-Los tumores óseos malignos primarios son relativamente poco frecuentes.

-Presentan una incidencia de 0,8/ 100.000 h

-Los tumores benignos más frecuentes son los encondromas, fibroma no osificante y displasia fibrosa cortical.

-Los tumores óseos malignos más frecuentes son las metástasis y mieloma múltiple, seguido por el osteosarcoma, condrosarcoma y sarcoma de Ewing.

Tumours of Bone(WHO classification, febrero 2013)

Tumours of Bone(WHO classification febrero 2013)

Chondrogenic tumours

The chondrogenic tumours of bone are now classified into benign, intermediate (locally aggressive), intermediate (rarely metastasizing) and malignant grades.

Benign

- Osteochondroma
- Chondroma
- Enchondroma
- Periosteal chondroma
- Osteochondromyxoma
- Subungual exostosis
- Bizarre parosteal osteochondromatous proliferation
- Synovial chondromatosis

Intermediate (locally aggressive)

- Chondromyxoid fibroma
- Atypical cartilaginous tumour / chondrosarcoma grade I

Chondrogenic tumours

Intermediate (rarely metastasizing)
Chondroblastoma

Malignant
Chondrosarcoma Grade I, II , grade III
Dedifferentiated chondrosarcoma
Mesenchymal chondrosarcoma
Clear cell chondrosarcoma

The osteochondromyxoma, subungual exostosis, bizarre parosteal osteochondromatous proliferation and synovial chondromatosis were added to the benign chondrogenic tumours of bone.

Chondromyxoid fibroma and atypical cartilaginous tumour / chondrosarcoma grade I are grouped together as intermediate (locally aggressive) tumours.

Chondroblastoma is classified as an intermediate (rarely metastasizing) tumour.
Enchondromatosis is discussed in the tumour syndromes chapter and not in this

Chondrogenic tumours

- **Osteochondromyxoma**

  Osteochondromyxoma is a newly added extremely rare entity to the group of chondrogenic tumours of bone, associated with Carney complex.

  Bizarre parosteal osteochondromatous proliferation

  Bizarre parosteal osteochondromatous proliferation is a benign entity that was added to the group of benign chondrogenic tumours of bone.

  Chondrosarcoma (grades I-III), including primary and secondary variants and periosteal chondrosarcoma
  Chondrosarcoma grade I (now officially termed atypical cartilaginous tumour) is reclassified as an intermediate (locally aggressive) tumour, better reflecting its clinical behaviour.

- Chondrosarcoma is subclassified into:

  Primary central chondrosarcoma

  Secondary central chondrosarcoma

  Secondary peripheral chondrosarcoma

  Periosteal chondrosarcoma

  Secondary chondrosarcoma is currently subdivided into central (arising in a pre-existing enchondroma) and peripheral (juxtaposed to the cartilaginous cap of an osteochondroma) types.

IDH1 and IDH2 mutations are found in primary, secondary central and periosteal chondrosarcomas as well as 50% of dedifferentiated chondrosarcomas. Mesenchymal chondrosarcoma carries a recurrent translocation resulting in a HEY1-NCOA2 gene fusion.

Osteogenic tumours

Benign
Osteoma
Osteoid osteoma
Intermediate (locally aggressive)
Osteoblastoma
Malignant
Low-grade central osteosarcoma
Conventional osteosarcoma
Chondroblastic osteosarcoma
Fibroblastic osteosarcoma
Osteoblastic osteosarcoma
Telangiectatic osteosarcoma
Small cell osteosarcoma
Secondary osteosarcoma
Parosteal osteosarcoma
Periosteal osteosarcoma
High-grade surface osteosarcoma

Other than the addition of osteoma to the benign osteogenic tumours category, the classification has not changed. I
n conventional osteosarcoma, the authors give less emphasis on the detailed histopathologic subclasses and indicate that currently, there is no evidence of any relationship between these subtype and prognosis.
IDH1 and IDH2 somatic mutations were found to be absent in all cases studied thus potentially helping to distinguish from chondrosarcoma where they are common.

Osteoclastic giant cell rich tumours

Benign

Giant cell lesion of the small bones

Intermediate locally aggressive, rarely metastasizing

Giant cell tumour of bone

Malignant

Malignancy in giant cell tumor of bone

Giant cell tumor of bone is now considered a locally aggressive, very rarely metastasizing lesion.

Fibrohistiocytic tumours

Benign

Benign fibrous histiocytoma / non-ossifying fibroma

Malignant fibrous histiocytoma of bone was removed from the current classification. MFH of bone was renamed undifferentiated high-grade pleomorphic sarcoma of bone.

Notochordal tumours

Benign

Benign notochordal tumour

Malignant

Chordoma

The expression of brachyury is specific for notochordal tumors. Recent studies showed that IDH1 and IDH2 mutations are not detected in chordomas. These findings help to distinguish chordomas from chondrosarcomas.

Vascular tumors

Benign

Haemangioma

Intermediate (locally aggressive rarely metastasizing)

epithelioid hemangioma

Malignant

Epithelioid hemangioendothelioma

Angiosarcoma

The new classification now recognizes epithelioid hemangioma as a separate entity that can occur, sometimes multifocal, in bone. EH can behave in a locally aggressive fashion, with (lymph node) metastases being rare.

Criterios Predictores del tipo histológico

A- Localización de la lesión:

-En el Eje Longitudinal: Metafisarias, epifisarias o diafisarias.

-En el Eje Transversal: central, cortical, yuxtacortical, parostal y periférica.

B- Edad de presentación.

C- Criterios de Imagen : muy importantes en la valoración del grado de agresividad

Criterios semiológicos de diferenciación B/M

1-Patrón: Lítico geográfico(1a,1b y 1c), apolillado , permeativo.

2- Zona de transición: Borde bien definido/ con o sin esclerosis o mal definido.

3- Alteración de la cortical: erosión endóstica, expandida, rota

Reacción perióstica: regular/ compacta / homogénea ó irregular/heterogénea (en capas de cebolla, triangulo de Codman, rayos de sol, en cepillo), en concha.

5- Masa de partes blandas.

Contribución de la TC

Valora mejor el componente osificado /calcificado

Contribución de la RM

Valora extensión local a partes blandas y Médula ósea

Contribución del Pet-TC

Agresividad de la lesión (captación de FDG)

Aporta criterios del TC

Extensión (ganglios, lesiones a distancia)

Metástasis

A recordar

- **Rx** imprescindible para localizar lesión , caracterización y criterios de agresividad

- **TC** para valorar mejor compoente óseo o calcificado de la lesión , rotura cortical y reacción perióstica

- **RM** ayuda localizar lesión , mejor valoración de extensión local a partes blandas e intramedular (intra ó extracompartimental) ,imprescindible para planificación quirúrgica y para control evolutivo de posible recidiva

- **PET-TC** Valoración de extensión a distancia (lesiones satélites, ganglionar , toraco-abdominal,) y control de respuesta al tto