Developmental Dysplasia of the Hip

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Developmental Dysplasia of the Hip

Comes in different flavors at different ages of kids

Common etiology: excessive laxity of the hip capsule with failure to maintain femoral head in the acetabulum
DDH: “The Spectrum”

- Normal Hip
- Dysplastic Hip: “shallow”
- Dislocatable Hip: “Barlow +”
- Reducible Hip: “Ortalani +”
- Fixed and Dislocated Hip

Good to Bad
What’s new with hip dysplasia?

To begin with, the name:

- **Was**: CDH (Congenital Dislocation/Dysplasia of the Hip)

- **Is**: DDH (Developmental Dysplasia of the Hip)
Why Change the Name to DDH?

1. As soon as we say “congenital” in a one-year old, we’re backpedaling

2. Evidence for true “developmental” dislocations:
   - No matter what the screening program, late dislocations occur
   - Documented late presentations after normal exam, X-ray, and now, even ultrasound
In 15 documented cases, subluxation or dislocation of the hip was discovered months or years after previous multiple normal physical examinations. The examiners were unique in that six were professors specializing in children’s orthopedics, four were board-certified orthopedists, and five were pediatricians. Delayed diagnosis of dislocation is not evidence that an inadequate physical examination was performed.

Clinical Orthopedics and Related Research 203:276, 1986
EXAMINATION: BOTH HIPS (INA (INFANT))

CLINICAL HISTORY: Possible hip dislocation.

FINDINGS: AP and frog leg views of the pelvis are compared side-to-side. Capital-femoral epiphyses have not yet appeared. There is no bony deformity or significant subluxation identified at this stage. The angle of the pelvis and amount of cartilaginous deposition is symmetric.

IMPRESSION: Capital-femoral epiphyses not yet developed. There is no obvious subluxation or bony deformity. If symptoms persist, follow-up study is advised.
Classification of Developmental Dysplasia of the Hip (DDH)

Typical

Teratologic
Teratologic Dislocations

Out in utero
Major changes in muscle, femoral head, acetabulum from birth
Associated with spina bifida, arthrogryposis, other conditions
Treatment is surgical (when indicated)
Typical Dislocations

Near normal in utero
Minor changes at birth
Muscle contractures develop secondarily
Major bone changes only after neglected a year
Incidence

Ethnic, cultural and geographic differences
- High: American Indians (188/1000 Manitoba), Caucasians
- Low: Chinese (Hong Kong 0.1/1000), African (Bantu 0%)

Dependent on definition
- Dislocation: 1/1,000
  - Female: 1/300 to 1/600
  - Males: 1/2,000 to 4,000
- Abnl newborn exam 1-2/100
- Abnormal ultrasound 8/100
Natural History

EARLY DIAGNOSIS AND TREATMENT OF CONGENITAL DISLOCATION OF THE HIP*

T. G. BARLOW, SALFORD, ENGLAND

From the Hope Hospital, Salford, Manchester

Barlow  JBJS 1962

9,289 newborns

139 w/ abnormal hip

(1.5%)

88 % normal in 2 mos
Natural History – Newborn

Barlow

- Around 1 in 100 infants have instability (positive Barlow)
- 60% stabilize in 1st week
- 88% stabilize in 2 months without treatment
- 12% become true dislocations and persist

Coleman (Navajo Children), *CORR 1968*

- 35 abnl hips <3 mn, 11/12 treated normalized
- 23 untreated, AI>40, lateralization of femoral beak
- 5 normalized, 9 dysplastic, 3 subluxed, and 6 dislocated at 3 years (78% abnl)

because not possible to predict outcome, all infants with instability should be treated
Long term Natural History

Subluxation predictably leads to degenerative joint disease and clinical disability

- mean age symptom onset 36.6 in females and 54 in men

Cooperman, *JBJS*

- 32 hips with CE angle < 20 without subluxation
- 22 years all had xray evidence of DJD
- no correlation between angle and rate of development
- concluded that radiologically apparent dysplasia leads to DJD but process takes decades
Etiology

Ligamentous laxity
Positioning
  • Prenatal
  • Postnatal
Genetics
Epidemiology

Risk Factors:
- First born
- Breech presentation
- Female
- Family History

Combined = 1:15
- Thus, heightened awareness is needed
- AAP now recommending routine imaging or referral
- However, it also means that 14/15 are normal
Etiology – Ligamentous Laxity

Female predilection
- Maternal hormones (relaxin crosses placenta)

Umbilical cord collagen
- Increased ratio of type III to I in pts with DDH

Autosomal dominant ligamentous laxity
Etiology - Prenatal Positioning

Breech

- 2% of births
- 17 to 23% of DDH
Etiology - Prenatal Positioning

Delivery Method and Breech Positioning

- Two studies have shown a lower incidence of DDH for babies delivered by C section vs vaginally in breech position

