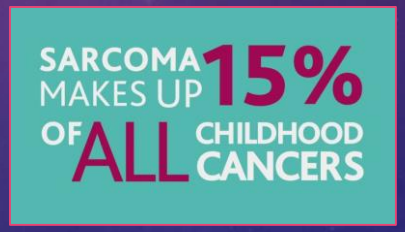

# 2020 ADULT AND PEDIATRIC SARCOMA:

## CONNECTIVE TISSUE, SOFT TISSUE, BONE, SOLID ORGANS, DIAGNOSIS, CLASSIFICATION, STAGING AND TREATMENT

2020-2021 FCDS ANNUAL EDUCATIONAL CONFERENCE SERIES

STEVEN PEACE, CTR

OCTOBER 29, 2020




# CDC & FLORIDA DOH ATTRIBUTION

"Funding for this conference was made possible (in part) by the Centers for Disease Control and Prevention. The views expressed in written conference materials or publications and by speakers and moderators do not necessarily reflect the official policies of the Department of Health and Human Services, nor does the mention of trade names, commercial practices, or organizations imply endorsement by the US Government."

FCDS would also like to acknowledge the Florida Department of Health for its support of the Florida Cancer Data System, including the development, printing and distribution of materials for the 2020 FCDS Annual Conference and the 2020-2021 FCDS Webcast Series under state contract CODJU. The findings and conclusions in this series are those of the author(s) and do not necessarily represent the official position of the Florida Department of Health.

# FLCCSC LMS – CEU QUIZ –FCDS IDEA

- Attendees must take and pass a 3-5 question CEU Quiz to get CEUs
- CEU Awards are Restricted to Attendees with a FLccSC LMS Account
- The CEU Quiz will be posted to FLccSC 1-2 hours after the webcast ends

- Only registered FLccSC Users will be given access to the CEU Quiz
- Florida attendees must have a Florida FLccSC Account to take the Quiz
- South Carolina attendees must have a South Carolina FLccSC Account
- New FLccSC States will follow similar instructions for the CEU Quiz

- Attendees can attend any of the live webcasts without receiving CEUs
- Recorded Sessions are also available for non-FLccSC Users – No CEUs

# 2020-2021 FCDS WEBINAR SERIES

Dear Program Host,

Thank you for your request for "Program Recognition" to support continuing education hours for the CTR Credential. Review of your applications by the NCRA Program Recognition Committee has determined the following.

| Program Title | Program # | CE Hours Awarded | Category A Hours |
|---|---|---|---|
| *Sarcoma - Pediatric and Adult Bone and Soft Tissue, GYN, Ewing Sarcoma/PNET, Other Sites* | 2020-140 | 2 | 2 |
| *Skin Cancers - Melanoma, Merkel cell, GYN sites, T-cell and B-cell Lymphoma, Kaposi Sarcoma, Sebaceous Gland Carcinoma, Dermatofibrosarcoma Protuberans* | 2020-141 | 2 | 2 |
| *Oral Cavity* | 2020-142 | 2 | 2 |
| *Pediatric Cancers* | 2020-143 | 2 | 2 |
| *Upper GI Tract - Esophagus, Stomach, Small Intestine, Pancreas, Liver and Biliary Tract* | 2020-144 | 2 | 2 |

Please ensure this information is listed on the formal program document you provide your

# PRESENTATION OUTLINE


- What is Connective Tissue?
- Is Bone and Cartilage Connective Tissue?
- Are muscles and tendons Connective Tissue?
- Is there Connective Tissue in the Nervous System?
- Introduction to Sarcoma – Incidence/Mortality – Causes/Risk Factors – Signs/Symptoms – WHO Classification
- Instructions for Coding Primary Site for Sarcoma Cases
- Anatomy/Physiology/Diagnosis/Classification/Molecular Genetics/Staging/Treatment
  - Soft Tissue and Connective Tissue – broad category with sarcomas of fat, blood vessels, veins and arteries, connective tissue, fascia, fibrous tissue, muscle, subcutaneous tissue, tendons, retroperitoneum, etc). C49.0-C49.9
  - Long and Short Bones, Cartilage - C40.0-C41.9
  - Muscle (including skeletal muscle and fascia) - C49.0-C49.9 – descriptions often mixed in soft tissue discussions
  - GYN Sarcoma – C54.1 or C54.2
  - GIST and Other Stromal Tumors – stomach or other stroma such as adnexa or endometrial stroma
- Treatment Options – New Chemotherapy Agents and Limb Salvage Procedures
- Questions

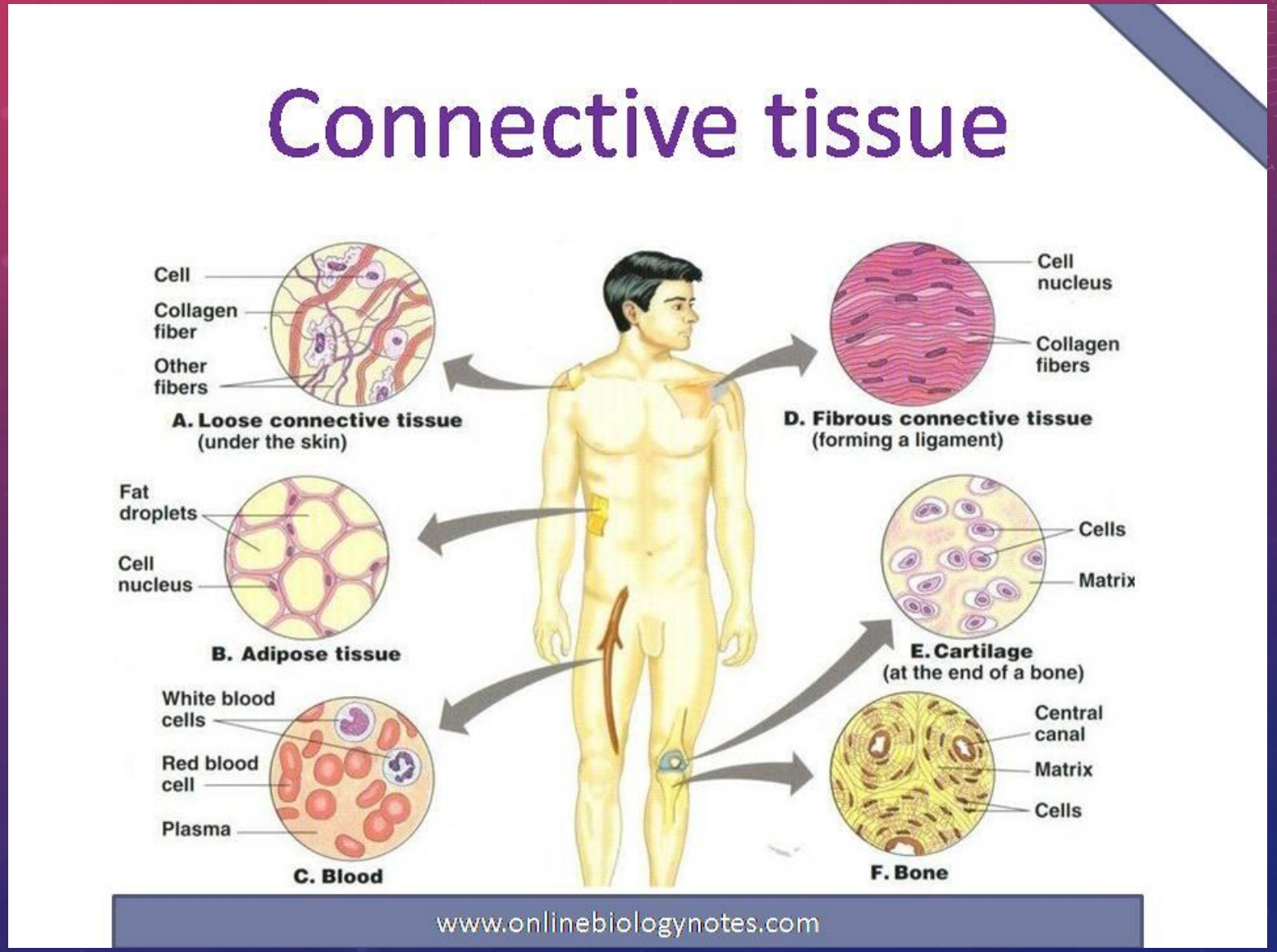
# WHAT IS CONNECTIVE TISSUE?

- **NCI Definition of Connective Tissue** is rather broad and includes: Tissue that supports, protects, and gives structure to other tissues and organs in the body. Connective tissue also stores fat, helps move nutrients and other substances between tissues and organs, and helps repair damaged tissue. Connective tissue is made up of cells, fibers, and a gel-like substance. Types of connective tissue include bone, cartilage, tendons, ligaments, fat, collagen, fibrous tissue, adipose tissue, blood, lymphatic tissue, and elastic tissues.

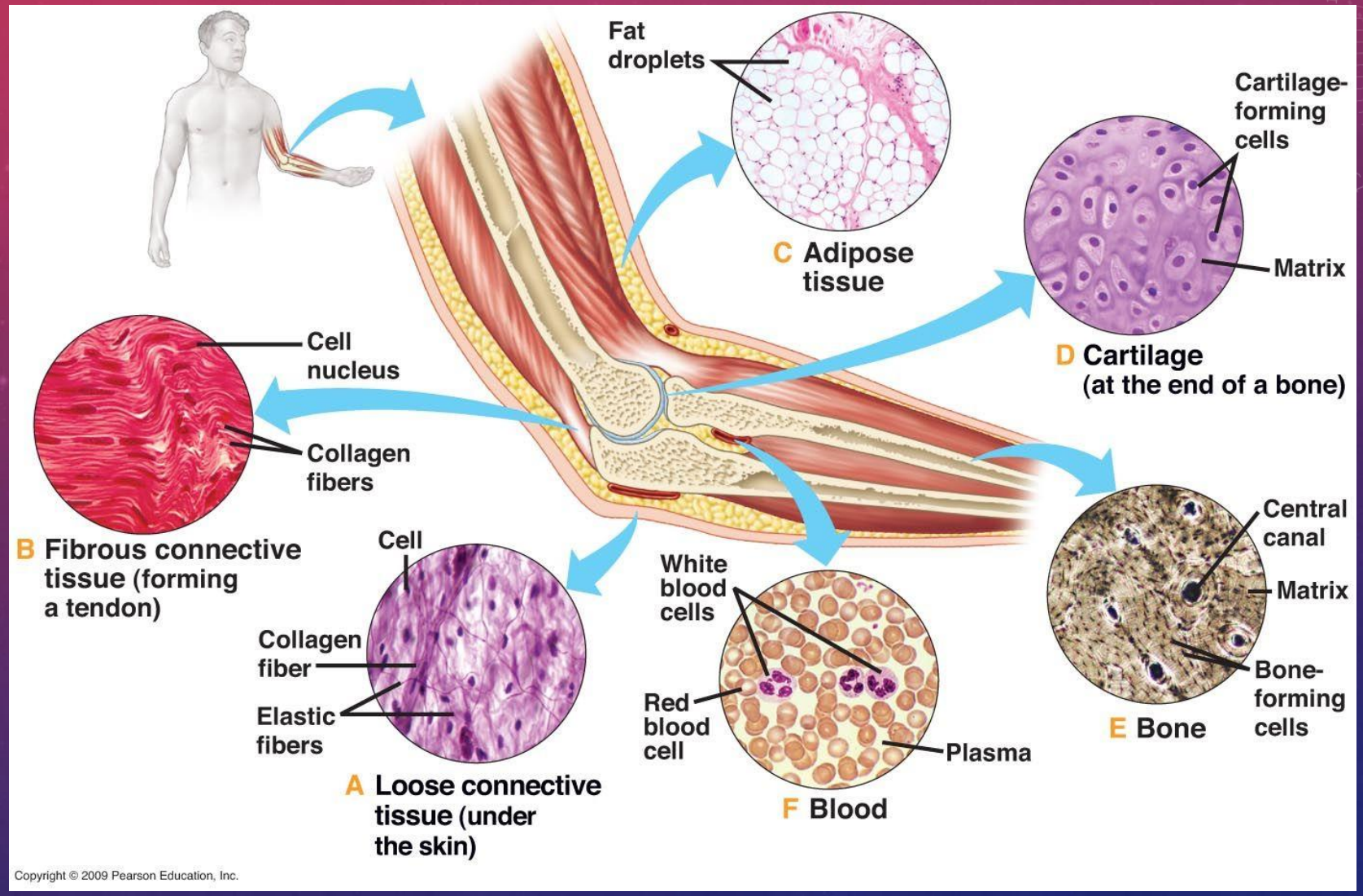
- **The Function of Connective Tissue** is equally broad: Connective tissue is the most abundant and widely distributed of the primary tissues. ... Major functions of connective tissue include: **1) binding and supporting, 2) protecting, 3) insulating, 4) storing reserve fuel, and 5) transporting substances within the body.** Connective tissues can have various levels of vascularity.

- And Another Definition of Connective Tissue: A material made up of fibers forming a framework and support structure for body tissues and organs. Connective tissue surrounds many organs. Cartilage and bone are specialized forms of connective tissue. All connective tissue is derived from mesoderm, the middle germ cell layer in the embryo.

# WHAT IS CONNECTIVE TISSUE?



# WHAT IS CONNECTIVE TISSUE?




https://courses.lumenlearning.com/ap1x94x1/chapter/examining-connective-tissue-under-the-microscope/

# WHAT IS CONNECTIVE TISSUE?

| | Types of connective tissue proper | | | | | |
|---|---|---|---|---|---|---|
| | Loose connective tissue proper | | | Dense connective tissue proper | | |
| Constituents | Areolar connective tissue | Reticular connective tissue | Adipose | Regular dense connective tissue | Irregular dense connective tissue | Elastic dense connective tissue |
| Cells | Fibroblasts, w/some macrophages, other white blood cells | Fibroblasts, w/many white blood cells | Adipocytes, w/some macrophages, other white blood cells. | Fibroblasts, w/ some macrophages, other white blood cells | Fibroblasts, w/ some macrophages, other white blood cells | Fibroblasts, w/ some macrophages, other white blood cells |
| Protein fibers | Collagen, elastic | Reticular | Collagen, some reticular | Thick collagen fibers, in parallel | Thick collagen fibers, no consistent arrangement | Elastic mainly, some collagen |
| Ground substance | Jelly-like, abundant | Jelly-like abundant | Smaller amounts due to abundant cells | Smaller amounts due to abundant fibers | Smaller amounts due to abundant fibers | Smaller amounts due to abundant fibers |
| Locations | Under epithelia of skin, mucous membranes, capillaries, organs | Spleen, lymph nodes, bone marrow | Under skin, surrounding organs, between muscle fibers, in pericardium. | Tendons, ligaments, aponeuroses | Capsules around joints and organs, dermis of skin | Walls of large arteries, walls of bronchial tubes, vertebral ligaments |



https://courses.lumenlearning.com/ap1x94x1/chapter/examining-connective-tissue-under-the-microscope/

