Endoscopic management of polyposis syndromes: European Society of Gastrointestinal Endoscopy (ESGE) Guideline

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Appendix 1s
Online content viewable at: https://doi.org/10.1055/a-0965-0605

MAIN RECOMMENDATIONS
ESGE recommends that individuals with hereditary gastrointestinal polyposis syndromes should be surveilled in dedicated units that provide monitoring of compliance and endoscopic performance measures.

Strong recommendation, moderate quality of evidence, level of agreement 90%.

ESGE recommends performing esophagogastroduodenoscopy, small-bowel examination, and/or colonoscopy earlier than the planned surveillance procedure if a patient is symptomatic.

Strong recommendation, low quality of evidence, level of agreement 100%.
Introduction

Colorectal cancer (CRC) is the fourth most incident cancer and is the second commonest cause of cancer-related death in Europe [1]. While the majority of CRC is sporadic, twin studies have shown that up to 35% of CRC cases have a familial component [2]. Approximately 2%–5% of CRC cases are genetically determined by mutations in the adenomatous polyposis coli (APC), MUTYH, DNA mismatch repair, or other predisposing genes [3].

Although hereditary CRC syndromes are rare, it is of great importance that clinicians recognize these syndromes so they can make appropriate management decisions for both the patient and their family members who may also be at risk. Because all patients with polyposis syndrome are at high risk of developing gastrointestinal (GI) malignancies, endoscopic surveillance and interventions are required to prevent the development of cancer or to detect cancer at an early stage. Current evidence is limited. Therefore, the aim of this evidence-based and consensus guideline, commissioned by the European Society of Gastrointestinal Endoscopy (ESGE), is to provide clinicians with a comprehensive overview of the management options regarding endoscopic surveillance and interventions for the most important polyposis syndromes, namely familial adenomatous polyposis (FAP), MUTYH-associated polyposis (MAP), Peutz-Jeghers syndrome (PJS), juvenile polyposis syndrome (JPS), and serrated polyposis syndrome (SPS) (overview shown in Table 1 [4–28]).

There are several other polyposis-associated genes, including PTEN, GREM1, POLE/POLD1, and biallelic NTHL1, that will not be discussed in this guideline because of their low prevalence. A second guideline will focus on the endoscopic management of familial and hereditary non-polyposis syndromes.

Methods

The ESGE commissioned this guideline (chair J.v.H.) and appointed a guideline leader (M.v.L.), who invited the listed authors to participate in the project development. The key questions were prepared by the coordinating team (M.v.L. and V.R.) and were then approved by the other members. The coordinating team formed task force subgroups, each with its own leader, and divided the key topics among these task forces (Appendix 1s; see online-only Supplementary Material).

The process of developing the guideline included telephone conferences, meetings, and online and face-to-face discussions among the guideline committee members from July 2018 to June 2019. Searches were performed in MEDLINE, Embase, and Cochrane. Articles were selected through title and abstract screening, followed by full-text screening. The results of the search were presented to all members of the guideline committee and statements were created by consensus. Evidence levels and recommendation strengths were assessed using the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system [29]. Further details on the methodology of ESGE guideline development have been reported elsewhere [30].

In May 2019, a draft prepared by M.v.L. and V.R. was sent to all group members. After the agreement of all group members had been obtained, the manuscript was reviewed by a member of the ESGE governing board and an external reviewer, and was sent for further comments to the ESGE national societies and individual members. After this, it was submitted to *Endoscopy* for publication.
This guideline was issued in 2019 and will be considered for update in 2024. Any interim updates will be noted on the ESGE website: http://www.esge.com/esge-guidelines.html.

As literature on polyposis syndromes is limited, a Delphi procedure was organized within the guideline committee, consisting of two rounds, in order to gain consensus [31]. All guideline committee members, except for the research fellow, were asked to complete the online Delphi questionnaire in isolation, and responses were anonymized to prevent participants from influencing each other [32]. In each round, all the guideline committee members were first asked to rate all the statements with their level of agreement using a seven-point Likert scale: “Very strongly agree,” “Strongly agree,” “Agree,” “Neither agree nor disagree,” “Disagree,” “Strongly disagree,” or “Very strongly disagree” [33]. If the statement was not their area of expertise, participants had the option to opt out. Secondly, participants were asked if the statement was clear and had the opportunity to write down their suggestions for improvement. After the first round of Delphi voting, all statements were discussed and adjusted if necessary during a face-to-face meeting. Consensus was reached when ≥80 % of the guideline committee members had voted either “Very strongly agree,” “Strongly agree,” or “Agree” during the second round of the Delphi procedure.

1 General recommendations for patients with a polyposis syndrome

**RECOMMENDATION**
ESGE recommends that individuals with hereditary gastrointestinal polyposis syndromes should be surveilled in dedicated units that provide monitoring of compliance and endoscopic performance measures.

Strong recommendation, moderate quality of evidence, level of agreement 90 %.
Management of patients with polyposis syndrome is challenging. Strict follow-up of these patients with high quality endoscopy and polypectomy is essential. It has been proven that provision of healthcare services is more effective when delivered in an organized and coordinated system [34].

Data from the Danish polyposis registry showed a significantly lower CRC risk in call-up cases compared with probands who were not under surveillance. The tracing and follow-up program increased life expectancy by 17.0 years [35]. For these reasons, polyposis patients should be followed in dedicated units (national registries, genetic counseling centers, or high risk cancer centers) where endoscopic surveillance recommendations are monitored and audited, in order to improve adherence and provide the highest quality of care.

