



COLUMBIA UNIVERSITY  
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# Pain management in Erdheim-Chester Disease

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# Disclosure

No relevant financial conflicts related to this subject

# **UNDERSTANDING PAIN IN ERDHEIM-CHESTER DISEASE**

# Taxonomy of Pain

- *Nociceptive*
  - Somatic
  - Visceral
- *Neuropathic*
- *Psychogenic*
- *Idiopathic*

**TABLE 91.1 Common Pain Syndromes in Hematologic Malignancies**

<b>Procedure-Related Pain</b>	
Deep somatic pain	Bone marrow aspiration, biopsy, and harvest Headache following lumbar puncture
Superficial somatic pain	Venepuncture (needle insertions) Central catheter placement/positioning
<b>Therapy-Related Pain</b>	
Deep somatic pain	Bone marrow expansion and/or sensitization by granulocyte colony-stimulating factor, osteoporosis (e.g., from corticosteroids use), myalgias (e.g., from corticosteroid withdrawal), myopathy
Superficial somatic pain	Oropharyngeal mucositis (e.g., from chemotherapy or radiotherapy)
Visceral pain	Enteritis, typhlitis, hemorrhagic cystitis
Neuropathic pain	Drug-related neuropathies (e.g., from chemotherapy agents)
Headache	Drug related (e.g., due to tretinoin)
<b>Pain From Hematologic Malignancy</b>	
Somatic pain	Bone infarct or necrosis, osteomyelitis, compression fracture, hemarthrosis
Visceral pain	Tumor involvement, splenomegaly, lymphadenopathy, or lymphadenitis
Neuropathic pain	Paraproteins with antimyelin properties, amyloid infiltration, peripheral nerve compression, spinal cord compression
Mixed pain	Headache, meningeal infiltration or infection, brain metastasis, or primary tumor

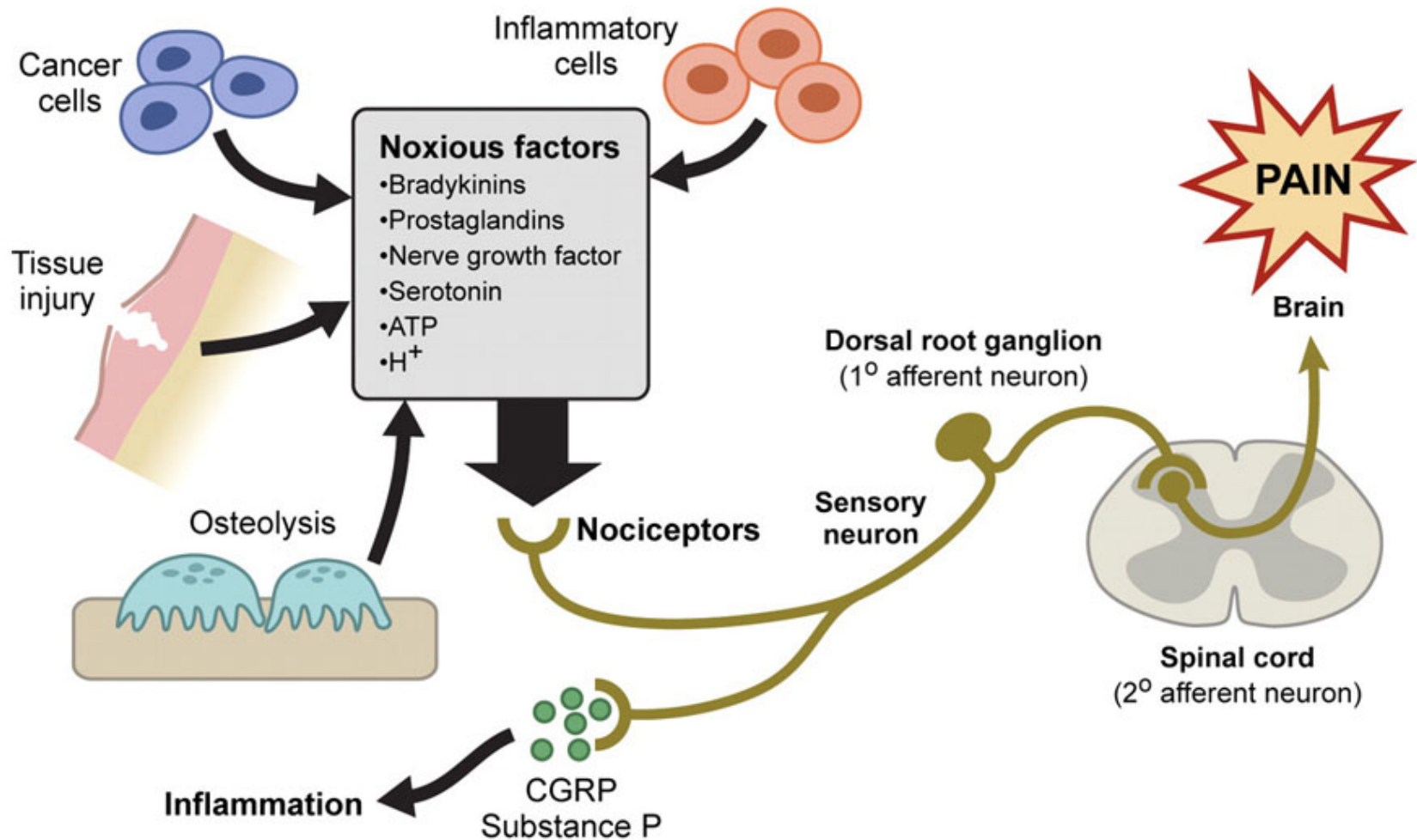
From Peterson SE, Selvaggi KJ, Fowler B, Blinderman CD. Pain Management and Antiemetic Therapy in Hematologic Disorders. Hoffman's Hematology, 5th edition, 2017

# Bone Pain

- Involvement of the skeleton occurs in up to 96% of ECD patients.
  - Affected bones are frequently the femur, tibia and fibula and less frequently the ulna, radius and humerus.
- However, bone pain occurs in only 50% of the cases.
  - Bone pain usually manifests around the knees and ankles.

