Legg Clave Perthes Disease

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Original description?

Condition possibly first described by an Austrian surgeon / physician: Prof Maydl in 1897
Waldenström - 1909

Johan Henning Waldenström
(1877-1972)

? Early tuberculosis
Legg – Calve – Perthes 1910

- American Arthur Legg (1874-1938) - 6 cases (February)
- Frenchman Jacques Calve (1875-1954) - 10 cases (July)
- German Georg Perthes (1869-1927) - 6 cases, 1 bilateral (October)
Observations:

- Limitations in abduction and hip rotation
- Acetabular and trochanteric overgrowth not uncommon
- Femoral head lateralisation in severe cases
- Stressed that immobilisation of the hip to be avoided

- Self-limiting non-inflammatory condition
- Degeneration and regeneration
Waldenström - 1923

Described the four stages:

- **Necrotic/Initial** - **Fragmentation/Resorption** - **Re-ossification/Healing** - **Remodelling/Residual**

Johan Henning Waldenström
(1877-1972)
What is Perthes?

- It is a self-limiting non-inflammatory idiopathic disorder of the femoral head leading to varying degree of necrosis in the growing child

- Synonyms:
  - Coxa Plana
  - Pseudocoxalgia (Calve)
  - Arthritis deformans juvenilis (Perthes)
  - Osteochondosis / Ostoechondritis of the hip
Anatomy

- Perthes pathologist in 1914 related to a vascular insult and described the femoral head vascular supply.
Epidemiology

- Most common age 4-9 years
- Boys : Girls   4 : 1
- Bilateral in 10% *(usually staggered)*
- Familial 8-12% *(Wansbrough et al. JBJS 1959, 41-A: 135-146)*

- Incidence: vary according to geography:
  - 3.8/100,000 Korea
  - 14.8/100,000 South India-Rural *(Joseph B et al Int J Epidemiol:1988 17(3): 603-7)*

? Relation to low socioeconomic class / poverty
The predisposed child

- Delayed bone age
- Retarded growth soon after diagnosis with later catch up growth
- Smaller size at diagnosis
  - (Wynne-Davies et al JBJS 1978;60B:6-14)
- Small hands and feet
  - (Hall et al 1988;70B:611-613)
- 30% have attention deficit hyperactivity disorder
  - (Loder et al JPO 1993;13:598-601)
Aetiology... The unanswered question!

- Multiple theories!
- Insufficient blood flow / Ischemic episodes
  - Arterial? Venous? Both?
- Fibrinolytic disorders
- Trauma
- Hyperactivity
- Genetic component / Environmental influences / Sequelae to synovitis?
Arterial Factors

- Repeated ischaemic episodes?

- Angiography
  - Definite obstruction to superior capsular arteries
    (Theron, Radiology 1980;135:81-92)
  - Decreased flow in medial circumflex artery
  - Intracapsular ring found to be incomplete
Venous Factors

- Hydrostatic Pressure theory
- Abnormal venous drainage
  - Increased intraossseous femoral metaphyseal pressure and congestion
- Disrupted venous drainage

(Heikkenen Acta Orthop Scand 1980;51:501-503)
Hyperviscosity

- First described in 1981 by Kleinman and Bleck (JPO)
- Further studies showed that Hypofibrinolysis in Perthes patients led to abnormalities in venous thrombus formation (Glueck 1994)

- 23/44 Perthes patients had thrombophilia
  - 19 protein C deficiency
  - 4 protein S deficiency
  - 7 high lipoprotein
  - Incidence of protein C or protein S deficiency in the population at large is 1/15000

Glueck et al JBJS 1996; 78A:3-13
Perthes and thrombophilia

- Further studies continued to investigate this

- 1999: 2 papers in JBJS Br - data did not confirm an aetiological role

- An association felt to be present in patients with
  - Factor-V Leiden mutation
  - Anticardiolipin antibodies

  Balasa, Glueck et al JBJS Am 2004; 86-A(12): 2642-7
Trauma

- Frequently proposed but no evidence
- First suggested by Legg 1910
- Lateral epiphyseal artery may be vulnerable to trauma
  Chung JBJS 1976;58A:961-970
- All parents recall a fall or sporting event that subsequently drew attention to their child’s hip
- Radiographs show a disease process that is not temporally related to the traumatic event
Hyperactivity ADHD

- 33% incidence of ADHD in Perthes cohort (24) looking at behavioural characteristics.