Surveillance intervals are provided in this guideline, but for patients with specific complaints, such as anemia, rectal blood loss, or abdominal pain, endoscopic interventions should be performed when indicated and not postponed to the next surveillance examination.

Table 2 and Table 3 provide a summary of all of the statements, including starting age and interval of endoscopic surveillance.

<table>
<thead>
<tr>
<th>Polyposis syndrome</th>
<th>Starting age</th>
<th>Surveillance interval</th>
<th>Treatment indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>(Attenuated) familial adenomatous polyposis</td>
<td>12 – 14 years</td>
<td>Every 1 – 2 years</td>
<td>Pre- and post-colectomy: remove all polyps &gt; 5 mm</td>
</tr>
<tr>
<td>MUTYH-associated polyposis</td>
<td>18 years</td>
<td>Every 1 – 2 years</td>
<td>Pre- and post-colectomy: remove all polyps &gt; 5 mm</td>
</tr>
<tr>
<td>Peutz–Jeghers syndrome</td>
<td>Baseline: 8 years Routine: 18 years</td>
<td>Baseline: if polyps found, every 1 – 3 years Routine: every 1 – 3 years</td>
<td>Elective polypectomy</td>
</tr>
<tr>
<td>Juvenile polyposis syndrome</td>
<td>12 – 15 years</td>
<td>Every 1 – 3 years</td>
<td>Elective polypectomy for polyps &gt; 10 mm</td>
</tr>
<tr>
<td>Serrated polyposis syndrome</td>
<td>NA</td>
<td>1 year: after ≥ 1 advanced polyp or ≥ 5 non-advanced clinically relevant polyps 2 years: after no advanced polyps or &lt; 5 non-advanced clinically relevant polyps</td>
<td>Clearing/approaching stage: remove all polyps ≥ 5 mm and all polyps of any size with optical suspicion of dysplasia</td>
</tr>
</tbody>
</table>

NA, not applicable.

RECOMMENDATION
ESGE recommends performing esophagogastroduodenoscopy, small-bowel examination, and/or colonoscopy earlier than the planned surveillance procedure if a patient is symptomatic.

Strong recommendation, low quality of evidence, level of agreement 100%.

2 Familial adenomatous polyposis and MUTYH-associated polyposis

2.1 Background

FAP is caused by an autosomal dominant mutation in the APC gene [36] (Table 1). The disease is characterized by the development of up to 100 – 1000 adenomas throughout the colon and rectum, and is also associated with extracolonic manifestations [4]. When the disease is left untreated, the cumulative risk of developing CRC is 100 % at a median age of 35 – 45 years [4]. Attenuated FAP (AFAP; arbitrarily defined as < 100 adenomas) is associated with a later onset of CRC and the absolute risk is thought to be lower than in those with a classical phenotype (> 100 adenomas) [5]. Duodenal adenomatosis is the most frequent extracolonic manifestation in FAP, and there are no robust data demonstrating that those with AFAP have a different duodenal phenotype to those with classical FAP. Approximately 10% – 30% of the patients with (attenuated) polyposis phenotype will remain without a detectable mutation. In these patients we suggest they be treated according to their clinical diagnosis.

There is no clear cutoff for referring an individual with a history of colorectal adenomatous polyps for genetic testing. The guideline of the American College of Gastroenterology advises referral for individuals with a history of 10 adenomatous polyps [37]. The Dutch guideline uses 10 or more colorectal adenomatous polyps in patients aged under 60 and 20 or more in those aged under 70 as a cutoff for referral [38].

The other main adenomatous polyposis syndrome is MAP, which is caused by a biallelic mutation in the MUTYH gene. Although there is significant phenotypic overlap with FAP, MAP is often associated with a lower number of colorectal polyps and a later age of onset, although significant phenotypic variation is observed [39, 40]. The lifetime risk for CRC in MAP patients ranges from 19% to 43% [6].

Table 2 Summary table of colonoscopy surveillance statements.
Compared with sporadic cancers, FAP is characterized by extremely early and multifocal carcinogenesis. However, the adenoma–carcinoma sequence is not accelerated, with adenomas taking up to 15 years to become malignant. Studies in patients with known **APC** mutation or clinical polyposis have shown that the median age of polyp development is 12–17 years [41–45]. In addition, the CRC rate below the age of 20 years is very low, approximately 1.3% [46].

Data also indicate that the **APC** mutation site may affect the severity of disease and cancer development. However, there is a wide spectrum of colorectal polyp burden in FAP and AFAP and care needs to be personalized [5]. Therefore, we recommend starting colonoscopy surveillance at age 12–14 years.

Active endoscopic surveillance is associated with a subsequent reduction of CRC incidence and mortality, mostly due to timely early surgical intervention. Studies showed that 47%–69% of symptomatic FAP patients were diagnosed with CRC, as opposed to 2%–4% of relatives with FAP in whom CRC was found during screening [47, 48].

