American College of Radiology ACR Appropriateness Criteria[®] Developmental Dysplasia of the Hip (DDH)–Child

<u>Variant 1:</u> Child, younger than 4 weeks of age. Equivocal physical examination or risk factors for DDH. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US hips	Usually Not Appropriate	0
Radiography pelvis	Usually Not Appropriate	\$ \$

<u>Variant 2:</u> Child, between 4 weeks to 4 months of age. Equivocal physical examination or risk factors for DDH. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US hips	Usually Appropriate	0
Radiography pelvis	Usually Not Appropriate	**

Variant 3: Child, younger than 4 months of age. Physical findings of DDH. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US hips	Usually Appropriate	0
Radiography pelvis	Usually Not Appropriate	\$

Variant 4:

Child, between 4 to 6 months of age. Concern for DDH. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography pelvis	Usually Appropriate	**
US hips	May Be Appropriate	0

Variant 5:

Child, older than 6 months of age. Concern for DDH. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography pelvis	Usually Appropriate	\$ \$
US hips	Usually Not Appropriate	0

<u>Variant 6:</u> Child, younger than 6 months of age. Known diagnosis of DDH, nonoperative surveillance imaging in harness.

Procedure	Appropriateness Category	Relative Radiation Level
US hips	Usually Appropriate	0
Radiography pelvis	Usually Not Appropriate	♦♥
CT pelvis with IV contrast	Usually Not Appropriate	\$\$\$\$
CT pelvis without and with IV contrast	Usually Not Appropriate	₸₽₽₽₽
CT pelvis without IV contrast	Usually Not Appropriate	***
MRI pelvis without and with IV contrast	Usually Not Appropriate	0
MRI pelvis without IV contrast	Usually Not Appropriate	0

DEVELOPMENTAL DYSPLASIA OF THE HIP-CHILD

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Summary of Literature Review

Introduction/Background

Developmental dysplasia of the hip (DDH) comprises a spectrum of abnormalities from hip instability to frank dislocation [1,2]. The mildest end of the spectrum overlaps with physiologic immaturity, therefore making it difficult to determine its true incidence, which is estimated to be 1.5 to 20 per 1,000 births, depending on the demographics of the study population and the inclusion criteria [3-5]. The diagnosis and monitoring of teratologic hips from neuromuscular or syndromic causes will not be covered.

The pathophysiology of DDH is multifactorial and not completely understood. The 2 leading causes are laxity induced by maternal hormones and limited in utero hip mobility. In infants with DDH, abnormally increased laxity of the hip capsule and surrounding ligaments have been attributed to the effects of maternal hormone relaxin [1] and a higher concentration of estrogen receptors [6]. In utero restriction to hip mobility can be encountered with oligohydramnios, first-born infants, and prolonged breech positioning. Breech fetal positioning produces extreme hip flexion with knee extension. This leads to shortening and contracture of the iliopsoas muscle, which promotes femoral head dislocation. Studies demonstrating increased prevalence of DDH among monozygotic twins as compared to dizygotic twins [7] and chromosomal analysis in familial DDH and population-based DDH suggest genetic predisposition to DDH [8-10].

The most important risk factors for DDH are female gender, breech positioning in utero, and a positive family history, carrying relative risks of 2.5, 3.8, and 1.4, respectively, in a large meta-analysis [8-11]. Another risk factor is infant swaddling [12,13]. First born, torticollis, foot abnormalities, and oligohydramnios have not been proven to increase the risk of DDH [11]. The left hip is three times more frequent to have DDH with a relative risk of 1.5 [9], theorized to be the result of more common left occiput anterior in utero position, which places the left hip against the mother's spine and limits its abduction. Preterm infants are not at an increased risk for DDH [3,14-16], and there is a lack of consensus on the association between multiparity and DDH [3,8,13].

