

Serrated Polyposis Syndrome

Key Points

- Serrated Polyposis Syndrome (SPS) is a syndrome in which multiple hyperplastic or serrated polyps are identified in the large bowel.
- It is estimated that I in 3000 people may have this condition.
- This syndrome has been identified in recent years to be associated with an increased risk of developing bowel cancer. Other cancers are not usually associated with this syndrome apart from possibly pancreatic cancer at older ages.
- Regular bowel screening by colonoscopy (with removal of polyps) can prevent bowel cancers developing
- Colonoscopy is usually performed yearly in the initial years after a diagnosis of SPS. This is to ensure that the polyps can be controlled by removal at colonoscopy. After a few years the interval between procedures may lengthen.
- Occasionally bowel surgery is required when polyps are multiple, large and show early signs of bowel cancer.
- Cigarette smoking is associated with increased polyp numbers.
- SPS is more common in individuals of European or Celtic descent
- Up to 50% of individuals with SPS may have a family history of bowel cancer suggesting a possible genetic or inherited cause but this has not been confirmed. Genetic testing is not available.
- First degree relatives of individuals with SPS i.e. mother, father, sister, brother, children, have been shown in studies to have a 5 fold increase in their life time risk of developing bowel cancer when compared with the general population.
- First degree relatives also have an increased risk of developing SPS
- First degree relatives are advised to have 5 yearly bowel screening by colonoscopy from the from the age of 40 years or from an age 10 years younger than the youngest age at which bowel cancer or SPS was identified in the family.
- The aim of the NZFGICS is to reduce the number of cancers occurring in families by facilitating the required bowel screening

New Zealand has a high incidence of bowel cancer with approximately 6% of New Zealanders developing bowel cancer by the age of 75 years. Most bowel cancer (90%) is diagnosed is people over the age of 50 years and in most individuals, it develops over a 5 to 10 year period from a bowel polyp called an adenoma. A polyp is an outgrowth from the bowel but there can be various types of polyps.

Approximately 20% of New Zealanders over the age of 50 would be identified at colonoscopy to have a small adenomatous polyp but the vast majority of these polyps, particularly if they are small in size and number and do not showing advanced changes under the microscope, would not develop into bowel cancer.

The other major type of bowel polyp called the hyperplastic or serrated polyp has been recognized for many years. Often small numbers of small (less than 5mm) hyperplastic polyps are seen in the distal bowel or back-passage. These have been considered to be of no concern. However, in recent years it has been recognized that some serrated polyps may progress to bowel cancer and this risk is increased if hyperplastic or serrated polyps are present throughout the whole length of the large bowel and not just confined to the distal bowel. Indeed on the basis of their appearance under the microscope, serrated polyps have now been sub categorized to try and identify the types of

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serrated polyps that may be associated with progression to bowel cancer. Often the "advanced" serrated polyps of concern occur in the proximal large bowel or right colon and they may be quite flat and difficult to detect at colonoscopy.

A diagnosis of the Serrated Polyposis Syndrome (SPS) is usually made when an individual is identified to have over 20 hyperplastic or serrated polyps throughout the colon or where there are at least five such polyps beyond the distal bowel and two are equal to or greater than 10 mm in size. Sometimes at an initial colonoscopy there may be suspicion that a diagnosis of SPS may be made after a future colonoscopy in a few years. For this reason the criteria for a diagnosis of SPS allow inclusion of the cumulated numbers of polyps.

It has been recognized that approximately 30 to 50% of patients with SPS present with bowel cancer. Unfortunately, we do not actually know the true risk for developing bowel cancer because although it is estimated that 1 in 3,000 people in the population have SPS, we do not know how many of those people would actually develop bowel cancer in their lifetime.



A number of studies have now documented the importance of frequent colonoscopy, with removal of polyps, in individuals identified to have SPS. Colonoscopy is usually offered one to two yearly, once control of the number of polyps has been achieved. Sometimes colonoscopy may be performed more frequently in the first instance to ensure removal of the majority of identified polyps.

On occasions it is not possible to remove all the bowel polyps at colonoscopy because of their size or because they are so numerous and sometimes the removed polyps, when examined under the microscope, are identified to be advanced serrated polyps. In these situations a small percentage of patients with SPS may be advised to have bowel surgery but this does not usually involve a stoma (colostomy/ileostomy).

A genetic predisposition to developing SPS is suspected because up to 50% of individuals with SPS may have a family history of bowel cancer. However, to date a genetic cause has not been identified and genetic testing is not available for this syndrome. It is possible that SPS may be "weakly inherited" requiring a number of minor genetic predispositions, from both parents, to be inherited together. SPS is certainly much more common in individuals of European or Celtic descent.

First degree relatives of individuals with the Serrated Polyposis Syndrome have been identified to have a five-fold increase in their lifetime risk for developing bowel cancer and for this reason, it is recommended that first degree relatives i.e. father, mother, sister, brother, children be offered five yearly bowel screening by colonoscopy from the age of 40 years or from an age 10 years younger than the earliest age at which SPS was diagnosed in the family.

Smoking increases the number of both adenomatous and hyperplastic or serrated polyps but the number of serrated polyps are particularly increased.

There are no known means by which we can reduce the development of the serrated polyps but we do recommend the standard healthy diet with maintenance of ideal body weight and the routinely advised exercise levels. <u>http://nzgg.org.nz/library_resources/94_bowel_cancer_consumer_resource</u> (page 8)

We would also recommend that smoking be avoided.

We are involved in the study seeking to identify the genetics associated with this condition and if you were diagnosed with the Serrated Polyposis Syndrome under the age of 70, you may be offered involvement in this study.

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