

RPA Newborn Care Guidelines

Royal Prince Alfred Hospital

Developmental dysplasia of the hip

Introduction:

Developmental dysplasia of the hip (DDH) describes a spectrum of abnormalities that affect the neonatal and infant hip joint. The central process is a disruption of the normal anatomical relationship that exists between the femoral head and acetabulum. This relationship is imperative, as it is the dynamic association of these two structures that provides the necessary stimulus for normal development of the joint in the first few months of life. DDH has superseded Congenital Dislocation of the Hip (CDH) as the appropriate term to be used. This is because as our knowledge of the natural history for both normal and abnormal hip development has improved it has become apparent that dislocation of the hip is a dynamic process encompassing the antenatal, perinatal and postnatal periods. DDH therefore incorporates:

- **Ligamentous laxity.** A transient ligamentous laxity that is thought to be an effect of transplacental maternal hormones lasting for a few days after birth.
- **Acetabular dysplasia.** A consequence of incomplete bony modelling leaving a shallow, flattened socket.
- **Subluxation.** An incompletely covered femoral head. The factors initiating this migration of the femoral head have not been clearly identified but central to it is the deficient cartilaginous roof of the acetabulum.
- **Frank dislocation.** Dislocation occurs when the femoral head loses contact with the acetabulum and rides postero-laterally over the fibro-cartilagenous rim.

Incidence

The published incidence of DDH in the literature varies considerably. This is because it depends on factors such as when the baby is examined and whether it is diagnosed on clinical examination alone or following ultrasonography. DDH by clinical examination occurs in 1 to 2% of all live births¹. There is an ethnic variation with higher rates, for example, in Scandinavian populations, and lower rates in Asian groups. This reflects both genetic factors and the type of infant swaddling applied, with legs held together in the former and legs held in wide abduction in the latter².

Incidence of dislocation with time of examination

Barlow³ in 1962 demonstrated that:

- On day 1, 1 in 60 babies examined had evidence of subluxation on clinical examination.
- By day 7, 68% of these were normal increasing to 88% at 2 months, leaving only 1.5 per 1000 still unstable after this time.

Risk factors

- **Females** In most series, there are about 5 to 8 girls affected for every boy (possibly reflecting the influence of female sex hormones).
- **Breech presentation** The treatment rate in the series from Coventry, UK⁴, which were universally screened with ultrasound, was 20/1000 for female breech presentation vs. 3/1000 for male breech vs 5.1/1000 for females with no risk and 0.28 for males with no risk.
- **First degree relative with DDH** The treatment rate in the series from Coventry which were universally screened with ultrasound was 12/1000 for males with a family history vs. 12/1000 for females with a family history⁴. There is a risk of 60/1000 with one affected sibling, 120/1000 with one affected parent, 360/1000 if there is both an affected parent and an affected sibling⁵.
- **Oligohydramnios, talipes, torticollis, birth weight >4kg.**

Table. Ultrasonographic abnormality and treatment rates From Bache⁴

Risk factor	US abnormality per 1000 hips	Treatment rate per 1000 births	95% confidence interval	Treatment rate per 1000 hips
Male - no risk	13	0.28	0.08 - 0.72	0.21
Male - all	14.5	0.53	0.23 - 1.05	0.43
Male - breech	39.4	3.75	0.46 - 13.49	2.81
Male - FH	61	12.2	1.48 - 43.36	9.15
Female - no risk	62.7	5.1	3.97 - 6.46	4.32
All females	66.5	5.86	4.68 - 7.26	4.99
All breechs	77.8	12.82	7.19 - 21.06	10.68
Female - breech	109.9	20.41	10.91 - 34.65	17.27
All - FH	133.5	11.87	3.24 - 30.11	10.39
Female - FH	202.3	11.56	1.4 - 41.14	11.56
BWt > 4Kg	61.1	5.39	2.37 - 10.75	4.58
BWt < 4 Kg	38.4	3.06	2.21 - 4.14	2.69
Male > 4 Kg	27.1	0	0 - 3.19	0
Male < 4 Kg	12.7	0.44	0.09 - 1.29	0.33
Female > 4 Kg	119.2	14.97	6.61 - 29.86	12.35
Female < 4 Kg	55.8	6.11	4.38 - 8.2	5.28

