PEDIATRIC HIP DISORDERS

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Topics

- Developmental dysplasia of the hips (DDH)
- Legg-Calve-Perthes disease
- Slipped capital femoral epiphysis (SCFE)
Case 1

- 6-week-old female born term via C-section for breech presentation found to have a “click” on examination of the left hip

- Ultrasound shows:
Case 2

- 7-month-old male with family history of DDH, family believes “one leg is longer than the other”
- Examination and x-ray show:
Case 3

- 2-year-old girl, previously received physical therapy for developmental delay, hypotonia and excessive lordosis
- Examination showed:
Cases

- 3 different scenarios
  - ALL DDH!
DDH - Objectives

- Understand the pathoanatomy and natural history of DDH
- Be able to identify hip dysplasia on ultrasound and x-ray
- “Briefly” review current screening guidelines
- Understand treatment modalities and surgical indications for DDH in children
DDH - Definition

- DDH refers to a spectrum of hip abnormalities ranging from a shallow acetabulum (socket) with instability to teratologic dislocations

- Old: Congenital dysplasia/dislocation of the hips (CDH)
  - Implies a condition existed at birth or shortly after.

- New: Developmental dysplasia of the hips (DDH)
  - Broader term including congenital conditions and conditions that may have developed after the newborn period.
DDH - Terminology

- **Dysplastic or shallow acetabulum**
  - Can occur with or without instability (subluxation/dislocation)

- **Hip subluxation**
  - **Incomplete contact** between femoral head and acetabulum

- **Dislocatable hip**
  - “Barlow positive”

- **Dislocated hip**
  - Reducible – “Ortolani positive”
  - Irreducible – teratologic (usually associated with other abnormalities)
The hip joint develops around the 11\textsuperscript{th} week of gestation

- Acetabulum and labrum grow during intrauterine life
- Normal hip joint development requires adequate positioning of the femoral head $\rightarrow$ ensures appropriate formation of the acetabulum
- Cartilaginous and other soft tissue structures (muscles/ligaments) play a role in development and stability
DDH - Epidemiology

- 1-2% newborn babies
  - Refers entire spectrum of DDH
  - 0.1% with hip dislocation
- M:F = 1:6
- Left hip more common
  - Left occiput anterior most common presentation
  - Left hip adducted against mother’s lumbosacral spine
DDH - Epidemiology

- Race/ethnicity
  - Higher incidence in central Europeans, Native Americans
  - Low incidence in Africans, Asians

- Genetics
  - 6-7% if siblings
  - 12% if one parent
  - 36% if one parent and one sibling
DDH - Etiology

- Mechanical factors—
  Limited hip motion in utero
  - Primagravida (50-60% DDH)
  - Oligohydramnios
  - “crowding phenomenon”
    - Torticollis, metatarsus adductus
    - Congenital knee dislocation
DDH Etiology

Major mechanical risk factor?
Breech presentation

- Restricts hip and knee intra-uterine motion
- Up to 20% of breech newborns have DDH
DDH Natural History

1. Shallow acetabulum $\rightarrow$ increased forces on cartilage $\rightarrow$ early osteoarthritis

2. Multiple studies link DDH and OA
   - Wiberg 1939, CEA defined as predictor of OA
   - Followed 19 pts with CEA < 20, all developed OA
Newborn exam

- Birth history and family history
- Complete MSK exam
  - Torticollis, knee dislocations
  - Other abnormal joints (Larsen syndrome, arthrogryposis)
- Hip exam
Newborn exam

Ortolani $\rightarrow$ dislocated hip

Barlow $\rightarrow$ dislocatable hip
Diagnosis

- Older infant

Clinical Findings in CDH (more obvious with growth and weightbearing)

Limitation of abduction caused by shortened and contracted hip adductor muscles
Diagnosis

- Older child
  - Increased lumbar lordosis
  - Waddling or Trendelenburg gait
  - + Trendelenburg sign
Imaging of the newborn hip
Imaging of the newborn hip

Ultrasound
- Newborns, useful up to 4-6 months
- Highly sensitive
  - Can’t make bad hip look good
- Less specific, technique dependent
  - Can make good hip look bad
Newborn ultrasound

- Diagnostic tool
- Monitor treatment in brace
- Newborn screening
  - Ideally done around 4-6 weeks of age
  - ? Abnormal exam
  - + breech +/
  - first born female, family history
Interpreting the newborn US

