Obscure Diagnoses in a Paediatric Pain Clinic

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When you hear hoofbeats, think of horses, not zebras

- Dr. Theodore Woodward

But in a pain clinic, you might find a Zorse!

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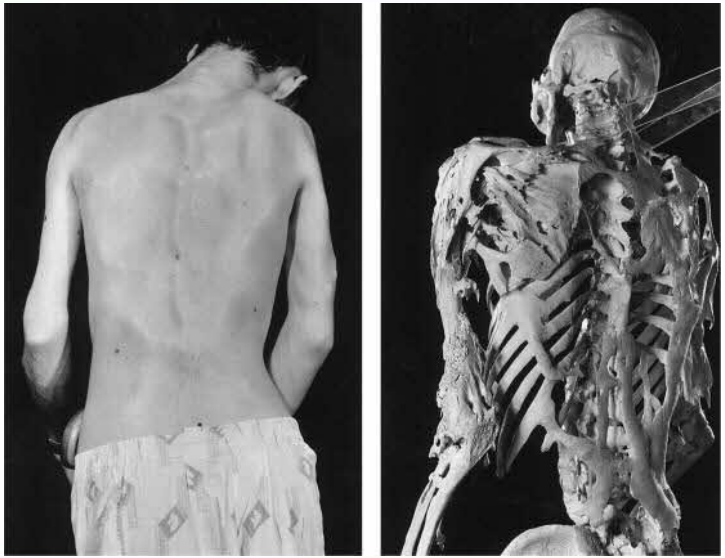
Stone Man Syndrome

Münchmeyer disease



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Fibrodysplasia Ossificans Progressiva

- An ultra-rare inheritable disorder of connective tissue characterized by congenital malformations of the great toes, and progressive heterotopic ossification (HO) in characteristic anatomic patterns (Kaplan et al., 2005; Pignolo et al., 2019).

- Heterotopic Ossification
- Skeletal muscles and soft connective tissues essentially transform into bone
- Progressive ankylosis (stiffness, limited movement, fusion)
- Neck, back, shoulders, elbows, hips knees, wrists, ankles, jaw

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Management

- Parents may be more knowledgeable about the disease than you
  - make yourself aware of the treatment recommendations
- Contact an IFOPA consultant eg Fred Kaplan
- Accelerated by trauma
  - intramuscular injections including immunisations, gastrointestinal endoscopy, airway manipulation, passive ROM exercises
  - avoid at all costs, treat empirically and as least invasively as possible.
- Flares are more common during the pubertal years
- Opioids are best avoided. NSAIDs are best avoided if epigastric pain is a symptom. Steroids can be used prophylactically or therapeutically.
- Warm hydrotherapy may be helpful
- Occupational Therapist involvement increasingly important as disease progresses

April 2021; August 2021 - updated pages 57-62, 64-68, 125; December 2021 - updated page 124; January 2022 - updated pages 64-69; May 2022 - updated pages 64-68

THE MEDICAL MANAGEMENT OF FIBRODYSPLASIA OSSIFICANS PROGRESSIVA: CURRENT TREATMENT CONSIDERATIONS

From The International Clinical Council on FOP (ICC) & Consultants:

Frederick S. Kaplan, Mona Al Mukaddam, Genevieve Baujat, Matthew Brown, Amanda Cali, Tae-Joon Cho, Corrie Crowe, Carmen L. De Cunto, Patricia Delai, Robert J. Diecidue, Maja Di Rocco, Elisabeth M.W. Eekhoff, Clive Friedman, Zvi Grunwald, Nobuhiko Haga, Edward C. Hsiao, Richard Keen, Joseph Kitterman, Charles Levy, Rolf Morhart, J. Coen Netelenbos, Christiaan Scott, Eileen M. Shore, Michael A. Zasloff, Keqin Zhang, Robert J. Pignolo

https://www.ifopa.org/for_medical_professionals

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Executive Summary Recommendations

- Activities: Activity is encouraged at all ages, but passive range of motion (motion performed by someone other than the patient) must be avoided. Singing, water exercises, and activities for respiratory health are encouraged. Avoid soft tissue injuries, contact sports, overstretching of soft tissues, muscle fatigue, biopsies, removal of heterotopic bone and all non-emergent surgical procedures.

