absence of extraintestinal manifestations.<sup>57–59</sup> Oesophageal CD is relatively more frequent in childhood onset disease compared with other locations. The prevalence of gastro-oesophageal symptomatic lesions of CD varies from 5% to 13% both in adults and in children. The identification of macroscopic and microscopic lesions at endoscopy varies from 30% to 64%, but the clinical significance of these findings remains uncertain. Upper GI endoscopy is indicated in the assessment of all paediatric and adolescent IBD patients to confirm a diagnosis of CD, and to differentiate between CD and ulcerative colitis [UC]. It is unclear if there is a need to perform routine upper GI endoscopy in asymptomatic adults.<sup>59,65</sup> Medical and surgical treatment of oesophageal lesions is only reported as case reports or small retrospective series highlighting the uncommon nature of these lesions. Classic immunomodulatory therapy is used for medical treatment, but complicated disease may necessitate a surgical approach. A few cases of oesophageal stricture and fistulas have been reported and patients typically report dysphagia, odynophagia, and retrosternal pain with eventual weight loss as the main symptoms. Most fistulas are connected with the bronchial system, either at the level of the bronchi or the level of the trachea, or to the pleural cavity. Some oesophagogastric fistulas have been described. Most fistulas will need surgery. The use of glue sealants has been reported, without any success. The successful use of covered stents has been described in very sporadic cases. Most patients with

fistulas will need an oesophagectomy which carries a risk of major morbidity and mortality. Oesophagogastric fistulas can be treated conservatively because they are mostly asymptomatic. Strictures are best treated conservatively by endoscopic dilatation. In rare cases, oesophagectomy may need to be performed.<sup>66–70</sup>

#### 4.1.4. Stomach and duodenum

##### 4.1.4.1. ECCO-ESCP Statement 3C

CD of the stomach typically affects the antrum and pylorus. If surgery is required, it usually includes distal gastrectomy and Roux-en-Y gastro-jejunostomy. Routine vagotomy is generally contraindicated in Crohn's patients [EL4]

##### 4.1.4.2. ECCO-ESCP Statement 3D

In CD of the antrum and duodenal bulb, stricturoplasty and Roux-en-Y bypass are considered effective surgical options. Strictures of the second and third duodenal portions are best treated, when technically possible, with stricturoplasty. Duodenal resection or pancreatoduodenectomy have to be considered as last resort. Routine vagotomy is contraindicated EL3

##### 4.1.4.3. ECCO-ESCP Statement 3E

When the stomach or the duodenum is non-diseased target of entero-enteric fistulas, these should be repaired with the most conservative technique possible [eg direct repair, tangential resection, stricturoplasty] [EL4]

Primary CD disease of the stomach and duodenum is an uncommon presentation, and the exact incidence and prevalence are unclear due to the lack of population-based studies. On the basis of clinical symptoms and radiographic findings, the incidence of gastroduodenal lesions is reported to occur in 0.5–5% of patients. In contrast, looking at endoscopic findings, macroscopic or microscopic inflammation is found in 60% and 70% of cases, respectively. The most common symptoms are upper abdominal pain, nausea and vomiting, weight loss, diarrhoea, haematemesis, and anaemia. Symptoms are initially managed with proton pump inhibitors [PPI] and anti-inflammatory drugs. Steroids, immunomodulators, and anti-TNF agents have been reported to be effective. The main indication for surgery in primary gastro-duodenal CD is for stricturing complications. The majority of the symptomatic patients eventually require surgery. The strictures are located in the distal stomach and duodenal bulb in 50–60% of patients, in the second and third duodenal portion in 25–30%, in the third and fourth duodenal portion in 10–15%; 10% of patients have strictures in multiple locations. Strictures in the antrum and pylorus are most commonly treated by a distal gastrectomy with Roux-en-Y reconstruction. An alternative to resection is a laparoscopic bypass. Duodenal strictures are best treated, when technically feasible, with stricturoplasty as there is no possibility of a blind loop, dumping, or anastomotic ulcerations. The incidence of recurrent CD in a stricturoplasty site is very low. Both the stomach and the duodenum have been described as targets of entero-enteric fistulas originating from a diseased segment of the colon or ileocolonic anastomosis. In these cases, it is important to know how clinically relevant the fistula is

in terms of electrolyte imbalance and malnutrition. Surgery must be as conservative as possible, preferably with wedge resection and primary closure. Postoperative complication of stricturoplasty and bypass has been high in older series [32–38%]. More recent data show rates below 15%. Five-year local surgical recurrence is 5–8% for gastroduodenal lesions, but since almost all these patients have concomitant jejunoileal and/or colonic disease, the overall recurrence of CD is considerably higher. Due to the small number of cases reported in literature, the best medical prophylaxis after surgery is not known.<sup>71–78</sup>

#### 4.2. Jejunum and ileum

It is important to have determined and documented the exact number, length, and features [inflammatory, stricturing, and fistulising] of all the lesions of the small intestine before embarking on surgery. In patients with multiple locations, not all the diseased segments present with the same level of inflammation or the same type of complication [stricture and fibrosis, perforation, or both]. The Montreal and Paris Classifications for CD are useful tools and should be used in the surgical reports, since they impact on the postoperative treatment and the long-term prognosis. Ultrasonography [US], computed tomography [CT], and magnetic resonance imaging [MRI] are all reliable tools for diagnosis of small bowel CD complications, with comparable diagnostic accuracy, but their overall sensitivity and specificity are reported to be 75–85% and 85–98%, respectively.<sup>24,40,79–85</sup> It is not infrequent to find intraoperatively more locations [mainly short skip lesions] than there were identified in the preoperative workup; a balloon catheter can be used to assess the luminal diameter and assist with the decision to perform a stricturoplasty. There is no role for 'preventive' stricturoplasty.

##### 4.2.1. Existing statements

###### 4.2.1.1. ECCO Statement 7C

Stricturoplasty is a safe alternative to resection in jejuno-ileal CD, including ileocolonic recurrence, with similar short-term and long-term results. Conventional stricturoplasty is advised when the length of the stricture is < 10 cm. However, in extensive disease with long strictured bowel segments where resection would compromise the effective small bowel length, non-conventional stricturoplasties may be attempted [EL3].

##### 4.2.2. ECCO-ESCP statements

###### 4.2.2.1. ECCO-ESCP Statement 3F

Diagnosis during childhood, jejuno-ileal location, and stricturing behaviour are independent risk factors for elevated risk of surgery and high surgical recurrence rates for small bowel CD [EL2]

Historically, patients with penetrating complications were believed to have more aggressive disease, with an elevated risk of clinical and surgical recurrence. Despite limited evidence for this assumption, in the Vienna, Montreal, and Paris Classifications patients with perforating disease are scored as having the highest level of disease complication. This may be true for terminal ileitis and Crohn's colitis, but young patients with concomitant jejuno-ileal location and a stenotic pattern have a surgical recurrence hazard ratio 13 times higher. Surgical recurrence rates as high as 50% and 70% at 5 and 10 years are reported in younger patients, whereas other patients have recurrence rates of only 10% and 25% at 5 and 10 years, respectively.<sup>86–89</sup>

**4.2.2.2. ECCO-ESCP Statement 3G**

After intraoperative identification of the number, the locations, and the characteristics of the small bowel lesions of CD, the treatment should be customised 'per-segment' on the basis of the characteristics of each single lesion [EL2]