# IS BONE AND CARTILAGE CONNECTIVE TISSUE?

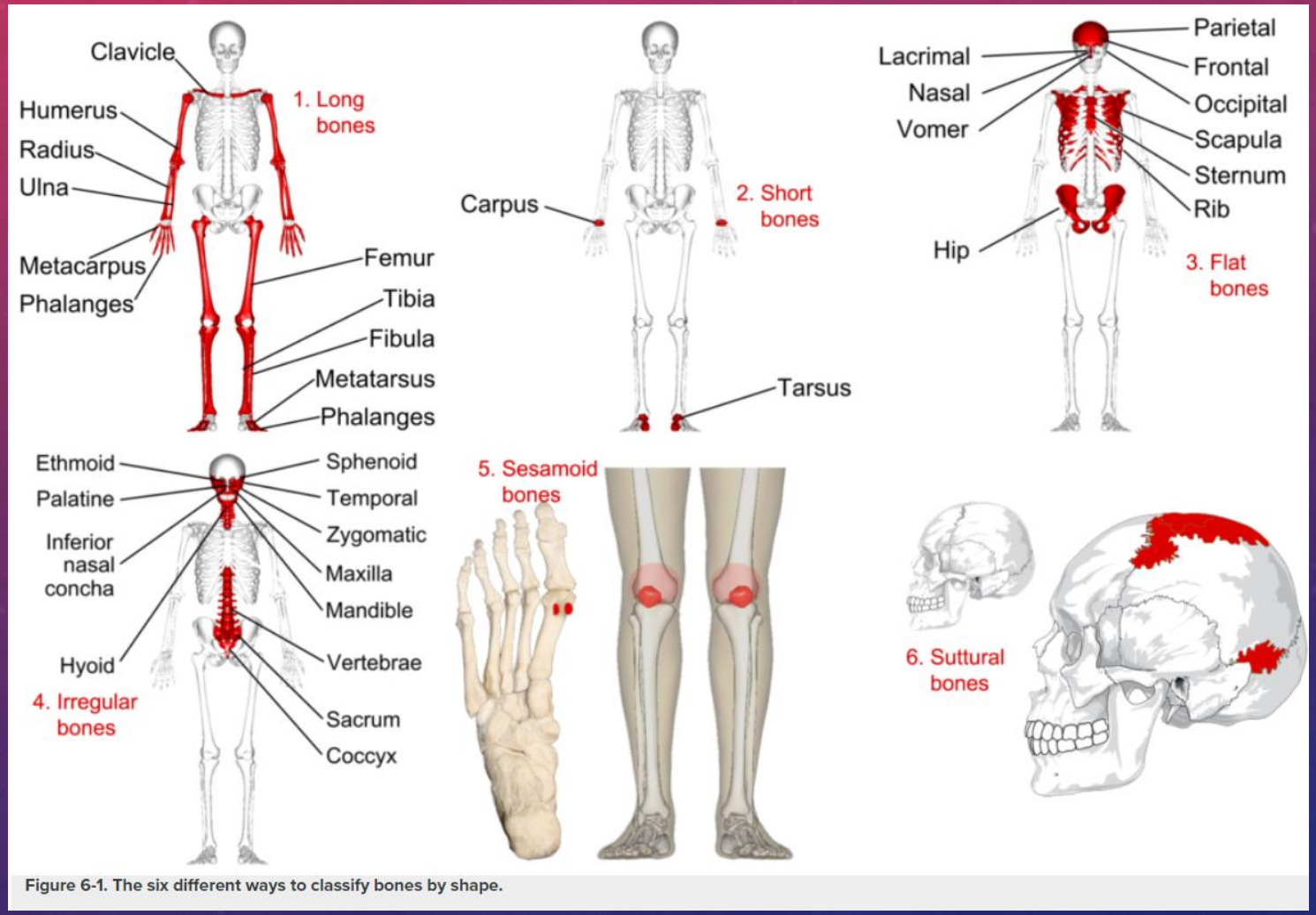
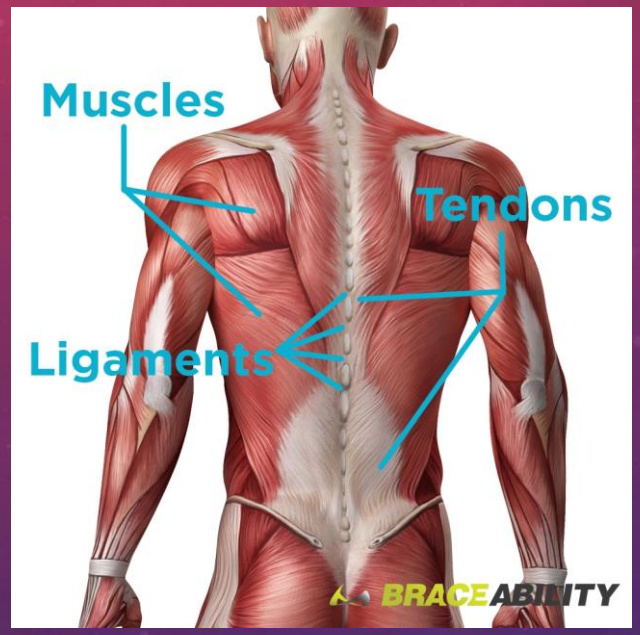


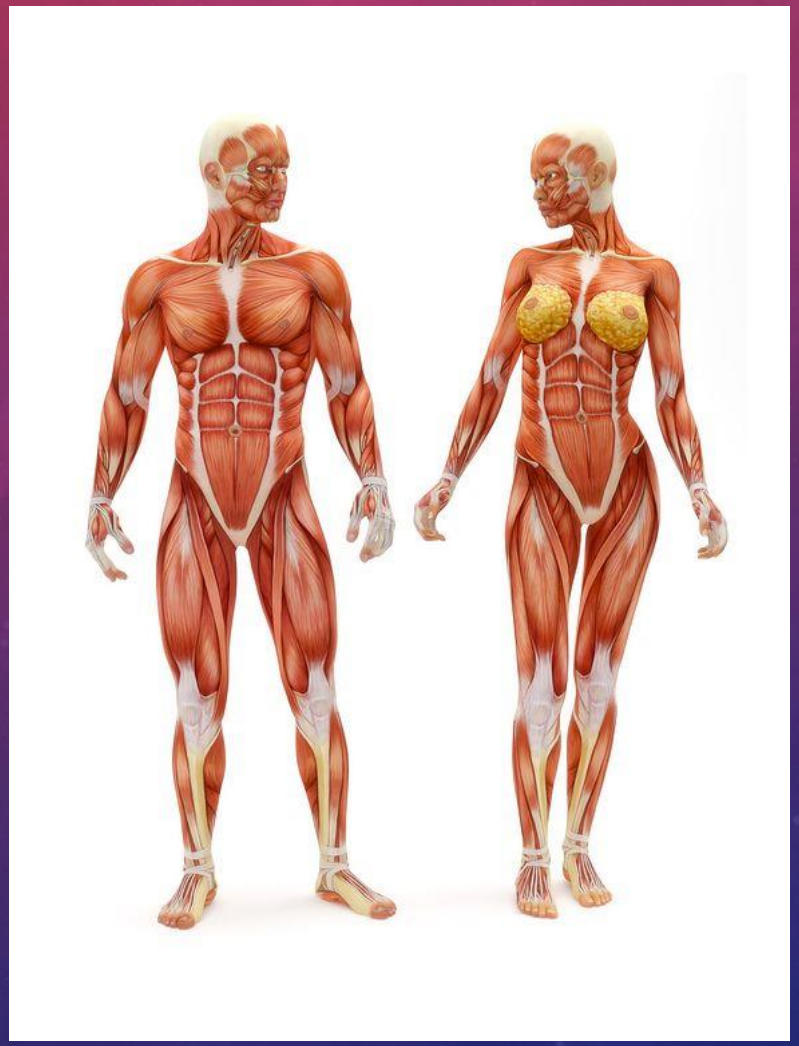

Figure 6-1. The six different ways to classify bones by shape.

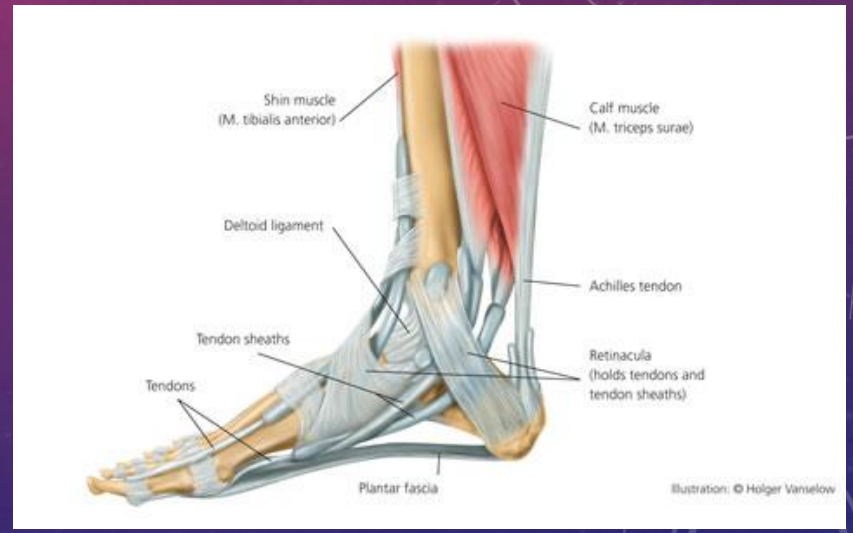


https://courses.lumenlearning.com/ap1x94x1/chapter/examining-connective-tissue-under-the-microscope/

# ARE MUSCLES AND TENDONS CONNECTIVE TISSUE?



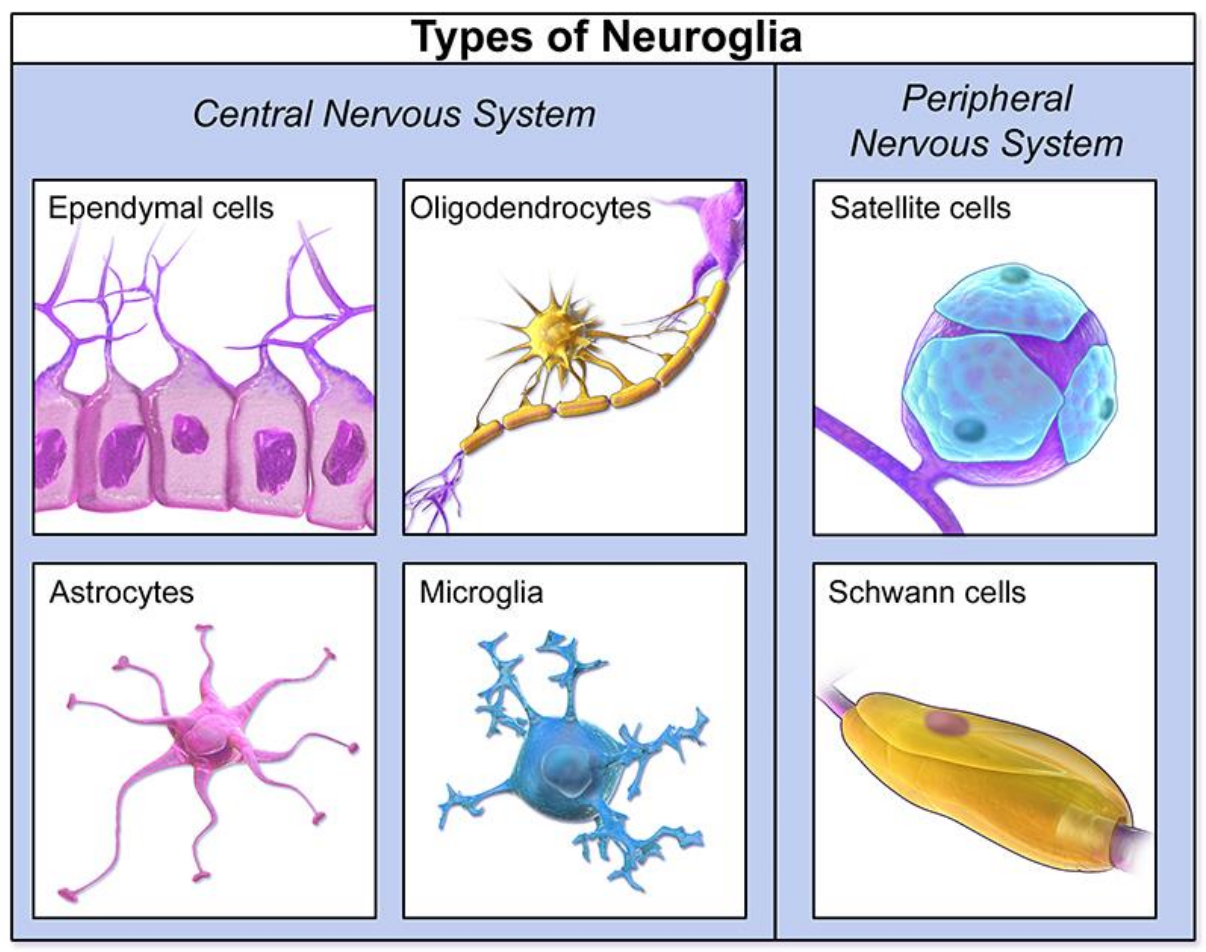








https://courses.lumenlearning.com/ap1x94x1/chapter/examining-connective-tissue-under-the-microscope/

# IS NERVOUS TISSUE CONNECTIVE TISSUE?






https://courses.lumenlearning.com/ap1x94x1/chapter/examining-connective-tissue-under-the-microscope/

# SUMMARY - WHAT IS CONNECTIVE TISSUE

- Bone and Cartilage – long bones and short bones
- Synovia – Joints and Fluids that Lubricate Joints
- Muscle – Cardiac and Smooth Muscle
- Muscle – Skeletal Muscle
- Fatty Tissue
- Fibrous Tissue
- Fascia and Mesentery
- Soft Tissue, NOS
- Blood vessel – Veins and Arteries
- Tendons and Ligaments and Bursa
- Subcutaneous Tissue – 'surface' sarcomas
- Connective tissue – Fascia and Adipose Tissue
- Neuron Protective Tissues – Astrocytes, Oligodendrocytes, Schwann Cells, Migroglia, Ependymal Cells
- BUT – but not neurons themselves – only the cells that protect the neurons
- Retroperitoneum/Peritoneum NOS

Most of tissues are found everywhere in the body.

If the sarcoma involves a solid organ – code primary site to that solid organ

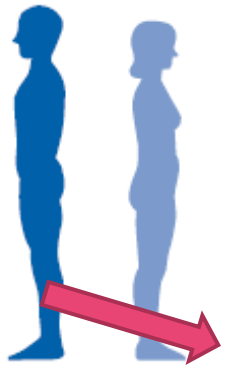
Otherwise, code the primary site to the bone or connective tissue site that describes the anatomic location of the connective tissue, regardless of type of connective tissue

# 2020 INCIDENCE & MORTALITY ESTIMATES

**Figure 3. Leading Sites of New Cancer Cases and Deaths – 2020 Estimates**

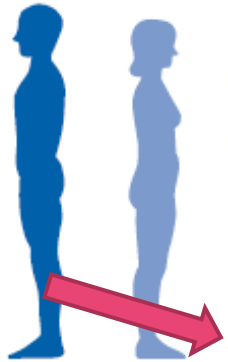
**Estimated New Cases**

| Male | | | Female | | |
|---|---|---|---|---|---|
| Prostate | 191,930 | 21% | Breast | 276,480 | 30% |
| Lung & bronchus | 116,300 | 13% | Lung & bronchus | 112,520 | 12% |
| Colon & rectum | 78,300 | 9% | Colon & rectum | 69,650 | 8% |
| Urinary bladder | 62,100 | 7% | Uterine corpus | 65,620 | 7% |
| Melanoma of the skin | 60,190 | 7% | Thyroid | 40,170 | 4% |
| Kidney & renal pelvis | 45,520 | 5% | Melanoma of the skin | 40,160 | 4% |
| Non-Hodgkin lymphoma | 42,380 | 5% | Non-Hodgkin lymphoma | 34,860 | 4% |
| Oral cavity & pharynx | 38,380 | 4% | Kidney & renal pelvis | 28,230 | 3% |
| Leukemia | 35,470 | 4% | Pancreas | 27,200 | 3% |
| Pancreas | 30,400 | 3% | Leukemia | 25,060 | 3% |
| All sites | 893,660 | | All sites | 912,930 | |



**Estimated Deaths**

| Male | | | Female | | |
|---|---|---|---|---|---|
| Lung & bronchus | 72,500 | 23% | Lung & bronchus | 63,220 | 22% |
| Prostate | 33,330 | 10% | Breast | 42,170 | 15% |
| Colon & rectum | 28,630 | 9% | Colon & rectum | 24,570 | 9% |
| Pancreas | 24,640 | 8% | Pancreas | 22,410 | 8% |
| Liver & intrahepatic bile duct | 20,020 | 6% | Ovary | 13,940 | 5% |
| Leukemia | 13,420 | 4% | Uterine corpus | 12,590 | 4% |
| Esophagus | 13,100 | 4% | Liver & intrahepatic bile duct | 10,140 | 4% |
| Urinary bladder | 13,050 | 4% | Leukemia | 9,680 | 3% |
| Non-Hodgkin lymphoma | 11,460 | 4% | Non-Hodgkin lymphoma | 8,480 | 3% |
| Brain & other nervous system | 10,190 | 3% | Brain & other nervous system | 7,830 | 3% |
| All sites | 321,160 | | All sites | 285,360 | |