- Versus 3-5% ADHD in general population

  Loder et al. JPO 1993;13:598-601
Genetic Component

- Familial association described
- X-linked recessive inheritance?
Environmental influences

- Lower socioeconomic status
- Related to poor nutrition?
- Smoking / passive smoking
Transient Synovitis

- Often the first symptom of Perthes disease
- Is tamponade 2° to synovitis responsible for ischaemia?
- However...
- Only 17 of 455 patients with synovitis developed Perthes

Pathogenesis – Waldenstrom’s stages

Radiographic staging
Pathogenesis - Waldenstrom’s stages

- Necrotic/ Initial
  - Necrosis
  - Crushing of trabeculae - microfractures
  - Degeneration of the basal layer of cartilage
  - Thickening of peripheral cartilaginous cap
  - Sclerotic femoral head
  - Shape of head is maintained
Pathogenesis - Waldenstrom’s stages

- **Fragmentation / Resorption**
  - Damage is done
  - Invasion of vascular connective tissue
  - Resorption of necrotic bone by osteoclasts
  - Repair leads to appearance of lateral fragmentation
  - Loss of epiphyseal height due to:
    - Collapse of trabeculae
    - Resorption of bone
Pathogenesis - Waldenstrom’s stages

- Reossification / Healing
  - Alteration in femoral head shape
  - Creeping substitution
  - Apposition of viable bone in dead trabeculae
  - Normalisation of density
Pathogenesis - Waldenstrom’s stages

- Remodelling / Residual
  - Replacement of biologically plastic bone
  - Change in shape of head – rugby ball shape

- Successful treatment:
  - Femoral head congruity

- Unsuccessful:
  - Femoral head incongruity – subluxation and deformation
Clinical Presentation

- Limping child
- Pain in groin or referred pain to knee
- Restriction in ROM (head shape):
  - Limited abduction
  - Limited Internal Rotation
  - In flexion – abduction and external rotation
Clinical Presentation

- Antalgic / Trendelenberg +ve gait
- Muscle wasting
- Limb shortening
Differential diagnosis

Unilateral
- Transient synovitis
- Septic arthritis
- JIA
- Blood disorders
  - Sickle cell / Thalassaemia / Leukaemia / Lymphoma / ITP / Haemophilia

Bilateral
- Hypothyroidism
- Systemic steroids
- Multiple Epiphyseal Dyplasia (MED)
- Spondyloepiphyseal Dysplasia (SED)
- Gaucher’s disease
Investigations

- **Bloods**
  - FBC, ESR, CRP, Blood Culture

- **Radiographs (AP/Frog Lateral)**

- **Ultrasound Scan**

- **Bone Scan**
  - Decreased bone scan uptake before radiographic changes
Investigations

- **MRI** *(Henderson et al. JPO 1990; 10: 287-297)*
  - Earlier diagnosis than plain radiography
  - More information regarding extent of necrosis than bone scanning

- **Arthrography**
Catterall’s Head-at-Risk Signs

- Lateral Subluxation
- Calcification lateral to the epiphysis
- Gage’s sign: V-shaped defect laterally
- Metaphyseal cysts
- Horizontal growth plate

Anthony Catterall
RNOH
Lateral subluxation and calcification of lateral epiphysis

Metaphyseal cyst formation

Gage sign
Classification Systems

- Walderstrom: Pathological process
- Catterall
- Salter & Thompson
- Herring
Catterall’s Classification

- Based on amount of femoral head involvement on AP and lateral X-ray
- Determined during the fragmentation phase
- May change during disease process
No metaphyseal Reaction
No sequestrum
No subchondral fracture line

Central anterior head involvement
Sequestrum present - junction clear
Metaphyseal reaction - antero lateral
Subchondral fracture line - anterior half

>25% head involvement – medial and lateral column intact
Sequestrum large - junction sclerotic
Metaphyseal reaction - diffuse - antero lateral area
Subchondral fracture line - posterior half

75% head involvement – intact medial column
Metaphyseal reaction - central or diffuse
Posterior remodelling

Whole head involvement
Based on the extent of the subchondral fractures
This correlates with eventual extent of resorption

Advantage: predicts what is going to happen to femoral head

Group A: <50% head involvement and intact lateral pillar
  - Good prognosis
Group B: >50% head involvement and involved lateral pillar
  - Bad prognosis
Antero-posterior

Lateral
Herring Lateral Pillar Classification

JPO 1992

Group A  Group B  Group C
Herring Group A
Herring Group B
Herring Group C
What can we do about it?
Principles of management

1. Maintain and obtain stable free ROM
2. Reduce forces through hip joint (JRF)
3. Prevent and correct head subluxation
4. Early union of subchondral fractures
Treatment

- Early Disease:
  - While head is elastic

- Late Disease:
  - When head is plastic
‘Stages of degeneration and regeneration’ Perthes 1913

- Can we stop degeneration?
  - We don’t know - ?Bisphosphonates

- Can we speed up regeneration?
  - We don’t know

- Can we influence femoral head deformation
  - We think so!
Treatment of early disease