The first step of each surgical procedure for complicated CD must be a complete assessment of the bowel. A complete and careful adhesiolysis is needed to detect entero-enteric fistulas or short skip lesions. Once all the CD locations have been identified, a tailored approach can be performed. The presence of complications, such as localised sepsis [fistula/abscess] and the absence of a residual lumen, are main indications for segmental resection. Fazio *et al.* demonstrated in a randomised controlled trial that CD surgical recurrence is unaffected by the width of resection margins from macroscopically and microscopically involved bowel.<sup>90</sup> Strictureplasties were first described in CD patients by Emanuele Lee and Nicos Papaioannou in 1982,<sup>91</sup> and then popularised by Victor W. Fazio<sup>92</sup> and John Alexander-Williams<sup>93</sup> few years later. Today several strictureplasty techniques have been described for any type of primary and recurrent stenosis of the small bowel and ileocolic region. Early and late outcomes are comparable to resectional surgery. Classical indications for strictureplasty are stricturing disease of a large portion of small bowel, previous bowel resection of more than 100 cm, short-bowel syndrome, strictures without an associated abscess or phlegmon, recurrent strictures at ileocolic anastomotic sites, strictures occurring within a year from previous surgery, and growth retardation in children. Strictureplasty can also be used as a first-line treatment whenever technically feasible. In most cases with multiple jejunoileal locations, the typical surgical procedure is based on a combination of small bowel resections and strictureplasties. For example, fistulising disease of the terminal ileum, even with concomitant phlegmon, is not a contraindication to perform strictureplasties in extensive jejunoileal locations. In patients treated with conservative surgery, 20-40% of the diseased segments are suitable for resection and the remaining for strictureplasty.<sup>86</sup> Due to the bowel wall characteristics, the vascularisation, and the potential of cancer development, strictureplasty of the colon is contraindicated.<sup>86,94-97</sup>

Conventional strictureplasties are the Heineke-Mikulicz and the Finney techniques. Ileo-ileal isoperistaltic strictureplasty [also referred to as the Michelassi strictureplasty], modifications of the conventional strictureplasty for complex stenosis, strictureplasties of the ileo-caecal region, and strictureplasties of the target bowel segment of fistula are named non-conventional strictureplasties.<sup>86,98-102</sup>

**4.2.2.3. ECCO-ESCP Statement 3H**

When surgery is indicated in primary, stricturing, and localised CD of the small bowel, it should be treated with either minimal bowel resection or strictureplasty. [EL2] The preferred treatment of multiple fibrotic strictures of the small intestine, when technically feasible, are strictureplasties [EL2]

**4.2.2.4. ECCO-ESCP Statement 3I**

For the treatment of stenotic segments up to 68 cm, the technique of choice is the HeinekeMikulicz strictureplasty [EL2]

**4.2.2.5. ECCO-ESCP Statement 3J**

Side-to-side, isoperistaltic strictureplasty [also known as Michelassi strictureplasty] is the preferred technique for the treatment of long stenotic segments or multiple and close strictures [EL2]

In a systematic review and meta-analysis, more than 1600 patients who had over 4500 strictureplasties were described.<sup>103</sup> The overall complication rate was low, ranging from 5% to 20%, and mortality rate was nil. Long-term recurrence rates ranged from 25% to 70%. Interestingly, half of the patients included in the meta-analysis of Campbell *et al.*<sup>102</sup> had strictureplasties as the first surgical procedure. The Heineke-Mikulicz technique is the most commonly [> 90%] used conventional strictureplasty. The Michelassi technique is the most commonly used non-conventional strictureplasty [> 80%]. The Finney technique has been reported to have a higher recurrence rate, probably due to the creation of a large, lateral diverticulum with faecal stasis and bacterial overgrowth.<sup>104</sup> The most common complications after strictureplasty are small bowel obstruction [2.6%], intra-abdominal septic complications and suture leakages [4.2%], and intra-luminal and intra-abdominal bleeding [3.2%], with a cumulative reoperation rate of 2.8%. The main risk factors claimed to influence postoperative complications are malnutrition, hypoalbuminaemia, unscheduled surgery, peritonitis, intra-abdominal septic complication with peritoneal contamination, anaemia, and older age. In contrast, steroid use, synchronous bowel resection, and number, site, or lengths of strictureplasties were not significant risk factors. Only five cases of adenocarcinoma of the small bowel arising at a strictureplasty site have been reported [0.3%.] Therefore, routine biopsy with frozen section before performing a strictureplasty is not advised.<sup>86,95,101-103</sup> The most intriguing observation of strictureplasties is the normalisation of the bowel wall and a very low site-specific recurrence rate, which has been reported as between 2% and 5% at 10 years. Repetitive surgery for recurrences typically shows macroscopically normal bowel segments previously treated by strictureplasty.<sup>105-107</sup>

**4.3. Terminal ileum****4.3.1. Existing statements****4.3.1.1. ECCO Statement 7A**

Surgery is the preferred option in patients with localised ileocaecal Crohn's disease with obstructive symptoms, but no significant evidence of active inflammation [EL4].

**4.3.1.2. ECCO Statement 7B**

Active small bowel Crohn's disease with a concomitant abdominal abscess should preferably be managed with antibiotics and percutaneous or surgical drainage, followed by delayed resection if necessary [EL3].

**4.3.1.3. ECCO Statement 7D**

Wide lumen stapled ileocolic side-to-side [functional end-to-end] anastomosis is the preferred technique [EL1].

**4.3.1.4. ECCO Statement 7E**

Terminal ileitis resembling Crohn's disease found at a laparotomy for suspected appendicitis should not routinely be resected [EL5].

#### 4.3.1.5. ECCO Statement 7F

A laparoscopic approach is to be preferred for ileocolic resections in Crohn's disease [EL 2] where appropriate expertise is available. In more complex cases or recurrent resection, there is insufficient evidence to recommend laparoscopic surgery as the technique of first choice [EL3].

#### 4.3.1.6. ECCO Statement 7H

Endoscopic dilatation is a preferred technique for the management of symptomatic and short anastomotic strictures. It should only be attempted in institutions with surgical back-up [EL3].

#### 4.3.1.7. ECCO Statement 8A

Current smoking [EL1], penetrating and stricturing disease behaviour [EL1], early steroid use [EL2], ileal disease [EL2], jejunal disease [EL3], and young age at diagnosis [EL3] are risk factors for surgery in Crohn's disease.

#### 4.3.1.8. ECCO Statement 8B

The following are considered predictors of early postoperative recurrence after ileocolonic resection: smoking, previous intestinal surgery, absence of prophylactic treatment [EL1], penetrating disease at index surgery, perianal location [EL2], granulomas in resection specimen [EL2], and myenteric proctitis [EL3].

#### 4.3.1.9. ECCO Statement 8C

Early treatment with thiopurines [EL2] is associated with reduced risk of first surgery. Treatment with anti-TNF reduces the risk of surgery [EL2].

#### 4.3.1.10. ECCO Statement 8D

Ileocolonoscopy is the gold standard in the diagnosis of postoperative recurrence, by defining the presence and severity of morphological recurrence and predicting the clinical course [EL2]. Ileocolonoscopy is recommended within the first year after surgery where treatment decisions may be affected [EL2].

#### 4.3.1.11. ECCO Statement 8E

Calprotectin, 'trans-abdominal' ultrasound, MR enterography, and small bowel capsule endoscopy [SBCE] are less invasive diagnostic methods emerging as alternative tools for identifying postoperative recurrence [EL3].

#### 4.3.1.12. ECCO Statement 8F

All patients with Crohn's disease should be informed of the risk associated with smoking and smoking cessation should be encouraged and supported [EL1].

#### 4.3.1.13. ECCO Statement 8G

Prophylactic treatment is recommended after ileocolonic intestinal resection in patients with at least one risk factor for recurrence [EL2].

To prevent postoperative recurrence, the drugs of choice are thiopurines [EL2] or anti-TNFs [EL2]. High-dose mesalazine is an option for patients with an isolated ileal resection [EL2]. Imidazole antibiotics have been shown to be effective after ileocolic resection but are less well tolerated [EL1].

#### 4.3.1.14. ECCO Statement 8H

Long-term prophylaxis should be recommended [EL2].