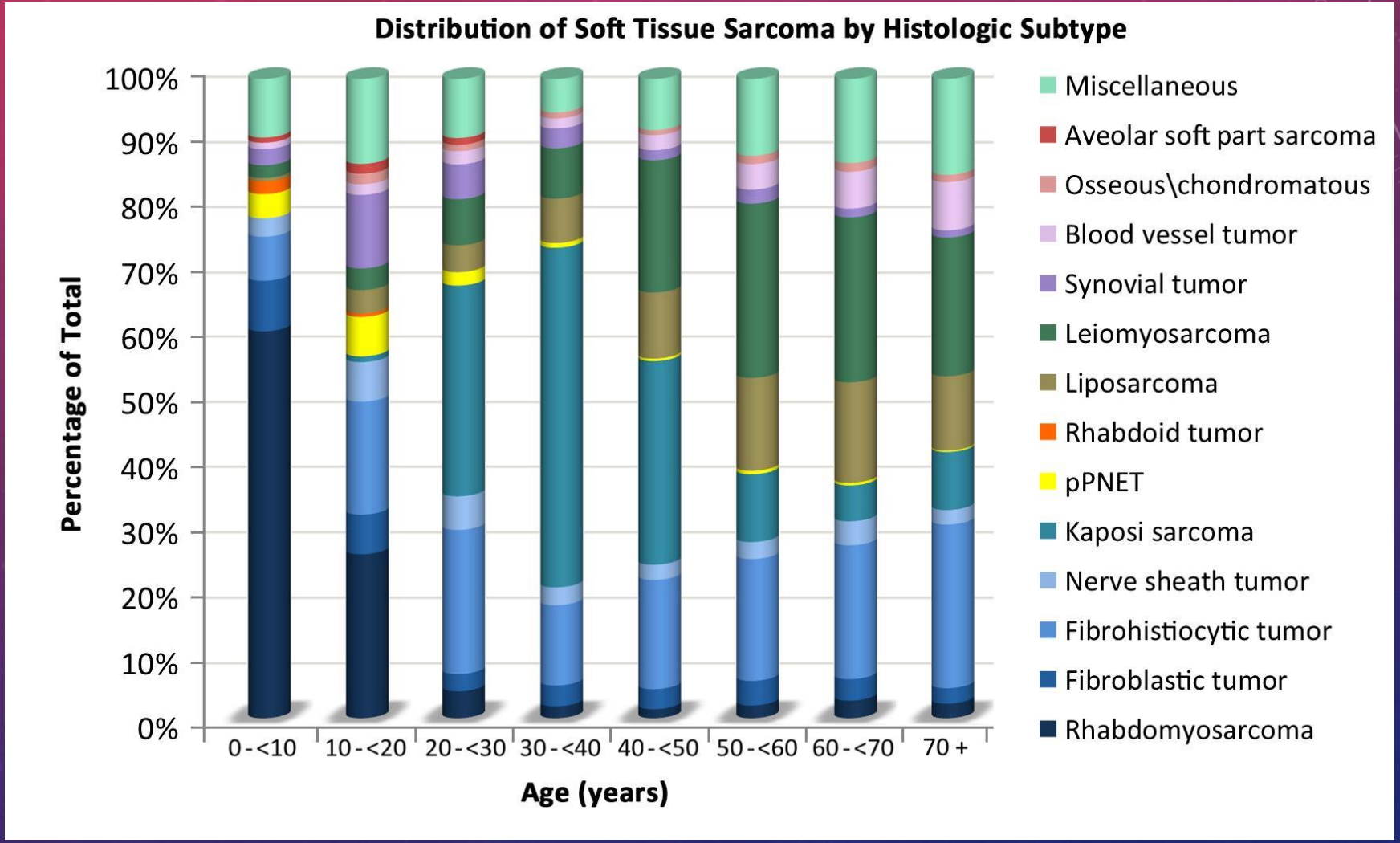
Estimates are rounded to the nearest 10, and cases exclude basal cell and squamous cell skin cancers and in situ carcinoma except urinary bladder. Estimates do not include Puerto Rico or other US territories. Ranking is based on modeled projections and may differ from the most recent observed data.