- Anesthesia: An expert anesthesiologist experienced in general anesthesia for FOP patients must be consulted pre-operatively in all cases. If general anesthesia is required, an awake intubation by nasotracheal fiber-optic technique should be performed because of the neck malformations, jaw motion limitations, sensitive airway and risk of an obstructing neck flare. Highly-skilled FOP-aware anesthesiologists should be present for all elective intubations.

Executive Summary Recommendations

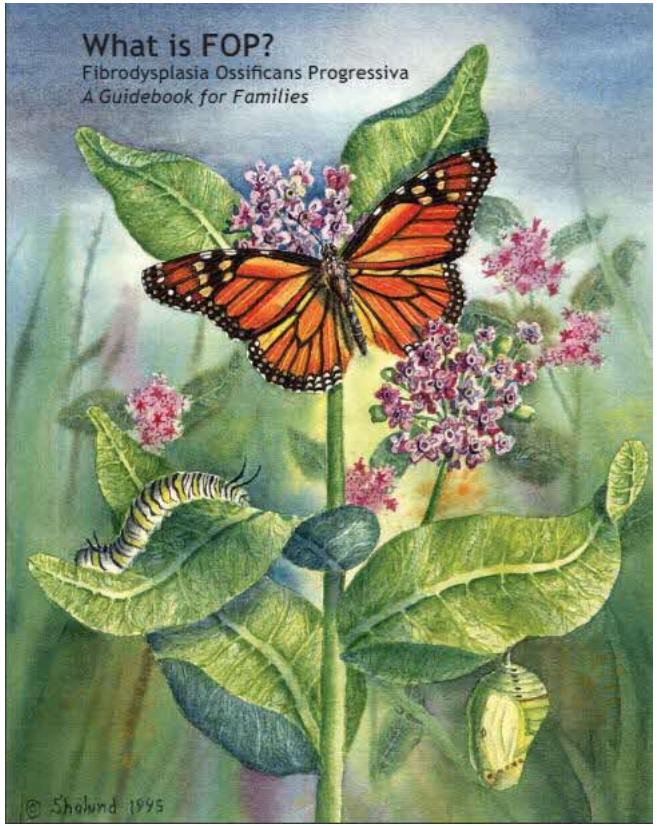
- Flare-up: (Back/chest): Consider non-steroidal anti-inflammatory medications or COX-2 inhibitors (oral or topical) with GI precautions. Use analgesics, muscle relaxants, and local applications of ice packs, as needed. Avoid narcotic analgesia.

- Flare-ups: (Limbs/throat/submandibular): Patients can present with inflammatory flare-ups with significant swelling and inflammation. These symptoms can be highly variable between patients and events. Prednisone – 2 mgs/kg once daily (up to 100 mgs daily) in AM (per oral) for four days (or equivalent corticosteroid); begin as early as possible after the onset of flare-up signs and symptoms. Keep prednisone on-hand as “pill-in-pocket” approach for emergencies. Alternatively, Pulse IV steroids may be used as directed. Avoid corticosteroids, if possible, for axial flares. Use oral and/or topical NSAID analgesics and/or muscle relaxants, as needed, with assiduous GI precautions. Local application of cool packs may also be helpful. Avoid narcotic analgesia whenever possible. FOP experts should be consulted with all submandibular flare-ups and the detailed guidelines should be assiduously followed.

- Flare-ups (Prophylaxis): Flare-ups often result from over-use and soft tissue injuries. Prednisone - 1-2 mgs/kg, (per oral) once daily for 3-4 days to prevent flare-up after severe soft-tissue injury. Do not use after minor bumps or bruises. Use prednisone prophylactically as directed for dental or surgical procedures.

Resources

- www.ifopa.org
  - The IFOPA is a nonprofit organization that provides hope to individuals with FOP and their families through education and support programs while funding research to find a cure and raising awareness for the rare genetic condition fibrodysplasia ossificans progressiva (FOP)
  - Expertise available in Japan, Seoul (Korea), Shanghai, Vellore (India)