Patients presenting with obstructive symptoms due to non-inflammatory disease confirmed by low C-reactive protein and calprotectin levels, ultrasound, or MRI can be treated with primary surgery. Surgery for stricturing disease has a long-term surgical recurrence rate less than 30% at 15 years.<sup>40,85,86,98,100,108-111</sup>

#### 4.3.1.15. ECCO-ESCP Statement 3K

A laparoscopic approach is to be preferred for ileocolic resections in CD [EL 1]. Where appropriate expertise is available, it should be attempted also in more complex cases or recurrent disease [EL2]

This statement clearly extends the role for laparoscopy in Crohn's disease in comparison with the position taken in the ECCO statement 7F. New evidence indicates that even in more complex cases, the laparoscopic approach can be performed safely. Three meta-analyses, including 15 studies [two of which were RCT with a 10 years' follow-up] and a US nation-wide registry study of 49 609 patients, showed consistent benefits of laparoscopy in terms of time to oral intake, time to bowel function, duration of hospital stay, perioperative complication rate, short- and long-term morbidity, mortality, reoperation for incisional hernia and adhesions, cosmesis, and body image scores. Moreover, open and laparoscopic resections share the same long-term recurrence rate of CD, eliminating the concern of missing occult segments in the upper gastrointestinal tract during laparoscopy. Reports from referral centres have shown safety and efficacy of laparoscopy also in recurrent disease and cases with complex fistulising disease.<sup>109-119</sup>

#### 4.3.1.16. ECCO-ESCP Statement 3L

In the presence of perforating disease, ileocaecal resection is strongly suggested. In stricturing disease, both ileocaecal resection and ileocolic stricturoplasties are valid options, with similar safety, efficacy, and long-term recurrence rates [EL2]

It is unclear whether the diseased bowel segment returns to normal function after stricturoplasty. Some studies report a normalisation of both the mucosa and the bowel wall. Perioperative results of stricturoplasty are similar to resectional surgery, and long-term recurrence rates are comparable. In cases of perforating disease with concomitant abscesses, percutaneous drainage is indicated together with optimisation of the nutritional status, antibiotics, and tapering of immunosuppressive treatment. Postponement of surgery for ongoing fistulising disease may lead to more complicated surgery and extensive resection.<sup>105,120,121</sup>

Two meta-analyses indicated that a stapled, functional, end-to-end anastomosis has lower leak rate and less postoperative complications compared with hand-sewn, end-to-end [but not side-to-side] anastomosis. It seems that wide anastomotic diameter is an important discriminating factor, whatever anastomotic technique is used.<sup>122,123</sup>

Endoscopic balloon dilatation is indicated in primary strictures  $\leq 4$  cm and in anastomotic recurrence. Repetitive dilatation is necessary in 20% and 50% of the patients at 1 and 5 years, respectively. Technical success is reported to be as high as 90% with a clinical success rate of 75%. Surgery rates of patients who eventually need surgery at 1, 3, and 5 years vary at 13–17%, 28–42%, and 36–42%, respectively. Perforation and bleeding rates are low [2–5%], and

no mortality has been reported. If the stricture occurs in the colon, malignancy must be ruled out. Most authors suggest that dilatation should preferably be attempted in institutions with surgical back-up.<sup>65</sup>

Postoperative recurrence may occur in weeks to months after surgery, and the most reliable tool to diagnose recurrence is endoscopy. Rutgeerts *et al.* developed an endoscopic scoring system based on the stratification of patients into five groups [i0-i4] according to mucosal inflammation activity.<sup>124</sup> Patients with a score of i0 or i1 have a low risk of clinical and surgical recurrence. Bowel wall thickening measured by ultrasound 1 year after surgery is also a strong independent risk factor for early postoperative recurrence.<sup>82,105</sup> Thiopurines and anti-TNF prophylaxis showed to be superior to placebo in preventing clinical and surgical recurrence. Anti-TNF agents may be the most effective prophylactic drug in preventing clinical and endoscopic recurrence.<sup>6,31,39,86,106,108,125-131</sup>

#### 4.3.1.17. ECCO-ESCP Statement 3M

Appendectomy of a macroscopically normal appendix in the presence of terminal ileitis has an elevated risk of intra-abdominal septic complications and fistulas [EL4]

The finding of terminal ileitis at emergency exploration for suspected appendicitis is non-specific, and it is difficult to differentiate intraoperatively between CD and infectious diseases. In addition, the ileitis could still be treated medically in the absence of stricturing or penetrating disease. It is preferable to decide to perform a resection after a full assessment and a multidisciplinary consultation. Appendectomy of a normal appendix in CD patients has been reported to have a considerable risk of intra-abdominal abscess formation and fistulisation.<sup>6,132</sup>

#### 4.4. Abdominal fistulising CD

##### 4.4.1. ECCO-ESCP Statement 3N

Abdominal fistulas are best classified on the basis of their anatomy, indicating the bowel segment of origin followed by the non-diseased target organ [eg. ileo-colonic, colovesical, entero-mesenteric]. Surgery is strongly recommended for enterourinary fistulas, fistulas associated with bowel stricture and/or abscess, fistulas causing diarrhoea and/or malabsorption [EL5]

##### 4.4.2. ECCO-ESCP Statement 3O

Entero-cutaneous fistulas are best classified on the basis of their output [low: < 200 ml/day; moderate: 200–500 ml/day; high: > 500 ml/day]. Surgery should be planned after stabilisation through the following four steps: fluid and electrolyte balance, control of sepsis [antibiotics and percutaneous drainage when appropriate], nutritional improvement, and skin care [EL4]

Abdominal fistulas are quite common in CD. MRI is the preferred tool for the diagnosis of enteric fistulas, with the best sensitivity and specificity. Most commonly, fistulas are classified on the basis of their anatomy, indicating the bowel segment of origin followed by the target organ. Entero-cutaneous fistulas are classified

also on the basis of their physiology, due to the impact of their output on nutrition and electrolyte balance. Surgery is indicated in entero-cutaneous fistulas with high output. In low-output fistula, proper counselling on the risk of surgery is even more important. Time must be reserved to optimise patient's nutrition and to control sepsis. Asymptomatic fistulas do not require surgical intervention. However, in fistulising disease it is important to monitor the effect of medication, because ongoing asymptomatic inflammation might result in more complicated disease in the long run.<sup>133-138</sup> Gastro-colic, duodeno-colic, jejuno-ileal or jejuno-colic, entero-biliary, and entero-urinary fistulas mostly require surgery because of the risk of severe malabsorption or faecal contamination of the upper GI tract, and the potential of severe urinary and biliary sepsis.<sup>75,139</sup>

## 5. Large bowel

### 5.1. Existing statements

#### 5.1.1. ECCO Statement 7G

If surgery is necessary for localised colonic disease [less than a third of the colon involved] then resection of the affected part only is preferable [EL3]. Two segmental resections can be considered for a patient with an established indication for surgery when macroscopic disease affects two separate segments of the colon [EL3]. Strictureplasty in the colon is not recommended [EL3].

A meta-analysis, including six retrospective cohort studies comparing segmental resection with subtotal colectomy for segmental colonic disease, demonstrates that both procedures are equally effective with no difference in recurrence, incidence of postoperative complications, or the need for a permanent stoma.<sup>140</sup> Earlier recurrence was seen in patients after segmental resection. The reduced recurrence-free survival after segmental resection was confirmed by Kiran *et al.*, without difference in stoma-free survival and quality of life between groups.<sup>141</sup> Since functional results of segmental resection are superior to subtotal colectomy, with fewer loose stools and better anorectal function, the conservative approach is generally supported.<sup>142</sup>

Most studies demonstrate that approximately one-third of patients need at least one re-resection. Female sex, a history of perianal disease, and young age have been identified as independent risk factors for re-resection.<sup>143,144</sup>

There is some support in the literature for separate segmental resection when macroscopic disease affects two widely separated segments of the colon, but subtotal colectomy with ileorectal anastomosis has been associated with lower risk of recurrence, and longer recurrence-free interval when two or more colonic segments are involved.<sup>140</sup> The majority of patients have good long-term functioning of the ileorectal anastomosis [> 70% after 10 years].<sup>145,146</sup> If the distal rectum is involved, proctocolectomy should be considered.