- **Conservative (supervised neglect):**
  - NSAIDS, analgesia, physio to maintain ROM, crutches (NWB), activity limitation, admission and traction for acute exacerbations

- **Containment:**
  - Bracing – controversial!
    - Parker – Broomstick cast in 1929
    - Eyre-Brook introduced traction in bed for >18-24 months
Early treatment concepts

- Bracing was based on TB treatment in 1940's and 1950's

Prolonged recumbency in many centres for up to 5 years

1966 - Harrison & Menon reported results of Broomstick Plaster (Petrie Cast)
Operative Containment (early)

- Theory: development of congruent joint dependant on maximal contact between immature femoral head and acetabulum

1962 - Salter osteotomy first used for Perthes
  - Increase pressure on femoral head and ‘contains’ it
  - Triple innominate – aggressive but better containments

1965 - Varus femoral osteotomy
  - Shortens and gives prominent GT
  - Can add rotation and extension
Pre-requisite for Varus Osteotomy

- Epiphyseal plate not too steep
- No major leg shortening
- Congruency between the femoral head and the acetabulum
- Ability to contain the femoral head in the acetabulum in abduction and internal rotation
- Only slight restriction of abduction
How does a femoral varus osteotomy alter the natural evolution of Perthes’ disease?

- Sphericity of the femoral head found to be significantly better in children who had a femoral varus osteotomy.

- Benefit most evident when performed in the stage of necrosis or in the early stage of fragmentation.

Arthrography to determine treatment

Neutral  Containable  Hinging

Neutral  Containable  Hinging

Abduction
Salvage Procedures (late)

- Valgus osteotomy
  - Hinge abduction with an enlarged head (coxa magna) extruding laterally and impingement

- Greater Trochanter epiphysiodesis
  - Often relative overgrowth GT, femoral neck shortening, Trendelenburg gait

- Shelf pelvic osteotomy
  - Older child, prevent subluxation and increase coverage

- Chiari pelvic osteotomy
  - Older child, no remodelling, increase WB area
Salvage Procedures (late)

- Near skeletal maturity: *(risk of affecting femoral head vascularity)*
- Treatment of hip impingement with head deformity
  - Anterior head neck debridement +/- pelvic osteotomy
  - Surgical hip dislocation and trochanteric distalisation +/- pelvic osteotomy
Does containment work and what’s the best method?

- Both Catterall (1971) and Herring (1994) reiterated the difficulty in comparing any of the published data on Perthes and containment due to the extreme variability of regimes and treatment methods.

- Catterall Group 1 & 2 / Herring A and B - good prognosis

- Catterall Group 2 & 3 / Herring B/C and C - Bad prognosis
LCPD Study Group – Herring, Kim & Browne
JBJS(A) 2004; 86:2121-2134

- 28 institutions - each institution used preferred uniform method of treatment
- 337 patients (345 hips) **all over chronological age 6** at diagnosis
- Followed to maturity

- **Herring A**
  - All Ages
  - All do well without treatment (but rare)

- **Herring B**: Bone Age < 6(8)
  - Uniform outcome irrespective of type treatment

- **Herring B**: Bone Age > 6(8)
  - Surgery (Femoral/Salter) > Brace > No Treatment

- **Herring B/C**: Bone Age > 6(8)
  - Surgery (Femoral/Salter) > Brace > No Treatment

- **Herring C**: All Ages
  - Poor outcome irrespective of type of treatment
Treatment guidelines by Herring

- **Patient < 6**
  - No evidence any form of treatment will alter outcome
    - Treat symptoms

- **Patients 6-8**
  - Group A: symptomatic
  - Group B: containment
  - Group C: ? Effect of treatment inconclusive

- **Patients 9<**
  - Group A: symptomatic
  - Group B & C: stronger case for operative containment but...
Poor Prognostic factors

- > age 6
- Females: mature earlier
- Catterall at risk signs
- Obesity
- Adduction contracture
- Loss of hip motion
- Flexion with abduction
- Advanced stage
- Recurrent episodes of stiffness

Congruency more important than sphericity
- Stulberg
Future

- More basic science understanding of pathophysiology
- Role of bisphosphonate therapy
- Surgical dislocations and soft tissue releases for severe later stage Perthes
- Stem cell therapy?
Conclusion

- A lot left to be discovered and understood

- Although Perthes disease cannot be prevented, progress has been made regarding risk stratification and minimising its deleterious effects on the hip, primarily via operative treatments

- Current basic science research efforts hold out the promise of providing us with answers and providing patients with even better long-term results in the future
‘If you were not confused at the start of the lecture I’m sure you are confused now!’

Any Questions?