JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Initial Surveillance for Developmental Dysplasia of the Hip (DDH) in the NICU Clinical Pathway

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Rationale

1. Surveillance for Developmental Dysplasia of the Hip (DDH) may allow early intervention and possible avoidance of surgery or future disability.
2. Breech presentation at birth is a risk factor for DDH and indication for evaluation. Evidence for evaluation of premature infants with fetal ultrasonographic documentation of breech positioning is unclear.

Background / Published Data and Levels of Evidence

Background: Developmental Dysplasia of the Hip (DDH) includes frank dislocation (luxation), partial dislocation (subluxation), hip joint instability, and/or malformed acetabula. *Developmental* is a more appropriate term than *congenital*, as dysplasia may develop in utero, perinatally, or in infancy/childhood. *Surveillance* is therefore a more appropriate term than screening, and as developmental hip laxity may resolve. The purpose of surveillance is to detect significant hip dysplasia to allow early intervention and possible avoidance of surgery or future disability (functional disabilities, leg length discrepancies, early onset osteoarthritis). Surveillance is recommended by all leading US and Canadian orthopaedic and pediatric physician organizations, although rated as “Inconclusive” by the US Preventive Services Task Force. The importance of the periodic examination cannot be overemphasized because other than female gender, most children with DDH do not have risk factors. Risk factors for DDH (current evidence): female sex, breech presentation, family history of DDH, history of improper lower-extremity swaddling. Selective hip ultrasound (US) screening may identify dysplastic hips that are clinically normal or with equivocal clinical findings. No surveillance or screening program completely eliminates the risk of late presentation of DDH.

Published Data and Levels of Evidence:

A. *Refer to AAP and AAOS publications; guidelines for surveillance for DDH in the term infant:*

1. AAP Clinical Report: Evaluation and Referral for Developmental Dysplasia of the Hip. Pediatrics, 138 (6), 2016. AAP Section on Orthopaedics.
2. American Academy of Pediatrics, Clinical practice guideline: early detection of developmental dysplasia of the hip. Committee on Quality Improvement, Subcommittee on Developmental Dysplasia of the Hip. Pediatrics. 2000;105(4 pt 1):896–905.
3. American Academy of Orthopaedic Surgeons. Detection and Nonoperative Management of Pediatric Developmental Dysplasia of the Hip in Infants Up to Six Months of Age. Evidence-Based Clinical Practice Guideline. Rosemont, IL: American Academy of Orthopaedic Surgeons; 2014.

B. *Selected evidence regarding risk for and investigation of DDH in preterm infants*:

1. Lee J, et al. Sonographic screening for developmental dysplasia of the hip in preterm breech infants: do current guidelines address the specific needs of premature infants? Journal of Perinatology, 2016, 36: 552-556.
    a. Prevalence of breech presentation increases as the gestational age (GA) decreases below 37 weeks, 1-3% of births are breech at term, 7% of births at 32 weeks, and 22% of births prior to 28 weeks.
    b. The fetus usually undergoes a spontaneous in utero version from breech to vertex by 32 weeks. Infants born prior to spontaneous in utero version may not undergo the sustained mechanical forces to predispose to DDH.
    c. 135 infants <32 weeks with breech presentation, 3 infants (2%) had abnormal US findings; all resolved on follow-up US. 183 infants 32-37 weeks GA and breech presentation, 17 (9%) had abnormal US findings; 13 resolved on follow-up US, 1 case was dysplastic, 3 cases lost to follow-up.
    d. Hip US performed prior to the recommended corrected age for screening (44-46 weeks PMA, post-menstrual age) may have a higher risk for false positive findings secondary to hip immaturity, and thus a higher risk of unnecessary treatment.