#### 5.1.2. ECCO Statement 7I

Patients with an [unsuspected] diagnosis of Crohn's disease after ileal pouch-anal anastomosis [IPAA] present markedly higher complication and failure rates. An IPAA may be discussed in highly selected and motivated patients with Crohn's colitis, pending proof of absent small bowel disease and no existing or previous evidence of perineal involvement. Intensive combined management by IBD physicians is mandatory to maintain an acceptable pouch function in those patients [EL4].

## 5.2. ECCO-ESCP statements

### 5.2.1. ECCO-ESCP Statement 4A

In carefully selected patients with no history of perianal or small bowel disease, a restorative proctocolectomy can be offered as a treatment option with comparable quality of life to those with ulcerative colitis, but the long-term pouch failure rate is increased [EL3]

Most series of restorative proctocolectomy with ileo-anal pouch anal anastomosis include some patients with CD.<sup>147</sup> Retrospective analyses show that these patients suffer higher complication rates, with pouch failure reported in up to 30%<sup>148</sup> compared with 10% in large series of ulcerative colitis at 10 years. Panis *et al.* were the first to describe a series of patients with CD but having a UC-behaving phenotype, undergoing restorative proctocolectomy<sup>149</sup> ileo-anal pouch surgery with acceptable early and late outcome. More recent studies have shown that the complication rate can be similar to that seen in ulcerative colitis in selected patients with no history of previous perianal or small bowel disease. Fazio *et al.* found in a series of more than 3000 IPAA that patients with an established Crohn's diagnosis have an increase of small bowel obstruction and up to 80% may have a functioning pouch 10 years after a primary IPAA regardless of when the disease was discovered, pre- or postoperatively.<sup>150,151</sup>

Patients with CD with no previous perianal or small bowel disease may be offered an IPAA with reasonable long-term results. Careful counselling to outline the risk of pouch failure, anorectal function, and pelvic septic complications has to be discussed thoroughly with the patient. In the presence of relative rectal sparing, where the rectal stump is too short for straight ileorectal anastomosis, an ileal pouch-rectal anastomosis can be considered.

The quality of life is comparable to those patients with an IPAA for ulcerative colitis. It is recommended to create a J pouch with a stapled pouch-anal anastomosis and no mucosectomy.<sup>151</sup>

### 5.2.2. ECCO-ESCP Statement 4B

Patients with a diagnosis of CD after ileo-anal pouch formation have markedly higher complication and failure rates. In this group of patients, multidisciplinary management is essential to maintain pouch function [EL3]

Patients diagnosed with CD after an IPAA have higher complications and failure rates than those with ulcerative colitis. These patients tend to have more anastomotic strictures and incontinence, and the pouch failure can be 6-fold more frequent when compared with ulcerative colitis and indeterminate colitis.<sup>152</sup> The development of Crohn's in the ileal pouch should indicate aggressive medical treatment. There is no role for rescue surgery in this group of patients; Defunctioning ileostomy or pouchectomy and definitive ileostomy is the only indicated surgery.

### 5.2.3. ECCO-ESCP Statement 4C

Revisonal pouch surgery is generally not recommended in the presence of CD [EL5]

Most patients with CD who develop pouch failure usually present with more than one complication among the following: pelvic sepsis, pouch fistulas, poor defaecation function, strictures, or small

bowel obstruction. These patients can be treated with diverting ileostomy, seton drainage, laparotomy and dilatation, but revisonal pouch surgery is not recommended as it is associated with poor outcomes, particularly pelvic sepsis.<sup>152</sup>

### 5.2.4. ECCO-ESCP Statement 4D

The treatment of choice for large bowel stricture is balloon dilatation or segmental resection. Strictureplasty is not recommended [EL3]

The occurrence of a colonic stricture harbours the risk of the presence of a tumour; furthermore, it hampers further colonoscopic surveillance of the proximal colon and there is the risk for acute colonic obstruction.

There are few data on strictureplasty for the management of large bowel strictures. The treatment of choice for short strictures of the large bowel is usually balloon dilatation.<sup>153-155</sup> In cases of failed balloon dilatation, surgical segmental resection should be considered.<sup>153</sup> Endoscopic stent placement<sup>27,156</sup> or an intralesional injection of infliximab for large bowel stricture can be considered, but there is insufficient evidence in the literature to support the routine use of these techniques.<sup>26,157</sup>

Generally, there is no place for strictureplasty in the large bowel, mainly due to the concern of potential risk for cancer.<sup>158</sup> Therefore, it is important to take biopsies at the time of the procedure. A single study reviewed 29 patients who underwent strictureplasty of the large intestine. Importantly, 40% of these were strictures of the ileocaecal junction or anastomotic strictures. The use of strictureplasty for colonic strictures, however, has not been further supported by other studies.<sup>159</sup>

### 5.2.5. ECCO-ESCP Statement 4E

In refractory pancolitis, a defunctioning ileostomy can be offered. [EL5]. After remission, either [sub]total colectomy with ileorectal anastomosis or further medical therapy can be considered [EL5]

The beneficial effect of faecal diversion has already been described in 1965.<sup>160</sup> It has been associated with acute clinical remission in the majority of patients with refractory Crohn's colitis, although subsequent reports question the long-term benefits. With the availability of new medical therapy, stoma closure can be discussed after remission, although the evidence for maintained remission is very limited. Most studies are of patients who have undergone [sub]total colectomy with ileorectal anastomosis or proctocolectomy after diversion, but in highly selected patients, surgery with intestinal diversion alone could offer a realistic alternative to major bowel resection.<sup>161</sup>

### 5.2.6. ECCO-ESCP Statement 4F

Defunctioning with a diverting stoma in situations where there is growth retardation has not been shown to have the same growth-promoting effect as resecting the affected segment [EL4]

### 5.2.7. ECCO-ESCP Statement 4G

Colonic resection in early puberty, where the disease is refractory to medical treatment, promotes catch-up growth [EL4]

### 5.2.8. ECCO-ESCP Statement 4H

The exact timing of colectomy in children is dependent on several interrelated factors: growth, educational stage, previous medical treatments, and disease phenotype [EL5]

Colectomy, but not a defunctioning stoma formation, has been shown to be a growth-promoting strategy in children. Resectional surgery, especially in early to mid puberty, has been demonstrated to allow catch-up height.<sup>162</sup> This has been demonstrated for all surgical resections in CD, most notably for ileo-caecal resection,<sup>34</sup> but this applies to colectomy as well.

Surgery in early or mid puberty in patients refractory to medical treatment has resulted in improved linear growth.<sup>162</sup> Surgery later in puberty has not been shown to have the same beneficial impact on height.<sup>163</sup> Growth promotion is one indication for surgery, but the timing of a colectomy for an adolescent should also take into account individual factors like the timing of their examinations, disease phenotype, and current and previous medical treatments.

