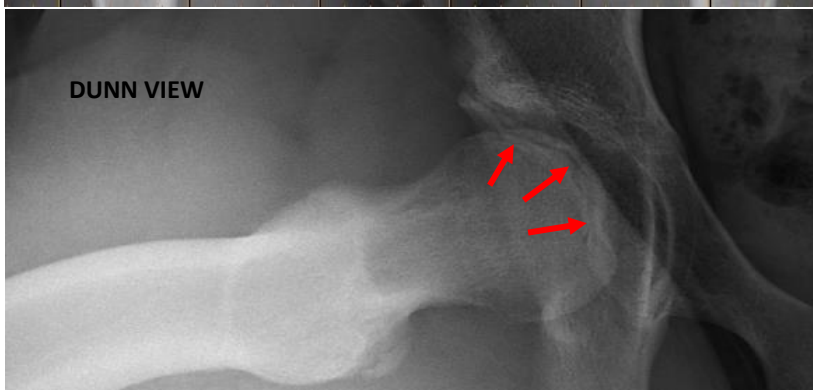
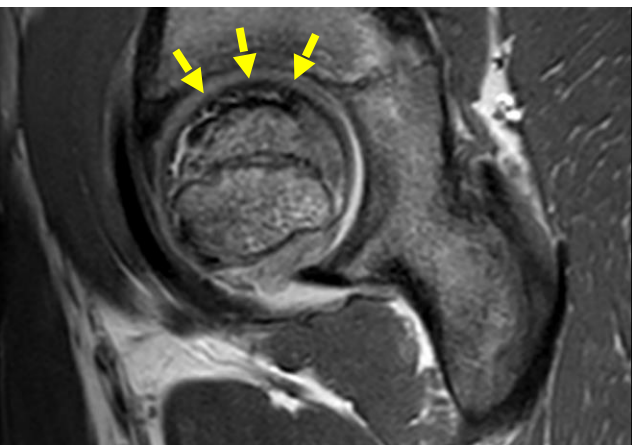
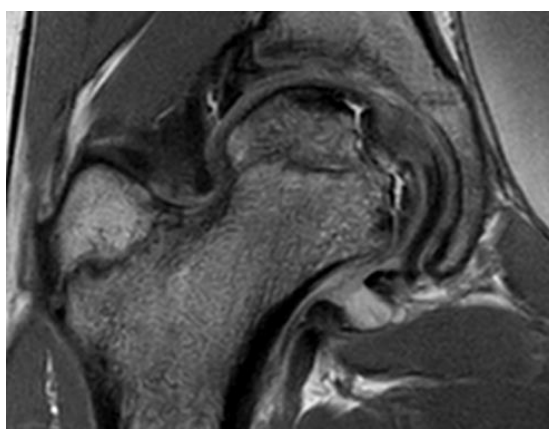
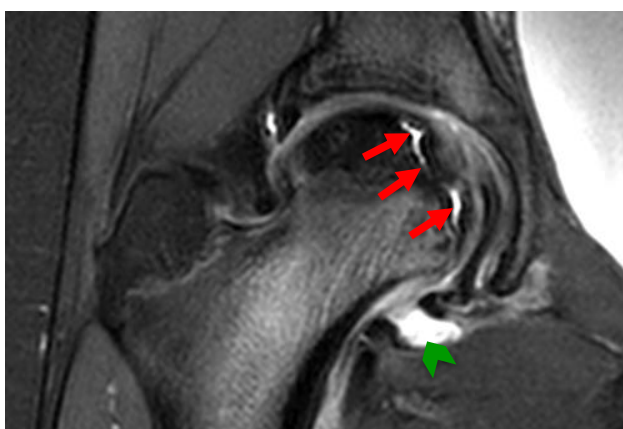


13 yo F. Dancer. 5 week history of right hip and groin pain with limp.