### Table 3: Summary table of gastric and small-bowel surveillance statements.

<table>
<thead>
<tr>
<th>Polyposis syndrome</th>
<th>Modality</th>
<th>Starting age</th>
<th>Surveillance interval</th>
<th>Treatment indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>(Attenuated) familial adenomatous polyposis</td>
<td>Esophagogastroduodenoscopy</td>
<td>25 years</td>
<td>According to Spigelman score, adjusted for appearance of the ampulla</td>
<td>Non-ampullary adenomas: consider endoscopic resection of adenomas ≥ 10 mm. Ampullary adenomas: consider discussing endoscopic treatment in a multidisciplinary setting for adenomas ≥ 10 mm, showing excessive growth, or with suspicion of invasive growth</td>
</tr>
<tr>
<td>MUTYH-associated polyposis</td>
<td>Esophagogastroduodenoscopy</td>
<td>35 years</td>
<td>According to Spigelman score, adjusted for appearance of the ampulla</td>
<td>Non-ampullary adenomas: consider endoscopic resection of adenomas ≥ 10 mm. Ampullary adenomas: consider discussing endoscopic treatment in a multidisciplinary setting for adenomas ≥ 10 mm, showing excessive growth, or with suspicion of invasive growth</td>
</tr>
<tr>
<td>Peutz–Jeghers syndrome</td>
<td>Esophagogastroduodenoscopy</td>
<td>Baseline: 8 years Routine: 18 years</td>
<td>Baseline: if polyps found, every 1–3 years Routine: every 1–3 years</td>
<td>Elective polypectomy</td>
</tr>
<tr>
<td></td>
<td>MRI studies or video capsule enteroscopy</td>
<td>8 years</td>
<td>Every 1–3 years</td>
<td>Elective polypectomy for polyps &gt; 15–20 mm, preferably using device-assisted enteroscopy</td>
</tr>
<tr>
<td>Juvenile polyposis syndrome: SMAD4 mutation carriers</td>
<td>Esophagogastroduodenoscopy</td>
<td>18 years</td>
<td>Every 1–3 years</td>
<td>Gastric management should be discussed in a multidisciplinary setting</td>
</tr>
<tr>
<td>Juvenile polyposis syndrome: BMPR1A mutation carriers</td>
<td>Esophagogastroduodenoscopy</td>
<td>25 years</td>
<td>Every 1–3 years</td>
<td>Gastric management should be discussed in a multidisciplinary setting</td>
</tr>
<tr>
<td>Serrated polyposis syndrome</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

MRI, magnetic resonance imaging; NA, not applicable.
In 16%–40% of the individuals with 20–100 adenomas in whom FAP was excluded, a MUTYH mutation was found [37]. Furthermore, biallelic MUTYH mutations are found in 7.5% to 12.5% of patients with >100 adenomas in whom a disease-causing APC mutation is not found [6]. Nieuwenhuis et al. demonstrated that colorectal polyposis was diagnosed at a mean age of 44.8 years in 254 biallelic MUTYH mutation carriers, while CRC was diagnosed in 58% of these individuals at an average age of 48.5 years [49]. Furthermore, these patients had an 11% risk of developing metachronous CRC within 5 years after surgery, suggesting that biallelic MUTYH mutation carriers may have accelerated carcinogenesis.

Patients with a monoallelic MUTYH mutation do not develop adenomatous polyposis. They do however seem to have a slightly elevated risk of developing CRC compared with the general population, although this is not sufficient to warrant enhanced surveillance. The management of these individuals should be the same as for those in the general population [50, 51].

2.3 Management of colorectal neoplasia in patients with an intact colon

ESGE suggests that endoscopic management of colorectal adenomas alone is not recommended in individuals with familial adenomatous polyposis/MUTYH-associated polyposis. It may be considered in individuals who have an attenuated phenotype, provided that high quality surveillance and robust recall systems are in place. Weak recommendation, low quality of evidence, level of agreement 60%.

There are no data indicating that endoscopic polypectomy alone is an appropriate management strategy for patients with FAP. (Laparoscopic) prophylactic surgery is considered the standard of care. Most studies reveal a very narrow window between the diagnosis of colonic polyposis and surgery [43, 45]. However, postponing surgery might be considered based on overall polyp burden, in particular in those with an attenuated phenotype. Some patients with mild polyposis may even be managed endoscopically.

Furthermore, colectomy with ileorectal anastomosis instead of proctocolectomy with ileo-pouch anal anastomosis can be considered if the polyp burden in the rectum is relatively limited (usually <20 adenomas). The choice of surgery should take into account a personal or family history of desmoid disease, and mutation site in the context of social, personal, and educational factors. Weak recommendation, low quality of evidence, level of agreement 90%.

RECOMMENDATION
ESGE recommends that colonoscopy surveillance should start at the age of 18 years in asymptomatic individuals with MUTYH-associated polyposis. Strong recommendation, low quality of evidence, level of agreement 90%.

RECOMMENDATION
ESGE suggests that, in individuals with familial adenomatous polyposis/MUTYH-associated polyposis who are not in need of immediate colectomy and are manageable by endoscopy, all polyps >5 mm be removed. Weak recommendation, low quality of evidence, level of agreement 90%.

RECOMMENDATION
ESGE suggests that the timing and type of surgery in individuals with familial adenomatous polyposis/MUTYH-associated polyposis should be discussed in a multidisciplinary setting, thereby taking into account the sex (fertility), polyp burden, extensiveness of rectal involvement, personal and family history of desmoid disease, and mutation site in the context of social, personal, and educational factors. Weak recommendation, low quality of evidence, level of agreement 90%.

RECOMMENDATION
ESGE recommends that colonoscopy surveillance should start at the age of 18 years in asymptomatic individuals with MUTYH-associated polyposis. Strong recommendation, low quality of evidence, level of agreement 90%.

