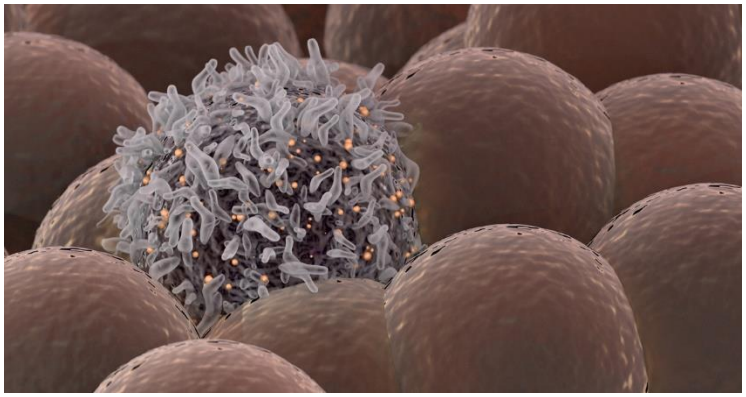
Cancer Association of South Africa (CANSA)

Research • Educate • Support

Fact Sheet on Sarcoma Cancer

Introduction

Cancer happens when cells start to divide uncontrollably and spread to other tissues. This creates masses called tumours.



[Picture Credit: Sarcoma vs Carcinoma}

Most cases of cancer involve either a carcinoma or a sarcoma. There are four other main types of cancer:

- Lymphomas – these affect cells in the lymph nodes
- Myelomas – these affect plasma cells in the bone marrow
- Melanomas – these affect melanocytes which produce pigments
- Leukaemias – these affect bone marrow cells and are responsible for various blood cancers

Sarcomas start in connective tissues, which are the supporting tissues of the body. Connective tissues include the bones, cartilage, tendons and fibrous tissue that support organs.

Sarcomas are much less common than carcinomas. They are usually grouped into 2 main types:

- bone sarcomas (osteosarcoma)
- soft tissue sarcomas

Altogether, these make up less than 1 in every 100 cancers (1%) diagnosed every year.

Differences Between Carcinoma and Sarcoma

The following table provides the main differences between carcinoma and sarcoma:

Researched and Authored by Prof Michael C Herbst
[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer; Medical Ethicist]
Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

	Carcinoma	Sarcoma
Where it Originates	Epithelium	Connective Tissue
Age Group	Older persons – usually over 50	Younger persons – mostly under 50
Vascularity	Less vascular	More vascular
Rate of Growth	Less rapid	More rapid
Means of spread	Early by lymphatics	Early by blood
Prognosis	Less worse	More worse
Microscopic	Cells arranged in groups	Cells arrange individually
Behaviour	Always malignant	Always malignant
Occurrence	More common	Less common
In situ phase	Yes	No
Occurrence in body	Lungs Liver Stomach Colon Rectum	Bone & Cartilage Muscle Fat Nerves Fibrous Tissue

Sarcoma Subtypes
The following is an indication of the most well-known subtypes of sarcoma:

- **ALVEOLAR SOFT-PART SARCOMA**
  This extremely rare sarcoma typically arises in the thigh or buttock of patients in their 20s. Men are much more commonly affected than women. This form of sarcoma is relatively resistant to standard chemotherapy. Despite its early spread, people with this diagnosis can live for 10-20 years or more after diagnosis, in some cases.

- **ANGIOSARCOMAS AND OTHER SARCOMAS OF BLOOD VESSELS**
  This uncommon group of sarcomas appear to arise from the lining of blood vessels (endothelial cells) or their precursors.

- **ATYPICAL FIBROXANTHOMA**
  An unusual and relatively less aggressive form of sarcoma that shows features of both fibroblasts and cells that retain fat (xanthomas). The primary treatment is surgical. Radiation is occasionally used to try and prevent tumour recurrence, and chemotherapy is largely ineffective for this diagnosis. These tumours metastasize (spread) very, very rarely.

- **BONE AND CARTILAGE SARCOMAS OF SOFT TISSUE**
  Some sarcomas that arise in soft tissue mimic those that typically arise in cartilage or bone.

- **CLEAR CELL SARCOMA**
  This unusual hybrid tumour appears to be biologically related to alveolar soft-part sarcoma. It is one of the rare tumours with features of both sarcoma and melanoma, including the ability to travel to

lymph nodes (typical of melanoma) and to lung (more common for sarcomas). Surgery and radiation of the primary tumour site provide the best chance for cure.

- **DERMATOFIBROSARCOMA PROTUBERANS**
  This tumour typically arises in the skin and must be excised by an appropriate expert, as less extensive resections often end in failure.