X-ray Findings:

- Relatively preserved femoral epiphyseal size
- Subtle subchondral lucency ('crescent sign' – red arrows) and subchondral sclerosis (immediately to the right of the red arrows). Both best seen on the Dunn view.
- As disease progress, x-rays may show
 - Early: May be occult
 - Established: Reduction in epiphysis size, subchondral lucency and sclerosis, fragmentation
 - Late: Flattening or asymmetric, circumferential enlargement of the femoral head (coxa magna)



MRI Findings:

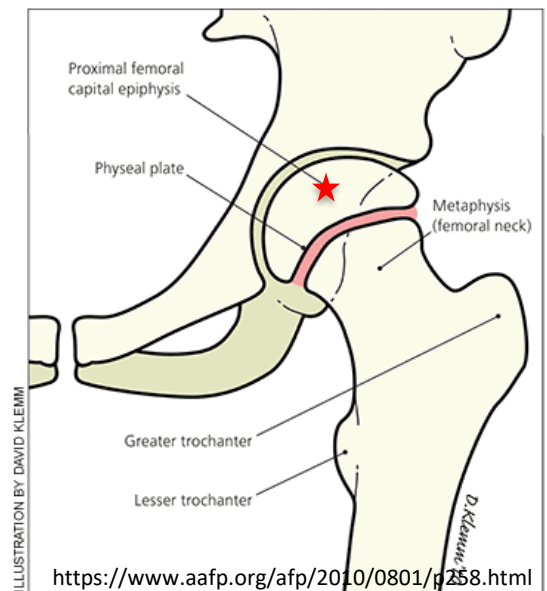
- Annotated T2 SPAIR (left above) and PD (right above) coronal sequences and PD (left) sagittal images demonstrate
 - Fragmentation of the femoral epiphysis with an undermining fluid signal cleft (red arrows)
 - Sclerosis (black / very low signal) of the femoral epiphysis with early flattening (yellow dashed arrows)
 - Small hip joint effusion (green arrowhead)

Annotated image (right) shows the anatomy of the proximal femur.

The epiphysis (affected region in Perthes) is indicated by a red star.

Discussion

- Overview
 - Idiopathic osteonecrosis of the upper femoral epiphysis (other causes of osteonecrosis should be excluded)
- Epidemiology
 - 5-15 per 100,000
 - Boys are 5x more likely to be affected than girls
 - Typically younger age than slipped upper femoral epiphysis. Peak presentation 4-8 years, range 2-14 years.
 - 15% bilateral
- Risk Factors
 - Positive family history
 - Low birth weight
 - Abnormal birth presentation
 - Second hand smoke
 - Asian, Inuit, and Central European decent
- Classification
 - Many different classification systems based on temporal evolution, severity / prognosis and healed stage deformity
- Aetiology
 - Idiopathic and controversial
 - Up to 75% of patients have some form of coagulopathy
 - Proposed mechanism of repeated subclinical trauma and mechanical overload
- Pathogenesis
 - Osteonecrosis occurs secondary to disruption of blood supply to femoral head
 - This is followed by revascularisation with subsequent bone resorption and later collapse
 - Creeping substitution (slow resorption of dead bone and replacement by new living bone made by osteoblasts) provides a pathway for remodelling after collapse
- Clinical
 - Classically atraumatic hip pain or limp
 - May have a coincidental trauma
- Differential diagnosis
 - Other causes of osteonecrosis should be excluded (sickle cell disease, leukaemia, corticosteroid administration, Gaucher disease)
- Management
 - Depends on severity (see also prognosis)
 - Non-operative: Clinical and radiographic follow up, activity restriction and protected/non-weightbearing, physical therapy to restore range of motion
 - Operative:
 - Femoral and/or pelvic osteotomy to correct more severe malalignment
 - Later in life, hip replacement may be required
- Prognosis
 - Influenced by age at time of presentation (more favourable prognosis in younger)
 - For the same age of presentation, prognosis is better in males due to less skeletal maturity
 - Percentage of femoral head involvement and degree of primary deformity of the femoral head (over 50% femoral head necrosis, consider operative management)



References & Further Reading:

1. El-Feky M, Gaillard F. Perthes disease. <https://radiopaedia.org/articles/perthes-disease>
2. Jin TY, Knipe H. Coxa magna. <https://radiopaedia.org/articles/coxa-magna>
3. Souder C. Legg-Calve-Perthes Disease. <https://www.orthobullets.com/pediatrics/4119/legg-calve-perthes-disease>