2. Lang et al. Population-Based Study of the Incidence of Congenital Hip Dysplasia in Preterm Infants from the Survey of Infants in Pomerania. BMC Pediatrics, 2017; 17:78.
    a. Study in Pomerania, Germany: US hip performed at 3-10 days of life (customary second well-child exam) in term infants
        1. 42 of 2,534 term infants had US suggestive of DDH
    b. Preterm infants <36 weeks GA screened at ≥ 36 weeks PMA
        1. 0 of 279 infants had US findings
        2. 3 of 97 infants 36-37 weeks GA had DDH

3. Sezer et al. Prevalence of Developmental Dysplasia of the Hip in Preterm Infants With Maternal Risk Factors. J Child Orthop. 2013. 7:257-261.
    a. 421 infants <37 weeks GA prospectively evaluated in Ankara, Turkey; only 1 DDH identified. 48 breech cases with no DDH. 159 oligohydramnios cases with no DDH.
    b. Concluded prematurity with or without maternal risk factors does not have an effect on DDH.

4. Quan T et al. Breech preterm infants are at risk of developmental dysplasia of the hip. Journal of Paediatrics and Child Health, 2013, 49: 658–663.
    a. Retrospective comparison of DDH in term infants versus <37 wks gestation.
    b. 3 of 129 infants <37 wks had DDH, 1 infant <32 wks (29 wks)

5. Hegde D, et al. Developmental dysplasia of the hip in preterm breech infants. Archives of Disease in Childhood, Fetal and Neonatal Edition, 2020; 105: F556–F558.
    a. Retrospective review of infants born in breech position at Perth Children's Hospital, Nedlands, Western Australia. Assessed by gestational age groups: 23–27 wks (n=112), 28–31 wks (n=254), 32–36 wks (n=552) and ≥37 wks (n=226).

b. Incidence of DDH by x-ray findings similar in gestational age groups: 23–27 wks (11.6%), 28–31 wks (9.4%), 32–36 wks (13.6%) and ≥37 wks (11.5%). Multiple logistic regression after correcting for potential confounders. Concluded that preterm infants born with breech presentation appear to have a similar incidence of DDH to term breech infants (p=0.40).
c. Subsequent intervention, non-surgical or surgical, was also similar in each gestational age group (p=0.44): 23–27 wks (2.7%), 28–31 wks (4.3%), 32–36 wks (5.8%) and ≥37 wks (6.2%).
d. Extrapolated results: 4.2% of infants <32 wks required intervention; 6.7% of infants ≥32 wks required intervention.
e. 51.7% of infants who required intervention had normal 6-week ultrasounds, emphasizing the role of physical examination in surveillance.
f. This study demonstrates that some premature infants <32 wks gestational age may be at risk for DDH. However, the utility of hip US screening remains unclear.

Clinical Management

A. JHACH DDH Surveillance and Referral Recommendations:
(in consideration of the 2000 AAP Clinical Practice Guideline, the 2016 AAP Clinical Report, the 2014 AAOS Clinical Practice Guideline, and available evidence regarding the premature infant; in conjunction with the JHACH Department of Orthopaedic and Scoliosis Surgery)

1. Surveillance hip examinations are recommended as a part of the routine newborn examination, and should include (see Appendix A, Figures 1-3):
    a. Ortolani test
    b. Barlow test
    c. Hip abduction
    d. Gluteal or major thigh crease asymmetry
    e. Leg length inequality (Galeazzi sign)
2. Newborns with a positive Ortolani or Barlow test should be referred to a pediatric orthopaedist by 2 weeks of age, with hip ultrasonography (US) requested at the time of referral.
    a. Unequivocally positive examination results in a *clunk.*
3. Orthopaedic referral or hip US are not recommended for softly positive findings on newborn examination (eg, laxity, but stable hip). Evidence supports initially observing, with physical examination followed for likely resolution by 2 weeks of age, at which time referral may be considered.
    a. Hip clicks without instability or dislocation are clinically insignificant.