- **DESMOPLASTIC SMALL ROUND-CELL TUMOUR (DSRCT)**
  It typically arises in young men between 15 and 35. The tumour can travel not only elsewhere in the abdominal or pelvic cavity where it starts, but it can also spread to liver, lung, or the space between the lungs (mediastinum).

- **EPITHELIOID SARCOMA**
  Typically affects the extremities (arms and legs) and tends to travel early to other sites of the body, affecting younger people more commonly than older people. Unlike other sarcomas, epithelioid sarcoma can travel to lymph nodes and cause side effects in lymph nodes and other body components.

- **EWING SARCOMA/PRIMITIVE NEUROECTODERMAL TUMOUR (PNET)**
  Typically occurs in children or young adults, although cases in people up to age 80 or more are occasionally seen. Ewing sarcomas more commonly affect bone in children and soft tissue in adults, and can be seen in any site of the body. They commonly recur in the lungs and bones.

- **(EXTRASKELETAL) MYXOID CHONDROSARCOMA**
  An unusual form of chondrosarcoma that shows a wide variety of features under the microscope and typically arises in people from 20 to 40 years of age. It grows relatively slowly but has a high risk of recurrence elsewhere in the body, such as the lung. It is largely insensitive to standard chemotherapy drugs.

- **EXTRASKELETAL OSTEOSARCOMA**
  This sarcoma (also called extraskeletal osteogenic sarcoma) arises in soft tissue but looks just like its counterpart in bone. Typically arises in older adults, not in children. It does not respond very well to the chemotherapy drugs used in osteogenic sarcoma of bone and is more commonly treated like other soft-tissue sarcomas.

- **EXTRARENAL RHABDOID TUMOUR**
  A very aggressive form of sarcoma that nearly always arises in childhood. It affects the kidneys and other structures in the abdomen and has a high risk of early spread to liver, lung, and other sites.

- **FIBROSARCOMA**
  Arises from fibroblasts or their precursors and forms a group of tumours that are difficult to diagnose correctly, given their relative scarcity. These tumours most frequently affect the extremity and trunk, and can metastasise to the lungs, like other sarcomas.

- **GASTROINTESTINAL STROMAL TUMOUR (GIST)**
  GIST is one of the most common types of sarcoma. It appears to arise from the interstitial cells of Cajal (or its precursors), which are the pacemaker cells of the intestines. The common places that GIST recur are in the abdominal cavity or in the liver.

- **GIANT CELL TUMOURS (GCT) OF TENDON SHEATH**
  Arises most commonly near the knee joint, but they can also affect large and small joints alike. They are initially removable with surgery, but some have a high risk of recurrence.

- **LEIOMYOSARCOMA**
  This is a tumour of smooth muscle (or its precursors), and can arise anywhere in the body. This is one of the most common types of sarcoma. Common initial sites for this tumour are the uterus, small intestine or stomach, or the wall of a blood vessel in the abdomen, extremity, or skin.

- **LIPOSARCOMA**
  A sarcoma that arises from fat cells or their precursors. There are three families of liposarcoma: well-differentiated and/or dedifferentiated (~50%), myxoid and/or round cell (~ 40%), and pleomorphic (10%). Each has its own specific biology and risk of recurrence or spread.

  - **MYXOID AND/OR ROUND-CELL LIPOSARCOMA**
    The second-most-common family of liposarcomas. This type of sarcoma is considered relatively chemotherapy-sensitive.

  - **PLEOMORPHIC LIPOSARCOMA**
    This is the least common form of liposarcoma, and it also tends to affect an extremity. It is often more aggressive than other liposarcomas and can spread to other sites of the body such as lung and soft tissue.

  - **WELL-DIFFERENTIATED AND/OR DEDIFFERENTIATED LIPOSARCOMA**
    This sarcoma typically arises in the abdominal cavity or in an extremity. It appears as a large painless mass. Primary therapy is surgical, although the recurrence risk in the abdomen is very high, at least 70-80% over 10 years. The less aggressive form of this tumour is termed "well-differentiated". The more aggressive version of this sarcoma is called "dedifferentiated", but is often less aggressive than other so-called "high-grade" sarcomas.

- **MALIGNANT PERIPHERAL NERVE SHEATH TUMOURS (MPNST)**
  Sarcomas that arise from the insulating cells that surround nerve endings.