1. Lowry – JBJS Br 2005
   3.7% incidence with premature c-section
   6.6% incidence with term c-section
   8.1% incidence with vaginal delivery

2. Fox – JBJS Br 2010
   US screening in patients – Graf 3+4 hips
   1.1% incidence with term c-section
   4.7% incidence with vaginal delivery

P<0.02
Etiology - Prenatal Positioning

DDH increased w/ oligiohydramnios

DDH assoc w/ other “packaging” problems
  • Metatarsus adductus (1 to 10%)
  • Torticollis (14 to 20%)
Intrauterine position

- Most common is L. occiput anterior
  - L hip adducted against sacrum
  - L. hip most commonly affected
Etiology – Post Natal Positioning

- Extended hip positioning: Cradleboards
  - Increased incidence of DDH

- Hips wrapped in flexed position or carried Astride the hip
  - Decreased incidence of DDH
Etiology – Genetics

Ethnic differences
- Low: African and Asian
- Higher: Caucasain and Native American

Identical twins 34%, Fraternal twins 3%

Increased risk w/ family history
Hip Development – 8 weeks

Cleft forms in mesenchymal hip analge
Hip Development – 16 weeks

Shape of the acetabulum follows the development of the round femoral head
Hip Development – Post Natal

Birth

5 Months

2 years
History - Newborn

Birth order, position, weight

Family history
History - Infant / Child

Toewalking
Painless persistent limp
Limb length inequality
Waddling
Swayback
Examiner must be *gentle*

Baby *must* be quiet

Neck, feet and spine

### TABLE 1. Relative and Absolute Risks for Finding a Positive Examination Result at Newborn Screening by Using the Ortolani and Barlow Signs

<table>
<thead>
<tr>
<th>Newborn Characteristics</th>
<th>Relative Risk of a Positive Examination Result</th>
<th>Absolute Risk of a Positive Examination Result per 1000 Newborns With Risk Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>All newborns</td>
<td>...</td>
<td>11.5</td>
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<tr>
<td>Boys</td>
<td>1.0</td>
<td>4.1</td>
</tr>
<tr>
<td>Girls</td>
<td>4.6</td>
<td>19</td>
</tr>
<tr>
<td>Positive family history</td>
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<td></td>
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<tr>
<td>Boys</td>
<td>1.7</td>
<td>6.4</td>
</tr>
<tr>
<td>Girls</td>
<td></td>
<td>32</td>
</tr>
<tr>
<td>Breech presentation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Boys</td>
<td>7.0</td>
<td>29</td>
</tr>
<tr>
<td>Girls</td>
<td></td>
<td>133</td>
</tr>
</tbody>
</table>
Examination - Newborn

Normal
Instability w/o dislocation ("rubbery hip")
Dislocatable ("Barlow positive")
Reducible ("Ortolani positive")
Fixed and dislocated

By 3 mos hip reduced or dislocated to exam
Examination – Newborn

Barlow = Dislocatable    Ortolani = Reducible
Examination

Usually Not Significant:

- Asymmetric thigh / gluteal folds (In infant)
- “Soft tissue clicks”
Examination - Infant and Child

Limited abduction

- Especially in bilateral dislocations!
Examination - Infant and Child

Positive Galeazzi’s sign
  •  (Apparent femoral shortening)
Examination - Infant and Child

Klisic’s Test
Imaging

X-rays
- Most helpful after 4 mos
- Appearance of ossific nucleus

Ultrasound
- Allows visualization in newborn
- May be too sensitive

Arthrography

CT

MRI
Radiographic Findings

Ossific nucleus high and wide
Increased Acetabular Index
Decreased Center-Edge angle
Break in Shenton’s line
Perkin’s Line

Hilgenreiner’s Line
Broken Shenton’s Line

NO!
Shenton’s Line

Intact Shenton’s Line

Broken Shenton’s Line
Ultrasound

Useful in diagnosis & monitoring treatment

If hip is stable: get first u/s at 4-6 weeks: eliminates ~ 90% of false + hips

Ultrasound quality is extremely technician and radiologist dependant → send to a good radiologist!