B. Selective Screening Hip Ultrasounds

1. *Selective screening hip US*, if indicated by risk factors (see below), should be deferred until **46 wks PMA** (postmenstrual age, corrected age), due to the high rate of false positives of immature hips.
    a. Risk factors for which selective screening hip US may be considered with a normal screening physical examination are:

i. breech position in the third trimester, *both* males and females, even if position changed by time of delivery
ii. family history of DDH
iii. history of improper lower-extremity swaddling

b. Selective screening hip US may also be considered for infants with a history of abnormal hip physical examination in the neonatal period, which subsequently normalizes.
c. Current evidence is conflicting regarding the risk of DDH in premature infants < 32 wks GA noted in breech presentation at delivery or at time of an antenatal ultrasound (see Lee et al.; Hedge et al). These premature infants born prior to the typical gestation of spontaneous in utero version (32 wks) may not undergo the sustained mechanical forces necessary to predispose to DDH. Serial surveillance physical exams should be performed as recommended by the AAP. Screening hip ultrasound at 6 weeks may be considered, but the risk of overdiagnosis of immature hips is high.
d. The AAP and AAOS do not recommend *universal ultrasound screening* due to the high risk of overdiagnosis of immature hips and because milder forms of DDH improve without intervention.

C. Screening Hip Radiographs

1. The femoral head ossification nucleus is visible radiographically at approximately 4 to 6 months of age. Radiographs are thus not recommended for DDH evaluation before 4 months of age.
2. Tudor, 2007. Seventy-four infants with ultrasound positive hips for acetabular dysplasia who met criteria for treatment received an AP pelvis radiograph. Of these 74 infants, 30 were found to have satisfactory acetabular indices and did not receive treatment.

D. Discharge Summary Documentation

1. Discharge Summary documentation should include notation of the infant's presentation at birth (*e.g. vertex, breech, etc*.) to convey potential risk factor.
2. Discharge physical examination should include Ortolani and Barlow tests, along with other relevant findings.
3. Additional Instructions for the pediatrician should include one of the following inclusions:
    a. **.hipexam (≥32 wks GA h/o breech presentation):** *"Breech presentation at delivery or at time of a third trimester antenatal ultrasound is a risk for Developmental Dysplasia of the Hip (DDH). As per AAP recommendations, it is suggested for pediatricians to perform serial surveillance physical exams and obtain a screening hip ultrasound at 46 weeks PMA".*
    b. **.hipexampremature <32 wks GA h/o breech presentation:** "*Current evidence is conflicting regarding the risk of Developmental Dysplasia of the Hip (DDH) in premature infants < 32wks GA noted in breech*

presentation at delivery. As per AAP recommendations, it is suggested for pediatricians to perform serial surveillance physical exams. As screening ultrasound of the immature hip is not reliable, it is also suggested to obtain an AP x-ray of the hips at 4-6 months."

Summary

1. DDH surveillance and the detection of significant hip dysplasia may allow early intervention and possible avoidance of surgery or future disability.
2. Surveillance hip examinations are recommended as a part of newborn examinations and all routine serial examinations for all infants.
3. Selective screening hip US should be deferred until **46 wks PMA** in at risk infants.
4. The role for screening hip US of premature infants <32 weeks GA with breech presentation and normal surveillance examinations is unclear. We recommend obtaining an AP x-ray of the hips at 4-6 months.
5. No surveillance or screening program completely eliminates the risk of late presentation of DDH.

Glossary

Developmental Dysplasia of the Hip (DDH)

Fetal breech positioning

Breech presentation at birth

Selective screening hip ultrasonography

References

American Academy of Pediatrics, Clinical practice guideline: early detection of developmental dysplasia of the hip. Committee on Quality Improvement, Subcommittee on Developmental Dysplasia of the Hip. Pediatrics, 2000;105(4 pt 1):896–905.

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AAP State-of-the-Art Review Article: Developmental Dysplasia of the Hip. Pediatrics, 2019; 143(1):e20181147.

American Academy of Orthopaedic Surgeons. Detection and Nonoperative Management of Pediatric Developmental Dysplasia of the Hip in Infants Up to Six Months of Age. Evidence-Based Clinical Practice Guideline. Rosemont, IL: American Academy of Orthopaedic Surgeons; 2014.