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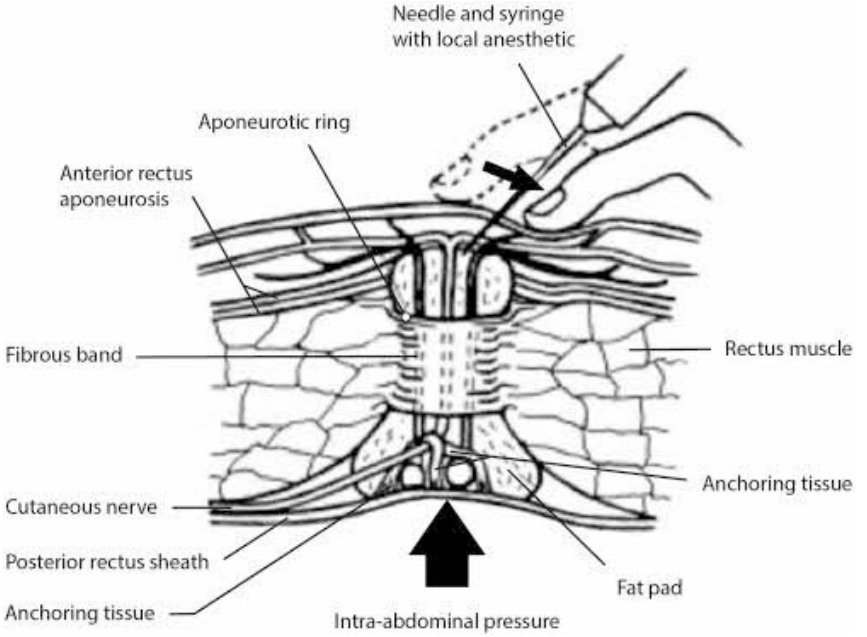
ACNE?

ACNE - Anterior Cutaneous Nerve Entrapment

- Entrapment of the cutaneous branches of the lower thoracoabdominal intercostal nerves at the lateral border of the rectus abdominis muscle, which causes severe, often refractory, chronic pain.
- Peripheral nerve entrapment occurs at anatomic sites where the nerve changes direction to enter a fibrous or osseofibrous tunnel or where the nerve passes over a fibrous or muscular band, mechanically inducing nerve irritation
- Muscle contraction at these sites may add additional insult by direct compression, although traction on the nerve from muscle activity also is likely.
- Mechanical irritation causes localized swelling that may injure the nerve directly or compromise the nerve's circulation.
- Tenderness of the main nerve trunk may be found proximal or distal to the affected portion (Valleix phenomenon).
- Proximal tenderness may result from vascular spasm or from unnatural traction on the nerve trunk against the point of entrapment.



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Anterior Cutaneous Nerve Entrapment

- Majority are female of young or middle age with a normal BMI
- Occurred spontaneously in either a sudden or gradual timeframe,
- Severe (NRS 6-8) abdominal pain
- Substantial delay in diagnosis

- Sensory disturbances at the painful abdominal area (78%)
- Positive pinch sign (78%), hypo/hyperaesthesia
- Positive Carnett's sign (87%)
  - Carnett's test is a simple clinical test in which abdominal tenderness is evaluated while the patient tenses the abdominal muscles.
  - It is useful for differentiating abdominal wall pain from intra-abdominal pain
- Positive response to a modified rectus sheath block (>50% pain reduction, 81%)

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Management

- In combination with systemic medication
- Lignocaine patch
- Trigger point injections (diagnostic and therapeutic)
- Ultrasound-guided blocks,
- Chemical neurolysis, and
- Surgical neurectomy,
- Radiofrequency ablation and neuromodulation

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Slipping Rib Syndrome vs Costochondritis

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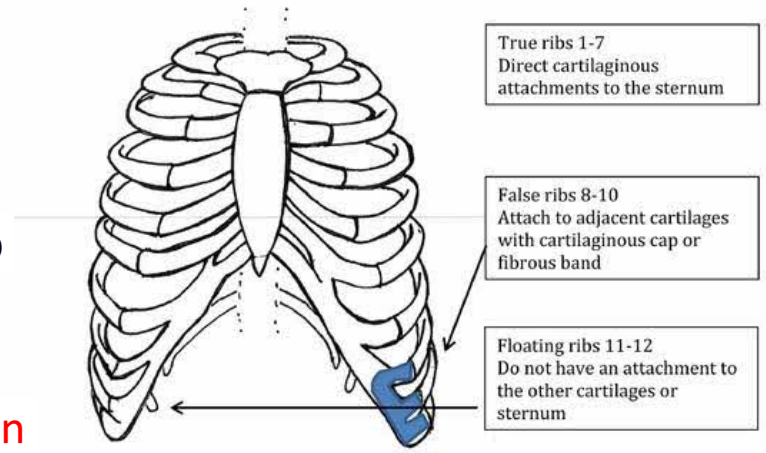
Slipping Rib Syndrome

- Caused by intercostal nerve impingement as a result of abnormal movement of false ribs (8-12) related to the medial fibrous attachments of the eighth, ninth, or tenth ribs being inadequate or ruptured or hypermobile
- Hypermobility of the anterior ends of the false rib costal cartilages then leads to slipping of the affected rib under the superior adjacent rib
- This slippage or movement can lead to an irritation of the intercostal nerve, strain of the intercostal muscles, sprain of the lower costal cartilage, or general inflammation in the affected area.