## 6. Perianal Crohn's disease

### 6.1. Introduction

Perianal disease in CD was first described by Penner and Crohn in 1938<sup>164</sup> and consists of a wide variety of entities, including both fistulising lesions [abscess, fistulas, or rectovaginal fistulas] and non-fistulising disease [eg fissures, deep ulcers, anorectal strictures, skin tags, or haemorrhoids]. The clinical impact of these entities may also vary significantly from asymptomatic and mild disease to severe and devastating scenarios. Whereas fissure, tags, or haemorrhoids will usually require only conservative management, abscesses and fistulas may require an aggressive medical and surgical intervention. In population-based studies,<sup>165-167</sup> the occurrence of perianal fistulas in CD varies between 14% and 23%, with cumulative incidence depending on time from diagnosis. Perianal disease may also precede intestinal symptoms, or may appear at the time of diagnosis, or later on. The risk for developing anal fistulising complications also depends on disease location. In case of colonic disease with rectal involvement, the prevalence of fistulising anal disease is reported as high as 92%.<sup>168</sup> The first population-based study was published by Hellers *et al.*<sup>168</sup> who analysed the natural history of fistulising CD among residents of Stockholm County, in Sweden, from 1955 to 1974; the cumulative incidence of anal fistula was reported to be 23%. After a mean follow-up of 9.4 years, 51% of patients were healed, 9% had an open fistula, and 40% had needed proctectomy. These figures reflect the natural history of perianal CD before the widespread use of new treatments [eg immunomodulators, anti-TNF], which are probably changing the natural course of the disease. Later, Lapidus *et al.*<sup>169</sup> followed 507 patients with 'colonic' or 'rectal' CD, again in Stockholm County, between 1955 and 1989, and they found that perianal or rectal fistulas occurred in 37% of patients. An update from Stockholm County residents<sup>167</sup> includes all the patients diagnosed with CD [regardless of the location and extent] between 1990 and 2001: perianal fistulising disease occurred 13.7% of patients [any time before or after the diagnosis of CD]. The impact of gender and age was also investigated: men were more likely to develop perianal disease than women [15.8% vs 11.6%,  $p < 0.05$ ]. The highest incidence was found among young adults [16-30 years] and a second peak in the elderly [76-90 years]. Results from Olmsted County cohort<sup>165</sup> showed a cumulative risk of 12%

after 1 year, 15% after 5 years, 21% after 10 years, and 26% after 20 years; 44% of these patients developed a fistula at or before the diagnosis of CD. Data similar to those were reported from Stockholm County.<sup>168</sup> Recurrent fistulas were uncommon, with 66% of patients experiencing only one fistula episode. Three-quarters of the perianal fistulas required surgery and one-third of patients required major surgery, including proctocolectomy and ileostomy, proctectomy and colostomy, or segmental resection of proximal disease. It is unknown how often drained perianal abscesses in patients with CD will eventually develop into fistulas. How the frequencies of this type of surgery exactly evolved in the era of biologics is unknown.

It is not stated how long after abscess drainage one should consider there is a fistula. Williamson *et al.* did a retrospective analysis of 48 patients treated over a 20-year period to assess the results of surgical abscess drainage in perianal CD;<sup>170</sup> 13 [27%] patients had recurrences after initial healing of their wounds. The mean time to recurrence after healing was 5.25 years. In cryptoglandular disease, one-third of the patients will have a perianal fistula after abscess drainage. Patients aged under 40 years and non-diabetic patients appeared to have a higher risk for fistula formation. Administration of perioperative antibiotics significantly reduced the rate of subsequent fistula formation.<sup>171</sup>

### 6.2. Existing statements

#### 6.2.1. ECCO Statement 9A

Contrast-enhanced pelvic magnetic resonance imaging [MRI] is considered the initial procedure for the assessment of perianal fistulising CD [EL2]. If rectal stenosis is excluded, endoscopic anorectal ultrasound [EUS] is a good alternative [EL2]. The specificity and sensitivity of both imaging modalities are increased when combined with examination under anaesthetic [EUA] [EL1]. Fistulography is not recommended [EL3]. If a perianal fistula is detected, EUA is considered the gold standard in the hands of an experienced surgeon. [EL5].

#### 6.2.2. ECCO Statement 9B

Since the presence of concomitant rectosigmoid inflammation has prognostic and therapeutic relevance, proctosigmoidoscopy should be used routinely in the initial evaluation [EL2].

#### 6.2.3. ECCO Statement 9C

There is no consensus for classifying perianal fistulas in CD. In clinical practice, most experts use a classification of 'simple' or 'complex' [EL5].

#### 6.2.4. ECCO Statement 9E

In an uncomplicated low anal fistula, simple fistulotomy may be discussed [EL5]. The presence of a perianal abscess should be ruled out, and if present should be drained [EL5].

#### 6.2.5. ECCO Statement 9F

Symptomatic simple perianal fistulas require treatment. Seton placement in combination with antibiotics [metronidazole and/or ciprofloxacin] is the preferred strategy. [EL3] In recurrent refractory simple fistulising disease not responding to antibiotics, thiopurines or anti-TNFs can be used as second-line therapy [EL4].

#### 6.2.6. ECCO Statement 9G

Seton placement after surgical treatment of sepsis is recommended for complex fistulas [EL2]. The timing of removal depends on subsequent therapy.

### 6.2.7. ECCO Statement 9H

Active luminal Crohn's disease should be treated if present, in conjunction with appropriate surgical management of fistulas [EL5].

### 6.2.8. ECCO Statement 9J

Imaging before surgical drainage is recommended. EUA for surgical drainage of sepsis is mandatory for complex fistulas [EL 4]. In complex fistulas, abscess drainage and loose seton placement should be performed [EL4].

### 6.2.9. ECCO Statement 9K

In evaluating the response to medical or surgical treatment in routine practice, clinical assessment [decreased drainage] is usually sufficient [EL2]. MRI [EL2] or anal endosonography [EL3] in combination with clinical assessment is recommended to evaluate the improvement of fistula track inflammation [EL5].

### 6.2.10. ECCO Statement 9L

Thiopurines [EL2], infliximab [EL1], or adalimumab [EL2], seton drainage, or a combination of drainage and medical therapy [EL3] should be used as maintenance therapy.

### 6.2.11. ECCO Statement 9M

Patients refractory to medical treatment should be considered for a diverting ostomy, with proctectomy as the last resort [EL 5].

### 6.2.12. ECCO Statement 9N

Entero-enteric and entero-vesical fistulae often require resective surgery [EL5]. Surgery is strongly recommended for entero-enteric fistulas if associated with abscess and bowel stricture and if they cause excessive diarrhoea and malabsorption [EL5].

### 6.2.13. ECCO Statement 9O

Asymptomatic low anal-introital fistulae do not need surgical treatment [EL5]. If a patient has a symptomatic recto-vaginal fistula, surgery is usually necessary [including diverting ostomy] [EL5]. Active CD with rectal inflammation should be treated medically before surgery, and after surgery to prevent recurrence [EL5].

## 6.3. ECCO-ESCP statements

### 6.3.1. ECCO-ESCP Statement 5A

The indications for surgery aiming to close a fistula-in-ano in CD include a symptomatic patient, with no concomitant abscess, with medically controlled proctitis, and a preferably anatomically defined fistula tract [EL3]

The appropriate surgical treatment of perianal [CD] must be individualised to each specific patient. Consideration must be given to the patient's presentation [as an emergency, with an associated abscess, or electively], the fistula's anatomy [location of openings, trajectory of tract, and complexity], the presence of concomitant proctitis,<sup>172</sup> and the severity of associated anal canal disease [ie stenosis]. In general, symptomatic fistulas require intervention. Before any form of treatment, be it medical or surgical, the sepsis must be drained using loose setons. This allows the inflammation around the tract to subside, and it prevents recurrence of the abscesses.

The most commonly used surgical techniques include simple fistulotomy, chronic seton, mucosal advancement flap, ligation of intersphincteric fistula tract [LIFT], fibrin glue, fistula plug, and video-assisted anal fistula treatment [VAAFT].