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Leonard SP et al. Developmental Dysplasia of the Hip is Not Associated with Breech Presentation in Preterm Infants. American Journal of Perinatology. doi: 10.1055/s-0042-1756139. Epub ahead of print, 2022.

Koob S et al. Is Prematurity a Protective Factor Against Developmental Dysplasia of the Hip? A Retrospective Analysis of 660 Newborns. European Journal of Ultrasound, 2022; 43: 177–180.

Outcome Measures

1. Longitudinal population outcomes of premature infants with breech presentation, incidence of DDH.
2. Audit of documentation in Discharge Summary documents.

Appendix 1: Ortolani and Barlow Tests (see Figure 1)

- **A. Ortolani Test**
  - a. Abducting the hips while applying anterior-directed pressure at the greater trochanters.
  - b. Considered positive if the femoral head relocates with a distinct “clunk”.
- **B. Barlow Test**
  - a. Adducting the hips to the midline and gently applying posterior force.
  - b. Considered positive test results when the femoral head subluxes, and a “clunk” is felt.

Appendix 2: Physical Examination (see Figure 2, Figure 3)

A. Limitations of physical examination tests

a. Dependent on skills of the provider
b. A severely dislocated hip that is not reducible may not have a Barlow or Ortolani positive result.

Appendix 3: Radiographic Imaging (see Figure 4, Figure 5)

Figure 1

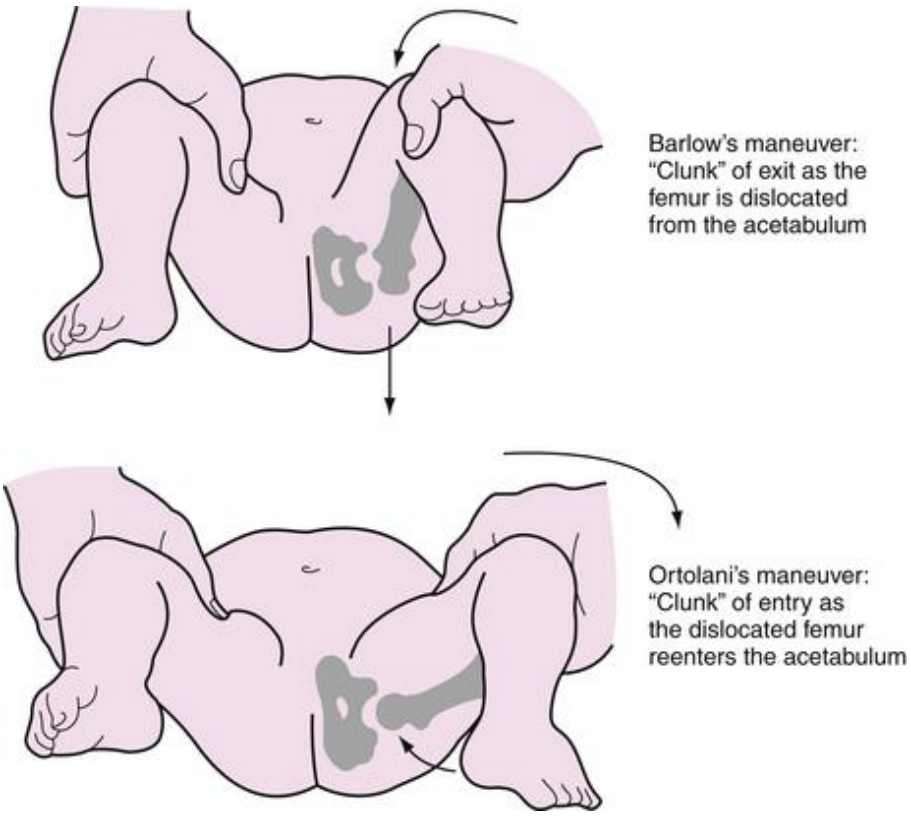


Figure 2: Gluteal or major thigh crease asymmetry

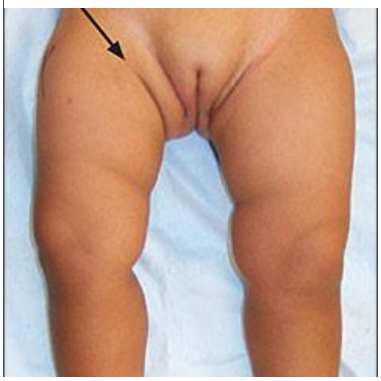


Figure 3: Leg length inequality (Galeazzi sign)

