Primary Bone Tumour and Pathological Fractures

Miss Gillian L Cribb
MBChB MRCS FRCS (Tr & Orth)
Consultant Orthopaedic and Oncological Surgeon

Primary Bone Tumour

- Since 2005
- 1 of 5 centres
  - RJAH Oswestry
  - ROH Birmingham
  - RNOH London
  - NOC Oxford
  - Newcastle Freeman
- £1,000,000 per annum

NHS England

Specialised Commissioning

Primary Musculo-Skeletal Tumours

- Rare
  - 10 / Million Malignant Primary Bone Tumours
- Highly Specialised Services
  - Diagnosis
  - Surgery
- Multi Disciplinary Team
  - Radiology
  - Pathology
  - Surgery
  - Oncology

Primary Bone Tumours

Symptoms

- Night Pain
  - Especially Young Adolescent
  - ‘Growing Pains’
- Non Mechanical Pain
- Swelling

Benign Bone Tumours

Benign Primary Bone Tx

- Many
- Chondroblastoma
- **Giant Cell Tumour**
- Osteoid Osteoma

Enneking Staging (1986)

Benign

- Latent
  - SBC, FD
- Active
  - Chondroblastoma, Osteoid osteoma
- Aggressive
  - GCT, ABC

Malignant Bone Tumours

Malignant Primary Bone Tumours

- **Osteosarcoma**
- **Ewing's**
- **Chondrosarcoma**

- Plasmacytoma / Multiple Myeloma
- Metastases
- Others

Risk Factors

- **High Risk**
  - Maffuci Syndrome
  - Ollier's Disease
- **Moderate Risk**
  - Diaphyseal Aclasia
  - Paget's Disease
  - Radiation Osteonecrosis
- **Low Risk**
  - Chronic Osteomyelitis
  - Osteonecrosis

Staging – Bone Tumour

Local Staging

- X-ray lesion
- MRI whole bone
- CT whole

Distant staging

- CXR / Chest CT
- Isotope bone scan / WB MRI

Biopsy

Enneking Staging (1986)

Plain radiograph

Malignant

1: Low grade without metastases

- A: Intra-compartmental
- B: Extra-compartmental

2: High grade without metastases

- A: Intra-compartmental
- B: Extra-compartmental

3: Metastases

- A: Intra-compartmental
- B: Extra-compartmental

Biopsy

- Specialist centre
- Usually GA
- Longitudinal incisions
- Excisable track
- Do NOT cross compartments
- Tourniquet
- No exsanguination
- Jamshidi needle

Diagnostic Aids

- Age of patient
  - Physes open or closed

- Location in the bone
  - Epiphyseal
  - Metaphyseal
  - Diaphyseal

Epiphyseal Tumours X3

Chondroblastoma

Giant Cell Tumour (physes closed)

Clear Cell Chondrosarcoma

Diaphyseal - Benign

Osteoid Osteoma

Eosinophil Granuloma

Fibrous Dysplasia

Diaphyseal - Malignant

Ewings

Adamantinoma

Diaphyseal Tumours - Age

< 10:	Eosinophil Granuloma
Teenage:	Ewing's
Adult:	Lymphoma
> 60:	Metastasis / Myeloma