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2.4 Surveillance and management of colorectal neoplasia after (procto)colectomy

**RECOMMENDATION**
ESGE recommends endoscopic surveillance of the rectum or pouch every 1–2 years in individuals with familial adenomatous polyposis/MUTYH-associated polyposis depending on the polyp burden.
Strong recommendation, low quality of evidence, level of agreement 90%.

Evidence on how to manage polyps in the rectal remnant or pouch, and the appropriate interval between endoscopies is scarce. Some experts have shown that, even in severe cases of rectal polyposis, polyp burden in the rectal remnant can be effectively reduced by cold snare polypectomies and endoscopic submucosal resections [63,64]. One study recommends the use of argon plasma coagulation, but without evidence of its effect on cancer prevention [65].

2.5 Duodenal surveillance and management

**RECOMMENDATION**
ESGE recommends that individuals with familial adenomatous polyposis start endoscopic duodenal surveillance at the age of 25 years.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE suggests duodenal polyps and the ampulla should be biopsied only if they are not amenable to endoscopic removal, either because they are too large or because there is a suspicion of invasive growth.
Weak recommendation, low quality of evidence, level of agreement 89%.

**RECOMMENDATION**
ESGE recommends endoscopic removal of all polyps >5mm during surveillance of the rectum or pouch in patients with familial adenomatous polyposis/MUTYH-associated polyposis.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE recommends endoscopic removal of all polyps >5mm during surveillance of the rectum or pouch in patients with familial adenomatous polyposis/MUTYH-associated polyposis.
Strong recommendation, low quality of evidence, level of agreement 90%.

**RECOMMENDATION**
ESGE suggests considering endoscopic resection of non-ampullary duodenal adenomas ≥10mm in patients with familial adenomatous polyposis/MUTYH-associated polyposis.
Weak recommendation, low quality of evidence, level of agreement 90%.

**RECOMMENDATION**
ESGE recommends thorough inspection and description of the duodenum and ampullary site at every surveillance esophagogastroduodenoscopy in individuals with familial adenomatous polyposis/MUTYH-associated polyposis. The duodenal surveillance interval should be determined on the basis of polyp characteristics.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE recommends endoscopic surveillance of the rectum or pouch every 1–2 years in individuals with familial adenomatous polyposis/MUTYH-associated polyposis depending on the polyp burden.
Strong recommendation, low quality of evidence, level of agreement 90%.

**RECOMMENDATION**
ESGE suggests duodenal adenomas are detected, with a lifetime risk approaching 100% [66–70]. However, only a minority of patients develop duodenal cancer, with a cumulative risk ranging from 4% to 10% by...
the age of 60 [66, 69–73]. The median age at duodenal cancer diagnosis varied from 52 to 67 years [67, 69, 74–76]. Regular duodenal surveillance and prophylactic surgery has resulted in a significantly improved prognosis in FAP patients [74].

During esophagogastroduodenoscopy (EGD), the severity of duodenal polyposis is assessed using the Spigelman classification system (Table 4). Scores for the number, size, histology, and grade of dysplasia of the duodenal adenomas result in a Spigelman stage varying from I to IV [77]. Several risk factors for developing duodenal cancer are acknowledged: age; Spigelman stage IV at first endoscopy; duodenal polyps ≥10 mm or containing high grade dysplasia; and amillary adenomas with high grade dysplasia, a (tubulo) villous component, or high grade dysplasia [67, 70, 74–76]. To obtain all components of the Spigelman score, pathology results are needed; however, routine biopsies of duodenal polyps may interfere with optical diagnosis and future endoscopic resection because of fibrosis. Therefore, taking routine biopsies is currently not recommended. If endoscopic removal is not necessary because the adenomas are small and there is no suspicion of invasive growth, the Spigelman stage should be determined based on previous pathology reports or optical diagnosis to determine the severity of duodenal polyposis and the surveillance interval. The site of the ampulla in particular should be evaluated and reported accurately, as this is a location of preference for adenoma and cancer development [78].

**Table 4** Spigelman Score, adapted from Spigelman et al. [77].

<table>
<thead>
<tr>
<th>Findings at duodenoscopy</th>
<th>1 point</th>
<th>2 points</th>
<th>3 points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of adenomas</td>
<td>1–4</td>
<td>5–20</td>
<td>&gt;20</td>
</tr>
<tr>
<td>Size, mm</td>
<td>1–4</td>
<td>5–10</td>
<td>&gt;10</td>
</tr>
<tr>
<td>Histology*</td>
<td>Tubular</td>
<td>Tubulovillous</td>
<td>Villous</td>
</tr>
<tr>
<td>Dysplasia*</td>
<td>Low grade</td>
<td>NA</td>
<td>High grade</td>
</tr>
</tbody>
</table>

* Based on pathology obtained for complete endoscopic removal of duodenal polyps or prior pathology results.

**RECOMMENDATION**

ESGE recommends starting endoscopic duodenal surveillance in individuals with MUTYH-associated polyposis at 35 years of age.