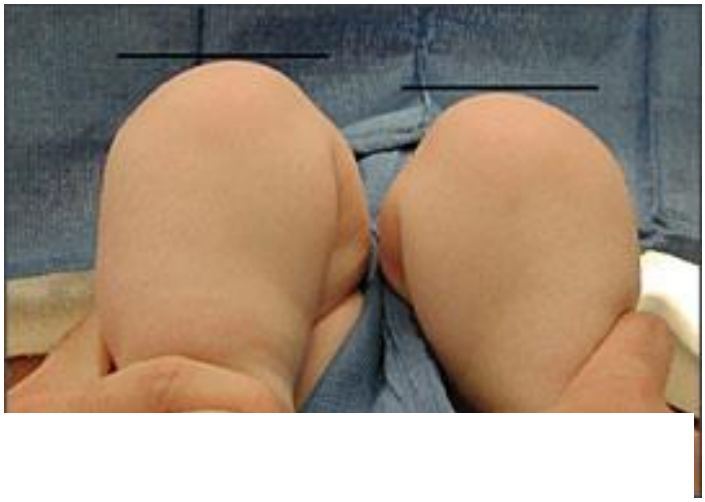


Figure 4: Ultrasound

Coronal ultrasound images of a 2-month-old dysplastic hip. Key ultrasound measurements include the α angle (which is formed by the bony ilium and the bony roof of the acetabulum), the β angle (which is formed by the bony ilium and the labral fibrocartilage), and the percentage of the femoral head covered by the bony roof of the acetabulum. The α angle has more clinical significance than the β angle. In this image, (1) the femoral head does not seat deeply in the socket, with <50% of the femoral head being covered by the acetabulum, and (2) the acetabulum is shallow (normal α >60°).