The natural history of DDH depends on the type and degree of abnormality. Most borderline "abnormal" hips during the neonatal period likely represent physiologic immaturity, as 60% to 80% identified by physical examination and more than 90% identified by ultrasound (US) spontaneously normalize at follow-up [12,17-21]. Late presentation is a major negative prognostic factor, with these patients more likely to require complex treatment [22], surgical intervention [4], and to experience long-term complications [22-24]. Unrecognized and untreated subluxation and dislocation inevitably lead to early degenerative joint disease. It is estimated that DDH is the cause of up to a third of all total hip arthroplasties performed in patients <60 years of age, which emphasizes the importance of proper screening, early diagnosis, and appropriate intervention [25].

The goals of an ideal screening program are early detection of patients who have DDH when therapy is typically noninvasive and often most effective and exclusion of patients without DDH for whom unnecessary treatment could be costly and potentially harmful. The most important screening method is a hip examination at every well-baby visit according to the recommended periodicity schedule for well-baby examinations (2–4 days for

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newborns discharged in <48 hours after delivery, by 1 month, 2 months, 4 months, 6 months, 9 months, and 12 months of age) [1,5]. The most serious complication of treatment is avascular necrosis [1,5,26], which is a predictor of poor prognosis [27,28]. Screening can be universal, when all neonates are evaluated, or selective, when only those at risk are evaluated [1,5,26].

Universal US screening for DDH in newborns is performed in some European countries [29], which increases the detection rate of "abnormal" hips. However, there is no evidence that it significantly decreases late diagnosis of DDH [1,5,16,30-32], and the higher rates of abduction splinting carry the risk of overtreatment and iatrogenic avascular necrosis [1,5,26,33]. For these reasons, the American Academy of Pediatrics (AAP) recommends selective screening [1,11] of children with risk factors [34,35] or based on physical examination findings [11]. A positive Barlow or Ortolani test implies an unstable femoral head that can be dislocated or relocated, respectively. Ortolani and Barlow tests are less sensitive after the age of 2 to 3 months because of increased tightening of the hip capsule. After that age, physical examination is less accurate, and the most important finding is limited hip abduction. Other findings may include asymmetric buttock creases and leg length discrepancy [7,11,36].

In the past 2 decades, imaging has become an integral part of screening, diagnosis, and monitoring of children with DDH [20]. A prospective 33-center United Kingdom Hip Trial [37] found that US of children with clinically detected hip instability allowed for a reduction in abduction splinting and was not associated with an increase in abnormal hip development or higher rates of surgical intervention [37].

Special Imaging Considerations

US

US is performed using a high-frequency linear array transducer [38]. Two techniques have emerged: a static acetabular morphology method proposed by Graf and a dynamic stress technique proposed by Harcke [39-42].

The Graf method uses coronal imaging of the hip joint. Graf developed a morphologic and geometric hip classification scheme (Types I–IV) using the alpha angle, which measures the osseous acetabular roof angle. The beta angle, which defines the position of the echogenic fibrocartilaginous acetabular labrum, was part of the initial classification but is now infrequently used in routine practice. Femoral head coverage method (Terjesen method) uses a 50% cutoff between normal and abnormal hips [43]. The different types can be broadly grouped into three major categories [40]:

- *Normal hip*: Type I hips are normal and require no treatment. The alpha angle is $\geq 60^{\circ}$.
- *Immature hip:* Type IIa hips are seen in infants <3 months of age. The hip is normally located, but the bony acetabular promontory is rounded, and the alpha angle is 50° to 59°. These patients have a small risk of delayed DDH. This group can be further divided to type IIa+ (alpha angle between 55° and 59°) and type IIa- (alpha angle between 50° and 54°). Most children with stable hips and Graf type IIa will have spontaneous normalization and only conservative management is recommended. The management of type IIa- is controversial as some children (up to 15%, mainly female) have abnormalities that will not resolve [44].
- *Dysplastic hip:* Type IIb has similar features to type IIa, but it is detected in children >3 months of age. Types IIc, IId, III, and IV represent progressively abnormal hips with frank subluxation in types III and IV. The alpha angle is <50° in types IIc and IId and <43° in types III and IV.