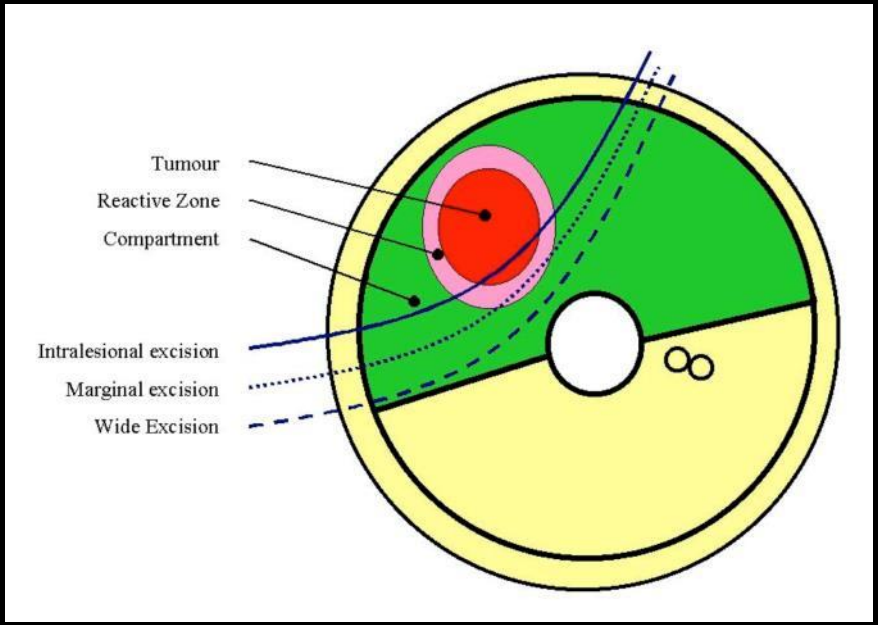
Metaphyseal Tumours

- All Others
- ABC
  - Primary (physes open)

Secondary ABC
- GCT
- Chondroblastoma

Surgical Margins

- **Intra-lesional**
  - Within lesion
- **Marginal**
  - Within reactive zone,
  - Extra-capsular
- **Wide**
  - Beyond reactive zone
  - In normal tissue
- **Radical**
  - Extra-compartmental



Malignant Primary Bone Tumours

- Neo-adjuvant chemotherapy
  - Osteosarcoma and Ewing's
- Radiotherapy
  - Ewing's
- Chondrosarcoma
  - **NOT** generally sensitive to radio- or chemo-therapy

Surgical Options
Malignant Primary Bone Tumours

- **Limb Salvage**
  - Excision Alone
  - Excision and Bone Graft
    - Autograft
    - Irradiated Autograft
    - Allograft
  - **Excision Endoprosthetic Replacement**
  - Excision and Arthrodesis
  - Rotationplasty
  - Excision and Bone Transport
- **Amputation**

Surgical Options
Benign Primary Bone Tumours

- **Observation**
- **Limb Salvage**
  - **Radiofrequency thermocoagulation**
  - **Curettage**
    - Alone
    - Reconstruction
      - Graft
      - Substitute
  - **Excision**
    - Alone
    - Reconstruction
      - Internal fixation
      - External fixation
      - Graft
      - Bone transport
      - Endoprosthesis
- **Amputation**

Osteosarcoma

- **Clinical Features**
  - Gradual onset pain & swelling
  - Commonly around the knee of adolescent / young adult
  - Often "contrived" history of trauma (no recognised trauma association)
  - Night / rest pain
- **Age**
  - Bi-modal age distribution
    - Peak 10-14 years – 70%
      - girls younger age than boys
    - Older >40 – 30%
- **Investigation**
  - X-ray
  - Refer to specialist centre for full staging and biopsy

Radiographic Features

- Metaphyseal
- Bone formation
- Codmans triangle
- Sunray spiculation
- Soft Tissue Mass

- **Treatment**
  - Chemotherapy (Neo–adjuvant)
  - Surgery
  - Further Chemotherapy

- **Prognosis**
  - 60-90 % 5 year survival
  - Limb salvage does not compromise survival