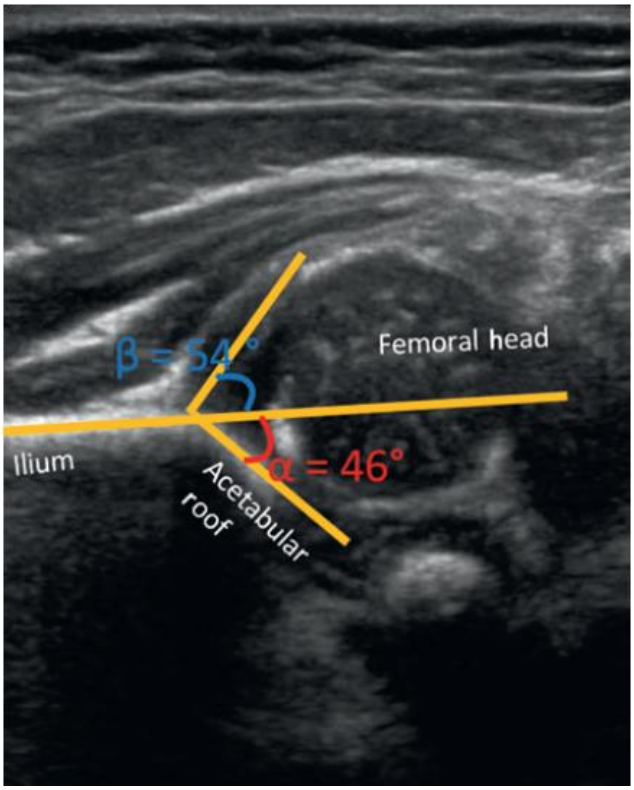
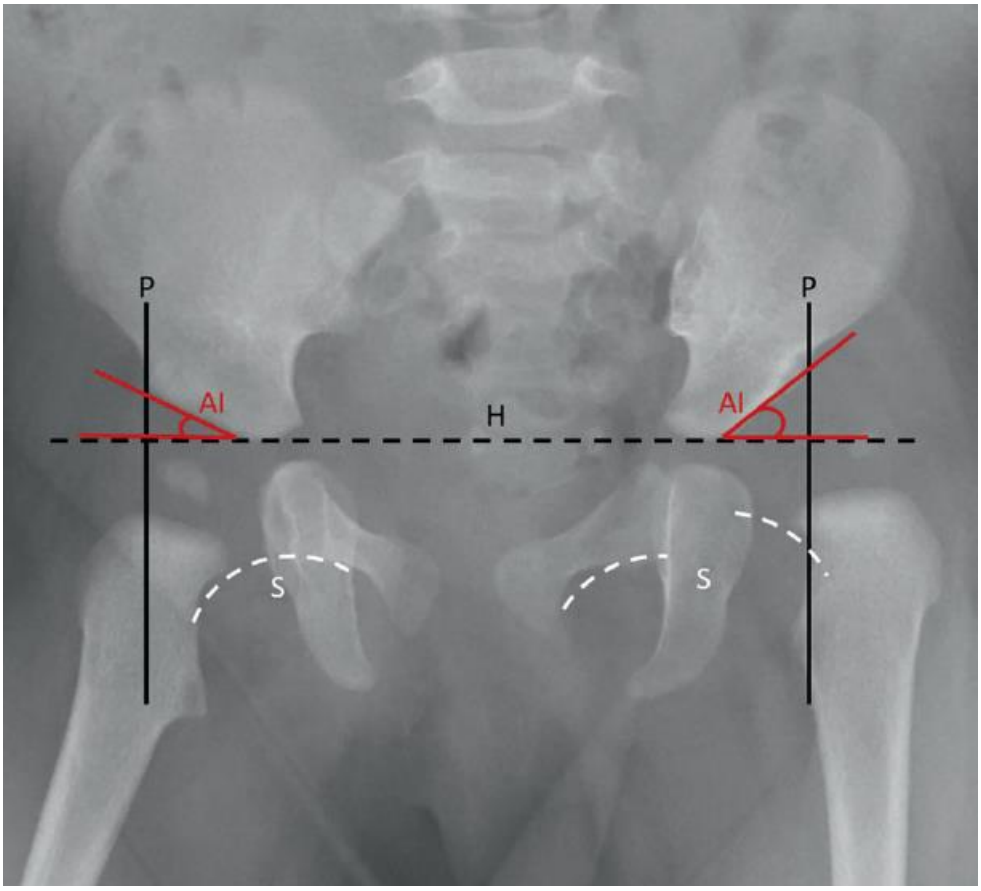


Figure 5: Radiograph

AP pelvis radiograph of a left-hip dislocation. On an AP pelvis radiograph, classic measurements include drawing several lines to help identify dysplasia. H is drawn as a horizontal line, connecting the bilateral acetabular triradiate cartilage. P is then drawn perpendicular to H at the lateral edge of the acetabulum. In the normal right hip, the ossific nucleus rests along the bottom-inner quadrant formed by the intersection of the 2 lines. In the dislocated hip, the ossific nucleus rests lateral to the intersection of the 2 lines. S should reveal a smooth arch from the obturator foramen to the inferior aspect of the femoral neck, as in the right hip. S is disrupted on the left hip, suggesting dislocation. The acetabular index is the angle formed along the acetabular roof and H, with steeper values indicating acetabular dysplasia. Notice also that the left femoral head ossific nucleus is smaller, and its appearance is more delayed compared with the nondysplastic side. AI, acetabular index; AP, anteroposterior; H, Hilgenreiner line; P, Perkins line; S, arc of Shenton.



Clinical Pathway Team

Initial Surveillance for Developmental Dysplasia of the Hip (DDH) in the NICU

Clinical Pathway

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