Harcke [45] developed the dynamic method, using US to attempt to visualize Barlow and Ortolani tests. This technique is performed in both coronal and transverse planes, with and without stress. The modified Barlow test is performed by holding the knee with the hip flexed at 90° and in adduction. The femur is pushed (pistoned) posteriorly. The <u>ACR-AIUM-SPR-SRU Practice Parameter for the Performance of the Ultrasound Examination for Detection and Assessment of Developmental Dysplasia of the Hip combines both static and dynamic techniques [42], which is the most commonly used imaging protocol practiced at most children's hospitals throughout the United States. The main disadvantage of US is that it has high interobserver variability, particularly for milder cases of dysplasia [39,46-48].</u>

Radiography

An anteroposterior radiograph of the pelvis with the hips in neutral position allows visualization of the femoral head ossific nucleus and acetabular morphology. Proper positioning is critical as both pelvic rotation and inclination can hinder diagnostic accuracy, producing false-positive and false-negative results.

The most commonly used measurement is the acetabular index [49]. This index is 30° in a newborn and decreases progressively with growth and maturation [49-51]. In dysplastic hips, the acetabular index is increased, which then decreases in response to successful treatment [52,53]. The position of the femoral head is evaluated based on the relationship of the ossific nucleus or proximal femoral metaphysis to the Hilgenreiner and Perkin lines and by evaluating for disruption of the Shenton arc [31]. A radiographic classification system has been developed by the International Hip Dysplasia Institute, which uses the midpoint of the proximal femoral metaphysis as a reproducible reference landmark [54].

Discussion of Procedures by Variant

Variant 1: Child, younger than 4 weeks of age. Equivocal physical examination or risk factors for DDH. Initial imaging.

For infants with equivocal physical examination or risk factors for DDH, there is evidence that the vast majority spontaneously normalize [21,55,56], and a short delay in intervention has no negative impact on outcome [22,57,58]. Therefore, the potential benefits of early diagnosis and treatment must be weighed against the risk of overtreatment and potential for iatrogenic complications [31]. Thus, the AAP recommends screening with US at the age of 4 to 6 weeks [1], and the American Academy of Orthopaedic Surgeons (AAOS) recommends pediatric orthopedic referral before 4 weeks of age [20].

US Hips

Although US can be performed shortly after birth, its high sensitivity for the detection of mild acetabular immaturity and minor degrees of hip laxity can suggest pathology, potentially leading to overdiagnosis (false-positives) and overtreatment [3,5,18]. Therefore, US is not recommended during the newborn period [59].

Radiography Pelvis

There is no relevant literature regarding the use of radiographs for screening of DDH in children <4 weeks of age.

Variant 2: Child, between 4 weeks to 4 months of age. Equivocal physical examination or risk factors for DDH. Initial imaging.

Although most physicians recommend the first imaging screening for nondislocated hips to be performed at 4 to 6 weeks of age, thus allowing time for normalization of neonatal physiologic immaturity and laxity, this remains an arbitrary time-point, balancing the risk of increased false-positive studies in early age with the potential benefits of early treatment. In a study of 5,170 infants screened at 1 month of age, 99.6% remained normal and 84% to 95% of Graf type II hips normalized at 3 months, indicating that the vast majority continue to normalize after the first month of life [60]. Because of the high false-positive rate of diagnosis with US, the AAP recommends selective US only in the highest risk group, girls with breech presentation at birth [1]. AAP also suggests US as an option in girls with a positive family history, boys with breech presentation, and when the physical examination is inconclusive [1].

US Hips

A prospective study using US screening was performed on 2,578 children with an unstable hip on physical examination or risk factors for developmental dysplasia. Screening US was shown to reduce the number of delayed diagnoses and decrease the rate of surgical intervention when compared to clinical screening alone [61]. Other studies have shown that US can help confirm the diagnosis of DDH, leading to a change in the clinical management [62,63].

Radiography Pelvis

Pelvic radiographs are limited for evaluation for DDH in the first 3 months of life. The ossific nucleus of the femoral head usually appears between 4 to 6 months (range 1.5–8 months), but for dysplastic hips, its appearance is often delayed [54]. The acetabular margin is also largely cartilaginous, hindering the assessment of acetabular morphology and femoral alignment [64,65].