In the setting of CD, additional factors must be considered when contemplating operative intervention with the aim of closing the fistula. More than two-thirds of patients have an abscess associated with their fistula, and this must be drained before considering operative intervention. As a result, CD patients are more likely to have setons placed, at least for a period, and less likely to undergo fistulotomy.<sup>172</sup> Complex fistulas are seen in 80% of patients and, with these fistulas, failure to heal and recurrence rates are increased.<sup>173-176</sup> Medical therapy to control disease-related inflammation is imperative to increase the likelihood of tract healing after surgery. The most conservative approach should be adopted to avoid soft tissue damage and prevent extensive scarring. Concomitant perianal skin tags should not be treated surgically as this can lead to chronic, non-healing ulcers.

### 6.3.2. ECCO-ESCP Statement 4B

The type of surgical procedure should be tailored to the anatomy of the fistula tracts and of the anus [exposure, strictures, access, previous surgery] and the expertise of the surgeon [EL5]

The objective of fistula treatment is to close the fistula tract in order to prevent recurrence and to preserve continence. A number of techniques can be applied to approach the fistula. Patients eligible for surgical treatment with Crohn's perianal fistula [no proctitis, simple fistula] aiming at closure differ with respect to anatomy of the fistula tracts, number of internal and external openings, scarring of the anus, and continence. The surgeon needs to choose the optimal strategy out of a variety of surgical options, eg chronic seton, fibrin glue, fistulotomy, advancement flaps [full-thickness or mucosal], plug, LIFT, and VAAFT. This means that not all techniques are applicable for the specific patient with perianal disease; for example, a mucosal advancement flap is not possible in a stenotic scarred anus.

The most frequently reported approaches to treat high perianal fistula aiming at closure are endorectal advancement flaps, fistula plugs, fibrin glue instillation, and the LIFT procedure. The greatest experience to date has been with the endorectal advancement flap. The least invasive surgical procedures may be considered as first options.

*Fistulotomy.* Conventional fistulotomy by laying open the tract and any side tracts, followed by debridement [with or without marsupialisation], is recommended for subcutaneous<sup>177</sup> or superficial<sup>178</sup> fistulas. Two studies elaborated on these indications by including patients with a submucosal, intersphincteric, or trans-sphincteric fistula in the lower third of the anal sphincter.<sup>179</sup> Suggested contraindications to fistulotomy in this setting include a Crohn's Disease Activity Index [CDAI] greater than 150<sup>178</sup> and evidence of perineal Crohn's disease involvement.<sup>177</sup>

*Setons.* Non-cutting or loose setons are primarily used to establish drainage of the fistula, minimising the risk of associated future abscesses and facilitating personal hygiene. Loose, fine, silastic setons are most commonly used.<sup>177,180</sup> The fistula tract running through the sphincter complex ending in the internal opening must

be identified and provided with a seton placed to maintain drainage, with the aim of preventing the development of a more complex fistula. If moderate to severe proctitis complicates a fistula, seton placement is the only sensible option. Medical therapy needs to be commenced to treat proctitis.<sup>172</sup> A loose seton may be a definitive treatment when used in combination with optimal medical therapy, with the seton being removed in up to 98% at a median of 33 weeks.<sup>181</sup>

**Endorectal advancement flap.** With the advancement flap procedure, first described by Noble *et al.* in 1902, the internal opening of the fistula is closed with a partial- or full-thickness rectal flap. It is a successful surgical option for Crohn's related fistulas unsuitable for fistulotomy [ie sphincter division would compromise continence], provided the tract has been simplified, associated sepsis has been eradicated, and there is no associated rectal inflammation.<sup>182</sup> Typically a rectal advancement flap is described as a second stage for high fistulae [the fistula tract is located in the upper two-thirds of the sphincter complex]. It is also the most common and successful procedure performed for Crohn's related rectovaginal fistulae [provided the sphincter complex is intact].<sup>177,183,184</sup> A recent systematic review reported closure rates of approximately 80% [range 24-100%] in patients with cryptoglandular fistulas.<sup>185</sup> Unfortunately, success rates for Crohn's fistulas are less favourable, with a weighted success rate of 64% [range 33-92%]. In Crohn's perianal fistulas, concomitant medical therapy with immunosuppressive is advised.

Poor wound healing occurs mainly in patients with active Crohn's disease in the rectum. Cigarette smoking is also well known to influence wound healing in various patient groups, and can have a significant negative impact on the results of the flap repair.<sup>186</sup> The experience of the surgeon, technical details like de-epithelisation of the internal opening, closure of the internal opening, drainage or excision of the extrasphincteric part of the fistula tract, perioperative administration of antibiotics, bowel preparation and bed rest, and type of flap are all factors contributing to success. Incontinence is described in 13% [0-35%] of patients with cryptoglandular fistulas and 9.5% [0-29%] in patients with Crohn's fistulas after endorectal advancement flap repair.<sup>179</sup>

**Anal fistula plug.** Advances in biotechnology have led to the development of fistula plugs. The plug is made of porcine small-intestinal submucosa or collagen and is inserted via the internal opening of the fistula. The reported indications in the literature include trans-sphincteric [single- or double-tract fistulas] and rectovaginal fistulas [lower two-thirds of the rectovaginal space] without evidence of perianal sepsis or severe inflammation.<sup>177,181</sup> Most fistulas are treated for 6-8 weeks with setons before plug insertion.<sup>177</sup> Success rates in literature vary from 24% to 88%. In a systematic review, the success rate of the plug was 55%.<sup>187</sup>

**Fibrin glue.** Fibrin glue is a rather new technique for fistula closure, with variable success rates reported in literature. It involves injecting the fistula with biodegradable glue to seal off the tract and to stimulate the migration, proliferation, and activation of fibroblasts. The few studies addressing the use of fibrin glue report fistula closure in between 38% and 50%.<sup>188</sup> However, follow-up in these studies is short and the number of patients is small.

**LIFT procedure.** Ligation of the intersphincteric fistula tract [LIFT] is a rather new technique in the surgical management of

[transsphincteric] fistulas. With an intersphincteric approach, the fistula is closed by ligating the tract. There are few data with respect to the application in Crohn's disease. A systematic review of 13 original articles, mostly retrospective and prospective case series, evaluating the success rate of the LIFT technique in cryptoglandular disease, was recently published. An overall mean healing rate of approximately 81% was reported.<sup>189</sup> Up till now, only one randomised controlled trial with a sample size of 36 patients [2:1] has been published comparing the LIFT procedure with an advancement flap for complex non-Crohn's perianal fistulas.<sup>190</sup> After a median follow-up of 19.2 months, there were three recurrences of which two were in the LIFT group. Complications between the groups were similar. In a cohort of 15 patients who had LIFT for Crohn's perianal fistula, the success rate was 67% at 12 months.<sup>191</sup> Because of the intersphincteric approach, LIFT is thought to be continence-saving. In order to be able to ligate the intersphincteric tract, the tract should be organised and suitable for ligation. A certain degree of chronicity is therefore important.

**VAAFT.** Only one study reported on the use of VAAFT in the setting of CD. The suggested benefit is that additional side tracts, not detected preoperatively, could be identified in 64% of cases.<sup>192</sup> The indications described included trans- or suprasphincteric and rectovaginal fistulas [fistulas of the lower two-thirds of the rectovaginal space] in the absence of proctitis, rectal stricture, or concomitant perianal sepsis.<sup>192</sup>

**Advanced, resectional approaches to Crohn's-related perianal fistulae.** For advanced perianal CD fistulae, circumferential transanal sleeve advancement flaps have been performed in select circumstances as an alternative to proctectomy.<sup>193</sup> The indications include major symptoms for at least 6 months, with multiple fistula tracts [including extension to the vagina and/or urethra]. Ulceration of the anal canal or anal canal stenosis is not a contraindication, but the eradication of perianal sepsis and the absence of a rectal stricture or proctitis are requirements.<sup>193</sup> Should a concomitant rectal stricture or proctitis be present in this setting, then the Turnbull-Cutait abdominoperineal pull-through is the procedure of choice.<sup>194</sup> However, it must be noted that the clinical experience and long-term outcome data with these procedures are limited.