Controversial as screening tool

• Never eliminate late presenters
Dislocatable Hip

Reduced in the resting position
Dislocatable with the Barlow maneuver
May stabilize spontaneously or with treatment
May convert to dislocated without treatment
Dislocated Hip

Dislocated in the resting position
May reduce with the Ortolani maneuver; this will be lost, usually by 8-12 weeks of age
Will not resolve without treatment
Dislocated Hip

Dislocated and irreducible in the resting position:

- Asymmetric thigh fold
- Limited abduction
- Positive Galeazzi
- Pistoning
- Trendelenburg gait
Goal:
Obtain, maintain and confirm a gentle, atraumatic concentric reduction
Treatment – 0 to 6 months

Pavlik Harness

- 95 to 99 % successful for “dislocatable” hips
- 50-80 % successful for fixed and dislocated
Pavlik Harness

Indications:

- Dislocatable hips
- Dislocated hips and:
  - Patients under 6 months of age
  - Femoral head points to triradiate cartilage with flexion, abduction
Pavlik Harness

Technique:

• Keep hip flexed 100-120° flexed, slightly abducted
• Document hip reduction by 4 weeks; if not reduced by that time, abandon harness
• Hold until stabilized
Treatment - 6 to 18 months

Closed vs Open Reduction

- Traction
- Not universal
- May increase success and decrease AVN
Treatment

6-18 months (or after Pavlik harness failure):

- Traction (Bryant’s or other balanced skin traction)
- Closed reduction
  +/− adductor tenotomy,
  +/− arthrogram
- Double hip spica cast for 3 months

**Open reduction if closed reduction fails**
Closed Reduction

Spica cast 3-6 months
6 weeks s/p spica

Initial CR
Open Reduction

Remove all impediments to reduction
Medial or Anterior
Treatment > 2 Years of Age

Open reduction
Capsulorrhaphy
Femoral shortening
Pelvic Osteotomy
Upper age limit

Unilateral: 8 years (perhaps higher!)

Bilateral: 6 years (perhaps lower!)

Depends on "skeletal maturity of patient and confidence and skill of surgeon"
- Routine u/s screening of all babies not recommended

- Recommendation from AAP to screen female breech babies and + FH with u/s
A

1. Physician performs newborn physical examination

2. Is physical examination positive?
   - No → 4
   - Yes → 3
      - 3. Refer to Orthopaedist

4. Is examination inconclusive?
   - No → 12
   - Yes → 5

5. Physician performs follow-up examination at 2 weeks

6. Is physical examination positive?
   - No → 8
   - Yes → 7
      - 7. Refer to Orthopaedist

8. Is examination inconclusive?
   - No → 10
   - Yes → 9
      - 9. Refer to Orthopaedist
         - *Ultrasound at 3 to 4 weeks
         - *Determine Risk Factors

10. Negative physical examination

11. Go to Box 14

12. Go to Box 14
**B**

14. **Negative examination**

15. Are risk factors present? 
   - Yes: Continue to follow periodicity schedule.
   - No: Follow periodicity schedule.

16. **RISK FACTOR**
   - **Girl**: Follow periodicity schedule.
   - **Family History and Boy**: Follow periodicity schedule.
   - **Family History and Girl**: Optional future imaging.
   - **Breech and Boy**: Optional future imaging.
   - **Breech and Girl**: Recommend future imaging.

17. Follow periodicity schedule

18. Is periodicity schedule follow-up positive?
   - Yes: Refer to Orthopaedist. 
     - Ultrasound < 5 months
     - X-ray > 4 months
   - No: Continue to follow periodicity schedule.
Hip dysplasia is here to make all good pediatricians and pediatric orthopaedists look like fools!
Stable hip, abnormal US

What is the problem?

• The exam missed the instability
  • Poor technique, baby crying
  • Hip initially unstable but only for a few days (or hours)
• The hip has some “dysplasia factor”
• Or the ultrasound is too sensitive
Stable hip, abnormal US: the exam

Who did the exam?
On what day of life was the exam done?
Was the baby crying or relaxed?
Does the baby have loose ligaments?
Most presume that the exam missed the instability
Hip was loose on the first few days of life, then stabilized
HOWEVER......
BABY “JONES”

Outside exam -> click
Age 2 weeks
Exam by resident-normal
Exam by me-normal
Return at 6 wks age for ultrasound
Ultrasound positive
Immediately repeated exam by me-normal
The exam was normal. Full abduction, no instability noted, child quiet and happy, doctor confused.
Ultrasound post 3 wks in Pavlik
BABY SMITH

Initial exam at birth unknown
My exam, my PA’s exam, my resident’s exam all negative at 3 weeks
Ultrasound at 6 weeks
Exam repeated-still normal
Normal exam, US dysplasia

Treatment - Pavlik harness, ultrasound done after 3 weeks in harness.
Response to Pavlik is rapid
CONCLUSIONS

• These hips probably have “subtle instability”
• They should be treated
• We need to overtreat some hips to include the ones which will subluxate or dislocate
• We should study this objectively (hard to do)
SUMMARY

Hip instability is best treated if detected early

Know the risk factors: breech, +FH, 1st born female

If hip is unstable → refer to pediatric ortho

If hip is stable on exam → u/s at 4-6 weeks if needed

>12 months old with chronic, nonpainful limp → THINK HIP DISLOCATION!