Frequently Asked Questions About Multiple Endocrine Neoplasia & Carcinoid

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What is carcinoid?

The term carcinoid actually originates from the German word Karzinoide. In 1907, a pathologist at the University of Munich named Siegfried Orbendorfer recognized an unusual type of tumor growing in the small intestine of certain patients. This tumor looked different under the microscope than a typical cancer (carcinoma), and at the time Dr. Orbendorfer thought the tumors were benign in their behavior, so he named them “carcinoma-like”, or carcinoid. Over 20 years later, Dr. Orbendorfer realized that the tumors were not always benign and could invade & spread like other malignancies, but by then the term carcinoid was well-established.

Is a carcinoid tumor the same thing as carcinoid syndrome?

No. Not every patient with a carcinoid tumor will experience carcinoid syndrome. Indeed, most patients with carcinoid tumor do not have carcinoid syndrome. Carcinoid syndrome occurs when a carcinoid tumor (usually arising in the small intestine) spreads to the liver. The liver normally metabolizes, or breaks down, hormones made by the carcinoid tumor. However, if the tumor spreads to the liver itself, then the liver cannot properly filter the hormones and the hormones can enter the main circulation of the body. As they circulate, they can affect different organs and cause various symptoms that, as a group, are called carcinoid syndrome: flushing, wheezing, and profound diarrhea. The hormones can also damage the right side of the heart by making the tissues of the heart (particularly the valves) abnormally stiff. As a result, some patients with carcinoid syndrome cannot move fluid through their hearts as they are supposed to, and they become visibly swollen as a consequence of right-sided heart failure.

What’s the difference between a carcinoid tumor and a neuroendocrine tumor?

There can easily be confusion in the terminology that is used to describe these tumors. Carcinoid tumors are neuroendocrine tumors, meaning they are often capable of making hormones. However, increasingly, there is a distinction made between carcinoid tumors and pancreatic neuroendocrine tumors (PNETs). These tumors, which arise in the pancreas, tend to behave in a more aggressive fashion than the carcinoids. In fact, it was the neuroendocrine tumors in the pancreas which Dr. Orbendorfer later realized had more malignant potential than the carcinoid tumors he had first identified in the small intestine. In very small sections under the microscope, carcinoid tumors and pancreatic neuroendocrine tumors look identical, so they have to be distinguished by their tissue of origin.
Do carcinoids act differently if they come from different parts of the body?

As a whole group, the carcinoids have less malignant potential than the pancreatic neuroendocrine tumors. Most carcinoid tumors arise in the small intestine. They can be very tiny and tend to grow extremely slowly. In fact, many people with carcinoid will never know that they have a tumor. By some estimates, 1 of every 250 appendectomies (surgery performed to remove the appendix) reveals the presence of a carcinoid tumor when the appendix is closely examined under the microscope after the operation, but the vast majority of these patients were completely unaware that the tumor was present. If intestinal carcinoids do become symptomatic, it is because they are causing pain, bleeding, or even intestinal blockage, but only rarely because they have spread elsewhere.

In contrast, carcinoid tumors that start growing in the stomach, rectum, or lungs are more likely to invade and spread.

If I have a carcinoid tumor, does that mean I have Multiple Endocrine Neoplasia?

No. Multiple Endocrine Neoplasia type 1 is a distinct genetic disorder. In any given year, roughly 5 out of every 100000 people will be diagnosed with a carcinoid tumor, and only a fraction of those patients were predisposed to develop carcinoid due to a lifelong inherited defect in the menin gene (the genetic error that defines MEN type 1).

If I have Multiple Endocrine Neoplasia, will I get a carcinoid tumor?

Not necessarily. Multiple Endocrine Neoplasia type I is strongly associated with neuroendocrine tumors of the pancreas (which, as above, are now distinguished from carcinoid tumors). MEN 1 patients are at higher risk than the rest of the population for having carcinoid tumors develop in the intestine, but also in sites outside the gut, such as the thymus and lung.