### 6.3.3. ECCO-ESCP Statement 4C

There is a real likelihood that a fistula will close after removal of the seton [EL3] in the absence of abscess, proctitis, or stenosis

**Seton drainage alone.** Closure rates of 13.6-100% are reported in patients who have their setons removed after being inserted for a variable amount of time [3 weeks to 40 months]. There is a large heterogeneity among the several studies.<sup>177,180,181,195,196</sup>

**Seton drainage with anti-TNF.** There are a number of retrospective studies addressing anti-TNF therapy with or without seton drainage, and seton drainage with or without anti-TNF. Although the evidence is weak, all of it directs towards better results if anti-TNF therapy and seton placements are combined.<sup>197-199</sup>

#### 6.3.4. ECCO-ESCP Statement 4D

The use of cutting setons in perianal CD is not recommended and may result in keyhole deformity and faecal incontinence [EL 3]

The use of the cutting seton was already described by Hippocrates in the 5th century BC. Applying this technique every 3 to 4 weeks, the seton is tightened, resulting in gradual fistulotomy. The success rate of closing of the fistula is almost 100%, but this technique is associated with a high risk of incontinence [57%], since transection and scarring of the anal sphincter occurs due to forced migration of the cutting seton.<sup>200,201</sup> This technique is therefore strongly disadvised.

#### 6.3.5. ECCO-ESCP Statement 4E

Fistulotomy [or lay-open] in the anterior perineum of a female patient should be avoided [EL5]

In fistulotomy, the primary tract of the fistula is laid open, which can be an efficient procedure for some type of fistulas. This procedure dates back to the medieval days. Literature shows healing rates as high as 100% for [low] fistulas.<sup>179</sup> Marsupialisation has been shown to improve healing and was therefore recommend by some guidelines. Although there is a lack of supporting data, there is consensus that because of the asymmetrical anatomy and consequently short anterior part of the anal sphincter in the female, fistulotomy of a low anteriorly located fistula has a high chance of jeopardising faecal continence.

#### 6.3.6. ECCO-ESCP Statement 4F

When treating perianal fistula with anti-TNF, seton removal is probably best done after the induction phase of anti-TNF is completed [EL5] and resolution of proctitis has been achieved

The optimal timing when to remove a seton is unclear. If it is removed too early, there is a high risk of the development of a recurrent perianal abscess. If the seton is left in place too long, the tract might epithelialise and the fistula might not close.<sup>202,203</sup>

In the ACCENT 2 study, all setons were removed by Week 2 and the overall new abscess rate was 15%. Several other authors reported the removal of the setons after a longer period of time.<sup>138,204</sup> According to a global consensus on the classification, diagnosis, and multidisciplinary treatment of perianal fistulising CD, it is recommended to keep the seton in place until at least the induction of the anti-TNF treatment period has been completed, that is approximately a month.<sup>172</sup>

#### 6.3.7. ECCO-ESCP Statement 4G

Faecal diversion is effective in reducing symptoms in perianal CD in two-thirds of patients and may improve quality of life, but only one-fifth of these patients are stoma-free in the long term [EL3]

A small percentage of patients with extensive and aggressive disease that is uncontrolled by medical management and long-term seton placement may require diversion or proctectomy to control

perianal sepsis.<sup>175,205–207</sup> Diversion is preferable with respect to proctectomy because perianal complications, chronic pelvic sinus, or impaired healing of a perianal wound may occur. Irrespective of aetiology, perianal Crohn's patients with asymptomatic fistulas and no signs of local sepsis require no surgical intervention. For patients with complex perianal CD, diversion rates range from 31% to 49%. Concomitant colonic disease, persistent anal sepsis, previous temporary diversion, faecal incontinence, and anal canal stenosis are reported as predictive factors. Despite optimal medical and minimally invasive therapy, 8% to 40% will require proctectomy to control symptoms.<sup>175,176,206,208,209</sup> Severe perianal disease [especially in the presence of rectal stenosis] and Crohn's colitis are strong predictors of permanent diversion. For this reason, primary total proctocolectomy or early completion proctectomy may be considered in this subgroup of patients.

There are indications that since the introduction of biologics, the permanent stoma rates have dropped from 60.8% to 19.2%. Univariate and multivariate analysis showed that the use of biologic drugs was significantly associated with an increased rate of rectal preservation [ $< 0.05$ ].<sup>210</sup>

The rate of perineal wound healing was assessed in a series of 112 patients undergoing proctectomy for Crohn's colitis. Healing was impaired in patients with a high fistula-in-ano or a recto-vaginal fistula preoperatively, faecal contamination, and postoperative perineal sepsis. Early healing [within 12 weeks] occurred in 63% of the patients. Healing was delayed by over 1 year in 21 patients [19%], and in 10 patients [9%] the perineal wounds never healed.

## 7. Special conditions

### 7.1. Colorectal cancer and Crohn's disease

#### 7.1.1. ECCO-ESCP Statement 5A

For colorectal cancer or high grade dysplasia in Crohn's colitis, proctocolectomy should be considered in patients who are fit for surgery, because of the multifocal nature of dysplasia and the high rate of metachronous cancer after segmental resection [EL2]. Close endoscopic surveillance or segmental resection could be discussed in case of low grade dysplasia or patients who are high risk for surgery [EL4]

Patients with inflammation of large bowel are at increased risk of development of colorectal cancer [CRC], which is described in statements 2A–2C of the European Evidence-based Consensus: IBD and Malignancies.<sup>211</sup> In a meta-analysis of population-based studies, the pooled standardised incidence ratio [SIR] for CRC was significantly increased [SIR, 1.9; 95% CI 1.4–2.5] in Crohn's disease.<sup>212</sup> In retrospective review of screening and surveillance colonoscopies of patients with Crohn's colitis, cancer or dysplasia was detected in 5.6% of patients with mean disease duration 17–37 years.<sup>213</sup> The risk of CRC in CD increases with longer disease duration, extent of colitis, a familial history of CRC, coexistent primary sclerosing cholangitis, and the degree and duration of inflammation.<sup>211,213,214</sup> Colorectal stenosis is the other risk factor for development of CRC, which was confirmed by Hungarian population-based study in which 640 incidental CD patients were analysed. Colorectal cancer developed in 6.5% [equalling 0.57/100person-years] with stenosis, even after a short disease duration.<sup>215</sup> CRC is important risk factor of increased mortality of IBD patients compared with the general population.<sup>216–218</sup> Two retrospective cohort series specifically focus on the

clinical, endoscopic, and pathological data to establish a decision-making process for patients with Crohn's colitis-associated dysplasia.<sup>219,220</sup> Both studies confirm the risk of cancer in these patients, with a cancer incidence up to 45% in patients undergoing colectomy for high grade dysplasia. In summary, CRC in IBD affects younger patients, arises more frequently in flat [non-visible] dysplasia, and tends to have higher histological grade and more often mucinous/signet-ring histological characteristics, and higher rates of synchronous dysplastic or neoplastic lesions than cancer in the general population.<sup>214</sup> Colorectal cancer in Crohn's disease presents at a more advanced stage and therefore has a less favourable outcome than sporadic colorectal cancer.<sup>218,221</sup>

Because of the multifocal nature of dysplasia in Crohn's colitis and the high rate of metachronous colon cancer after segmental surgical resection, proctocolectomy is recommended in fit patients with preoperative diagnosis of cancer or high grade dysplasia.<sup>220,222</sup> Prophylactic colectomy decreases the incidence of CRC.<sup>211</sup> In specific circumstances, particularly in the less healthy surgical candidate or in the setting of low grade dysplasia [LGD] and in absence of dysplasia in the rectum, segmental resection or subtotal colectomy with close postoperative endoscopic surveillance may be discussed.<sup>211,216,220,222</sup> Patients with colorectal cancer in Crohn's colitis should be operated according to the rules of oncological surgery, with adequate lymphadenectomy.<sup>222,223</sup> Principles of oncological surgery should be considered in cases of colonic stenosis and long-lasting extensive Crohn's colitis, because the preoperative confirmation of cancer is not common.<sup>213,219,222</sup> Strictureplasty is not recommended.