2020 Cancer Facts and Figures – American Cancer Society

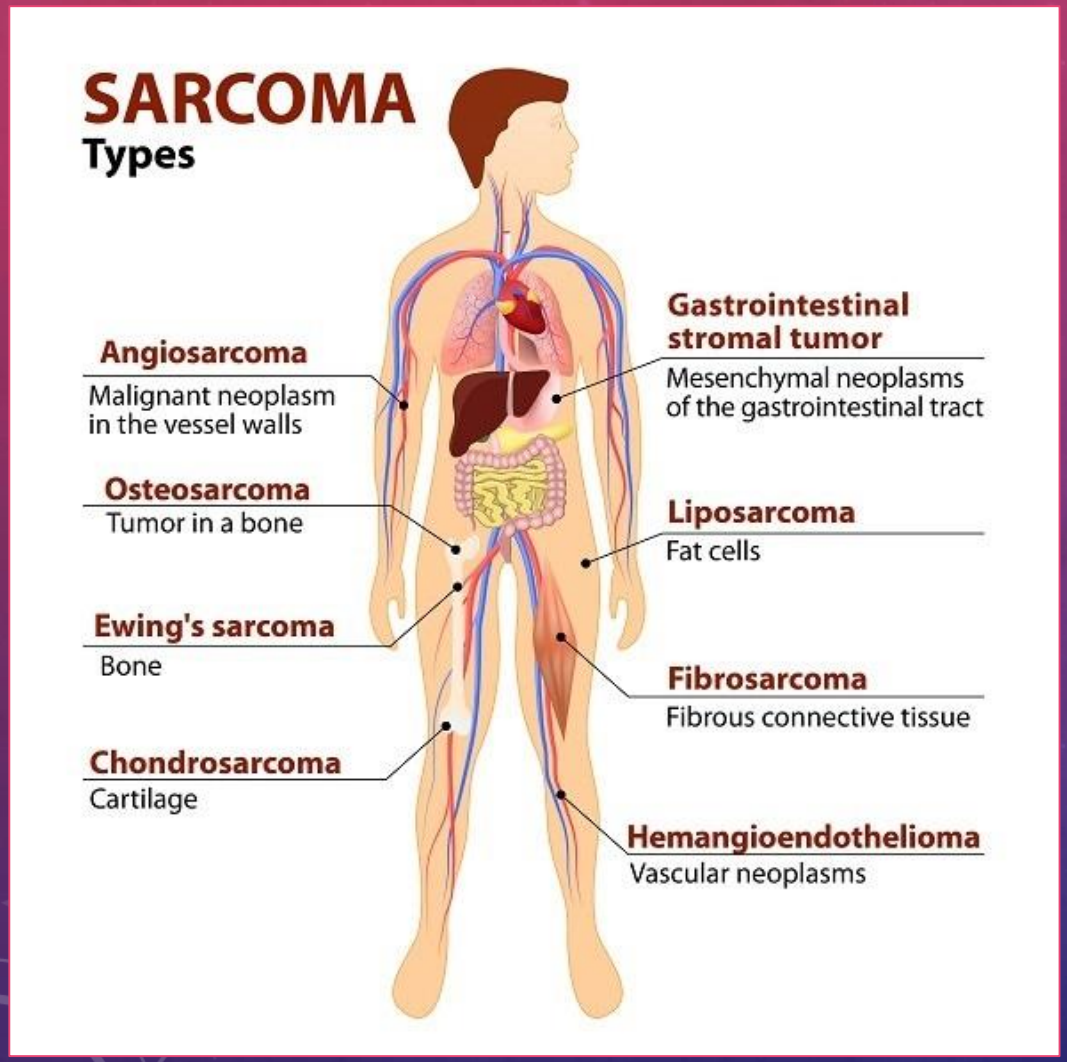
# 2020 INCIDENCE & MORTALITY ESTIMATES

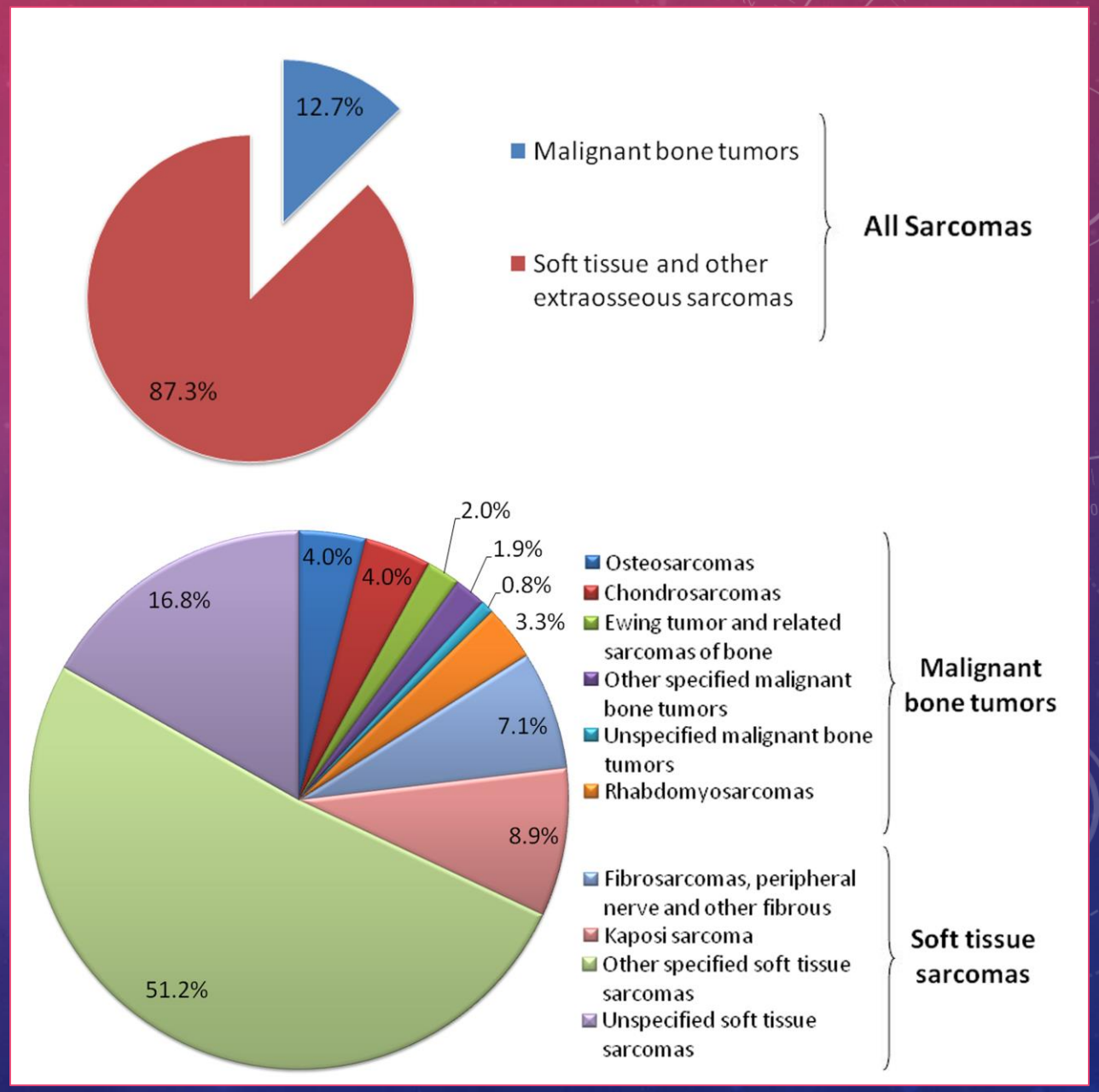





NCI PDQ Childhood Soft Tissue Sarcoma Treatment

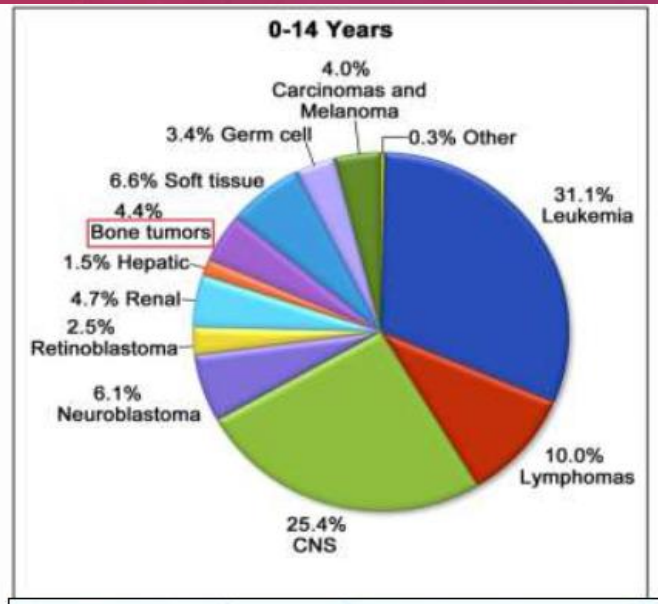
# INTRODUCTION

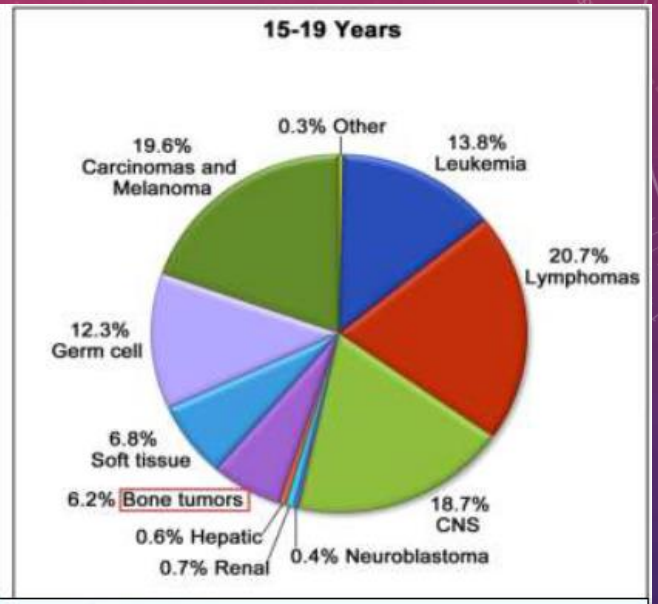


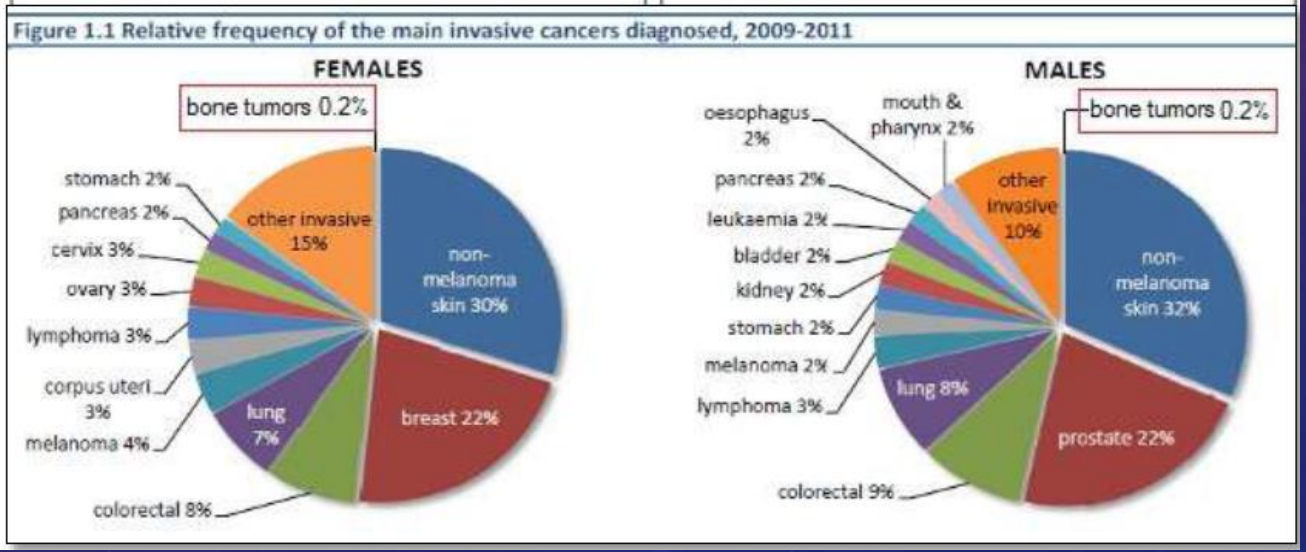



# INTRODUCTION

Bone tumor frequency according to age



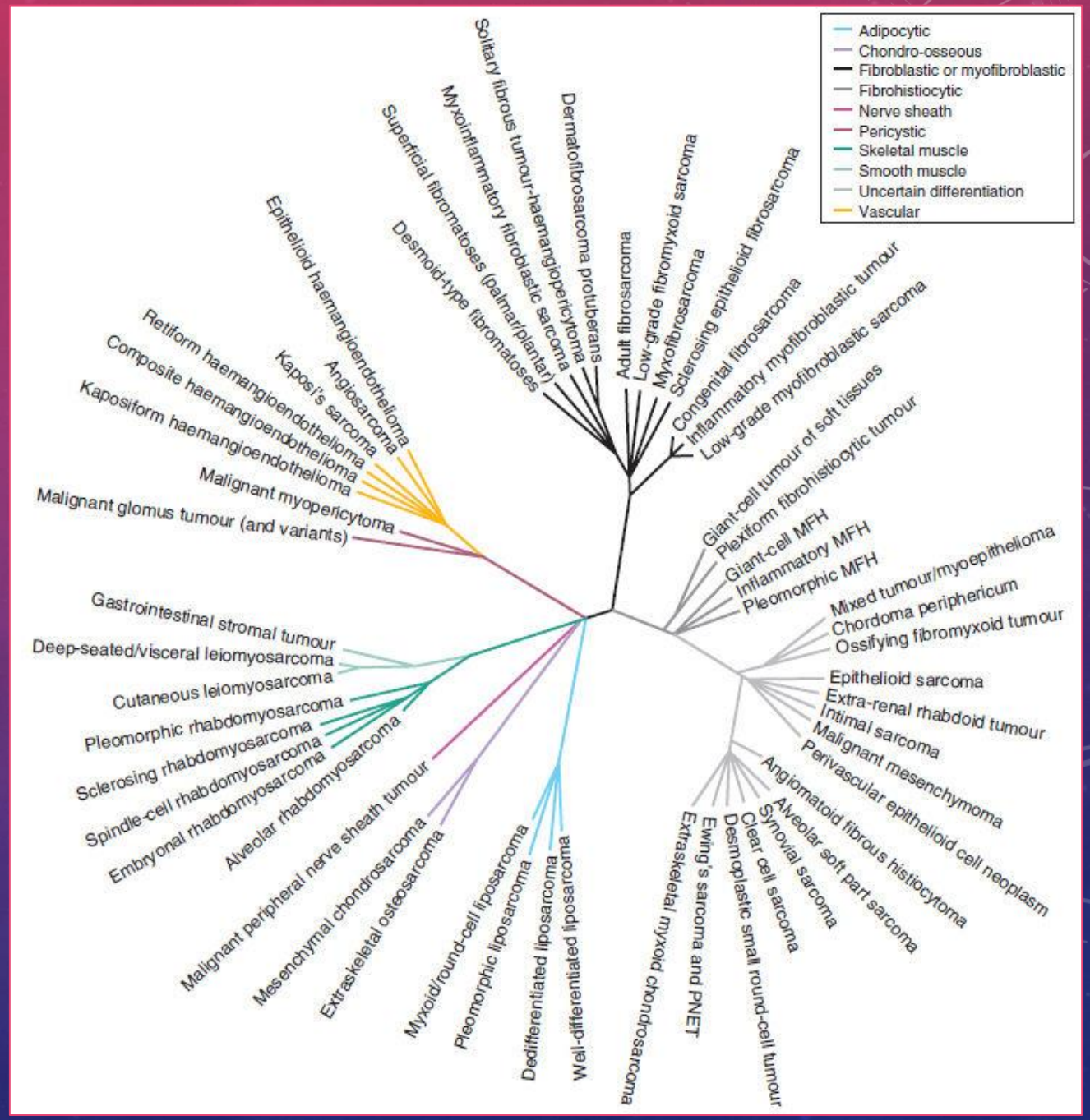



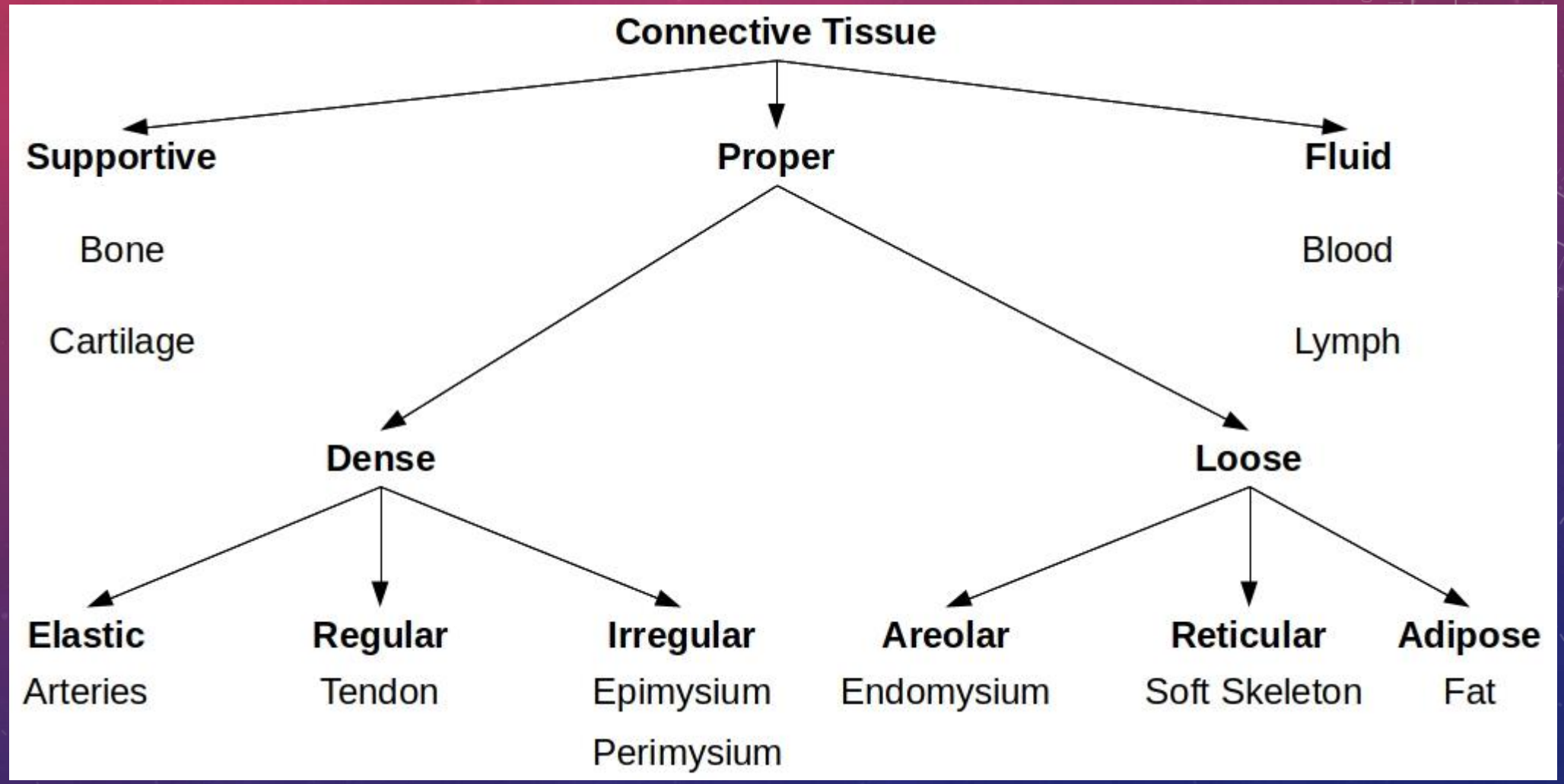

Bone Tumor Staging Systems – Dr Abdullah K Ghafour and Dr Ali Abdulnabi Alwan

# INTRODUCTION

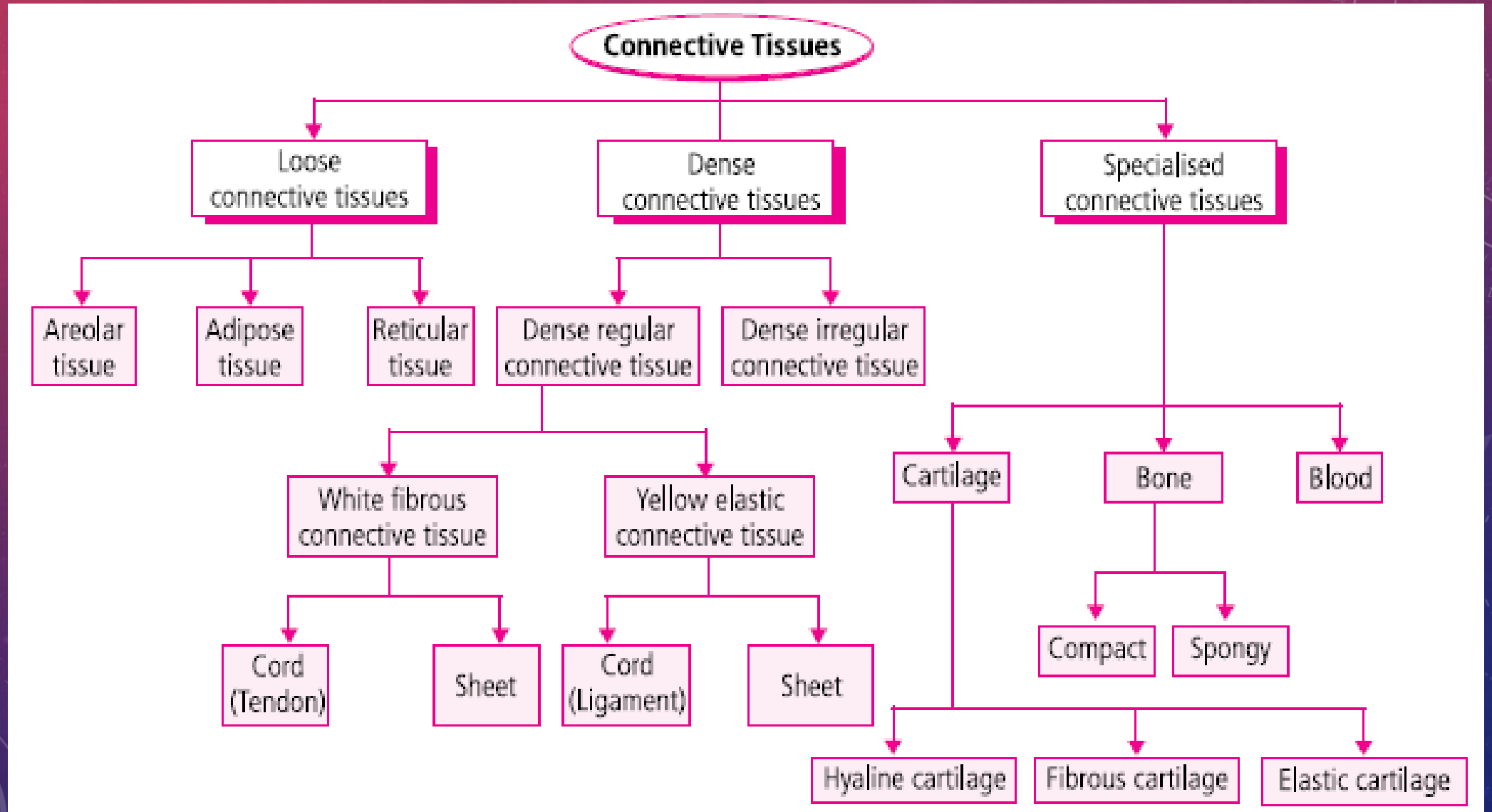
- Angiosarcoma
- Kaposi Sarcoma
- Fibrous Tumors
- Soft Tissue Tumors
- Fatty Tumors (Lipomatous)
- Chondroid Tumors (Bone and Cartilage)
- Peripheral Nerve Sheath Tumors
- Rhabdomyosarcoma
- Schwannoma
- Spindle Cell Tumors
- Myofibroblastic Tumors
- Fibromyxoid Tumors
- Hemangioma
- Lymphangioma
- Epithelioid Sarcoma
- Extraskeletal Chroncdrosarcoma
- Mixed Tumors



# INTRODUCTION



# INTRODUCTION



# CAUSES AND RISK FACTORS

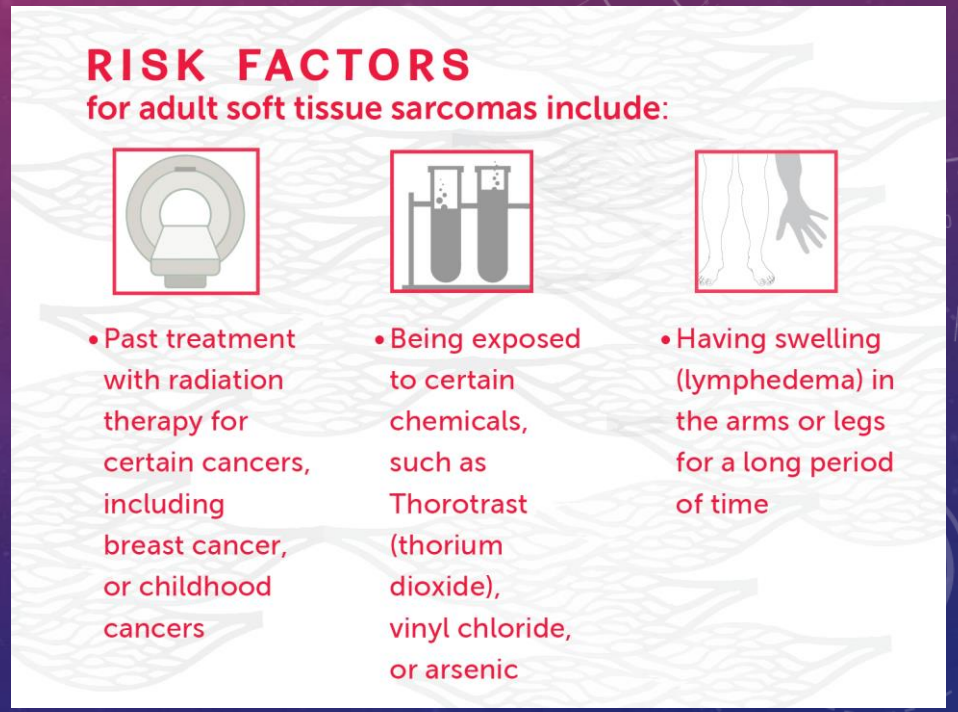
- **The etiology of most benign and malignant tumours of soft tissue is unknown**. In rare cases (< 10%), genetic and environmental factors, irradiation, viral infections, and immunodeficiency have been found to be associated with the development of usually malignant soft tissue tumours.

- There are also isolated reports of sarcomas arising in scar tissue, at fracture sites, and close to surgical implants.

- Some angiosarcomas arise in chronic lymphoedema. However, the large majority of soft tissue sarcomas seem to arise de novo, without an apparent causative factor.

- Multistage tumorigenesis sequences with gradual accumulation of genetic alterations and an increasing histological degree of malignancy have not yet been clearly identified in most tumours of soft tissue.

- Four main types of etiological agents have been implicated in the literature: **chemical carcinogens, radiation, viral infection and immunodeficiency, and genetic susceptibility**.

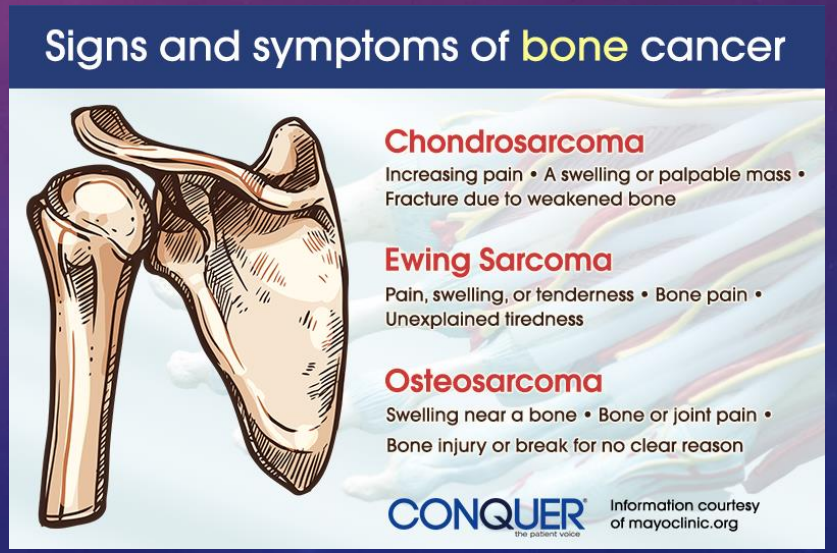


# SIGNS AND SYMPTOMS

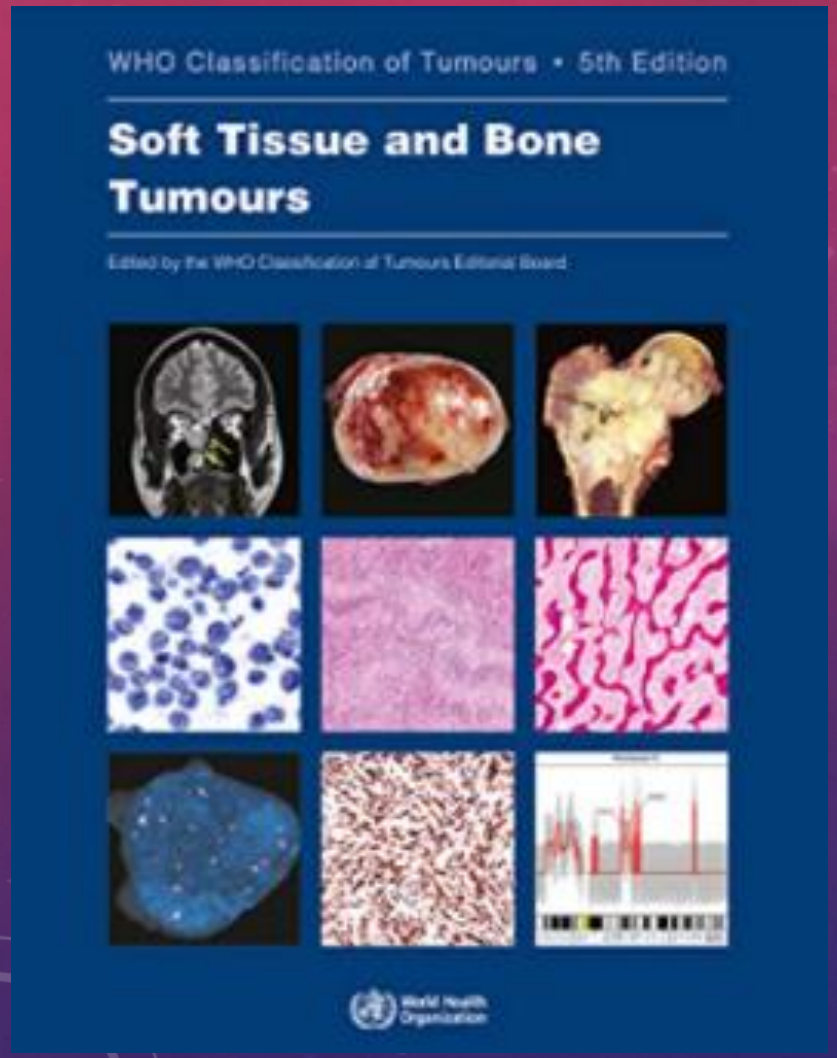
**Table 1. Distribution of Soft-Tissue Sarcoma.***

