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Carcinoid syndrome

What is carcinoid syndrome?

Carcinoid syndrome is a group of symptoms and signs associated with carcinoid tumours. Development of these symptoms and signs usually signifies metastases of a malignant carcinoid tumour (i.e.: cancer has spread to the liver and possibly to other organs). The main characteristics of carcinoid syndrome include [flushing](#), diarrhoea, intermittent abdominal pain, wheezing, heart palpitations, and low blood pressure.

The main cutaneous feature of carcinoid syndrome is flushing, which occurs in 75% of cases. This is caused by transient dilation of blood vessels.

- Flushing occurs in the face, head, neck, chest and epigastric areas
- It may last for 2–5 minutes or several hours (particularly in the later stages of the disease)
- The skin may be diffusely affected or mottled, with or without accompanying oedema (swelling)
- Recurrent flushing may lead to long-lasting skin changes, including telangiectasia (broken blood vessels) and plethora (persistent redness)
- Flushing may be precipitated by stress, or eating hot foods, chocolate, alcohol or cheese.

Dermatitis may also occur and appears identical to that seen in [pellagra](#). The signs are reddened skin with superficial scaling in areas exposed to sunlight.

What is the cause of carcinoid syndrome?

Carcinoid syndrome develops when carcinoid tumours spread to the liver. Vasoactive amine chemicals produced by a carcinoid tumour are not broken down by the liver and so gain access into the bloodstream. There are wide variations in the substances secreted by carcinoid tumours. One hormone that is commonly implicated is serotonin. Serotonin is broken down to 5-hydroxyindole acetic acid (5-HIAA) and excreted in urine.

How is the diagnosis made?

Flushing accompanied by diarrhoea, wheezing and an enlarged liver, in association with a carcinoid tumour points towards the diagnosis of carcinoid syndrome. An elevated urinary level of 5-HIAA confirms this.

Patients with carcinoid syndrome need to undergo thorough medical and physical examinations, and appropriate blood tests and X-rays to follow the growth and activity of the carcinoid tumour(s).

What is the treatment for carcinoid syndrome?

The main goal of treatment is to completely remove the carcinoid tumour. Removal of the entire tumour will result in permanent cure. If the entire tumour cannot be removed then removing large portions of the tumour can relieve the symptoms of carcinoid syndrome. This partial removal known as surgical debulking can effectively relieve the symptoms by decreasing the amount of harmful hormones being produced and entering the bloodstream. There are many different surgical techniques that can be used to treat tumours that have spread to the liver or other organs.

For advanced carcinoid tumours that cannot be removed surgically, several different medical treatments are available.

- Octreotide – anti-hormone injection that in some cases may inhibit and reverse the growth of tumours. In most cases symptoms of carcinoid syndrome are improved.
- Interferon – immune-modulator injection used to suppress or stop tumour growth. Often given in conjunction with octreotide where octreotide given alone has failed.
- Specific drugs that either suppress production of vasoactive amines or block their effects. These include alpha-adrenergic blocking drugs, cyproheptadine and H2-receptor blockers (antihistamines), and serotonin antagonists.
- One of several combinations of chemotherapy may relieve symptoms of carcinoid syndrome. If one combination doesn't work, another combination may be effective. Between 20–30% of patients benefit from chemotherapy.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Cutaneous markers of internal malignancy](#)

Other websites:

- emedicine dermatology, the online textbook
 - [Paraneoplastic Diseases](#)
 - [Malignant Carcinoid Syndrome](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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