## 7.2. Small bowel cancer and Crohn's disease

### 7.2.1. ECCO-ESCP Statement 5B

The treatment recommendation in localised small bowel cancer is wide resection with the corresponding mesentery, right hemi-colectomy for cancer of the distal ileum, and pancreaticoduodenectomy for lesions of duodenum [EL4]

Patients with CD are at increased risk of development of small bowel cancer, which is described in statements 2G-2I of European Evidence-based Consensus: IBD and Malignancies.<sup>211</sup> In meta-analysis of population-based studies, the pooled standardised incidence ratio [SIR] for small bowel cancer was 27.1 [95% CI 14.9–49.2]. However, the absolute risk of developing small bowel cancer in CD remains low, because of the low incidence in general. Risk factors for small bowel adenocarcinoma in CD appear to be long duration of disease, ileal localisation, small bowel stenosis, and previous bypassed segments and young age at diagnosis.<sup>211,224,225</sup> In retrospective review of 29 surgical patients with CD and small bowel cancer, 55% had long periods of quiescent disease before diagnosis [7–45 years] and 55% of patients were operated for small bowel obstruction resistant to medical therapy.<sup>226</sup> Small bowel cancer is usually found postoperatively and often accidentally at histological examination.<sup>211,224–226</sup> Small bowel cancer in CD has a poor prognosis. Two-year survival rates have been found to be as low as 27%.<sup>227</sup>

Localised small bowel cancer is best managed with wide segmental surgical resection of the small bowel segment and corresponding mesentery and lymph nodes. Pancreaticoduodenectomy is recommended for lesions of the second or third portion of the duodenum, and right colectomy for carcinoma of the distal ileum would be required.<sup>227,228</sup>

## 7.3. Perianal Crohn's disease and cancer

### 7.3.1. ECCO-ESCP Statement 5C

Malignant transformation in perianal CD is rare. Symptoms, endoscopy, and imaging all have a low sensitivity. Evaluation under anaesthesia and biopsies of suspicious areas are indicated [EL4]. Surgical treatment should be in accordance oncological surgery [EL5]

Anal cancer arising from perianal fistulas is rare; incidence [0.2/1000 patient-years] and symptoms were described in the European Evidence-based Consensus: IBD and Malignancies, statements 2D-2E.<sup>211</sup> In a systematic review of case series, 61 cases were identified, most of them were adenocarcinomas [59%] or squamous cell carcinomas [31%].<sup>228</sup> In anal stenosis, cancer was found in 2% of cases.<sup>229</sup> Diagnosis is usually made in advanced stage, when prognosis of conservative and surgical treatment is poor.<sup>211,230,231</sup> No specific recommendation of surgical treatment exists; therefore surgical treatment should be in accordance to the principles of oncological surgery.

## 7.4. Short bowel syndrome.

### 7.4.1. ECCO-ESCP Statement 5D

The most frequent causes of short bowel syndrome are repetitive surgical resection, surgery for postoperative complications [EL3], and aggressive small bowel disease [EL4]. Preoperative optimisation, reduction of postoperative complications, and bowel-sparing tactics decrease risk of short bowel syndrome [EL4]

Patients with Crohn's disease are at 5–10% risk of developing short bowel syndrome.<sup>232</sup> The most frequent causes are repetitive surgical procedures, surgery for complications, inadequate preoperative diagnosis [pouch surgery and CD recurrence in previously diagnosed ulcerative colitis], and penetrating extensive small bowel disease.<sup>232–235</sup> In a French retrospective group of 38 patients with CD and chronic intestinal failure requiring home parenteral nutrition, severity of small bowel disease was a more important risk factor of intestinal failure than surgical procedures.<sup>235</sup> Preoperative optimisation, good nutritional preparation, preoperative modification of medical therapy,<sup>236–239</sup> minimal invasive surgery,<sup>240</sup> bowel-sparing surgery, and enhanced recovery pathways of perioperative care<sup>55,241</sup> reduce postoperative complications. Short small bowel resection and stricturoplasties in optimally prepared patients save the length of the small bowel.<sup>241,242</sup>

## 7.5. Stoma care

### 7.5.1. ECCO-ESCP Statement 5E

Specialised stoma care of IBD patients improves quality of life and decreases costs [EL3]

Some authors have reported a lower incidence of stomas in patients with CD in the biologics era.<sup>210</sup> Appropriate preoperative assessment and adherence to acceptable stoma construction techniques will avoid stoma-related complications.<sup>243</sup> Specialised stoma care and pre- and postoperative counselling improve quality of life of patients and significantly reduce re-hospitalisation, complications, and costs.<sup>244</sup>

## Conflict of Interest

ECCO has diligently maintained a disclosure policy of potential conflicts of interests [CoI]. The conflict of interest declaration is based on a form used by the International Committee of Medical Journal Editors [ICMJE]. The CoI statement is not only stored at the ECCO Office and the editorial office of *JCC*, but also open to public scrutiny on the ECCO website [<https://www.ecco-ibd.eu/about-ecco/ecco-disclosures.html>], providing a comprehensive overview of potential conflicts of interest of authors.

## Disclaimer

The ECCO Consensus Guidelines are based on an international consensus process. Any treatment decisions are a matter for the individual clinician and should not be based exclusively on the content of the ECCO Consensus Guidelines. The European Crohn's and Colitis Organisation and/or any of its staff members and/or any consensus contributor may not be held liable for any information published in good faith in the ECCO Consensus Guidelines.

## Acknowledgments

The following ECCO National Representatives participated in the review process of this consensus: Austria: Christoph Högenauer; Bosnia and

Herzegovina: Ante Bogut; Croatia: Brankica Mijandrušić-Sinčić; Cyprus: Ioannis Kaimakliotis; Denmark: Brynskov Jörn; Estonia: Karin Kull; France: Arnaud Bourreille; Germany: Britta Siegmund; Greece: Ioannis Koutroubakis; Hungary: Peter Lakatos; Italy: Anna Kohn; Moldova: Svetlana Turcan; Poland: Edyta Zagorowicz; Romania: Mihai Mircea Diculescu; The Netherlands: Marieke Pierik.

In addition, the following Additional Reviewers from ECCO also participated in the revision of the statements: Brazil: Paulo Gustavo Kotze; Croatia: Iva Hojsak; Germany: Matthias Brüwer; Italy: Livia Biancone, Stefano Scaringi, Giuseppe Sica; UK: Marco Gasparetto.

The following ESCP National Representatives participated in the review process of this consensus: Bulgaria: Dragomir Dardanov; Egypt: Waleed Omar; Finland: Anna Lepistö; Iceland: Tryggvi Stefansson; Israel: Nir Wasserberg; Poland: Zbigniew Lorenc; Slovenia: Pavle Kosorok; Sweden: Pamela Buchwald; Turkey: Dursun Bugra; UK: Karen Nugent.

In addition, the following Additional Reviewers from ESCP also participated in the revision of the statements: Austria: Irmgard E. Kronberger; Denmark: Alaa El-Hussuna; Germany: Peter Kienle; Italy: Cesare Ruffolo; Spain: Eloy Espín Basany; Sweden: Pär Myrelid; UK/USA: Charlotte Kvasnovsky.

## References

References for this paper are available as supplementary data at *ECCO-JCC* online.