Variant 3: Child, younger than 4 months of age. Physical findings of DDH. Initial imaging.

For the purpose of this variant, positive physical examination is defined as a positive Barlow or Ortolani test, which implies an unstable femoral head that can be dislocated or relocated, respectively [66].

US Hips

The AAP guideline published in 2000 [1] did not recommend US screening after a positive physical examination. However, recent studies have shown that 41% to 58% of abnormal findings from a physical examination were

false-positive findings when correlated with US, thus leading to unnecessary treatment [1]. A prospective 33center United Kingdom Hip Trial [37] addressed the value of selected US screening in infants following a positive physical examination. It found that US examinations in infants with clinically detected hip instability allowed for a reduction in abduction splinting and was not associated with an increase in abnormal hip development or higher rates of surgical treatment [37]. This policy was found to reduce costs [37]. A 15-year longitudinal observation study found the sensitivity, specificity, and positive predictive value of clinical screening for diagnosing DDH to be 62%, 99.8%, and 24%, as opposed to US screening at 77%, 99.8%, and 49%, respectively [7].

Radiography Pelvis

There is no relevant literature regarding the use of radiographs in children <3 months of age with physical findings of DDH, which is in part because the ossific nucleus of the femoral head usually appears between 4 to 6 months (range 1.5–8 months) [54].

Variant 4: Child, between 4 to 6 months of age. Concern for DDH. Initial imaging.

Late-presenting DDH, defined as diagnosis after 3 months of age, is uncommon, occurring at an estimated rate of 0.22 per 1,000 births [55]. At this age, the clinical assessment is less reliable and imaging is often required to confirm the diagnosis. By 8 to 12 weeks of age, the capsule laxity decreases, muscle tightness increases, and the Barlow and Ortolani maneuvers may not be positive regardless of the status of the femoral head. Thus, the finding of limited hip abduction becomes the most important screening method in older children. However, currently there is no consensus on the reliability of this test for diagnosing DDH [5], with one study demonstrating a positive predictive value of 40% for DDH that can increase up to 55% after 8 weeks of age [36], while another study demonstrated no correlation between a positive abduction test and an abnormal acetabular angle [66]. Other screening methods, such as the findings of asymmetric skin folds in the proximal thigh and shortening of the limb on the dislocated side, lack specificity for the diagnosis of DDH [66]. These inconsistencies among various studies may reflect differences in patient selection or inclusion, expertise of the examiners, and the defined gold standard [1,67-69].

US Hips

The AAP and AAOS do not advocate the use of US for the screening of DDH after 4 to 5 months [1,20]. There is limited evidence on the use of US for screening of DDH beyond 4 months. A study that obtained anteroposterior radiographs in patients who are 4 to 6 months of age with positive US found that US overdiagnosed DDH in 40% of patients [58].

Radiography Pelvis

Shortly after the appearance of the ossific nucleus, pelvic radiography becomes the preferred confirmatory imaging modality, as it allows for the assessment of the femoral head ossific nucleus, the development of the proximal femur, and bony acetabular morphology [1]. Normal pelvic radiograph at 4 months can reliably exclude DDH in children with risk factors [56] and decrease the need for treatment in infants who are 4 to 6 months of age with positive US by 40% [58]. This eliminates unnecessary serial follow-ups and potential for iatrogenic treatment-related complications. Thus, for infants with suspected hip dysplasia, a radiograph is often obtained between 4 to 6 months of age [21,56-58]. However, there are a few limitations to pelvic radiograph. The timing for the appearance of the ossific nucleus varies widely, from 1.5 to 8 months of age [65], and in dysplastic hips, its appearance is often delayed and, when it does appear, is often eccentric [54,70].

Variant 5: Child, older than 6 months of age. Concern for DDH. Initial imaging.

Clinical assessment at this age is often limited, as the traditional physical examination findings—such as the Ortolani and Barlow tests, hip abduction, asymmetric proximal thigh skin folds, and limb length—lack sensitivity and specificity [5,22,36,66]. This leads to a great reliance on imaging for the confirmation and monitoring of DDH.