- Coronal views
Interpreting the newborn US

Axial views

Stable hip

Unstable

Stress views
X-rays
X-ray landmarks

- Hilgenreiner’s (H) line
- Perkin’s (P) line
- Dysplasia suspected if metaphyseal beak lateral
X-rays
DDH Classifications

- **Tonnis**
  - Based on AP pelvis

- **IHDI**
  - Tonnis classification but using ossified metaphysis
- Acetabular index
  - Helpful for tracking dysplasia

<table>
<thead>
<tr>
<th>Age</th>
<th>AI</th>
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<tbody>
<tr>
<td>Birth</td>
<td>&lt;35</td>
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<tr>
<td>1 year</td>
<td>&lt;25</td>
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<td>2-3 years</td>
<td>&lt;20</td>
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<td>6-7 years</td>
<td>&lt;15</td>
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<tr>
<td>10 years</td>
<td>&lt;12</td>
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<tr>
<td>15 years</td>
<td>&lt;10</td>
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- **Shenton’s line** –

  - Most reliable after 6-7 yrs of age
- Older patients
  - CEA
  - 10 yr < 25
- Less reliable in young kids
  - Acetabular ossification
Treatment Overview

- By age (at presentation)
  - Birth to 6 months: Pavlik harness or abduction brace
  - 7 to 18 months: Closed reduction attempted
  - 18+ months: Open reduction
Newborn: Pavlik harness

- Ortolani + = reducible hip
- Femoral head locates & stabilizes
- Monitor with exam and ultrasound
Pavlik harness

- Anterior straps—flexion
- Posterior straps—limit adduction
- Outcome
  - Barlow + near 100% success
  - Ortolani + 80-90% success
- Complications
  - Nerve palsy: Femoral, Brachial plexus
  - AVN
Pavlik Harness Treatment

- Full time until hip stable: 6-8 weeks
- Nights only 4-6 weeks
- Continued follow-up to age 5 years
- Success rate: 90-96%
Infant DDH Treatment

- May substitute abduction orthosis
Older infants/toddlers (7-18 mo)

- Fixed dislocation
  - Limited abduction
  - +Galeazzi if unilateral
  - -Ortolani (irreducible when awake)

- X-rays:
  - Superolateral femoral head
  - Delayed ossification
  - Acetabular deficiency
Older infants/toddlers

- Age 7-18 mo
- Infants that failed Pavlik
- Closed reduction
Arthrogram
Adductor tenotomy
90° flex & <50° ABD
Closed reduction

- Assess stability
- 3D imaging: CT vs MRI
MRI post reduction

Can also use for perfusion assessment
- Casting period
  - Varies among surgeons
  - 12-18 weeks
- If unsuccessful proceed to open reduction
Older age at diagnosis

- Presentation > 18 mo
  - Limp/waddle
  - Limb length discrepancy

Treatment → Open reduction
Extra articular
- Psoas tendon
- Adductor musculature
- Long adductor and hamstrings
- Abductors and extensors short and adherent to capsule
Open reduction

- > 18 months
- Or younger if failed closed reduction
- Anterior approach
  - “bikini line” incision
    - Sartorius/tensor interval
Complications

- Redislocation

- AVN/growth disturbance
  - Can occur with treatment
  - Rates highly variable
  - Iatrogenic
    - Excess compression on femoral head
    - Damage to blood vessels during open reduction

- Role of ossification
A note on swaddling
Does swaddling lead to DDH?

- Studies from Turkey, Saudi Arabia, N.Americans
  - Traditional methods immobilizing hips/knees in extension

- JBJS 2012
- Wearable blankets
  - Sleep Sac
  - Mothercare
  - Grobag
  - Woombie
  - Snoozie
  - Sleep Bag
Screening the Newborn for Developmental Dysplasia of the Hip

Now What Do We Do?

Richard M. Schwend, MD,* Perry Schoenecker, MD,†** B. Stephens Richards, MD,‡
John M. Flynn, MD,§ and Michael Vitale, MD¶

To Screen or Not to Screen? A Decision Analysis of the Utility of Screening for Developmental Dysplasia of the Hip

By Susan E. Mahan, MD, MPH, Jeffrey N. Katz, MD, MS, and Young-Jo Kim, MD, PhD

Investigation performed at Children's Hospital Boston, Boston, Massachusetts
Recommendations differ from:

