

Carcinomatosis

What is carcinomatosis?

Carcinomatosis is described as a condition in which multiple carcinomas develop simultaneously, usually after dissemination from a primary source. It implies more than spread to regional nodes and even more than just metastatic disease. The term is usually taken to mean that there are multiple secondaries in multiple sites.

Strictly, it should be used only for epithelial cancers or carcinomas and not sarcomas or lymphomas but has been extended to include all types of cancer which have spread. The word is now used to describe any widespread dissemination of cancer in the body, and also conditions with specific anatomical spread, as in:

Leptomeningeal carcinomatosis^[1] ^[2]

- Involvement of leptomeninges through seeding via the cerebrospinal fluid, which occurs either by direct spread or via the bloodstream.
- It is an uncommon and usually late complication of cancer.
- Any cancer can cause this but adenocarcinomas are most commonly involved. Breast cancer is one of the most common causes.
- Presentation is very variable, and may be nonspecific (for example, headache or confusion) or with focal or multifocal neurological deficits (for example, cranial nerve palsies).
- Diagnosis is based on careful history and neurological examination, cerebrospinal fluid cytology and neuroimaging (preferably gadolinium-enhanced MRI).

Pulmonary lymphangitic carcinomatosis^[3]

- Diffuse infiltration of the lungs with obstruction of the lymphatic channels.
- May occur with a variety of different cancers, most commonly breast, stomach, pancreas, lung, and prostate.
- Lymphangitic carcinomatosis can also affect the kidneys and may present with acute kidney injury (AKI).^[4]

Peritoneal carcinomatosis^[5]

- Spread of metastases into the peritoneum, usually from gynaecological (usually ovarian) and gastrointestinal cancers.
- Peritoneal carcinomatosis carries a poor prognosis.

Symptoms of carcinomatosis (presentation)

Carcinomatosis may be a progression of known disease. It may be the presentation of recurrence or it may be the primary presenting feature. Presentation will depend upon where is affected.

- In the lungs it may present as shortness of breath and haemoptysis.
- In the liver it often presents as jaundice.
- In the brain there may be headaches, vomiting and focal neurological features.
- In bones there may be pain or pathological fracture.
- Peritoneal involvement may present with varying symptoms including ascites, pain, nausea, cachexia and bowel obstruction.

Differential diagnosis

When these features present, the question is whether this is part of the known disease or something else. For example, whether jaundice is due to metastatic carcinoma in the liver or to gallstones.

When carcinomatosis is the presenting feature it is usual to seek a primary tumour through imaging and histology.

Diagnosing carcinomatosis (investigation)

The purpose of investigations is to confirm the nature of the disease and to assess its severity and extent.

- Blood tests: in cases of unknown primary, FBC may show iron deficiency suggestive of gastrointestinal malignancy, microscopic haematuria may reveal occult genitourinary malignancy and occult blood may point to a colorectal cause. Depending on the presentation, certain tumour markers may be helpful, eg PSA in men with bony metastases. In cases where the primary is known, FBC, U&E, creatinine and LFTs may indicate severity.
- Imaging: newer imaging techniques such as ultrasound, CT and MRI scanning as well as older investigations, such as CXR, provide very good information although an exploratory laparotomy may sometimes be required for peritoneal carcinomatosis. As above, gadolinium-enhanced MRI is the preferred first-choice investigation for suspected leptomeningeal involvement.^[2]
- Biopsy: it may be desirable to obtain tissue for histology. Transbronchial biopsy provides definitive diagnosis for pulmonary lymphangitic carcinomatosis.^[3] Histological analysis helps guide management choice and is one of the factors to consider in deciding on whether aggressive treatment with surgery and/or chemotherapy is appropriate. The evolution of molecular pathology has allowed it to contribute significantly to management decisions.^[6]

Management of carcinomatosis^[5] ^[7]

As this condition may occur from numerous aetiologies, specific management strategy options are also numerous and may be targeted towards the specific cancer. In many cases there is no realistic hope of curative therapy, although chemotherapy and radiotherapy may have a palliative effect.

Surgery may be palliative and 'debulking' of the tumour before chemotherapy may be helpful. Resection of liver metastases secondary to colorectal cancer has had some success in limited disease. There are some subgroups of patients who do relatively well with treatment.

Multimodality treatment (palliation with surgery, radiation, and/or chemotherapy, administered either systemically or directly into the cerebral spinal fluid) may be used in patients with leptomeningeal metastases secondary to breast cancer although prognosis remains poor.

A multimodality approach (of cytoreduction surgery followed by hyperthermic intraperitoneal chemotherapy) is also used in selected people with peritoneal carcinomatosis and may improve prognosis significantly in some cases.

Chemotherapy

- Peritoneal carcinomatosis can in some cases be treated with intraperitoneal and/or intravenous chemotherapy. Treatment can be started postoperatively or chemotherapy drugs can be instilled in the abdominal cavity during or after surgery. In ovarian cancer with peritoneal spread, chemotherapy is more standard than in other primaries, and prognosis is often significantly improved.
- Intrathecal trastuzumab may in some cases be an effective option for the treatment of HER2-positive breast cancer patients with leptomeningeal involvement but is still under investigation.^[8] ^[9] ^[10] Methotrexate (intra-CSF) is a more established chemotherapy option for breast cancer patients with leptomeningeal involvement.

Embolisation^[11] ^[12]

- Transcatheter arterial chemoembolisation (TACE) has resulted in a successful outcome, particularly in patients with neuroendocrine tumours and colorectal metastases. It is increasingly used in treating malignant liver tumours. A microcatheter is inserted into the hepatic blood supply and a combination of chemotherapeutic agents and embolic agents is injected. Radio-embolisation promises to serve an expanding role in the treatments available for treating and managing metastatic disease but further trials, particularly comparisons to chemotherapy regimes, are needed.

Radiotherapy^[13]

- Palliative radiotherapy can often be used to:
 - Reduce or eliminate pain from bone metastases.
 - Palliate brain metastases.
 - Relieve spinal cord compression or compressive symptoms from visceral metastases (eg, airway or gastrointestinal obstruction).
 - Control bleeding - eg, haemoptysis or haematuria.
- Various ablative techniques have been used to destroy liver metastases, including freezing, microwaves, lasers and the use of alternating current within the radiofrequency range.

Surgery

- Although palliative surgery for malignant bowel obstruction from carcinomatosis can benefit patients, it comes at the cost of high mortality and morbidity relative to the patient's remaining survival time.^[14]
- Current evidence on the efficacy of cytoreduction surgery (CRS) followed by hyperthermic intraoperative peritoneal chemotherapy (HIPEC) for peritoneal carcinomatosis shows some improvement in survival for selected patients with colorectal metastases, but evidence is limited or equivocal for other types of cancer.^{[15] [16]}
- There may be a role for surgical treatment of some bone metastases to improve life expectancy and the quality of life.^[17]

For patients who are incurable, a frank and honest discussion must take place. This may require more than one session and the skills for [breaking bad news](#) are required. Other considerations may be [dying at home](#) and [dyspnoea in palliative care](#). [Pain control in terminal care](#) and [nausea and vomiting in palliative care](#) may also warrant attention.

Good palliative care is likely to require a multidisciplinary team approach. Early referral to a palliative care team is essential for specialist help with symptom control and emotional concerns for patients and relatives.^[18]

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Last updated by: Dr Doug McKechnie, MRCGP 26/06/2024	
Peer reviewed by: Dr Philippa Vincent, MRCGP 26/06/2024	Next review date: 25/06/2027

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