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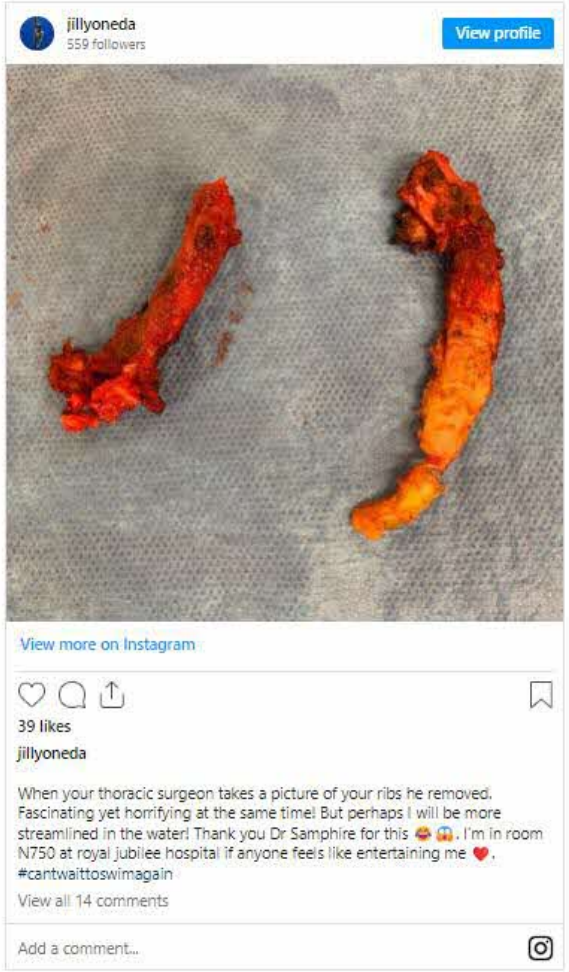
Slipping Rib Syndrome

1. Intense unilateral pain in the lower chest or upper abdomen, commonly left > right
2. May be associated with a popping or clicking sensation with activity that was associated with pain
3. Pain is often sharp in nature and can progress to a dull pain that can last for hours to weeks, may be also be described as burning
4. Exacerbated by certain movements such as stretching, twisting the trunk, bending, carrying heavy loads, or doing sports such as swimming, running
5. Reproducible pain on palpation of affected cartilage (rib), usually at the border of the rib
6. May exhibit chest wall asymmetry, or thinner rectus abominis on affected side
7. “Hooking” technique (Heinz and Zavala, 1977)
   - hooking the fingers under the costal margin and pulling the rib superiorly and anteriorly
   - Pain or clicking indicates positive test

Management of SRS

- Conservative
  - activity modifications
  - topical and oral analgesia with NSAIDs and/or opioids,
  - osteopathic manipulation (OMT)
  - intercostal nerve blocks (botulinum, LA)
- Surgical
  - costal cartilage excision (most common)
  - ± vertical rib plating
  - minimally invasive rib fixation



Differentials

	Costochondritis	Tietze
Age Group	Usually above 40yo	More common under 40yo
Gender predisposition	Females	
	Autoimmune conditions	Autoimmune conditions
Location of pain	2^{nd} to 5^{th} ribs	2^{nd} or 3^{rd}
Associated signs	No swelling	Swelling
Purported cause	Multiple	Microtrauma
Management	Rest, NSAIDs, Inj Steroids	Rest, NSAIDs, Inj Steroids

