Legg Calvé Perthes Disease

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Legg Calve Perthes Disease

- Idiopathic avascular insult(s) to developing femoral head
- Age range 4 to 8 years commonly, has been reported 2 to 15 years
- M:F = 5:1
- Bilateral 10%
- Not familial
Demographics

- Lower SES
- Later born children
- Higher parental age
- Skeletal maturation delay ~ 2 years
- Rare in Blacks, Native Americans, Aborigines
Anthropometry

- Smaller than normal in all dimensions other than head circumference

- Indicates definite growth disturbance
Perthes

- Aetiology
- Radiology
- Classification
- Clinical features
- Treatment
- Outcome
Aetiology

- Unknown

- Excluded:
  - Infectious agent
  - Trauma alone
  - Hypothyroidism
  - Transient synovitis (only 1 to 3%)
Aetiology

Current theories

- Vascular embarrassment
- Multiple infarction
- Susceptible child
- Venous hypertension
- Protein C and Protein S deficiency, hypofibrinolysis leading to a tendency to thrombosis - present in 70% of patients with Perthes and 1:15000 population
Aetiology – Unifying Hypothesis

‣ “Lack of thrombolysis in the venous drainage of the femoral neck increases pressure in the femoral head circulation, resulting in AVN. Antecedent trauma might precipitate the avascular event”

Tachdjian
Histology

- Very few specimens
- Thickened cartilage over head
- Hypercellularity of epiphyseal cartilage
- Fibrocartilage
- Proliferative tongues of cartilage in metaphysis
- Similar changes in unaffected hip
Perthes

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Radiology

- All agree on stages of evolution
- Waldenstrom
  - Initial
  - Fragmentation
  - Reossification
  - Healed

2 to 5 years to pass through the stages
Initial

- Negative radiograph
- Joint space widening
  - (continued chondral growth)
- Smaller, more dense epiphysis
- Crescent sign
Initial

Perthes
Fragmentation

- Increased and decreased density
- Alteration of femoral head shape
- Classify at this time
- Treatment effective at this time
Fragmentation  

Crescent
Reossification

- Healing occurring
- Deformity can be readily appreciated

Healed

- Normal bone density
- Can assign Stulberg classification
Residual Deformity

- Coxa magna, Coxa breva
- Central arrest
- Lateral arrest
- Flattening
- U – deformity
  - Hinge abduction
- Osteochondritis dissecans
Classification

- Confusing
- Meant to be of prognostic value

- Catterall
- Salter and Thompson
- Herring
  - Lateral Pillar
Catterall

1. Anterior head involvement only
2. Anterolateral, sequestrum, lateral column in tact
3. 3/4 of head involved, including lateral column
4. Total head involvement

90% 1&2 good, 90% 3&4 bad
Head at Risk Signs

- Gages sign (radiolucent V in lateral epiphysis)
- Calcification lateral to epiphysis
- Lateral subluxation
- Horizontal epiphysis
- Metaphyseal erosions
- Loss of ROM
Salter and Thompson

- Groups A and B
- More or less than half head involvement, involvement of lateral pillar
- Crescent sign predictive of % involvement
Herring Lateral Pillar Classification

- A  Not involved
- B  Height > 50%
- C  Height < 50%

- Best interobserver reliability
- Prognostic
- Base treatment on this classification
Modified Lateral Pillar Classification

- **A** Not involved
- **B** Height > 50%
- **B/C border**
- **C** Height < 50%

Narrow lateral column
Poorly ossified lateral column
Exactly 50%

Herring 2004

k 0.81
Modified Lateral Pillar Classification

- A very narrow lateral pillar (2-3mm) >50% original height
- Lateral pillar with very little ossification with at least 50% of original height
- Lateral pillar with 50% of original height that is depressed relative to the central pillar
Lateral Pillar
Age

- Most predictive prognostic factor in all studies

- Related by most to remodelling potential
  - In DDH the head remodels the acetabulum
  - In Perthes we expect the acetabulum to remodel the head
Stulberg Classification