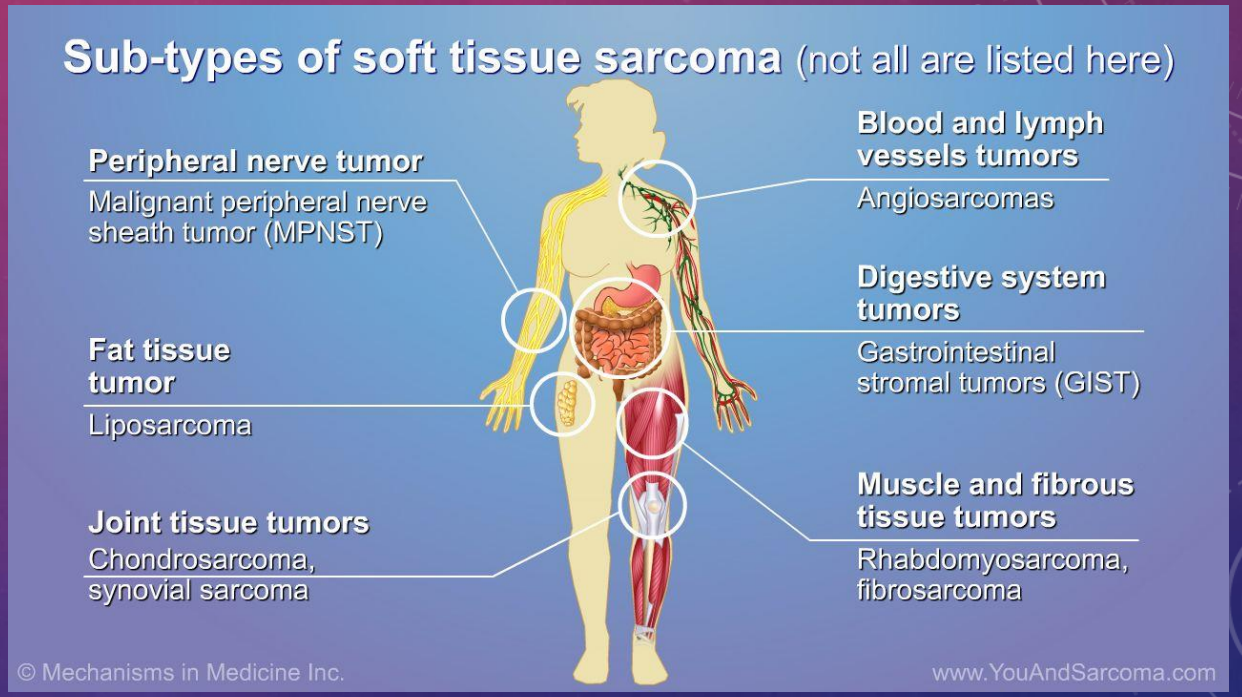
| Site | Incidence |
|---|---|
| | % |
| Lower limb and girdle | 40 |
| Upper limb and girdle | 20 |
| Retroperitoneal and intraperitoneal sites† | 20 |
| Trunk | 10 |
| Head and neck | 10 |

* Percentages are approximate.
† These sites include gastrointestinal stromal tumors.

# 2020 WHO CLASSIFICATION – 5TH EDITION



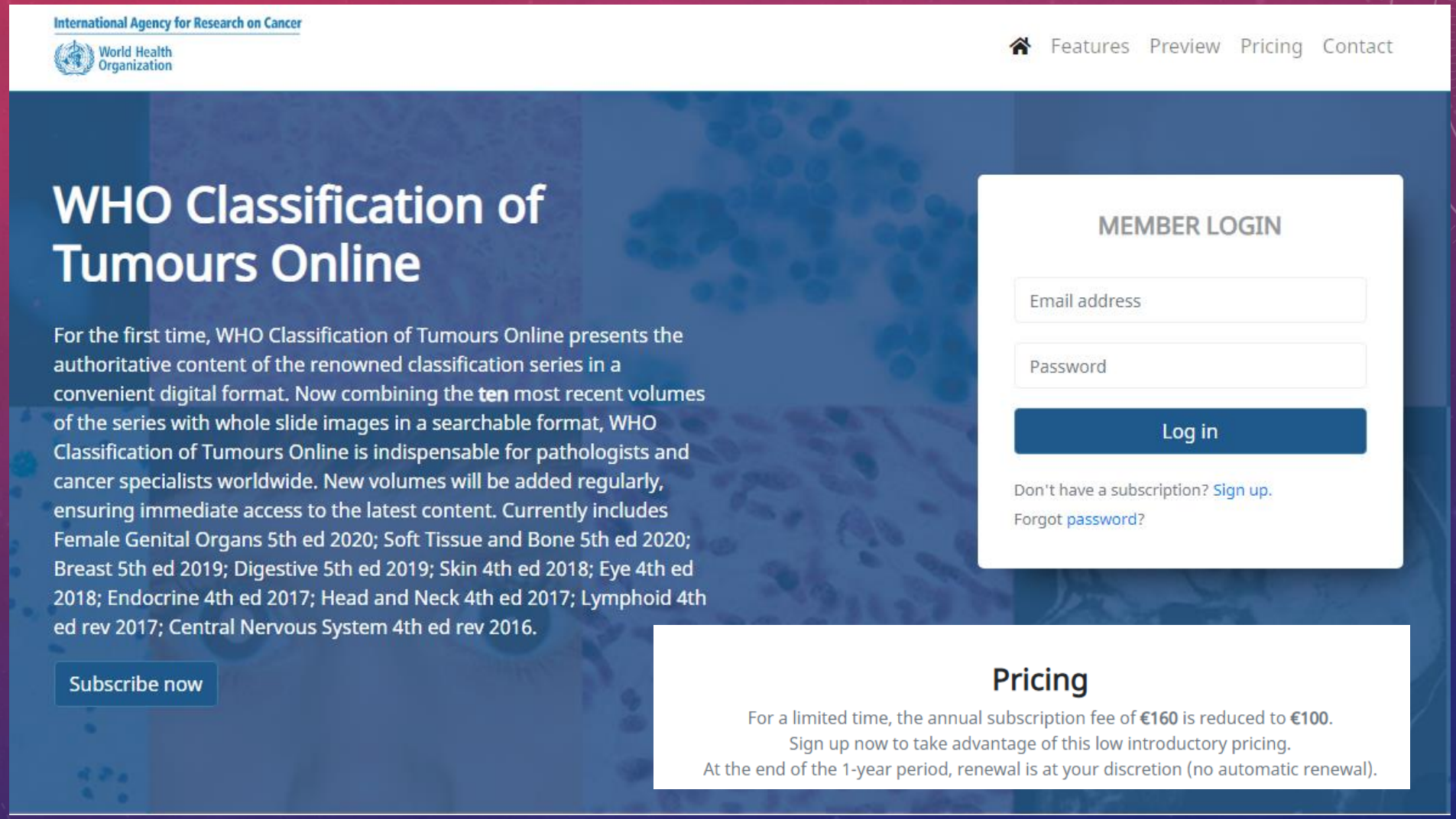




https://tumourclassification.iarc.who.int/welcome/

# 2020 WHO CLASSIFICATION – 5TH EDITION

- The book contains major modifications to the previous one through the addition of new chapters, inclusion of new entities as well as deletion and reclassification of others.
- It also demonstrated minor changes such as renaming of some tumours and updates of molecular and genetic findings.
- 159 authors from 24 different countries contributed to this book
- Includes the addition of three new chapters (gastrointestinal stromal tumours, nerve sheath tumours and undifferentiated high-grade pleomorphic sarcoma of bone).
- Incorporates more cytogenetic and molecular genetics data than previous editions
- 2 basic new terms have been promulgated: “Locally Aggressive” and “Rarely Metastasizing”
- This book will certainly serve as the basis for the classification of soft tissue and bone tumors for the next decade

# 2020 WHO CLASSIFICATION – 5TH EDITION




https://tumourclassification.iarc.who.int/welcome/

# COMPARING 4TH AND 5TH EDITION CATEGORIES

- GENERAL CLASSIFICATION GROUPS INCLUDE
  - Adipocytic Tumors – 15 distinct histologic types
  - Fibroblastic and Myofibroblastic Tumors – 25 distinct histologic types
  - So-Called Fibrohistiocytic Tumors – 4 distinct histologic types
  - Vascular Tumors – 15 distinct histologic types
  - Pericytic (perivascularo) Tumors – 3 distinct histologic types
  - Smooth Muscle Tumors – 4 distinct histologic types
  - Skeletal Muscle Tumors – 6 distinct histologic types
  - Gastrointestinal Stromal Tumor – 4 distinct histologic types – ALL NOW REPORABLE for 2021+
  - Peripheral Nerve Sheath Tumors – 11 distinct histologic types
  - Tumors of Uncertain Differentiation – 21 distinct histologic types
  - Undifferentiated Small Round Cell Sarcomas of Bone and Soft Tissue – 4 distinct histologic types

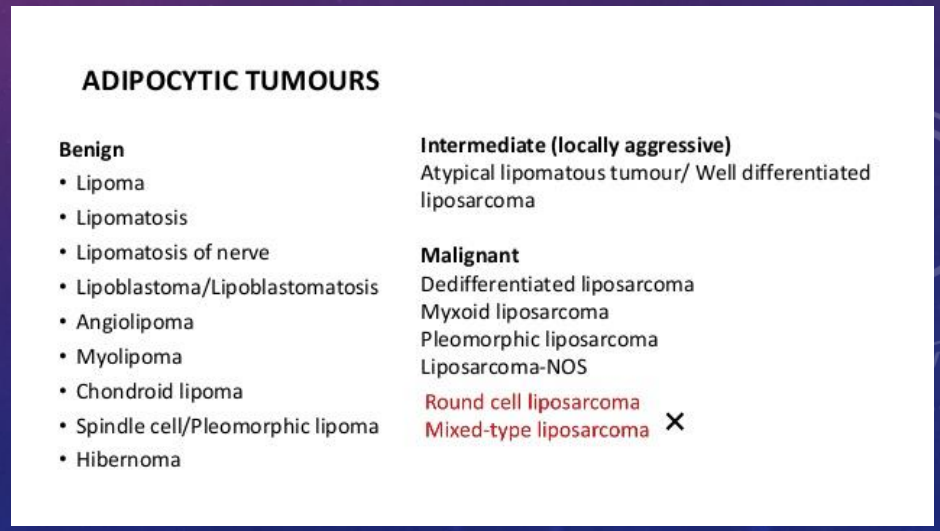
# COMPARING WHO SARCOMA CATEGORIES 2013 VS 2020

| 2013 Classification - WHO 4th edition | | 2020 Classification - WHO 5th edition |
|---|---|---|
| Angiosarcoma | | Adipocytic Tumors |
| Kaposi Sarcoma | | Fibroblastic and Myofibroblastic Tumors |
| Fibrous Tumors | | So-Called Fibbrohistiocytic Tumors |
| Soft Tissue Tumors | | Vascular Tumors |
| Fatty Tumors (Lipomatous) | | Pericytic (perivascular) Tumors |
| Chondroid Tumors (Bone and Cartilage) | | Smooth Muscle Tumors |
| Peripheral Nerve Sheath Tumors | | Skeletal Muscle Tumors |
| Rhabdomyosarcoma | | Gastrointestinal Stromal Tumor |
| Schwannoma | | Peripheral Nerve Sheath Tumors |
| Spindle Cell Tumors | | Tumors of Uncertain Differentiation |
| Myofibroblastic Tumors | | Undifferentiated Small Round Cell Sarcomas of Bone and Soft Tissue |
| Fibromyxoid Tumors | | |
| Hemangioma | | |
| Lymphangioma | | |
| Epithelioid Sarcoma | | |
| Extraskeletal Chroncdrosarcoma | | |
| Mixed Tumors | | |

# 2020 WHO CLASSIFICATION – 5TH EDITION

- **Adipocytic tumours**
  - Lipoma
  - Lipomatosis
  - Lipomatosis of nerve
  - Lipoblastoma and lipoblastomatosis
  - Angiolipoma
  - Myolipoma of soft tissue
  - Chondroid lipoma
  - Spindle cell lipoma and pleomorphic lipoma
  - Hibernoma
  - Atypical spindle cell / pleomorphic lipomatous tumour
  - Atypical lipomatous tumour / well-differentiated liposarcoma
  - Dedifferentiated liposarcoma
  - Myxoid liposarcoma
  - Pleomorphic liposarcoma
  - Myxoid pleomorphic liposarcoma

**Adipocytic tumors** are the most common mesenchymal **neoplasms**, liposarcoma accounting for approximately 20% of soft tissue sarcomas. The differential diagnosis between benign and malignant **tumors** is often problematic and represents a significant proportion of consultation cases.



# 2020 WHO CLASSIFICATION – 5TH EDITION

## Fibroblastic and Myofibroblastic Tumours

| | |
|---|---|
| · Nodular fasciitis | · Acral fibromyxoma |
| · Proliferative fasciitis and proliferative myositis | · Gardner fibroma |
| · Myositis ossificans and fibro-osseous pseudotumour of digits | · Palmar fibromatosis and plantar fibromatosis |
| · Ischaemic fasciitis | · Desmoid fibromatosis |
| · Elastofibroma | · Lipofibromatosis |
| · Fibrous hamartoma of infancy | · Giant cell fibroblastoma |
| · Fibromatosis colli | · Dermatofibrosarcoma protuberans |
| · Juvenile hyaline fibromatosis | · Solitary fibrous tumour |
| · Inclusion body fibromatosis | · Inflammatory myofibroblastic tumour |
| · Fibroma of tendon sheath | · Low-grade myofibroblastic sarcoma |
| · Desmoplastic fibroblastoma | · Superficial CD34-positive fibroblastic tumour |
| · Myofibroblastoma | · Myxoinflammatory fibroblastic sarcoma |
| · Calcifying aponeurotic fibroma | · Infantile fibrosarcoma |
| · EWSR1-SMAD3-positive fibroblastic tumour (emerging) | · Adult fibrosarcoma |
| · Angiomyofibroblastoma | · Myxofibrosarcoma |
| · Cellular angiofibroma | · Low-grade fibromyxoid sarcoma |
| · Angiofibroma of soft tissue | · Sclerosing epithelioid fibrosarcoma |
| · Nuchal-type fibroma | |

**Fibroblastic/myofibroblastic tumors** are divided into 4 main groups: benign, intermediate (locally aggressive), intermediate (rarely metastasizing), and malignant.

Many **fibroblastic/myofibroblastic** lesions are moderately or highly cellular and show mitotic activity; thus, they can be easily mistaken for sarcoma.

# 2020 WHO CLASSIFICATION – 5TH EDITION

| Vascular tumours | Pericytic (perivascular) tumours |
|---|---|
| · Haemangiomas | · Glomus tumour |
| · Synovial haemangioma | · Myopericytoma, including myofibroma |
| · Intramuscular angioma | · Angioleiomyoma |
| · Arteriovenous malformation/haemangioma | |
| · Venous haemangioma | **Smooth muscle tumours** |
| · Anastomosing haemangioma | · Leiomyoma |
| · Epithelioid haemangioma | · EBV-associated smooth muscle tumour |
| · Lymphangioma and lymphangiomatosis | · Inflammatory leiomyosarcoma |
| · Tufted angioma and kaposiform haemangioendothelioma | · Leiomyosarcoma |
| · Retiform haemangioendothelioma | |
| · Papillary intralymphatic angioendothelioma | **Skeletal muscle tumours** |
| · Composite haemangioendothelioma | · Rhabdomyoma |
| · Kaposi sarcoma | · Embryonal rhabdomyosarcoma |
| · Pseudomyogenic haemangioendothelioma | · Alveolar rhabdomyosarcoma |
| · Epithelioid haemangioendothelioma | · Pleomorphic rhabdomyosarcoma |
| · Angiosarcoma | · Spindle cell / sclerosing rhabdomyosarcoma |
| | · Ectomesenchymoma |

A vascular tumor is a tumor of vascular origin; a soft tissue growth that can be either **benign** or **malignant**, formed from **blood vessels** or **lymph vessels**.

