Carcinoid Syndrome

Carcinoid syndrome is a group of symptoms (the main one is flushing) caused by tumors called carcinoids. Carcinoids secrete hormones (*serotonin*) and chemicals that cause the symptoms. This happens after they have spread (*metastasized*). The tumors can occur anywhere. But about 70% of them start out in the appendix or small bowel. Carcinoid syndrome usually affects adults ages 50 to 70, and affects both sexes equally. Carcinoids grow slowly and are often not cancerous (*benign*). But they can spread and are then cancerous (*malignant*). In a few cases, carcinoid cells spread to other body parts. There they produce secondary, hormone-producing (serotonin) tumors.

CAUSES

There are no preventive measures which can be taken to prevent this illness. But smoking, increasing age, and a family history for this type of tumor seem to be connected with increased occurrence.

Symptoms are caused by secretions of the tumor. Some of the triggers which cause the tumor to release the hormone and bring on symptoms are:

- Heavy exercise.
- Tomatoes.
- Pineapple.
- Alcohol.
- Plums.
- Walnuts.
- Bananas.
- Avocados.
- Cheese.

SYMPTOMS

Many people have no symptoms. The main symptom is flushing (like a hot flash) of the head and neck. Others may have symptoms that include:

- Flushed skin on the head and neck.
- Diarrhea with abdominal cramps.
- Irregular heartbeat.
- Low blood pressure.
- Unexplained weight loss.
- Watery eyes.
- Respiratory symptoms similar to asthma.
- Nausea and vomiting.
- Sexual dysfunction in men.
- Disease of the heart valves.

DIAGNOSIS

Your caregiver will do a physical exam and ask questions about your symptoms. A number of medical tests will be done.

- **Nonfunctioning carcinoids. This is a carcinoid which is not producing hormones or symptoms.** These can be detected similarly to other growths by angiography, CT, or MRI (all specialized x-rays), depending on the site. Small-bowel carcinoids may show abnormalities on barium x-ray studies. A final diagnosis is made by taking a part of the growth out and looking at it under a microscope.
- **Functioning carcinoids. These produce hormones and problems.** They are suspected on the basis of the symptoms. The diagnosis is confirmed by a urine check which shows increased amounts of the serotonin breakdown product, 5-hydroxyindoleacetic acid (5-HIAA). A test is performed after the patient has stayed away from serotonin-containing foods (for example, bananas, tomatoes, plums, avocados, pineapples, eggplant, and walnuts) for 3 days to avoid false-positive results. On the 3rd day, a 24-h urine sample is collected for testing. Normal excretion of 5-HIAA is < 10 mg/day (< 52 µmol/day). In patients with carcinoid syndrome it is usually > 50 mg/day (> 260 µmol/ day).
- Tests with calcium gluconate, catecholamines, pentagastrin, or alcohol have been used to cause flushing. These may help in the diagnosis in some patients but must be performed with care.
- Localization of the tumor may require an extensive evaluation, including laparotomy. A liver scan may help show spreading (*metastases*).
- The rare tumors mentioned above must be excluded by the correct examinations.

The tests first help diagnose the cancer and then determine if it has spread (staging).

TREATMENT

Treatment varies and depends on:

- The location and size of the tumor.
- Any spread of the cancer.
- Your health, age, and preferences.

Treatment may include:

- Surgery.
- Anticancer medications (*chemotherapy*).
- Other medications.

Treatments listed below may be used to control symptoms:

- Antidiarrheal medications.
- Medications to prevent serotonin production.
- Cortisone medications to reduce inflammation.
- Medications to prevent flushed skin.
- Anticancer medications (they do not cure these tumors, but may help symptoms).
- Avoid foods that trigger symptoms.
- Include at least 2 servings of protein a day.
- Multivitamins and niacin supplements.
- Do not drink alcohol.
- Resume your normal activities once symptoms improve. Avoid strenuous exercise.

Surgery is curative if the entire tumor can be removed. Sometimes it can spread to local lymph nodes (your glands), which is cured with surgical removal. If surgery is not completely successful, the symptoms can often be helped with the medications listed above. Chemotherapy or medical treatment of the tumor is usually unsuccessful.

COMPLICATIONS

- High blood pressure.
- Bowel obstruction.
- Disease of the heart valves.
- Renal failure.
- Risk for stroke, blood clots, and similar disorders.
- Heart failure.
- Hives (*angioedema*).

If the carcinoid tumor can be removed completely, there can be a cure. Even removing a large part of a tumor that has spread, helps cut down on symptoms. This is because there is less tumor size to produce the hormone causing the symptoms.

Information courtesy of the NIH.
To learn more, contact the National Cancer Institute at 800-4-CANCER.
website: www.cancer.gov.

Document Released: 08/01/2005 Document Re-Released: 03/16/2010
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