**Strong recommendation, low quality of evidence, level of agreement 90%**.

In MAP, the prevalence of duodenal adenomas is lower than in individuals with FAP, with 17%–34% at a median age of 50 years [90, 91]. Only 6% of these patients with MAP developed ampullary disease [90]. Because duodenal polyposis occurs later in life and with a slower progression than in individuals with FAP, duodenal surveillance may commence at a higher age. Walton et al. showed that only 8 of 92 MAP patients (9%) underwent an endoscopic intervention, starting at 38 years [90]. In this series, two duodenal cancers were diagnosed in
patients with MAP over the age of 60 years who were not undergoing surveillance [90]. Duodenal cancers in MAP patients can often occur without significant duodenal polyp burden [90, 92].

RECOMMENDATION
ESGE recommends performing thorough gastric assessment at the time of duodenal surveillance. If gastric adenomas are suspected, endoscopic resection is recommended, or surgical resection if endoscopically unresectable.

In patients with FAP, fundic gland polyps are reported in 20%–88% [99, 100]. Fundic gland polyps are thought to have little tendency for malignant transformation. On the other hand, gastric adenomas are considered to have a premalignant potential, given that 8%–14% of gastric adenomas harbor high grade dysplasia [101, 102]. Historically, the risk of developing gastric cancer among Western FAP patients was not found to be higher than the general population [102–104]. However, two recent series from Western countries, described 17 cases of gastric cancer, with a median age at diagnosis between 50 and 60 years [102, 103]. In both series, the proximal cancers were associated with carpeting fundic gland polyposis, which can make identification of the premalignant adenoma extremely difficult. These findings suggest that identification and resection of gastric adenomas are important to prevent the development of gastric cancer, but currently there are no data as to whether or not this is effective.

RECOMMENDATION
ESGE suggests treatment for individuals with familial adenomatous polyposis/MUTYH-associated polyposis who have ampullary adenomas ≥ 10 mm showing excessive growth or suspicion of invasive growth should be discussed in a multidisciplinary setting.

Weak recommendations, low quality of evidence, level of agreement 100%.

Duodenal polyps in FAP and MAP often occur in the region of the ampulla [78]. To prevent ampullary cancer, endoscopic ampullectomy can be performed in individuals with adenomatous changes of the ampulla. However, ampullectomy is associated with severe complications, therefore benefits and harms should be weighed in an experienced multidisciplinary setting. The effect of endoscopic ampullectomy has been evaluated in three small observational studies, including 8–28 FAP patients [93–95]. In these series, complication rates such as pancreatitis (19%–20%), bleeding (4%–13%), and abdominal pain (8%) were high [93, 94]. Recurrence at the site of ampullectomy occurred in 0–67% of the cases after a follow-up ranging from 53 to 85 months with no evidence of ampullary cancer [93–95]. In one study of 15 FAP patients, two (13%) required surgery after multiple repeated endoscopic resections [93].

Finally, if endoscopic ampullectomy is indicated but not possible in an expert center, the patient should be referred for surgical intervention.

RECOMMENDATION
ESGE suggests that endoscopic ultrasonography should not be routinely performed in the pretherapeutic evaluation of ampullary adenomas in individuals with familial adenomatous polyposis/MUTYH-associated polyposis. It may be considered for assessment of large or suspicious ampullas to help exclude invasive cancer.

Weak recommendation, low quality of evidence, level of agreement 89%.

In the literature, endoscopic ultrasonography (EUS) for the pretherapeutic staging of ampullary tumors has focused mainly on advanced ampullary cancers. One study focusing on ampullary adenomas in 38 FAP patients showed no EUS utility, with no information on duct involvement [93]. A comparison of preoperative staging of ampullary tumors showed comparable accuracy of EUS and intraductal ultrasound (IDUS), with an accuracy of 63% (EUS) and 78% (IDUS), in particular for advanced stages [96]. On the other hand, over-staging at EUS/IDUS occurred in 25%–40% of cases of benign adenoma or early cancers [96–98]. Therefore, EUS and IDUS present limitations in the pretherapeutic evaluation of ampullary tumors, with over-staging of early and even benign lesions.
3 Peutz–Jeghers syndrome

3.1 Background

PJS is characterized by the development of hamartomatous polyps [3]. PJS is diagnosed using clinical criteria (Table 1) or by a pathogenic germline mutation in the serine threonine kinase 11 tumor suppressor gene (STK11/LKB1) gene, which is found in 80%–94% of PJS patients [7]. Individuals with perioral or buccal pigmentation and/or two or more GI hamartomatous polyp(s) or a family history of PJS should be referred for genetic testing [37].

The predominant clinical feature of PJS is GI polyposis, most often found in the small bowel (60%–90%), where they may cause bleeding, anemia, and intussusception [108, 109]. The cumulative risk of GI cancers (excluding pancreatic cancer) has been reported to be around 33% at the age of 60, increasing to 57% at the age of 70 years [8]. However, data are often historical, retrospective, and subject to bias that probably overestimates the cancer risk. Surveillance of the GI tract in PJS patients has two purposes: (i) to detect GI polyps that may cause complications (bleeding, anemia, intussusception) and should be removed (in particular small-bowel polyp-related complications are the predominant clinical problem) [110, 111]; (ii) to detect cancer (mainly occurring in adults) at an early stage [9].

3.2 Esophagogastroduodenoscopy and colonoscopy surveillance

RECOMMENDATION
ESGE recommends a baseline esophagogastroduodenoscopy and colonoscopy at the age of 8 years in asymptomatic individuals with Peutz–Jeghers syndrome.
Strong recommendation, low quality of evidence, level of agreement 100%.