Conventional (central) osteosarcoma

Osteoblastic Osteosarcoma

Telangiectatic Osteosarcoma

Chondroblastic Osteosarcoma

Surface Osteosarcoma

Parosteal Osteosarcoma

Osteosarcoma

Intra-osseous well-differentiated (low grade) osteosarcoma

Secondary Osteosarcoma

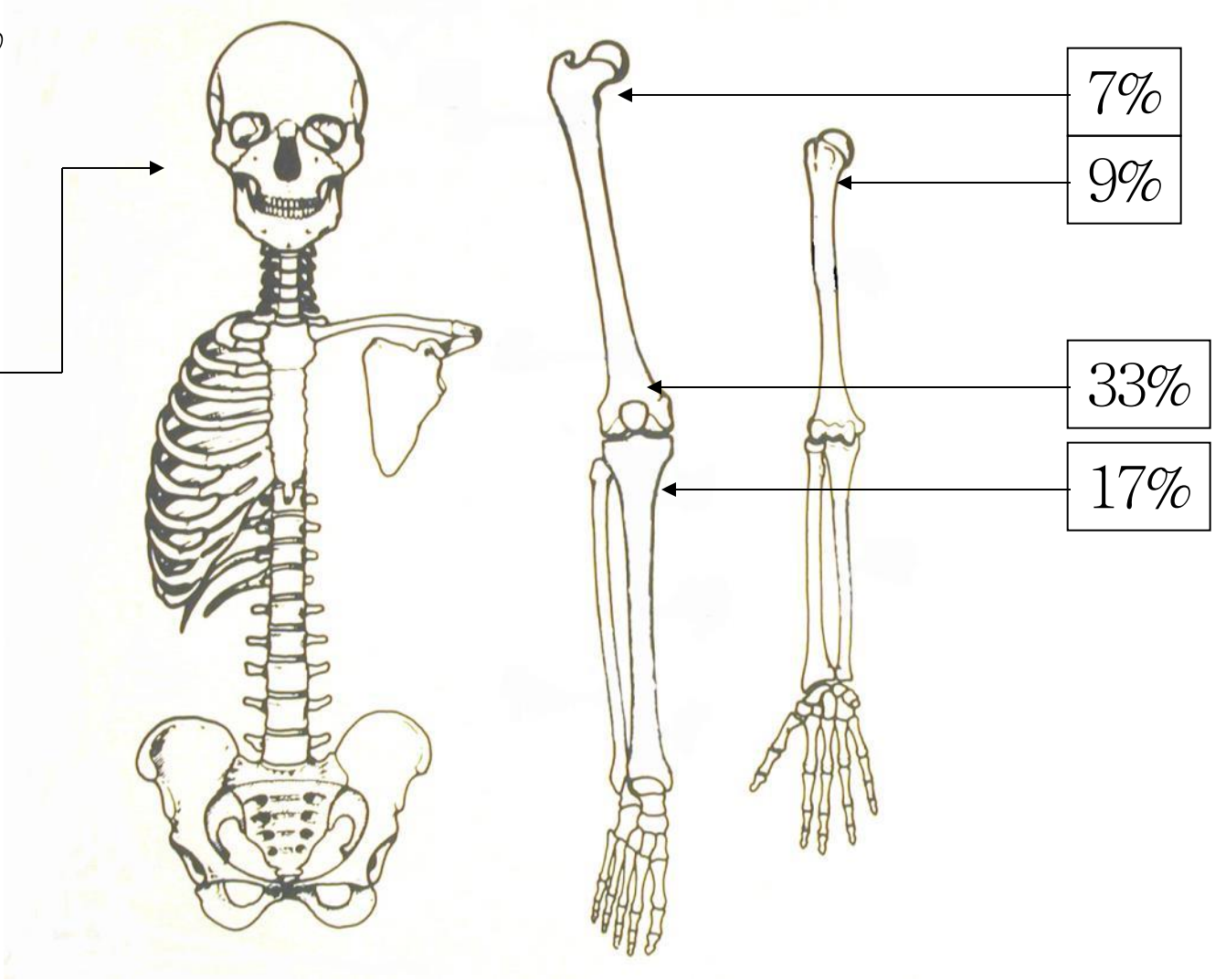
Pagets

Radiation

Osteosarcoma

Age:
10-30 = 75%

Sex:
M:F = 2:1



Ewing's Sarcoma

- **Clinical Features**
  - Tumour of childhood and adolescence (though can occur all age groups)
  - Pain & swelling
  - Symptoms increasing over months
  - Systemic symptoms (fever & malaise) common