Three major types of muscle tumours are leiomyomas, rhabdomyomas, and **rhabdomyosarcomas**.

**Smooth muscle tumors** occur infrequently in the skin. They consist of a diverse group of lesions representing hamartomas as well as benign and malignant **neoplasms**. They may arise from arrector pili **muscle**, specialized **smooth muscle** of the genitalia, or vascular **smooth muscle**.

# 2020 WHO CLASSIFICATION – 5TH EDITION

**Undifferentiated small round cell sarcomas of bone and soft tissue**

- Ewing sarcoma
- Round cell sarcoma with EWSR1-non-ETS fusions
- CIC-rearranged sarcoma
- Sarcoma with BCOR genetic alterations

**Peripheral nerve sheath tumours**

- Schwannoma
- Neurofibroma
- Perineurioma
- Granular cell tumour
- Dermal nerve sheath myxoma
- Solitary circumscribed neuroma
- Ectopic meningioma and meningothelial hamartoma
- Benign triton tumour / neuromuscular choristoma
- Hybrid nerve sheath tumour
- Malignant peripheral nerve sheath tumour
- Malignant melanotic nerve sheath tumour

| Tumours of uncertain differentiation |
| --- |
| · Intramuscular myxoma |
| · Juxta-articular myxoma |
| · Deep (aggressive) angiomyxoma |
| · Atypical fibroxanthoma |
| · Angiomatoid fibrous histiocytoma |
| · Ossifying fibromyxoid tumour |
| · Myoepithelioma, myoepithelial carcinoma, and mix |
| · Pleomorphic hyalinizing angiectatic tumour of soft |
| · Haemosiderotic fibrolipomatous tumour |
| · Phosphaturic mesenchymal tumour |
| · NTRK-rearranged spindle cell neoplasm (emerging) |
| · Synovial sarcoma |
| · Epithelioid sarcoma |
| · Alveolar soft part sarcoma |
| · Clear cell sarcoma of soft tissue |
| · Extraskeletal myxoid chondrosarcoma |
| · Desmoplastic small round cell tumour |
| · Extrarenal rhabdoid tumour |
| · PEComa, including angiomyolipoma |
| · Intimal sarcoma |
| · Undifferentiated sarcoma |

# SARCOMA RULES DO NOT EXIST OR ARE LIMITED

- THE ONLY SARCOMA ‘RULES’ OR ‘INSTRUCTIONS’ AVAILABLE ARE FOR STAGING
- THE PRIMARY SITE CODING RULES FOR SARCOMA ARE ANCIENT
- THERE ARE NO SOLID TUMOR MULTIPLE PRIMARY RULES FOR SARCOMA
- THERE ARE NO HISTOLOGY CODING RULES FOR SARCOMA
- THERE ARE SCHEMA-SPECIFIC GRADE RULES FOR SARCOMA
- THERE ARE ONLY 3 SITE SPECIFIC DATA ITEMS IN ALL SARCOMA SCHEMA

# TUMOR GRADE AND OTHER PROGNOSTIC FACTORS

**Fédération Nationales des Centres de Lutte Contre le cancer; High Power Field**

| Tumor differentiation | Definition |
|---|---|
| Score 1 | Sarcomas that closely resemble normal adult mesenchymal tissues |
| Score 2 | Sarcomas for which histologic typing is certain |
| Score 3 | Embryonal and undifferentiated sarcomas, synovial sarcoma and sarcomas of uncertain differentiation |
| Mitotic count | |
| Score 1 | 0-9 mitosis/ 10 hpf |
| Score 2 | 10-19 mitosis/ 10 hpf |
| Score 3 | 20≥ mitosis/ 10 hpf |
| Tumor necrosis | |
| Score 0 | No necrosis |
| Score 1 | <50% tumor necrosis |
| Score 2 | ≥50% tumor necrosis |
| Histologic grade | Tumor differentiation + mitotic count + tumor necrosis |
| Grade 1 (Low grade) | Total score: 2 or 3 |
| Grade 2 (Intermediate grade) | Total score: 4 or 5 |
| Grade 3 (High grade) | <50% tumor necrosis |



https://www.sciencedirect.com/science/article/abs/pii/S0972978X18302587

# CODING THE PRIMARY SITE FOR SARCOMA CASES

- When a Sarcoma involves/originates in a solid organ such as stomach or lung or breast – code the solid organ as the primary site, regardless of sarcoma type.
- Even when you know the sarcoma is of blood vessel or fat cell or fibrous tissue origin – code the solid organ
- When you cannot identify a solid organ of origin then you look at histology and anatomic location.
- When a Sarcoma involves a bone – code the bone and laterality if applicable (C40.0-C41.9)
- When a Sarcoma involves cartilage – code the bone cartilage and laterality if applicable (C40.0-C41.9)
- When a Sarcoma involves soft tissue – code the area of soft tissue in the C49.0-C49.9 series
- When a Sarcoma involves muscle – code the area of the muscle in the C49.0-C49.9 series
- When a Sarcoma involves the uterus – code to myometrium not endometrium or uterus, NOS
- When a Sarcoma involves the lung – be sure this is not lung metastasis with multiple nodules
- When a Sarcoma involves the meninges – code to meninges C70.0 (NOT C70.9)
- Most Sarcoma of Skin Sites are actually Subcutaneous Soft Tissue - use the site/histology validation list
- Rarely some sarcoma are coded to skin such as Kaposi Sarcoma – use the site/histology validation list
- Unknown Primary Sarcoma is rare but does occur – code primary site to C80.9 in these rare cases

# CODING THE PRIMARY SITE FOR SARCOMA CASES

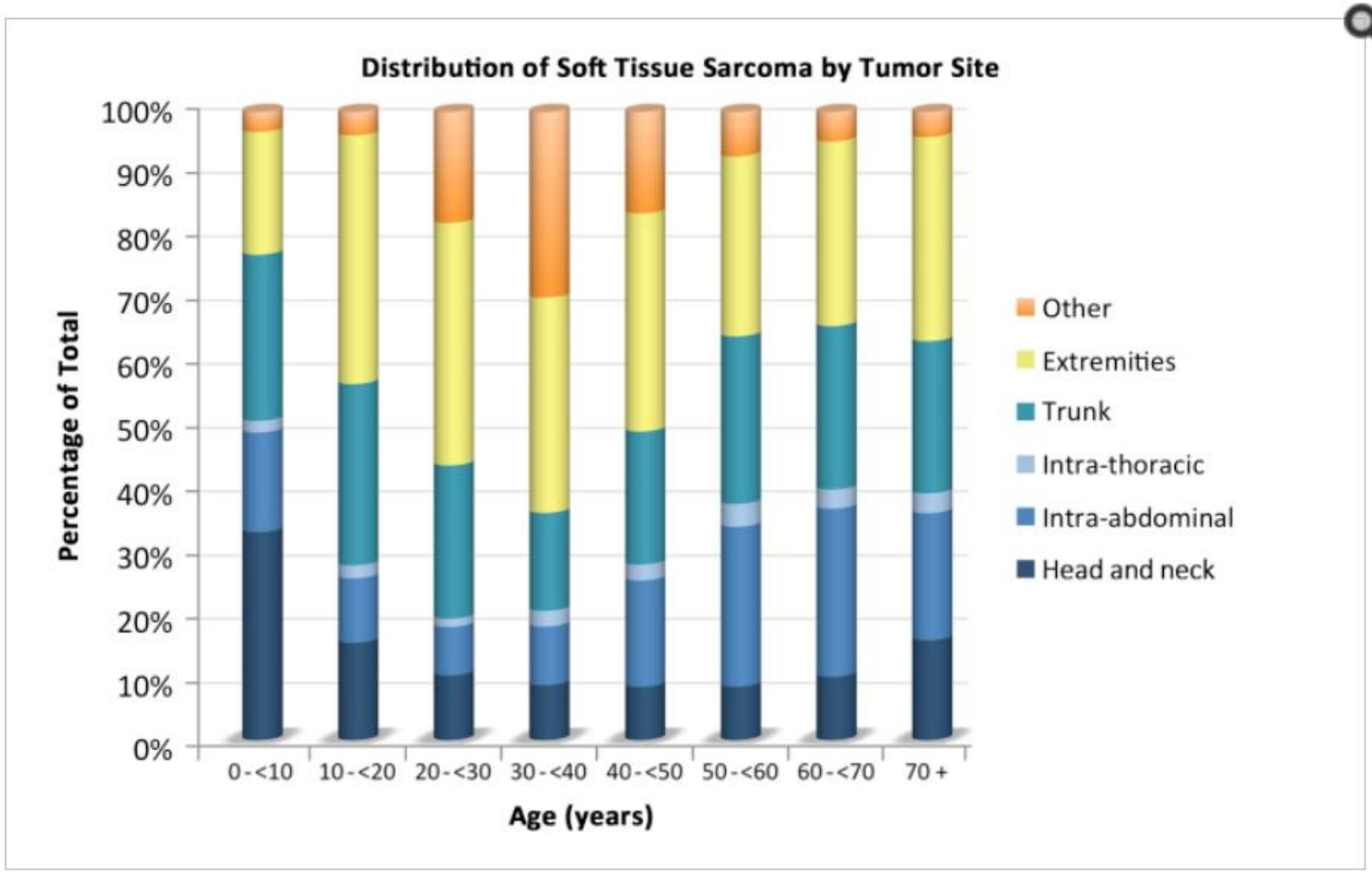



Figure 3. The distribution of nonrhabdomyosarcomatous soft tissue sarcomas by age according to tumor site.

https://www.ncbi.nlm.nih.gov/books/NBK65923/

# BIOLOGICAL POTENTIAL DESCRIBED SPECIFICALLY

- **Benign** - Most benign soft tissue tumours do not recur locally. Those that do recur do so in a non-destructive fashion and are almost always readily cured by complete local excision.
- **Intermediate (locally aggressive)** - Soft tissue tumours in this category often recur locally and may be associated with an infiltrative and locally destructive growth pattern. Lesions in this category very rarely if ever metastasize but typically require wide excision with a margin of normal tissue in order to ensure local control.
- **Intermediate (rarely metastasizing)** - Soft tissue tumours in this category are often locally aggressive (see above) but, in addition, show the well-documented ability to give rise to distant metastases in occasional cases. The risk of such metastases appears to be < 2% and is not reliably predictable on the basis of histomorphology. Metastasis in such lesions is usually to lymph node or lung.
- **Malignant** - In addition to the potential for locally destructive growth and recurrence, malignant soft tissue tumours known as soft tissue sarcomas) have a substantial risk of distant metastasis, ranging in most instances from 20% to almost 100%, depending on histological type and grade. Some (but not all) histologically low-grade sarcomas have a metastatic risk of only 2–10%, but such lesions may advance in grade in a local recurrence and thereby acquire a higher risk of distant spread (e.g. myxofibrosarcoma and leiomyosarcoma).
- **IMPORTANT NOTE:** the intermediate categories do not correspond to histologically determined intermediate grade in a soft tissue sarcoma (see below), nor do they correspond to the ICD-O /1 behaviour category described as uncertain whether benign or malignant. The locally aggressive subset of entities with no metastatic potential, as defined above, are generally given ICD-O /1 codes, and the rarely metastasizing lesions are given ICD-O /3 codes

# MOLECULAR GENETICS AND TUMOR MARKERS

- Tumor Markers and Molecular Genetics are used for:
  - Diagnose, stage, and/or classify cancer
  - Estimate prognosis
  - Select an appropriate treatment (eg, treatment with a targeted therapy)
  - Assess response to treatment
  - Detect residual cancer after treatment
  - Monitor whether cancer has become treatment resistant
  - Detect recurrence or progression after treatment

# MOLECULAR GENETICS AND TUMOR MARKERS

- There are very few Tumor Markers or Molecular Genetic Mutations or Tests Available for Sarcoma
- Sarcomas with simple/single genome may be identified with specific molecular marker(s)
- However, sarcomas with complex genome show multiple, non-recurrent molecular alterations
- There are more than 80 sarcoma types/subtypes making markers and genetic testing difficult for sarcoma

Diagnostic, Prognostic and Predictive Molecular Markers in Sarcoma

| Recurrent rearrangements | Fusion oncogenes | Sarcoma-type specific (reviewed by Mertens)[4] | Diagnostic / Prognostic |
|---|---|---|---|
| Point mutations or small *indels* | *KIT / PDGFRA, SDHA / B* | GIST | Predictive / Diagnostic |
| | *CTNNB1* | Desmoid tumor | Diagnostic |
| | *IDH1, IDH2* | Enchodroma / chondrosarcoma | Diagnostic |
| | *SUZ12, EED* | Malignant peripheral nerve sheath tumor | Diagnostic |
| | *PIK3CA* | Myxoid liposarcoma | Predictive |
| | *KDR* | Angiosarcoma | Diagnostic |
| | *NRAS, KRAS, HRAS, FGFR4* | Embryonal rhabdomyosarcoma | Diagnostic / Predictive |
| | *MYOD1* | Spindle cell rhabdomyosarcoma | Diagnostic |
| | *MED12* | Leiomyoma (and small subset of leiomyosarcoma) | Diagnostic |
| | *NF1* | MPNST and others | Diagnostic |
| Copy number gain / Amplification | *MDM2, CDK4* | WD/DDLPS | Diagnostic |
| | *CDK4* | | Predictive |
| | *MYC* | Postradiation sarcoma | Diagnostic |
| | *MYOCD* | Leiomyosarcoma | Diagnostic |
| Copy number loss / Deletion | *TP53* | Osteosarcoma, leiomyosarcoma, and others | Prognostic |
| | *SMARCB1* | Rhabdoid tumor, epithelioid sarcoma | Diagnostic |
| | *SMARCA4* | SMARCA4-deficient thoracic sarcomas | Diagnostic |
| | *CDKN2A* | MPNST, fibrosarcomatous DFSP, advanced GIST | Predictive |
| | *RB1* | Spindle cell lipoma, and others | Diagnostic |
| | | High-grade sarcomas with complex karyotype | Predictive |
| | *NF1* | MPNST and others | Diagnostic / Predictive |

Tumors with Simple Genome Include:

- Ewing Sarcoma
- GIST
- Undifferentiated Pleomorphic Sarcoma
- Osteosarcoma

However, as these tumors mature, additional point mutations may occur with potential for great variability in genomically-complex sarcomas – these suggest tumor progression



https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4988391/

# MOLECULAR GENETICS AND TUMOR MARKERS

**Table 2.** Selected Sarcomas With Distinct Cytogenetic and Molecular Alterations of Diagnostic Relevance

| Sarcoma Type | Cytogenetic Alteration | Molecular Alteration |
|---|---|---|
| Alveolar rhabdomyosarcoma | t(2;13)(q35;q14)<br>t(1;13)(p36;q14), double minutes<br>t(2;2)(q35;p23)<br>t(X;2)(q35;q13) | *PAX3-FOXO1* fusion<br>*PAX7-FOXO1* fusion<br>*PAX3-NCOA1* fusion<br>*PAX3-AFX* fusion |
| Alveolar soft part sarcoma | t(X;17)(p11.2;q25) | *TFE3-ASPSCR1* fusion |
| *BCOR*-rearranged sarcoma | Inv(X)(p11p11)<br>?<br>? | *BCOR-CCNB3* fusion<br>*BCOR-MAML3* fusion<br>*ZC3H7B-BCOR* fusion |
| *CIC*-rearranged sarcoma | t(4;19)(q35;q13) or t(10;19)(q26;q13)<br>t(X;19)(q13;q13.3) | *CIC-DUX4* fusion<br>*CIC-FOXO4* fusion |
| Clear cell sarcoma | t(12;22)(q13;q12)<br>t(2;22)(q32.3;q12) | *EWSR1-ATF1* fusion<br>*CREB1-EWSR1* fusion |
| Dedifferentiated liposarcoma | Ring and giant marker chromosomes | Amplification of 12q13-15: *MDM2, CDK4, HMGA2* |
| Desmoplastic small round cell tumor | t(11;22)(p13;q12) | *EWSR1-WT1* fusion |
| DFSP | Ring form of chromosomes 17 and 22 | *COL1A1-PDGFB* fusion |
| Endometrial stromal sarcoma, low grade | t(7;17)(p15;q21)<br>t(6;7)(p21;7p15)<br>t(6;10)(p21;p11)<br>t(1;6)(p34;p21)<br>t(X;17)(p11;q21) | *JAZF1-SUZ12* fusion<br>*PHF1-JAZF1* fusion<br>*EPC1-PHF1* fusion<br>*MEAF6-PHF1* fusion<br>*MBTD1-CXorf67* fusion |
| Endometrial stromal sarcoma, high grade | t(10;17)(q22;p13)<br>t(X;22)(p11;q13) | *YWHAE-NUTM2* fusion<br>*ZC3H7B-BCOR* fusion |
| Epithelioid hemangioendothelioma | t(1;3)(p36;q25)<br>t(X;11)(p11;q22) | *WWTR1-CAMTA1* fusion<br>*YAP1-TFE3* fusion |
| Epithelioid sarcoma | Deletion 22q<br>t(8;22)(q22;q11)<br>t(10;22) | *SMARCB1* inactivation |
| Ewing sarcoma | t(11;22)(q24;q12)<br>t(21;22)(q12;q12)<br>Others | *EWSR1-FLI1* fusion<br>*EWSR1-ERG* fusion<br>Others |
| Extraskeletal myxoid chondrosarcoma | t(9;22)(q22;q12)<br>t(9;17)(q22;q11)<br>t(9;15)(q22;q21)<br>t(3;9)(q11;q22)<br>t(9;17)(q22;q11) | *EWSR1-NR4A3* fusion<br>*TAF2N-NR4A3* fusion<br>*TCF12-NR4A3* fusion<br>*TFG-NR4A3* fusion<br>*RBP56-NR4A3* fusion |
| GIST | Deletion 14q, 22q, 1p, 15q | *KIT* or *PDGFRA* mutation (85% of cases); *NF1, SDHX, BRAF* mutation, *SDHC* hypermethylation, other |
| Inflammatory myofibroblastic tumor | t(1;2)(q22;p23)<br>t(2;19)(p23;p13)<br>t(2;17)(p23;q23) | *TPM3-ALK* fusion<br>*TPM4-ALK* fusion<br>*CLTC-ALK* fusion |



https://ascopubs.org/doi/pdf/10.1200/JCO.2017.74.9374

# CRITICAL STAGING FACTORS

- SEER Summary Stage 2000 – Uses Basic Concepts of Local, Regional Direct Extension, Regional Nodes, Combined Extension and Nodes and Distant Disease – nodes are rarely involved limiting  Summary Stage to Local, Regional Direct, or Distant
  - Bone
  - Gastro-Intestinal Stromal Tumors (GIST)
  - Heart, Mediastinum and Pleura
  - Retroperitoneum
  - Soft Tissues