RECOMMENDATION
ESGE recommends starting routine esophagogastroduodenoscopy and colonoscopy surveillance at the age of 18 if the baseline endoscopy is negative.
Strong recommendation, low quality of evidence, level of agreement 100%.

RECOMMENDATION
ESGE recommends an interval of 1–3 years based on phenotype for small-bowel surveillance.
Strong recommendation, moderate quality of evidence, level of agreement 100%.

RECOMMENDATION
ESGE recommends either MRI studies or video capsule enteroscopy for small-bowel surveillance.
Strong recommendation, moderate quality of evidence, level of agreement 89%.

Most studies about cancer risk in PJS patients are single-center cohort studies and rather small, which may overestimate the cancer risk because of ascertainment bias. Giardiello et al. performed a systematic review including 210 PJS patients from six studies and reported a cumulative risk of gastric cancer of 29% at 15–64 years of age, with a relative risk (RR) of 213 (95% confidence interval [CI] 96–368) compared with the general population [112]. The average age of gastric cancer diagnosis was 30–40 years [9, 113]. The cumulative risk of colon cancer was 39% at 15–64 years of age, with an RR of 84 (95% CI 47–137) [113–115].

There are no prospective studies evaluating the effect of surveillance strategies for gastric cancer, duodenal cancer, or CRC. Furthermore, there is no evidence regarding the type and frequency of surveillance and starting/stopping age. Hamartomas are predominantly found in the small bowel and colon and only seldomly give rise to complications in the esophagus or stomach. Latchford et al. evaluated 28 PJS patients who had undergone one or more surveillance endoscopies by the age of 18 [111]. In 17 patients a significant gastroduodenal or colonic polyp was found, including 20 gastroduodenal polyps over 10 mm [111]. In this series, no PJS patients were observed to develop GI cancer. Furthermore, dysplasia or atypia was very rarely observed.

3.3 Small-bowel surveillance

RECOMMENDATION
ESGE recommends small-bowel surveillance from the age of 8 years in asymptomatic individuals with Peutz–Jeghers syndrome.
Strong recommendation, moderate quality of evidence, level of agreement 100%.
Symptoms related to small-bowel polyps are frequent and intussusception is seen by the age of 10 in 33% and by the age of 20 in 50% of PJS patients [110]. The cumulative risk of small-bowel cancer was 13%, with an RR of 520 (95%CI 220 – 1306) [113]. The average age of diagnosis of small-bowel cancer was 37–42 years [9, 113]. However, it is difficult to interpret these data because of the small studies, which may overestimate cancer risk due to ascertainment bias, and misinterpretation of pseudoinvasion as cancer.

Currently, magnetic resonance imaging enteroclysis/enterography (MRI-E) and video capsule endoscopy (VCE) are the most used imaging modalities for detection of polyps in the small bowel [109, 116–119]. There are four studies that have compared MRI-E and VCE, including a total of 47 patients with PJS [118–121]. Gupta et al. [118] did not find a significant difference between the two modalities for the detection of clinically relevant polyps (> 10 mm), as opposed to Urquhart et al. [119], who showed superiority for VCE over MRI-E. Both modalities do miss clinically relevant polyps (> 15–20 mm or smaller polyps that do give rise to symptoms). Based on the current literature, both VCE and MRI-E are reasonable options for small-bowel surveillance.

3.4 Management of small-bowel polyps

**RECOMMENDATION**

ESGE recommends that elective polypectomy should be performed for small-bowel polyps >15–20 mm to prevent intussusception. In a symptomatic patient, smaller polyps causing obstructive symptoms should be removed.

Strong recommendation, low quality of evidence, level of agreement 90%.

**RECOMMENDATION**

ESGE recommends device-assisted enteroscopy for the removal of polyps. Based on phenotype, intraoperative enteroscopy could be considered.

Strong recommendation, moderate quality of evidence, level of agreement 89%.

In a cohort study including 110 PJS patients, 69% developed at least one intussusception at a median age of 16 years [110]. The intussusception occurred in the small bowel in 95% of the cases. Based on the histology of 37 cases, intussusception occurred owing to polyps with a median diameter of 35 mm (15–60 mm). In almost all publications, the indication for balloon enteroscopy is set at polyps over 10–15 mm on VCE or MRI-E, although some studies used a threshold of 20 mm [109]. Several studies have shown that polypectomy of relevant small-bowel polyps can prevent the need for emergency surgery [108, 122, 123].

Balloon-assisted enteroscopy facilitates polypectomy in almost all patients with clinically relevant polyps [109]. Single-balloon and double-balloon enteroscopy (DBE) have been shown to be effective for the removal of polyps up to 60 mm [124] and 100 mm [125], respectively. Prior abdominal surgery is not a contraindication for balloon enteroscopy. For individuals with too many small-bowel polyps, or large or high risk polyps, laparoscopically-assisted DBE or intraoperative enteroscopy can be performed [123].

The effect on cancer reduction is not known. Only one T2N0 adenocarcinoma in the jejunum has been detailed in the DBE literature, which has reported more than 3000 polypectomies [109, 111, 126].

4 Juvenile polyposis syndrome

4.1 Background

The diagnosis of JPS is based on clinical criteria [10] (Table 1). Individuals with five or more juvenile polyps in the colorectum or any juvenile polyps in other parts of the GI tract should undergo genetic testing [37]. A germline mutation in SMAD4 or BMPR1A is identified in around 40%–60% of those with a clinical diagnosis. Germline mutations in these genes result in two relatively different phenotypes [127]. SMAD4 mutation carriers present with colonic and gastric involvement, in combination with hereditary hemorrhagic telangiectasia (HHT), whereas BMPR1A mutation carriers mostly develop a colonic phenotype [11, 12]. JPS is associated with an increased GI cancer risk varying from 39% to 68% [10, 13].