- **Investigation**
  - X-ray (appearances vary especially in adults)
  - Full staging
  - Biopsy in specialist centre
  - This tumour is a great mimic of other pathologies (infection)
  - Abnormal haematology (raised ESR, leucocytosis, anaemia

Radiographic Appearances

- Diaphyseal
- Periosteal reaction
- Onion-skinning
- Soft tissue mass

MRI Features

- Large soft tissue mass

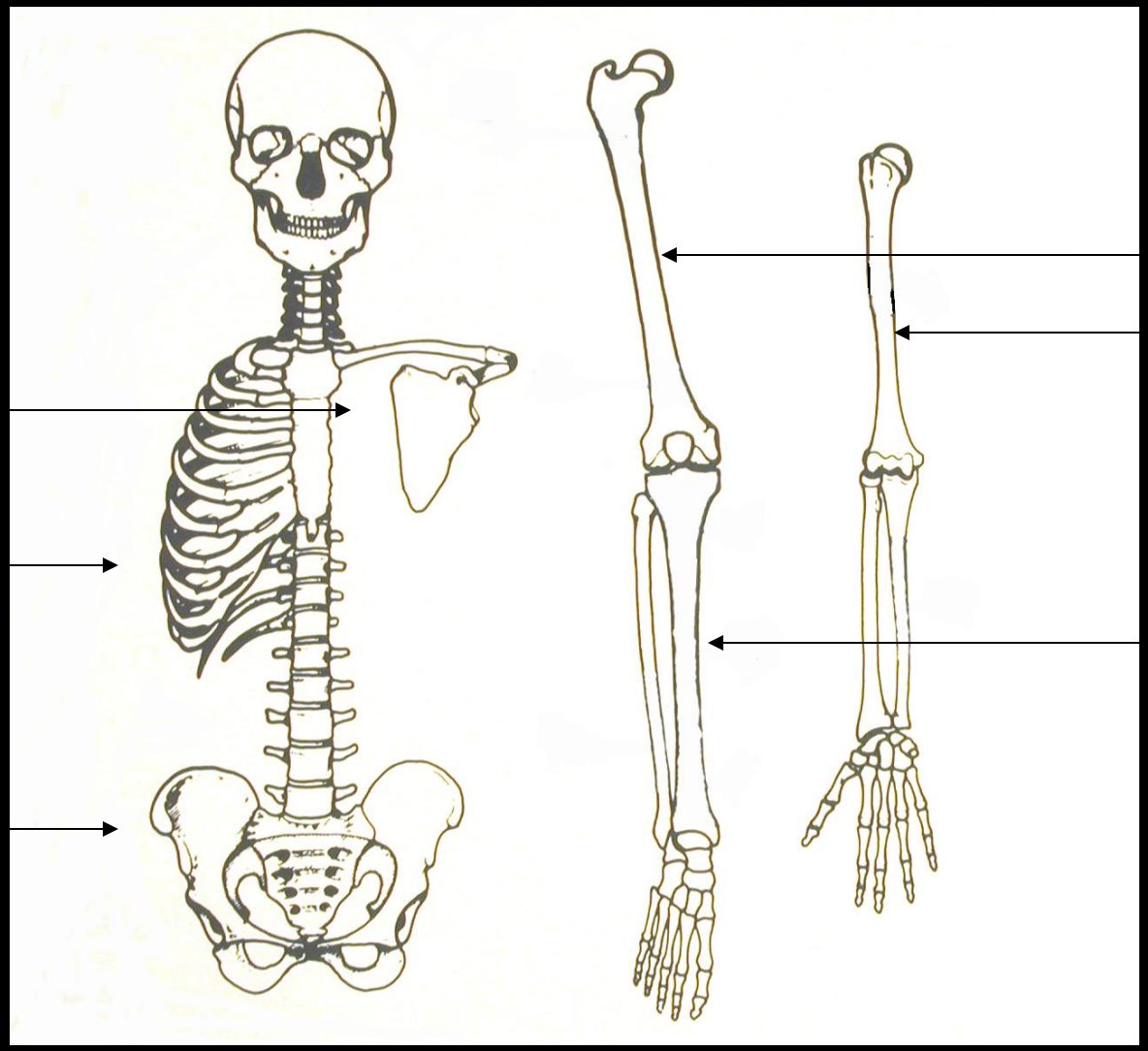
Ewing's Sarcoma

- **Treatment**
  - National Ewings MDT
  - Both chemo- and radio- therapy are effective
  - Role of limb salvage surgery increased in recent times
- **Prognosis**
  - Much improved with modern chemotherapy (and radiotherapy)
  - Central / peripheral
  - Tumour volume
  - 70 % 5 year survival

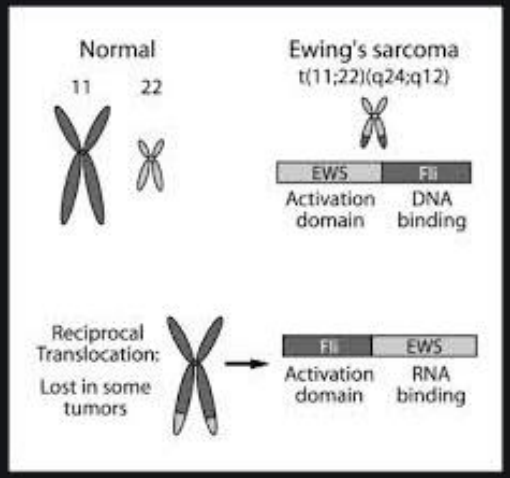
Ewing's sarcoma

Age:
< 20 = 75%

Sex:
M:F = 4:3



Translocation	Fusion gene	% of tumors exhibiting EWS gene rearrangement
t(11;22)(q24;q12)	*EWSR1-FLI1*	85
t(21;22)(q22;q12)	*EWSR1-ERG*	10
t(7;22)(q22;q12)	*EWSR1-ETV1*	rare
t(17;22)(q21;q12)	*EWSR1-ETV4*	rare
t(2;22)(q35;q12)	*EWSR1-FEV*	rare



Chondrosarcoma

- **Clinical Features**
  - Typically a tumour of middle age
  - Slow growing tumours may be very large at presentation
  - Relatively mild local discomfort

Radiographic Features

- Metaphyseal
- Scalloping
- Mineralisation
  - ‘popcorn’
- Extra-osseous mass

Chondrosarcoma

- **Investigation**
  - Staging and biopsy in specialist centre
  - MR – high signal
  - CT scan essential – mineralisation, cortical breach
  - Image guided biopsy preferred (permeation)

Chondrosarcoma

- **Treatment**
  - Wide surgical resection
  - No role for chemo- or radio- therapy in primary treatment

Histology

- Enchondroma
- Atypical enchondroma
- Grade 1 chondrosarcoma
- Grade 2 choondrosarcom
- Grade 3 chondrosarcoma
- Dedifferentiated chondrosarcoma

