Lynch syndrome is the most prevalent cause of inherited colorectal cancer with a lifetime risk of CRC of up to 50%. There is also an increased risk of other cancers, including endometrial, ovarian, gastric and small bowel. Gastric cancer was initially reported as the most common cancer after CRC and endometrial cancer with up to an 11% lifetime risk. Consequently, there was the recommendation that surveillance gastroscopy be considered in individuals with Lynch syndrome.

The incidence of gastric cancer in the general population has shown a marked decline over the last 30 years due to a number of reasons, including the identification and eradication of H. pylori. This decline is mirrored in the Lynch syndrome population, and more recent data suggests the risk of gastric cancer is lower, between 1–6% depending on genotype.1

Previously, based on published risks, NZFGICS recommended annual surveillance gastroscopies in Lynch syndrome patients with an MLH1 and MSH2 mutation. We wanted to re-assess the appropriateness of these recommendations and therefore undertook an audit of the outcome of surveillance gastroscopy in these patients.

Between 1 May 2011 and 1 November 2014, 320 gastroscopies were performed in 177 mutation-positive individuals across New Zealand. There were two cancers detected at surveillance. One adenocarcinoma at the GO junction arising within a short segment of Barrett’s oesophagus and one duodenal cap tumour. Both of these individuals underwent curative resection. In summary, 160 gastroscopies were needed to identify one resectable upper GI cancer.

As a result of these reassuring results as well as recently published international Consensus Guidelines2–3 and aware of the resource-constrained environment in which we practice, from mid-2016 we have changed our recommendations with regards to gastroscopy surveillance in Lynch syndrome.

As a result, we now recommend only a single gastroscopy at the age of 35 years in Lynch syndrome patients carrying the MLH1 or MSH2 mutation. This should include 1) inspecting the distal duodenum as 50% of small bowel tumours occur in the duodenum and 2) eradicating H. pylori if present. If there are significant findings (such as extensive intestinal metaplasia), further surveillance gastroscopy will be considered in three years.

There are no specific recommendations for gastroscopic surveillance in Lynch syndrome patients with the other MMR mutations. If there is a history of gastric cancer in a particular family, specific recommendations may be made by the Service.

Familial adenomatous polyposis—update on surveillance of the anal canal and retained rectum

Familial adenomatous polyposis patients who have undergone proctocolectomy and ileorectal anastomosis require ongoing surveillance of the anal canal as there remains a risk of adenoma and potentially carcinoma development. The risk of neoplastic development at this site is reduced if a mucosectomy is performed in preference to a double-stapled pouch-rectal anastomosis, and consideration may be given to this at the time of pouch formation. Adenomas may also develop in the ileum of the pouch and should be biopsied or removed.
Therefore we recommend annual ‘pouchoscopy’ for all patients with an ileal pouch, up to the age of 75 years, with careful examination of the pouch-anal anastomosis (including retroflexion). A paediatric colonoscope may be preferable in this regard. Examination of the pouch can be usefully combined with upper GI endoscopy to examine the duodenum for adenomatous polyp formation according to recommendations for upper GI surveillance in FAP patients.

In individuals who have undergone an ileorectal anastomosis and have a retained rectum with increasing polyp burden, the frequency of surveillance may be increased. Chemoprotective agents including Sulindac or Celecoxib may reduce the polyp burden but do not change the recommendations. If the polyp burden is uncontrolled endoscopically, consideration should be made for completion proctectomy +/- pouch formation.

**Serrated polyposis syndrome (SPS)**

There are three criteria as defined by the WHO (2010) for the diagnosis of serrated polyposis syndrome (SPS)

1. Five or more serrated polyps proximal to sigmoid colon (2>10mm)
2. Any number of serrated polyps proximal to the sigmoid colon in an individual who has a first-degree relative with serrated polyposis
3. >20 serrated polyps of any size throughout the colon (not confined to rectum). Note this polyp count is cumulative over repeated colonoscopies.

As hyperplastic polyps are simple serrated polyps, the term hyperplastic polyposis syndrome has been replaced by the term serrated polyposis syndrome.

Although the initial reports of colonoscopy surveillance in SPS cancer reported cumulative colorectal risks during surveillance of 6.5–7%, more recent studies have been provided evidence that colonoscopy surveillance is safe. However, control of the initial polyp burden is essential as is caution in extending the surveillance interval, particularly in those with higher than 20 pan colonic polyps. Consequently, in line with international recommendations, because the initial polyp burden in some patients with SPS can be high, we advise that an initial colonoscopy may be required every 3–6 months to clear all polyps (or at least ones >10mm). Once control of polyp burden is achieved, annual surveillance colonoscopy is recommended with removal of all lesions >5mm and smaller as time allows. In our cohort of 96 patients with SPS (polyps alone at presentation) followed for a median of 4.8 years, the majority of patients have an average of fewer than 10 polyps at the fourth colonoscopy procedure. This refers to colonoscopies performed, after diagnosis of SPS, at intervals appropriate to the polyp burden.

Based on these findings we advise that patients who have had two consecutive annual colonoscopies meet the following criteria:

1. Less than 10 polyps where the majority of polyps are less than 5mm in size
2. All right-sided polyps have been removed
3. No histology of concern such as SSPs with dysplasia
4. Good bowel preparation (particularly in the right colon).

Then an extension of the surveillance interval to two-yearly can be considered.

However, a return to annual surveillance should be considered if the polyp burden exceeds these criteria at any procedure. A possible algorithm of clinical and endoscopic management in SPS has recently been proposed in GUT by Hassan and is in keeping with the above recommendations, although we have been more conservative with regards to polyp size with regards to extending the interval.

There will be some patients with SPS whose polyps are not adequately controlled by colonoscopy and polypectomy, even if colonoscopy is initially performed at 3–6 monthly intervals—in these patients surgery may be the best management option. It would appear from our data that a review of the polyp findings at the fourth colonoscopy could alert clinicians to which patients may not be achieving adequate polyp control within an appropriate time frame—such patients could then be kept under close review with consideration being given to the option of surgical management.

Risk factors for colorectal cancer in SPS have also been identified and are summarised below.
These factors should be taken into consideration when determining surveillance intervals in SPS.

- Any proximal polyp SSP with high-grade dysplasia
- Two proximal SSA/P’s
- 1 serrated polyp (SP) with dysplasia or advanced adenoma
- Fulfilment of both WHO criteria 1 and 3

Patients who only meet WHO criterion 2 for SPS while still at increased risk of CRC should undergo surveillance colonoscopy on a five-yearly basis or as dictated by their polyp burden. If these patients eventually meet criterion 1 or 3 in their own right, they should enter appropriate SPS surveillance.

Two studies4,11 have documented that first-degree relatives of individuals with SPS have a five-fold increase in their lifetime risk of developing bowel cancer and a higher risk of developing SPS themselves. For this reason, it is recommended that first-degree relatives be offered five-yearly bowel screening by colonoscopy from the age of 40 to 75 years if they are otherwise well.

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Nil.

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