4.2 Colonoscopy surveillance

**RECOMMENDATION**

ESGE recommends that colonoscopy screening in asymptomatic individuals with juvenile polyposis syndrome starts at the age of 12–15 years.

Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**

ESGE recommends an interval of 1–3 years based on phenotype for routine colonoscopy surveillance in individuals with juvenile polyposis.

Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**

ESGE recommends that colorectal polyps >10 mm should be removed in individuals with juvenile polyposis syndrome to prevent complications and the development of colorectal cancer.

Strong recommendation, low quality of evidence, level of agreement 90%.
Almost all patients with SMAD4 and BMPR1A germline mutations present with colonic hamartomas, with a wide range of disease expression from a few polyps to over 100 polyps [128–130]. Very young patients with symptomatic polyposis have been reported (4–12 years) [129, 130]. In the largest published series of 84 cases fulfilling the clinical criteria for JPS, from the Johns Hopkins’ hospital, 8 of the 84 patients (9.5%) developed CRC between the ages of 30 and 58 years, with a lifetime calculated risk of 37% [13]. In another retrospective series from Baltimore, the frequency of colectomy was 49% [128]. Besides classical cases, a much more severe phenotype has been described in patients harboring a microdeletion in chromosome 10 that involves both the BMPR1A and PTEN genes [131].

4.3 Esophagogastroduodenoscopy surveillance

**RECOMMENDATION**
ESGE recommends that esophagogastroduodenoscopy surveillance should start at the age of 18 years in asymptomatic individuals with a SMAD4 mutation.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE suggests that esophagogastroduodenoscopy surveillance should start at the age of 25 years in asymptomatic individuals with a BMPR1A mutation.
Weak recommendation, low quality of evidence, level of agreement 90%.

**RECOMMENDATION**
ESGE recommends an interval of 1–3 years depending on phenotype for esophagogastroduodenoscopy surveillance in individuals with juvenile polyposis syndrome.
Strong recommendation, low quality of evidence, level of agreement 90%.

RECOMMENDATION
ESGE recommends gastric management (polypectomy, surgery, surveillance) be discussed in expert multidisciplinary teams as no clear algorithm can be proposed based on the available data.
Strong recommendation, low quality of evidence, level of agreement 100%.

The lifetime risks of extracolonic cancers, including stomach, pancreas, and small intestine, are difficult to quantify owing to a lack of good quality data. Risks that have been reported vary from 20% to 60% [132]. However, these are likely to be influenced by overestimation of risk due to ascertainment bias.

4.4 Small-bowel surveillance

Small-bowel involvement in JPS is rare and, if present, predominantly located in the duodenum [127, 128, 130]. Wain et al. found a prevalence of 29% for duodenal polyps in SMAD4 mutation carriers [130]. Involvement of the distal duodenum in JPS is not described [134, 135]. In addition, no cases of jejunal or ileal carcinoma have been reported. Therefore, EGD seems to be sufficient for small-bowel surveillance in JPS patients. Finally, the association of SMAD4 mutation with HHT suggests that, in expert centers, management of iron deficiency anemia unexplained by EGD and colonoscopy could be an indication for small-bowel evaluation with VCE. In patients with evidence of HHT, screening for vascular lesions in other organs should be performed.

5. Serrated polyposis syndrome

5.1 Background

SPS has emerged as the most frequent form of polyposis, with an estimated prevalence of up to 1:111 (0.9%) of individuals in fecal occult blood test-based screening cohorts and up to 1:238 (0.42%) in primary screening cohorts [14–17]. SPS is often grouped with the hereditary polyposis syndromes although no underlying gene defect has been identified yet. SPS is diagnosed using clinical criteria defined by the World Health Organization criteria, recently revised [18, 136].

The prevalence of CRC in patients with SPS has been estimated to range between 15% and 30% and there is an increased risk for CRC prior to or at the time of SPS diagnosis and treatment [14, 19–22]. In one prospective and three retrospective cohorts, the cumulative 5-year incidence of CRC under endoscopic surveillance ranged between 0 and 7.0% [14, 19, 20, 137].

**RECOMMENDATION**
ESGE does not recommend small-bowel surveillance in asymptomatic individuals with juvenile polyposis syndrome.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE recommends gastric management (polypectomy, surgery, surveillance) be discussed in expert multidisciplinary teams as no clear algorithm can be proposed based on the available data.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE recommends esophagogastroduodenoscopy surveillance should start at the age of 18 years in asymptomatic individuals with SMAD4 mutation.
Strong recommendation, low quality of evidence, level of agreement 100%.

**RECOMMENDATION**
ESGE recommends esophagogastroduodenoscopy surveillance should start at the age of 25 years in asymptomatic individuals with BMPR1A mutation.
Weak recommendation, low quality of evidence, level of agreement 90%.

