

Gastrointestinal Tumors in NF Type 1

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What is a GIST?

Gastrointestinal stromal tumors (GISTs) are one of the most common tumors in the GI tract.

What causes a GIST and what are the symptoms?

They can occur in the stomach, small intestine and large intestine. These tumors can produce bleeding which can result in obvious blood in patients' bowel movements or can be more subtle and result in anemia or low blood count. Sometimes these tumors can produce abdominal pain, vomiting and even bowel obstruction (requiring surgery).

In someone with NF1, new pain, worsening of existing pain and especially pain that wakes one up from sleep, should never be ignored!

How are GISTs diagnosed?

Unfortunately, GISTs are not necessarily seen in colonoscopy or endoscopy and so they may go undetected in many patients. Specialized CT scans seem to be the test technique for detecting them.

How is a GIST treated?

However, once a GIST is found, it is not clear whether they should be treated. There have been only a few reports of GISTs in NF1. It seems clear that patients with NF1 are at substantially increased risk of GISTs. Estimates vary from 1% to as much as a 33% risk. The risk may also increase with age. Those few reports also indicate substantial differences in GISTs in NF patients compared to non-NF patients. Because of these differences as well as lack of experience with significant numbers of adult NF1 patients, patients with NF1 are often offered a wide range of treatment opinions.

What is the future of GIST research?

We are currently reviewing our experience to try to determine whether there are significant differences in the tumors in NF1 patients and where those differences can tell us something about the best way of monitoring patients and whether specific treatments are needed.