- U.S. Preventive Services Task Force
- Canadian Task Force on Preventative Care
- American Academy of Pediatrics
- American Institute of Ultrasound in Medicine
- American Academy of Orthopaedic Surgeons
Independent panel of physicians charged by Congress to review scientific evidence for clinical preventive services and develop evidenced-based recommendations for the health care community
"The literature evidence concerning screening is insufficient to recommend for or against this service."
CANADIAN TASK FORCE ON PREVENTATIVE CARE

- Serial clinical examinations to age 12 mos
- Supervised period of observation for newborns with diagnosis of DDH
• Serial clinical examinations of hips and record findings
• Imaging for female infants born in breech position (U/S at 6 weeks or xray at 4 mos)
• Optional imaging for breech boys or girls with family history
American Institute of Ultrasound in Medicine

- Female with breech presentation
- Family History in parent and sibling
- Abnormal findings on clinical exam
- Oligohydramnios
American Academy of Orthopaedic Surgeons

9 recommendations - 7 with limited evidence

- Moderate evidence
  - Against Universal Screening
  - For imaging infants with risk factors:
    - Breech
    - Family History
    - Clinical Instability

AAOS Guidelines 2014
Economics

- Ultrasound hips: $790
- Xray hips: $175

- 4 million babies born in US/year
- 4% breech = 160,000 babies
- 160,000 babies = 80,000 girls
- 80,000 ultrasounds = $63 million
- 4,000,000 ultrasounds = $3.1 billion
Screening infants for DDH

2006: USPSTF not enough evidence to support screening
MOST DDH occurs in infants with NO risk factors
Physical exam—
  At birth and throughout development
Ultrasound if family history or history of breech
No Screening:

- More children retain a preventable disability
- Increase in surgical treatment
- Potential higher liability for primary care doctors
SCFE
Case

- 14-year-old football player with a 4-month history of bilateral knee pain.
- Seen by PMD after 1 month, diagnosed with muscle strain. Now with increased pain, R>L.
- Pain over distal thighs
Physical Exam

- BMI=33
- Unable to ambulate
- Knee exam unremarkable
  - Symmetric, full ROM
  - No effusion
  - Normal ligamentous exam
  - No Meniscal signs
Physical Exam continued

- Hip ROM in prone position
  - Right hip
    - Internal rotation: lacking 15 degrees
    - External rotation: 45 degrees
  - Left hip
    - Internal rotation: 20 degrees
    - External rotation: 45
Radiographs
Radiographs
Diagnosis?
Slipped Capital Femoral Epiphysis (SCFE)

- Displaced proximal femur epiphysis relative to the metaphysis
- Results from shear stress on vulnerable physis in early adolescents
SCFE

- Incidence: 1/50,000
- Around age 13 in boys, 11 in girls
- Risk factors: Obesity!, endocrine disorders (hypothyroid, hypopit), metabolic disorders (rickets)

- Treatment:
  - in situ pinning
  - Osteotomy surgery to improve alignment?

- Natural history: risk of osteoarthritis increases with increasing severity of slip
PERTHES
Objectives

- Review historical treatment and radiographic classification
- Data on cast/brace therapy
- Bisphosphonates
- Other medications being studied
Legg-Calvé-Perthes

- Described in 1910

Arthur Legg  |  Jacques Calvé  |  Georg Perthes
Etiology theories

- Early theories
  - Infection, Inflammation
  - Trauma
  - Congenital

- Current theories
  - Vascular Insult

From Chung, JBJS 1976
Vascular Etiology

- Anatomy: Age/Sex
- 2 Infarction Theory
- Hyperviscosity
- Thrombophilia
Prognosis

- Catterall
- Herring’s Lateral Pillar Classification

Lovell and Winters, 2006
Outcome Classification

- **Stulberg**
  1. Normal hip
  2. Spherical head, congruent
  3. Aspherical oval head, congruent
  4. Flat head, congruent
  5. Flat head, incongruent

- **Mose Sphericity Scale**

Lovell and Winters, 2006
CASTING AND BRACING FOR LEGG-CALVÉ-PERTHES SYNDROME
Non-containment

- Early Treatment:
  - No weight-bearing
  - Long-term bedrest +/- traction

Katz, JBJS, 1967
Containment Treatment

- Petrie and Bitenc, 1971
  - Abduction casting
  - Required inpatient stays
Containment Treatment

Waldonstrom’s Radiographic stages

1. *Initial
2. *Fragmentation
3. Reossification
4. Healed
Abduction Orthoses

- Removable braces
  - Containment without hospitalization

Curtis et al. JBJS 1974
Abduction Orthoses

- Scottish Rite Atlanta
Abduction Orthoses

- Goal: contain femoral head until patients enter reossification phase
- Average wear: 6-18 mo
Efficacy of Brace Treatment