- **MESENCHYMAL CHONDROSARCOMA**
  Another unusual version of chondrosarcoma more common in the soft tissues rather than in the cartilage. Primary therapy is typically surgery and radiation, and some doctors advocate the use of chemotherapy in primary treatment of this tumour, given its distant kinship with Ewing sarcoma. When such tumours are treated successfully with chemotherapy, they often leave behind a less aggressive version of the tumour, which should be surgically removed to obtain the best overall outcome for patients with this very rare diagnosis.

- **RHABDOMYOSARCOMA**
  This rare sarcoma typically affects children. At most, 20% of rhabdomyosarcomas occur in adults. Rhabdomyosarcomas are themselves a separate family of sarcomas, with several recognized subtypes, including Embryonal, Botryoid, Alveolar, and Pleomorphic. Treatment for these sarcomas nearly always involves surgery, radiation, and chemotherapy. Cure rates are better for children than for adults, for unclear reasons. This is one form of sarcoma that can travel to lymph nodes, though it can also travel to lungs and other sites.

- **SOLITARY FIBROUS TUMOUR**
  This sarcoma is an uncommon tumour that is found in the chest cavity, the orbits (which contain the eye), the covering of the brain (dura mater) or the pelvis. There are less aggressive and more aggressive versions of this tumour, which can easily grow to a size of 15-50cm or more in size.

- **SYNOVIAL SARCOMA**
  Synovial Sarcoma is usually seen in patients between the ages of 15 and 35, and often affects the leg, foot, and hand, although other unusual sites such as the chest cavity are seen. It is often a chemotherapy-sensitive form of sarcoma. Therapy for a primary leg tumour often involves surgery, radiation, and sometimes chemotherapy. The lungs are the most common site of recurrence for synovial sarcomas. This is the subtype of sarcoma about which there is the most interest in using immunotherapy for treatment.

- **UNDIFFERENTIATED PLEOMORPHIC SARCOMA (UPS)**
  UPS used to be called MFH (malignant fibrous histiocytoma) by pathologists. It tends to affect people over 50 years of age in the leg, trunk, or arm. The most common place for MFH to recur is in the lungs.

Sarcoma Treatment Modalities

Because of their rarity and the frequent need for multimodality treatment, evaluation and management of soft tissue sarcomas (STS) should ideally be carried out in a centre with expertise in the treatment of sarcomas, including surgical oncology, orthopaedic surgery, plastic surgery, adult or paediatric medical oncology, and radiation oncology. The multidisciplinary team approach to care of STS optimizes treatment planning, minimizes duplication of diagnostic studies, and reduces the time to implementation of the definitive therapeutic protocol.

Medical Disclaimer

This Fact Sheet is intended to provide general information only and, as such, should not be considered as a substitute for advice, medically or otherwise, covering any specific situation. Users should seek appropriate advice before taking or refraining from taking any action in reliance on any information contained in this Fact Sheet. So far as permissible by law, the Cancer Association of South Africa (CANSA) does not accept any liability to any person (or his/her dependants/estate/heirs) relating to the use of any information contained in this Fact Sheet.

Whilst the Cancer Association of South Africa (CANSA) has taken every precaution in compiling this Fact Sheet, neither it, nor any contributor(s) to this Fact Sheet can be held responsible for any action (or the lack thereof) taken by any person or organisation wherever they shall be based, as a result, direct or otherwise, of information contained in, or accessed through, this Fact Sheet.

Sources and References Consulted and/or Utilised

CureSarcoma
https://www.curesarcoma.org/patient-resources/sarcoma-subtypes/

Sarcoma
https://www.cancercenter.com/community/blog/2017/11/whats-the-difference-carcinoma-and-sarcoma
https://health.clevelandclinic.org/whats-the-difference-between-a-sarcoma-and-carcinoma/
https://www.healthline.com/health/carcinoma-vs-sarcoma
https://www.cancerresearchuk.org/what-is-cancer/how-cancer-starts/types-of-cancer
https://www.uptodate.com/contents/overview-of-multimodality-treatment-for-primary-soft-tissue-sarcoma-of-the-extremities-and-chest-wall

Sarcoma Alliance
https://sarcomaalliance.org/resources/sarcoma-subtypes/

Sarcoma vs Carcinoma
https://www.cancercenter.com/community/blog/2017/11/whats-the-difference-carcinoma-and-sarcoma
https://pediaa.com/difference-between-cancer-and-sarcoma/
https://www.slideshare.net/Himesharo/spread-of-tumours
https://quizlet.com/206153065/hemeonc-week-3-flash-cards/
https://doctorlib.info/anatomy/staging-atlas-oncoanatomy/50.html
https://health.clevelandclinic.org/whats-the-difference-between-a-sarcoma-and-carcinoma/