US Hips

There is insufficient evidence to recommend the use of US as the evaluation may be inadequate because of the suboptimal visualization of the anatomy of the hip joint from decreased acoustic penetration.

Radiography Pelvis

Shortly after the appearance of the ossific nucleus, pelvic radiography becomes the preferred imaging modality as it facilitates the assessment of the femoral head ossific nucleus and the development of the proximal femur and bony acetabular morphology [1]. There is ongoing debate regarding the necessity of serial radiographic studies for

mild acetabular dysplasia in the setting of normal clinical examination and normalized US findings, with one study reporting that up to 17% had radiographic signs of dysplasia at 6 months [71], another study concluding only 5% at 12 months and none at 21 months of age, and a different study citing 24% at a mean follow-up of 9 years [72]. These inconsistencies likely reflect differences in patient selection/inclusion, but the long-term implication of these imaging findings remain unknown.

Variant 6: Child, younger than 6 months of age. Known diagnosis of DDH, nonoperative surveillance imaging in harness.

The treatment algorithm for DDH varies among practices but typically includes a trial of nonoperative management using abduction splinting, often with a Pavlik harness. The efficacy of the Pavlik harness decreases with age. It is most effective if the harness is applied before 6 weeks of age, and the harness can be used up to 6 months of age. The overall success rate of the harness ranges from 67% to 83% [28]. Surgical intervention is typically reserved for children with severe dysplasia or dislocation, late presentation or diagnosis, or failed nonoperative management [73].

US Hips

In children who are undergoing nonoperative treatment with Pavlik harness, US can be used to confirm concentric hip reduction [74,75], assess treatment response [73,76-79], and identify signs of therapy failure [80]. Predictors of failure include low postreduction alpha angle and <20% femoral head coverage [81]. US is typically performed without applied stress to the hips and with the child either in or out of brace, depending on the discretion of the referring provider.

Radiography Pelvis

Radiography is not the preferred modality for monitoring children undergoing nonoperative treatment using a harness [76,82] because of the delay in the appearance of the femoral head ossific nucleus and suboptimal patient positioning (within the harness). Rather, radiographs are often obtained at or near the conclusion of the treatment to document bony acetabular development and to provide a baseline for future surveillance [59].

CT Pelvis

There is no relevant literature regarding the use of CT to monitor children with DDH who are being treated nonoperatively using a harness.

MRI Pelvis

There is no relevant literature regarding the use of MRI to monitor children with DDH who are being treated nonoperatively using a harness.

Summary of Recommendations

- Variant 1: Imaging is not recommended for the initial imaging of children younger than 4 weeks of age with an equivocal physical examination or risk factors shown for DDH.
- Variant 2: US of the hips is usually appropriate for the initial imaging of children between 4 weeks to 4 months of age with an equivocal physical examination or risk factors shown for DDH.
- Variant 3: US of the hips is usually appropriate for the initial imaging of children younger than 4 months of age with physical findings of DDH at initial imaging.
- Variant 4: Radiographs of the pelvis is usually appropriate for the initial imaging of children between 4 to 6 months of age with a concern for DDH at initial imaging.
- Variant 5: Radiographs of the pelvis is usually appropriate for the initial imaging of children older than 6 months of age with a concern for DDH.
- Variant 6: US of the hips is usually appropriate for children younger than 6 months of age with a known diagnosis of DDH during nonoperative surveillance imaging in harness.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at <u>https://acsearch.acr.org/list</u>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents go to <u>www.acr.org/ac.</u>

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk- benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria[®] Radiation Dose Assessment Introduction document [83].

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
•	<0.1 mSv	<0.03 mSv
**	0.1-1 mSv	0.03-0.3 mSv
***	1-10 mSv	0.3-3 mSv
****	10-30 mSv	3-10 mSv
$\mathbf{\hat{x}}\mathbf{\hat{x}}\mathbf{\hat{x}}\mathbf{\hat{x}}\mathbf{\hat{x}}$	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."

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The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.