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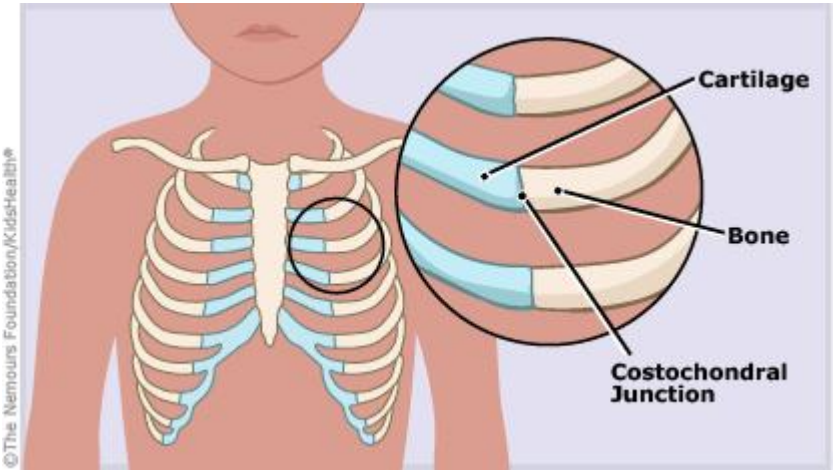
Costochondritis

- Tenderness on palpation of the chest wall makes the diagnosis
- Costosternal or costochondral joints can be affected
- Differential diagnoses include acute coronary syndromes, pancreatitis, pulmonary embolism, pericarditis, pleuritis and neoplasia
- Pathogenesis unclear
  - lower respiratory infection
  - chest wall trauma
  - extreme coughing, sneezing or laughter
  - Fibromyalgia
  - inflammatory joint disease
  - microtrauma from costovertebral dysfunction
- Self-limiting and benign



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Management of Costochondritis

- Local application of heat (heat pack)
- Nonsteroidal anti-inflammatory drugs (Oral, Topical)
- Lidocaine patches
- Capsaicin cream
- Physical therapy
- High-energy Flux Density Extracorporeal Shock-wave Therapy
- Acupuncture
- Recalcitrant cases - corticosteroid injections, surgery

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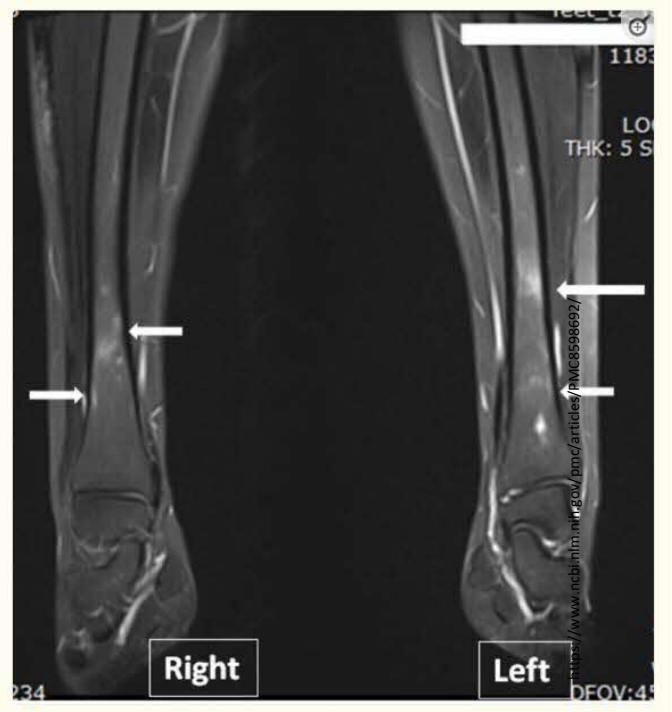
CRMO

Chronic Recurrent Multifocal Osteomyelitis

- Autoinflammatory disorder, commonly seen in peripubertal/adolescent females
  - Mean age at diagnosis is 11 years (range : 3 - 17)
- Characterised by involvement of metaphysis of long bones rather than axial involvement
- Diagnosed based on clinical, radiologic, pathological data
  - Dx of exclusion (exclude malignancy and infection)
  - Often exhibits periodic flairs and phases of remission
  - characterized by lytic, sclerotic, and hyperostotic lesions
  - MRI may demonstrate patchy abnormal marrow signals in metaphysis and/or periphyseal regions
- Most common sites are the long bone (femoral, tibial) metaphysis, though pelvis, spine, clavicle, and mandible may also be involved



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Management

- No cure – aim is to reduce inflammation
- First line (keystone) : NSAIDs
- First line (in vertebral involvement) : Bisphosphonates
- Second-line :
  - Bisphosphonate
  - anti-TNF agents
- Rescue :
  - Colchicine
- Physiotherapy : Hydrotherapy, TENS
- Surgery : vertebral lesions

Any questions?

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