I  Normal hip
II Spherical Congruity, Coxa magna
III Ovoid head, Ovoid Acetabulum
IV Flat head, Flat Acetabulum
V  Flat head, Round Acetabulum
Modified Stulberg Classification

- **2** Round fitting within 2mm of a circle on 2 views
- **3** Out of round by >2mm on either view
- **4** At least 1cm of flattening of weight bearing surface
Perthes

- Aetiology
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Clinical Features

- Limp
- Marked pain not a feature
- Mild groin pain or anteromedial thigh or knee
- Usually activity related, relieved by rest
- History of trauma \( \approx 20\% \)
Clinical Features

- Limited ROM
  - Internal rotation
  - Abduction
  - Fixed flexion present
- Positive Trendelenburg
- Limb length inequality
wasting
Examination - abduction
Radiology

- AP (? Standing)
- Frog laterals
- Bone Scan
  - Diagnostic
  - Not prognostic
MRI

- Sensitive
- Specific
- Role undetermined
- Gadolinium
Differential Diagnosis

- Primary bone dysplasia (bilateral)
- DDH sequelae
- Sickle cell, Gauchers
- Traumatic AVN
- Transient Synovitis
- Sepsis
Initial Treatment

- Rest +/- traction to improve ROM
- NSAID
- No therapy effective in absence of good ROM
- Can’t get ROM
  - EUA
  - Adductor tenotomy
  - Petrie cast
Treatment

◆ Possibilities:
  – Hattage
  – Continued ROM
  – Crutches
  – Petrie casts
  – Bracing
  – Femoral osteotomy
  – Pelvic osteotomy
Hattage

- Wave your hat over the patient
- Observe and take radiographs
- Deal with sequelae as they occur
- Therapeutic Nihilism
Continued ROM

- Readmit the child for bed rest if lose ROM
- Restrict impact

Weight Relief (Crutches, Caliper)

- Never been shown to be beneficial
- Compliance ?
Arthrogram
Non - Operative Containment

- Petrie casts
- Abduction orthoses

- Recent publications show this is ineffective in changing natural history
Operative Containment

- **Varus osteotomy**
  - May stay varus in older child
  - Adds to LLD
  - Changed mechanical axis
Operative Containment

- **Salter osteotomy**
  - ?Enough coverage
  - Restores some leg length
  - Possible ↑ pressure
Shelf procedure

- Increases the acetabulum to cover the enlarged femoral head

- Lower morbidity than Salter or Femoral osteotomies
Chung - Varus extension
Multicenter Study Recommendations

- Herring et al JBJS 2004;86-A:2121-34
MULTICENTER STUDY RECOMMENDATIONS

- No difference between no treatment, bracing, ROM therapy
- No difference between femoral & innominate osteotomy
- Treatment in onset <8y (ca) & 6 (sa) no difference
Multicenter Study Recommendations

- In B & B/C border >8y (6y sa) surgery significantly better than non-operative
- In B <8y no difference btw op & non-op
- C worse with no difference btw op & non-op

- Lateral pillar & age are strong prognostic factors
- Females significantly worse than males if onset >8y
Treatment Summary

- Pain relief
- Restore ROM
- Leave or Treat

Arthrogram

ROM

Operate

Pelvis

Femur
Late Treatment

- Osteochondritis dissecans (3%)
  - Loose body removal
- Hinge abduction and pain
  - Cheilectomy
  - Valgus / extension osteotomy
- Trendelenburg gait
  - Trochanteric transfer
Final Salvage

- Arthrodiatasis
- Arthrodesis
Natural History

- 20 to 40 years, 70% to 90% pain free
- 93% Iowa hip scores > 80
- 50 year follow up (Weinstein)
- 40% Iowa hip scores > 80
- About half have had arthroplasties
Summary

- Aetiology
- Classification
- Head at risk signs
- Presentation
- Treatment
  - Non-operative
  - Operative
- Outcome