- AJCC Chapters for Sarcoma – stated not to be used for pediatric sarcoma (but, they still are in many cases)
  - Bone
  - Gastro-Intestinal Stromal Tumors (GIST)
  - Orbital Sarcoma
  - Soft Tissue Sarcoma – Unusual Histologies and Sites
  - Soft Tissue Sarcoma – Abdomen and Thoracic Visceral Organs
  - Soft Tissue Sarcoma – Head & Neck
  - Soft Tissue Sarcoma – Retroperitoneum
  - Soft Tissue Sarcoma – Trunk and Extremities

# CRITICAL STAGING FACTORS

- Basis for BOTH AJCC AND SEER Summary Stage Rests on Very Simple Criteria
  - Tumor Location
  - Tumor Size
  - Tumor Extension to Adjoining Structures
  - Tumor with Invasion into Specific Adjoining Structures
  - Presence or Absence of Positive Regional Lymph Node(s) - rare
  - Presence or Absence of Distant Metastasis (usually lung, bone, or liver)
  - Tumor Grade and/or Mitotic Rate or Score (low mitosis/high mitosis)
  - Tumor Necrosis
  - Histologic Grade
- The hope is to eventually develop Risk Stratification Scoring Models based on above to guide and manage treatment, recurrence, progression, and new therapies.

# CRITICAL STAGING FACTORS

- SEER Summary Staging Manual 2020 – GUIDELINES BY STAGE – page 10-20 of manual
  - Provides the general descriptions for each general stage
    - Localized
    - Regional by Direct Extension
    - Regional Lymph Nodes Only
    - Regional by BOTH Direct Extension and Regional Lymph Node Involved
    - Distant (Disseminated)
    - Benign/Borderline
    - Unknown/Unstaged
  - There is no such thing as an in-situ sarcoma.

# CRITICAL STAGING FACTORS

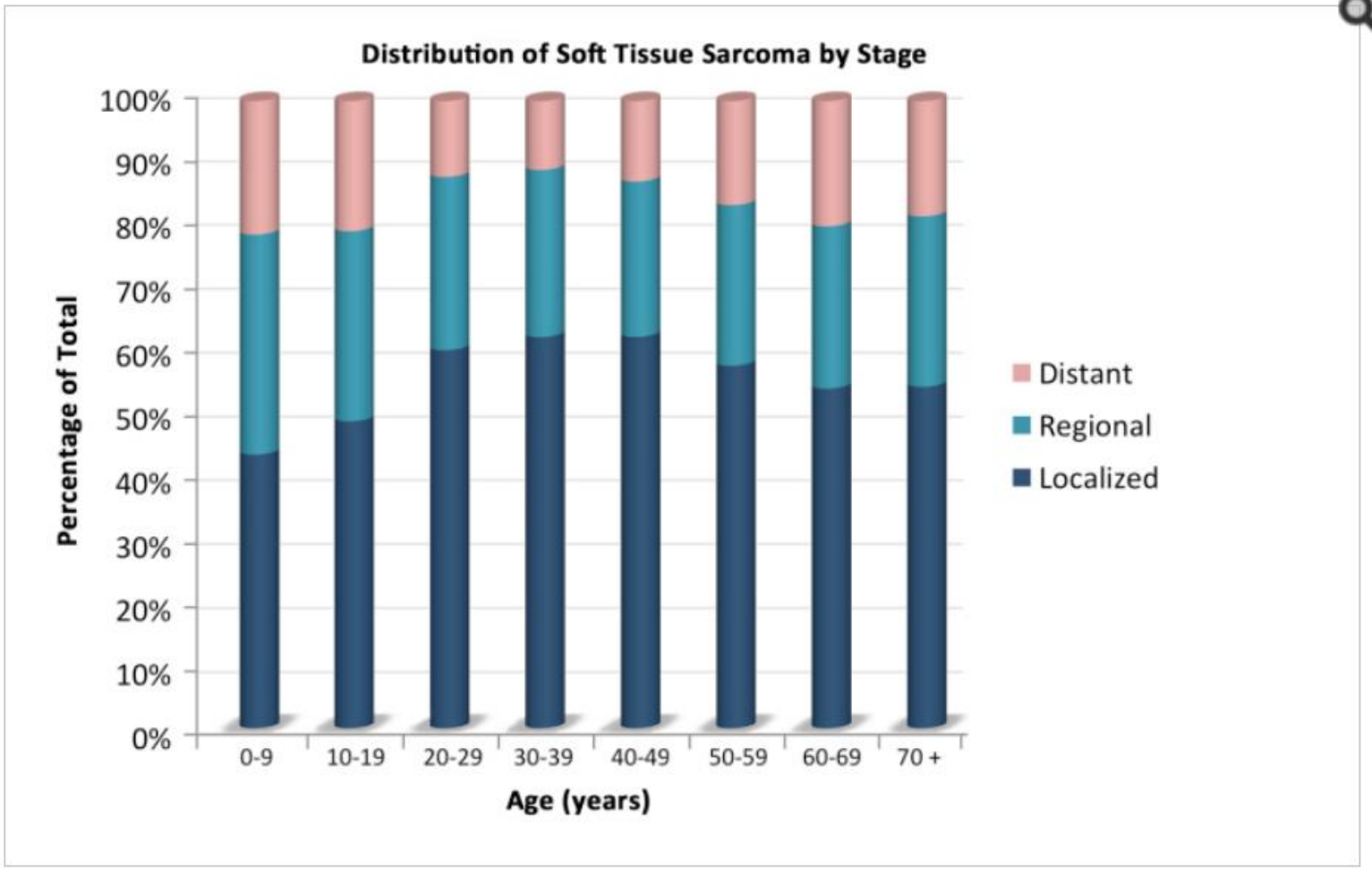



Figure 1. The distribution of nonrhabdomyosarcomatous soft tissue sarcomas by age according to stage.

https://www.ncbi.nlm.nih.gov/books/NBK65923/

# SOFT TISSUE AND CONNECTIVE TISSUE

- Soft Tissue Sarcomas is a broad group of many types of sarcoma that can develop in soft tissues like fat, muscle, nerves, fibrous tissues, blood vessels, or deep skin tissues. They can be found in any part of the body. Most of them start in the arms or legs. They can also be found in the trunk, head and neck area, internal organs, and the area in back of the abdominal cavity or retroperitoneum.
  - Adult Fibrosarcoma
  - Alveolar Soft-Part Sarcoma
  - Angiosarcoma [hemantgiosarcoma (blood vessel) or lymphangiosarcoma (lymph vessel)]
  - Clear Cell Sarcoma
  - Desmoplastic Small Round Cell Tumor
  - Epithelioid Sarcoma
  - Fibromyxoid Sarcoma, low grade
  - Kaposi Sarcoma (HHV8)
  - Leiomyosarcoma
  - Liposarcoma
  - Malignant Mesenchymoma
  - Malignant Peripheral Nerve Sheath Tumors (neurofibrosarcoma, malignant schwannoma, neurogenic sarcoma)
  - Myxofibrosarcoma, low-grade
  - Rhabdomyosarcoma
  - Synovial Sarcoma
  - Undifferentiated Pleomorphic Sarcoma

C49.0-C49.9 includes smooth muscle and skeletal muscle, connective and soft tissues, tendons, nerves, fat, fibrous tissue, subcutaneous, skin, etc.

# LONG AND SHORT BONES AND CARTILAGE

- **Benign tumors** are more common in the cartilage than malignant ones. These are called enchondromas.
- **Another type of benign cartilage tumor** is a bony projection of cartilage called an osteochondroma.
- **Osteosarcoma** (also called osteogenic sarcoma) is the most common primary bone cancer. It starts in the bone cells. It most often occurs in young people between the ages of 10 and 30, but about 10% of osteosarcoma cases develop in people in their 60s and 70s. It's rare in middle-aged people, and is more common in males than females. These tumors develop most often in bones of the arms, legs, or pelvis.
- **Chondrosarcoma starts in cartilage cells**. It's the second most common primary bone cancer. It's rare in people younger than 20. After age 20, the risk of getting a chondrosarcoma goes up until about age 75. Chondrosarcomas can start anywhere there's cartilage. Most develop in bones like the pelvis, legs, or arms . Sometimes chondrosarcoma starts in the trachea, larynx, or chest wall. Other sites are the scapula (shoulder blade), ribs, or skull. Subtypes include Clear Cell Chondrosarcoma, Mesenchymal Chondrosarcoma, and De-Differentiated Chrondrosarcoma – the last two are usually high grade tumors.
- **Ewing Tumor is the third most common primary bone cancer,** and the second most common in children, teens, and young adults. It's rare in adults older than 30. This cancer (also called Ewing sarcoma) is named after Dr. James Ewing, who first described it in 1921. Most Ewing tumors develop in bones, but they can start in other tissues and organs. The most common sites for this cancer are the pelvis, the chest wall, and the long bones of the legs or arms.
- Ewing tumors occur most often in white people and are very rare among African Americans and Asian Americans.
- **Malignant fibrous histiocytoma (MFH) most often starts in soft tissue (connective tissues such as ligaments, tendons, fat, and muscle); it's rare in bones.** This cancer is also known as pleomorphic undifferentiated sarcoma, especially when it starts in soft tissues. When MFH occurs in bones, it usually affects the legs (often around the knees) or arms. This cancer most often occurs in elderly and middle-aged adults.

# MUSCLE (INCLUDES SKELETAL MUSCLE & FASCIA)

- **Most Sarcoma of Muscle are Described in the General Section Soft Tissue/Connective Tissue**
- **Rhabdomyosarcoma** (RMS) is a type of sarcoma made up of cells that normally develop into skeletal (voluntary) muscles. These are muscles that we control to move parts of our body. Well before birth, cells called rhabdomyoblasts (which will eventually form skeletal muscles) begin to form. These are the cells that can develop into RMS. Because this is a cancer of very early forms of muscle cells, it is much more common in children, although it does sometimes occur in adults.
- Embryonal rhabdomyosarcoma (ERMS) is one subtype of rhabdomyosarcoma that usually affects children in their first 5 years of life, but it can occur at older ages as well. ERMS tends to occur in the head and neck area, bladder, vagina, or in or around the prostate and testicles.
  - There are 2 subtypes of ERMS, **botryoid and spindle cell rhabdomyosarcomas** which tend to have a better outcome
- The second subtype of rhabdomyosarcoma is **alveolar rhabdomyosarcoma** or ARMS
- ARMS typically affects all age groups equally. It makes up a larger portion of RMS in older children, teens, and adults than in younger children. ARMS most often occurs in large muscles of the trunk, arms, and legs.
- ARMS tends to grow faster than ERMS, and it usually requires more intense treatment.

- **Anaplastic rhabdomyosarcoma or pleomorphic rhabdomyosarcoma** and **undifferentiated sarcoma** are more rare cancers however, they are both high grade and require intensive treatment.

# GYNECOLOGICAL SARCOMA

- Coding Primary Site for GYN Sarcoma can be tricky – some originate in endometrium, some in myometrium.
- Cancers that start in epithelial cells, the cells that line or cover most organs, are called carcinomas of endometrium.
- More than 95% of uterine cancers are carcinomas.
- Sarcomas start from tissues like muscle, fat, bone, and fibrous tissue (tendons and ligaments) may be either.
- Most uterine sarcomas are put into categories, based on the type of cell they start in:
- But, one type of mixed sarcoma of the GYN that starts in the uterus is called **carcinosarcoma**. These cancers start in the endometrium and have features of both sarcomas and carcinomas. These cancers are also known as malignant mixed mesodermal tumors or malignant mixed mullerian tumors. The primary site is Endometrium not Myometrium.
- The Two Types of Uterine Sarcoma that originate in myometrium or connective tissue (stroma) are:
  - **Uterine Leiomyosarcoma** – starts in muscular wall of uterus – most common type – grow and spread quickly
  - **Endometrial Stromal Sarcoma** – starts in stroma of the lining of the uterus (endometrium) – more rare and often low-grade. However, may be high-grade if it is large, growing quickly or has spread and hard to treat
- **Undifferentiated Sarcoma may start in either endometrium or myometrium and have poor prognosis**

# GIST AND OTHER STROMAL TUMORS

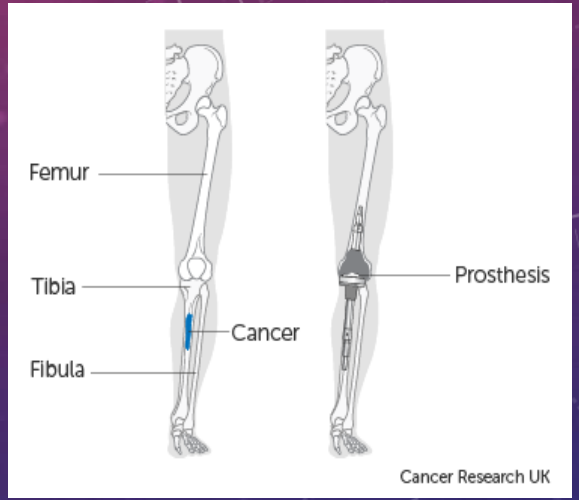
- Gastrointestinal stromal tumors (GISTs) start in very early forms of special cells in the wall of the GI tract called the interstitial cells of Cajal (ICCs). ICCs are sometimes called the “pacemakers” of the GI tract because they signal the muscles in the GI tract to contract to move food and liquid along.
- More than half of GISTs start in the stomach. Most of the others start in the small intestine, but GISTs can start anywhere along the GI tract. A small number of GISTs start outside the GI tract in nearby areas such as the omentum or the peritoneum.
- GIST is likely to grow and spread quickly, such as:
  - The size of the tumor
  - Where it's located in the GI tract
  - How fast the tumor cells are dividing (its mitotic rate)
- **ALL GIST ARE REPORTABLE STARTING 1/1/2021 regardless of the specified criteria now outdated.**
- Other more rare types of cancer in the GI tract include different types of soft tissue sarcomas such as:
  - Leiomyosarcomas: cancers of smooth muscle cells
  - Angiosarcomas: cancers of blood vessel cells
  - Malignant peripheral nerve sheath tumors (MPNSTs): cancers of cells that support and protect nerves

# TREATMENT OPTIONS

- Surgery (including Limb Salvage and Reconstruction)
- Radiation (Cause and Cure)
- Chemotherapy (Limited Agents)
- Anti-Angiogenesis Agents – block new blood vessel formation
- Immunotherapy (Experimental - promising)
- Targeted Therapy (Experimental - promising)
- Combination Therapy (Surgery and Various Types of Radiation)
- Other Therapy ( May add Chemo, Immuno or Experimental for Advanced Disease
- Other Therapy (May add Chemo, Immuno or Experimental for Recurrence/Progression