- Martinez, Weinstein, Dietz, JBJS, 1992
  - 31 patients: Caterall III or IV
  - Average age = 6 y
  - Treated with abduction orthosis >6 mo
  - Stulberg:
    - Class II—41%
    - Class III-VI—53%
    - Class V—6%
Efficacy of Brace Treatment

- Meehan et al, JBJS, 1992
  - 35 patients with LCPS Caterall III or IV or Salter-Thompson B
  - Average age = 7y 8mo
  - Treated with abduction orthosis >4 mo
  - Stulberg:
    - Class II—9%
    - Class III-VI—88%
    - Class V—3%
Non-operative Containment

- Aksoy et al JPO 2004
  - Tachdjian’s ischial-bearing brace
  - Brace vs. no brace: no difference in outcomes

- Kim et al JPO B, 2006
  - “New Pogo Stick” ischial weight-bearing brace
  - 20 patients → 17 Stulberg I-II
Efficacy of Brace Treatment

- Cooperman and Stulberg, CORR, 1986
  - 248 patients reviewed at 5 hospitals
    - Crutches
    - Scottish Rite type brace
    - Newington brace
    - Femoral varus osteotomy
  - Patients <6y did well
  - Patients over 8y with whole head involvement → Better with surgery
Efficacy of Brace Treatment

- Wang et al, CORR, 1995
  - 124 patients reviewed
    - Non-weightbearing
    - Scottish Rite type brace
    - Petrie casts
    - Femoral varus osteotomy
    - Salter osteotomy
  - Patients over 8y with whole head involvement seemed to do better with surgery
Efficacy of Bracing

Prospective study

- 438 patients (451 hips) at 5 centers
  - No treatment
  - Brace
  - ROM exercises
  - Femoral osteotomy
  - Innominate osteotomy
Psychosocial effects of bracing

- Price et al, JPO, 1988
- Found deficits in brace group in social, academic and sexual behavior
PHARMACOLOGIC AGENTS FOR THE TREATMENT OF LEGG-CALVÉ-PERTHES AND FEMORAL HEAD OSTEONECROSIS
Bisphosphonates: history

- Pyrophosphates
  - Body’s “water softener”

- Bisphosphonates
  - Low doses: inhibit bone resorption
  - High doses: inhibit calcification

Russell, Pediatrics, 2007
Bisphosphonates

- Inhibit osteoclast function
- Inhibit osteoclast formation
- May protect osteocytes and osteoblasts from steroid-induced apoptosis

Russell, Pediatrics supp, 2007
Bisphosphonate Research

- Kim et al, JBJS, 2002
- Tied ligatures around the femoral necks of piglets
- Found osteolytic changes, fragmentation, flattening of femoral heads
Bisphosphonates Research

- Injected labeled ibandronate into femoral head
- Epiphyseal quotient was greater in ibandronate group
- Ibandronate stayed in femoral head
RANK-L Inhibition

- Kim et al, J Bone Min Res, 2006
- Administered sub-Q labeled OPG to piglets with ligature induced osteonecrosis
- Osteoprotegrin (OPG) binds RANK-L and prevents its interaction with RANK
- OPG group had better preservation of femoral head sphericity and less osteolysis
Bisphosphonates Research

- Astrand, BMC Musculoskeletal Disorders, 2006
- Put bone grafts in bone chambers into the tibiae of rats
- Animals given systemic zoledronate had less resorption of the grafts and less resorption of newly formed bone
Bisphosphonates Research

- Little et al, J Ortho Res, 2005
- SHR rats
  - Weekly sub-Q Zoledronate
- Zoledronate group
  - bone volumes
  - increased sphericity
  - increased epiphyseal quotient
Bisphosphonates Research

- Lai et al, JBJS, 2005
- Randomized, prospective study
- 40 patients (54 hips) with non-traumatic femoral head osteonecrosis
  - Alendronate group: 4 surgeries
  - Control group: 21 surgeries
Prenylates GTP-binding protein which is needed for normal osteoclast production and f’n...
Statin Medications

- Pritchett et al, CORR, 2001
- Looked retrospectively at 284 patients taking statins and high-dose steroids.
- 3 patients (1%) developed osteonecrosis (as opposed to 3-20% in the literature)
- Conflicting results for studies on statins and fracture risk