6. Conclusion

The lifetime risks of extracolonic cancers, including stomach, pancreas, and small intestine, are difficult to quantify owing to a lack of good quality data. Risks that have been reported vary from 20% to 60% [132]. However, these are likely to be influenced by overestimation of risk due to ascertainment bias.
In SPS patients, successful endoscopic treatment at diagnosis (the so-called “clearing phase”) can be achieved in the majority of patients [14, 20, 138]. However, clearing in some cases requires commitment, time, and expertise to perform a large number of polypectomies in one or more procedures [138]. Accordingly, these patients should be managed in dedicated units with expert endoscopists in order to prevent unnecessary surgery. Studies with expert endoscopists have shown that EMR of large serrated lesions is easy, safe, and has a lower recurrence rate than for adenomas [139].

The risk of developing CRC during endoscopic surveillance following diagnosis and clearing of the initial polyp burden seems to be low. Based on two large retrospective cohort studies, the cumulative incidence during surveillance varied from 0 to 3.1% after 3–5 years [14, 20]. The median interval between surveillance colonoscopies in these cohort studies varied between 12 and 19 months [14, 19–22, 138, 140]. Although the CRC risk during surveillance is low, one retrospective and one prospective cohort study reported that the incidence of advanced neoplasia during surveillance is as high as 34%–42% after 3 years of surveillance [19, 22].

1 Advanced polyps: (tubulo)villose adenomas, adenomas with high grade dysplasia, adenomas ≥10 mm in diameter, traditional serrated adenomas, serrated lesions with dysplasia, serrated lesions ≥10 mm in diameter.

2 Non-advanced clinically relevant polyps: any adenoma or serrated polyp that does not meet the criteria for an “advanced polyp,” with the exception of hyperplastic polyps <5 mm in diameter (which can be left in situ).
use of conventional chromoendoscopy improves polyp detection and could be considered in the surveillance of SPS patients. However, its routine use must be balanced against practical considerations.

Finally, a recent RCT evaluated the usefulness of Endocuff-assisted colonoscopy in the surveillance of SPS [146]. In this study, with 123 SPS patients included, no statistical differences were found between Endocuff-assisted colonoscopy and HD-WLE colonoscopy for the detection of overall polyps, sessile serrated lesions, sessile serrated lesions, and adenomas.

5.4 Screening of first-degree relatives

Most SPS cases seem non-familial. However, the presence of the disease in family members has been described in previous reports [137,147,148]. Moreover, various studies have described an increased incidence of CRC in relatives of patients with SPS. Boparai et al. investigated the risk of CRC in 347 first-degree relatives of 57 patients with SPS; they established an absolute risk of CRC of 8% and an RR of 5.4 (95%CI 3.7–7.8) [149]. Two other studies reported an absolute risk of CRC of 12.2–15.4 in first-degree relatives [150,151]. The age at diagnosis of CRC in relatives ranged from 55 to 62 years in these studies [148–150]. During follow-up of these first-degree relatives of patients with SPS, retrospective studies [148,152,153] found a high risk of CRC and advanced polyps. Hazewinkel et al. prospectively investigated the yield of screening colonoscopy in 77 first-degree relatives of patients with SPS in whom no CRC was found, with significant polyps being present in 43% of patients [154].

Discussion

The management of patients with polyposis syndromes is challenging. The various types of polyposis syndrome have variable risks for a large spectrum of cancers. In addition, the phenotype may differ among individuals having a specific germline mutation, and even within/between family members carrying the same mutation. Furthermore, in a proportion of patients with clinical polyposis, no germline mutation can be identified. This guideline gives a framework on how these patients should be endoscopically managed according to the current literature and expert opinion (Table 2 and Table 3).

The ESGE aligns with the European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) guidelines on polyposis syndromes in children and young adults [155–157]. The ESPGHAN guideline differs from this guideline with regard to the colonoscopy interval for FAP patients with intact colon, with this being 1–3 yearly in the ESPGHAN guideline and 1–2 yearly in our guideline [156]. We have chosen to align the FAP and MAP surveillance intervals to make it less confusing for endoscopists. Again, the interval should mainly be based on phenotype and the endoscopist may lengthen the surveillance interval based on adenoma characteristics (number, size, and degree of dysplasia). The main difference with the American College of Gastroenterology (ACG) guideline is the proposed endoscopic management for gastric and duodenal adenomas in (A)FAP and MAP patients [37]. In contrast with the ACG guideline, the ESGE guideline does not recommend random sampling of fundic gland polyps during EGD surveillance. Furthermore, the ESGE advises endoscopic polypectomy of duodenal adenomas of ≥10 mm.

Disclaimer


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Competing interests

E. Dekker was an advisory board chair for Cancer Prevention Pharmaceu
ticals (2019) and is a co-editor for Endoscopy. M. F. Kaminski has
received speaker’s, teaching, and consultancy fees from Olympus
(2017 to present) and speaker’s and teaching fees, and a loan of equipment from Fujiﬁlm (2019). H. Neuman has provided consultan
cy services to Fujiﬁlm and Pentax (2012 to present). M. Pellissi has re
cieved consultancy fees from Norgine Iberia (2019), speaker’s fees from Casen Recordati (2017–2019), Olympus (2017), and Jansen
(2018), and is a co-editor for Endoscopy; her department has received an equipment loan from Fujiﬁlm (2017 to present) and a research do
nation from Fujiﬁlm (2019). J. E. van Hootf has received lecture fees from Medtronics (2014–2015) and Cook Medical (2019), and consul
tancy fees from Boston Scientiﬁc (2014–2017); her department has re
ceived research grants from Cook Medical (2014–2018) and Ab

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Appendix 1s: Key questions and task force members/leads.

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