- PERMEATION

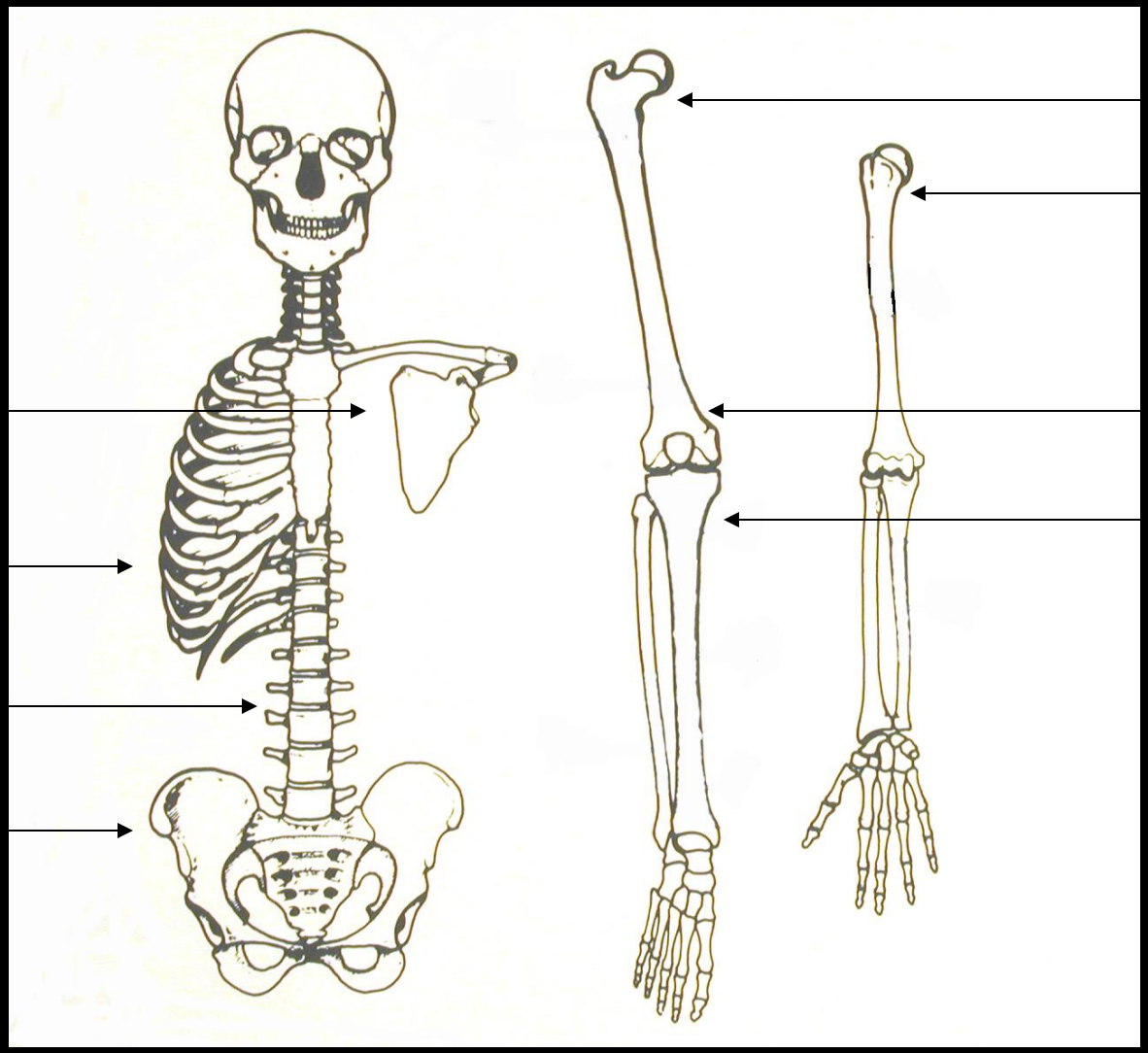
Prognosis

- Depends on resectability and grade
  - 5 year survival
    - Grade 1 – 89%
    - Grade 2 and 3 – 53%
  - Dedifferentiated
    - 20% survival at 2 years
- Metastases to lung

Typical chondrosarcoma

Age:
30-60 = 60%

Sex:
M:F = 5:4



Secondary Chondrosarcoma

- Olliers and Maffucci

- Diaphyseal aclasia

Giant Cell Tumour

- **Clinical Features**
  - 4-5% benign bone tumours
  - Slight female preponderance
    - ?association with pregnancy
  - Pain, increasing over months
  - Swelling
  - Pathological fracture 5-10%
  - Malignant transformation <1%
  - Chest disease
    - Rare

- **Investigation**
  - X-ray
  - MR and CT
  - Needle biopsy
  - CXR

Radiographic Features

- Epiphyseal
- Eccentric
- Lucent
- Expanded
- ‘Soap-bubbles’

Treatment

- Excision
- Curettage, cement and plate
- Excision and endoprosthetic replacement
- Excision, autograft and arthrodesis
- Denosumab