# Cause of Pain

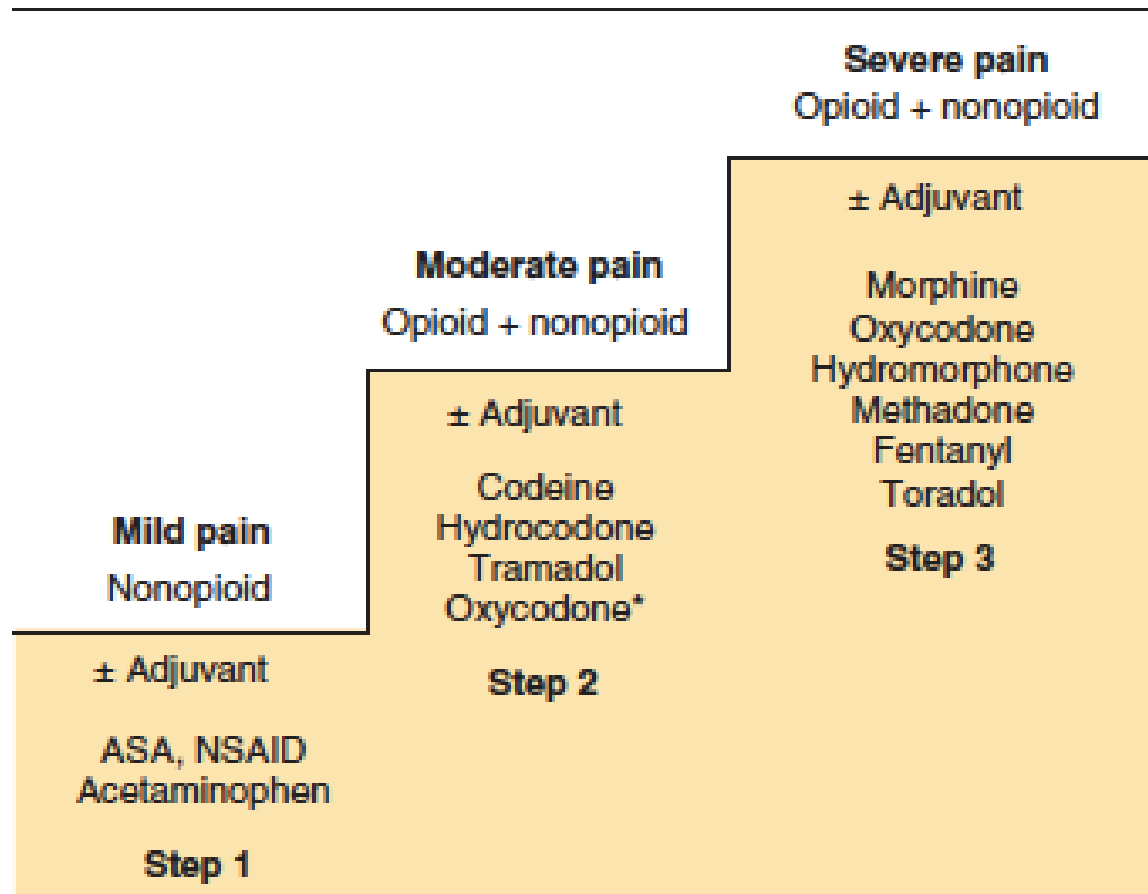
- Bone expansion from histiocytosis
- Osteosclerosis
- Lytic lesions
  - 30% of ECD cases exhibit osteolytic lesion involvement Oweity T, et al. J Neurosurg 2002, 96:344–351.
- Bone infarct or necrosis





# **PAIN MANAGEMENT**

## WHO ANALGESIC LADDER



From Peterson SE, Selvaggi KJ, Fowler B, Blinderman CD. Pain Management and Antiemetic Therapy in Hematologic Disorders. Hoffman's Hematology, 5th edition, 2017

# Pharmacological Management for Hematologic Bone Pain Syndromes

- Adjuvants for bone pain:
  - NSAIDs
  - Corticosteroids
  - Bisphosphonates (e.g. pamidronate and zoledronate)
    - For MM and hematologic malignancies with painful bone lesions
  - Denosumab (Xgeva)
    - RANKL inhibitor-- Receptor Activator of nuclear factor K-B ligand
    - decreases osteoclast activity.
    - reduces bone fractures in cancer
    - Need Ca and Vit D supplementation
  - Radiopharmaceutical strontium chloride (89Sr) and Samarium 153-lexidronan
  - Miacalcin spray (evidence lacking)
- Opioids
  - for moderate to severe pain
- Cannabinoids (emerging evidence and experience)

Peterson SE, Selvaggi KJ, Fowler B, Blinderman CD. Pain Management and Antiemetic Therapy in Hematologic Disorders. Hoffman's Hematology, 5th edition, 2017

# Take Home Points

- Bone Pain is the primary cause of pain in EDC
- Understanding of bone pain pathophysiology in EDC is extrapolated from other underlying pathophysiology in hematologic and oncologic diseases
- A number of adjuvant pharmacological therapies used in hematologic-oncologic bone pain syndromes may have benefit
- Opioids should be considered when pain is moderate to severe and adjuvant medications have not proven to be effective
- Need for more clinical studies on pain management in hematologic diseases

# Questions