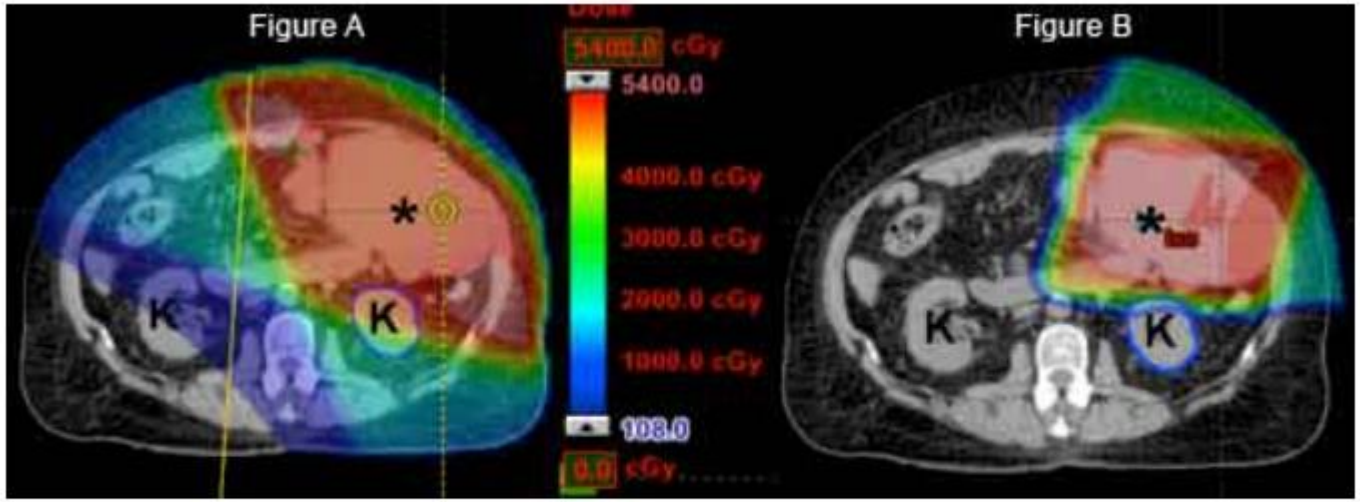
# SURGERY OF PRIMARY SITE, LIMB SALVAGE & RECONSTRUCTION

- **Who Has Surgery**
  - Surgery works best for solid tumors that are contained in one area. It is a local treatment.
  - Sometimes surgery will be the only treatment you need. But most often, you will also have other cancer treatments. The usual treatment following surgery for sarcoma is radiation therapy as initial course of tx.
- **Remove the entire tumor**
  - Surgery removes cancer that is contained in one area.
- **Debulk a tumor**
  - Surgery removes some, but not all, of a cancer tumor. Debulking is used when removing an entire tumor might damage an organ or the body. Removing part of a tumor can help other treatments work better.
- **Ease cancer symptoms**
  - Surgery is used to remove tumors that are causing pain or pressure.




**Limb Salvage with Reconstruction** are procedures that are an alternative to amputation. Limb salvage procedures replace a diseased bone and reconstructs a functional limb by using a metal implant, a bone graft from another person (allograft), or a combination bone graft and metal implant (allo-prosthetic composite) to spare the patient's limb and provide functional limb support where amputation may be the only other surgical option. These procedures are only performed for long-bones such as leg or arm. They allow for a more functional outcome and improved qualify of life in sarcoma patients.

# RADIATION THERAPY




*Dose distribution comparison between a conventional radiation plan (Figure A) and a proton therapy plan (Figure B) in a patient with an abdominal sarcoma (*). Note the difference in dose to the kidneys (K).*



https://www.floridaproton.org/cancers-treated/sarcoma/sarcoma-treatment

# SYSTEMIC THERAPIES
# CHEMOTHERAPY AND TARGETED THERAPIES

**Soft Tissue Sarcoma – FDA Approved Agents**

- Cosmegen (Dactinomycin)
- Dactinomycin
- Doxorubicin Hydrochloride
- Eribulin Mesylate
- Gleevec (Imatinib Mesylate)
- Halaven (Eribulin Mesylate)
- Imatinib Mesylate
- Pazopanib Hydrochloride
- Tazemetostat Hydrobromide
- Tazverik (Tazemetostat Hydrobromide)
- Trabectedin
- Votrient (Pazopanib Hydrochloride)
- Yondelis (Trabectedin)
- Drug Combinations (VAC)

**Bone Cancer Agents – not just Sarcoma**

- Cosmegen (Dactinomycin)
- Dactinomycin
- Denosumab
- Doxorubicin Hydrochloride
- Methotrexate
- Trexall (Methotrexate)
- Xgeva (Denosumab)

Three New Agents for Sarcoma FDA Approved in 2016 & 2020

- **Lartruvo (olaratumab)** with doxorubicin for soft tissue tumors. Lartruvo is a platelet-derived growth factor (PDGF) receptor-alpha blocking antibody.
- **Avapritinib (Ayvakit)** GIST – PDGFRA/D842V mutations
- **Tazemetostat (Tazverik)** – epithelioid sarcoma – only agent available for this type of sarcoma which represents <1% sarcoma
- New agents are slow to market for sarcoma of any type – these 3 brand new agents are the first new breakthrough agents including immunotherapies in many years for ANY sarcoma.

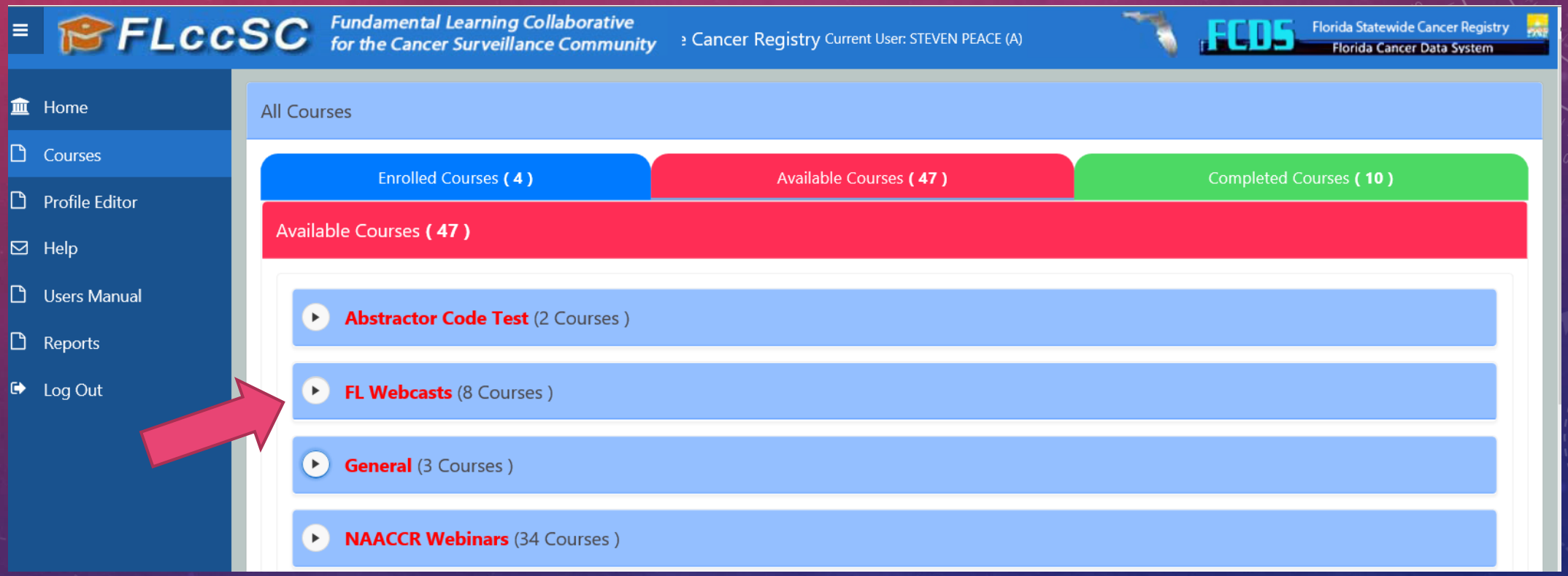
# RECURRENCE AND PROGRESSION OF DISEASE


- For all sarcoma patients, local tumor control ranges from 30% to 100%, but that data represents a broad spectrum of sarcoma subtypes and treatments.
- Certain tumors that are high grade, large, and/or located deep under the skin are associated with the highest risk for recurrence.
- Also, compared to younger individuals (i.e., those <55-60 years old), tumors in older patients tend to relapse more often.

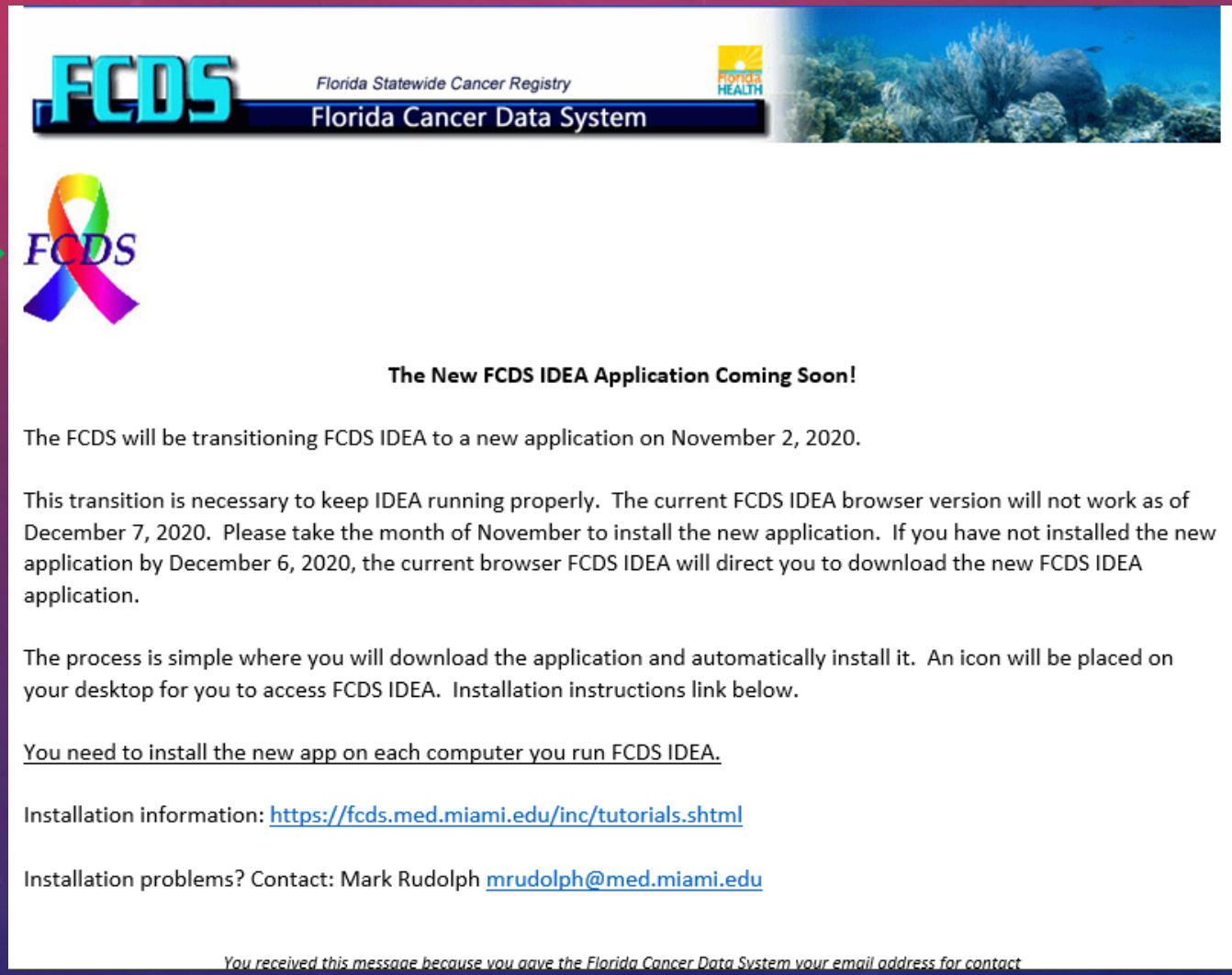


https://www.floridaproton.org/cancers-treated/sarcoma/sarcoma-treatment

# CEU QUIZ IS ACTIVATED NOW



# FCDS IDEA APPLICATION ANNOUNCEMENT




**The New FCDS IDEA Application Coming Soon!**

The FCDS will be transitioning FCDS IDEA to a new application on November 2, 2020.

This transition is necessary to keep IDEA running properly. The current FCDS IDEA browser version will not work as of December 7, 2020. Please take the month of November to install the new application. If you have not installed the new application by December 6, 2020, the current browser FCDS IDEA will direct you to download the new FCDS IDEA application.

The process is simple where you will download the application and automatically install it. An icon will be placed on your desktop for you to access FCDS IDEA. Installation instructions link below.

You need to install the new app on each computer you run FCDS IDEA.

Installation information: https://fcds.med.miami.edu/inc/tutorials.shtml

Installation problems? Contact: Mark Rudolph mrudolph@med.miami.edu

*You received this message because you gave the Florida Cancer Data System your email address for contact*

# 2021 STANDARDS – UPDATES, MANUALS, NEW ITEMS

1. 2021 Implementation of Reporting Requirements
   a. Retire Flat File Format – will allow flat file submissions/data transmissions until 6/30/2021
   b. XML File Format – 7/1/2021 (tentative)/optional for submissions starting Jan 2021
   c. 2021 New Reportable Criteria – ALL GIST, ALL Thymoma, Evolving Melanoma –
   d. 2021 Not Reportable Criteria – Need to remove NIFTP, EFVPTC, EFVPTC and other histologies from thyroid
   e. Schema ID – changes to criteria and one new schema added
   f. 2021 New Data Items Required
      i. Name-Birth Surname – Replaces Maiden Name which will be Retired
      ii. Medicare Beneficiary ID
      iii. Grade Post Therapy Clinical (yc)
      iv. Gleason Pattern Clinical
      v. Gleason Pattern Pathological
      vi. Gleason Score Clinical
      vii. Gleason Score Pathological
      viii. Gleason Tertiary Pattern
   g. Changed Data Items – LDH
      i. HER2 Overall Summary added to Esophagus and Stomach Schemas 00161, 00169, 00170
      ii. Radiation Modality New Codes
      iii. Changes to SSDIs
      iv. FIGO Stage
      v. Grade – schema specific
   h. Grade Field Conversions
   i. Stage Conversions
   j. SSDI Conversions
   k. 2021 Retired Data Items – TNM 6 & 7, Maiden Name
   l. 2021 FCDS DAM Revision – by March 31, 2021
   m. FCDSv21 EDITS Metafile – under review
   n. FCDSv21 Updates to Abstractor Code Test & Review of Existing Q&A
   o. 2021 Grade Manual
   p. 2021 SSDI Manual
   q. 2021 Heme Updates
   r. 2021 Solid Tumors – 2 newly revised chapters and changes to all 8 other chapters
   s. 2021 ICD-O-3 Updates
   t. 2021 STORE Manual
   u. 2021 Site/Type Validation Table

# REFERENCES AND RESOURCES


- WHO Classification of Neoplasms of Soft Tissue and Bone, 5th edition, January 2020
- WHO Classification of Neoplasms of Soft Tissue and Bone, 4th edition, February 2013

- American Cancer Society Cancer Series
  - Bone Cancer
  - Ewing Tumors
  - Gastrointestinal Stromal Tumor
  - Osteosarcoma
  - Rhabdomyosarcoma
  - Soft Tissue Sarcoma
  - Uterine Sarcoma

- Cancer.Net (ASCO) – Sarcoma, Soft Tissue – May 2019

# REFERENCES AND RESOURCES

- Bone Tumor Staging Systems – Dr Abdullah Ghafour and Dr Ali A Alwan
  - AJCC TNM, 8th edition – 8 chapters
  - SEER Summary Stage 2000 – 4 chapters
  - Enneking Staging System – soft tissue and bone – 1 system

- CAP Protocols for Sarcoma
  - Primary Sarcoma of Uterus
  - Primary Tumors of Bone
  - Gastrointestinal Stromal Tumor (GIST)
  - Pediatric Ewing Sarcoma
  - Pediatric Rhabdomyosarcoma

# REFERENCES AND RESOURCES


- NCCN Clinical Practice Guidelines
  - Bone Cancer – version 1.2020
  - AIDS-Related Kaposi Sarcoma – version 3.2020
  - Soft Tissue Sarcoma – version 2.2020
  - Uterine Neoplasms

- FDA - Drugs Approved for Soft Tissue Sarcoma – October 2020
- FDA Announcement Accelerated Approval - New Treatment for Advanced Soft Tissue Sarcoma – 1/2019
- FDA – Two New Soft Tissue Sarcoma Treatments Approved by the FDA – January 28, 2020 – K. Honey

- "Contemporary Sarcoma Diagnosis, Genetics, and Genomics" – J Clin Oncol 36:101-110. © 2017 by American Society of Clinical Oncology
- "Malignant Bone Tumor" – Resection and Reconstruction– Orthopaedic Oncology - Dr P. Agarwal – 2019

# QUESTIONS ……