FH = Family history of DDH; BWt = Birth weight

Aetiology and Pathophysiology

The acetabulum is derived from mesoderm appearing at the end of the 4th week of intra-uterine life. It initially is a shallow socket, deepening as a result of the pressure applied by the spherical femoral cartilaginous head. The constant defect in DDH is a deficient development of the osseous buttress found at the postero-superior rim of the acetabulum. This lies along the axis of weight transference and is a feature of animals whose hind limbs have a postural function.

Consequences of dislocation

- **Bones.** The acetabulum is shallow, occupied by an overgrowth of fibro-cartilage with the femoral head positioned in a depression in the dorsal ilium. The femoral head is flattened, usually with delayed ossification, and the neck of the femur is shortened and anteverted such that after reduction the patella appears medially directed due to the internal rotation.
- **Soft tissue.** The capsule becomes a suspensory ligament for the pelvis, having to support body weight, with predictable hypertrophy and stiffness.
- **Muscles.** Shortening of the hip adductors and hamstrings can contribute to the difficulties in reducing the joint operatively when surgery is required for a missed case of established hip dislocation.

- **Clinical consequences** of undiagnosed hip dislocation are mainly moderate to severe osteoarthritis. This may occur as soon as the second decade.⁶ Other consequences include back pain, knee pain, limb length discrepancy and gait abnormalities.

Clinical Diagnosis

In the neonatal period, the diagnosis of DDH is made by physical examination. Frank dislocation is not common and manifests on examination as hips that are difficult to abduct. On routine newborn examination, you are most likely to find hips in which you can feel movement or that are dislocatable over the posterior margin of the acetabulum.

Neonatal Clinical Screening: for DDH should be done as part of the routine examination of the newborn, ideally after day 2. The baby should be warm and relaxed on a firm surface. The hips are assessed using the Barlow and Ortolani manoeuvres³. The essence of the examination is to try and dislocate the flexed hip with a postero-lateral movement of the proximal femur (Barlows Manoeuvre). Then to feel the movement of the reduction of the dislocated hip back into the acetabulum (Ortolani) by moving the femoral head anteriorly whilst the hip is abducted

Infant Clinical Screening: DDH is dynamic process and examination may be normal in the newborn period and become abnormal later. Examination of the hips should be a routine part of all infant screening examinations. The physical signs change as the infants grows and after the age of three months the Barlow and Ortolani tests may be unreliable. Other physical signs and symptoms should be sought and include:

- Asymmetric thigh or gluteal folds
- Shortened Leg
- Prominent greater trochanter
- Limited abduction
- Abnormalities of walking or gait.

Diagnostic imaging

- **X-ray.** The predominantly cartilaginous nature of the bones make x-rays an unsuitable means of assessing structure in the first few months after birth, although frank dislocation will be apparent on x-ray. However after the first 4 months a number of useful

measurements can be made as well as assessment of femoral epiphysial ossification which is characteristically delayed in DDH.

- **Ultrasound** is now established in the static and dynamic analysis of the neonatal hip. The position of the femoral head, degree of acetabular coverage, stability on dynamic testing as well as confirmation of a satisfactory location for a splinted hip are all achieved with non-invasive ultrasonography. There are several established techniques for assessment of the neonatal hip.⁷ The method commonly used at the RPA is measurement of the percentage bony coverage (PBC) of the femoral head.

At birth, the mean percentage bony coverage is 55% in girls and 57% in boys. The lower limits of normal (-2SD) was 44% in girls and 47% in boys.⁸ This study of Tejerson et al⁸ also highlighted how the percent coverage improves in almost all cases over the first 6-8 weeks. By 2 months only 2 of 53 babies, who had had normal clinical examination at birth but low percentage coverage on ultrasound, still had abnormal ultrasound findings.

Screening

Detection of DDH is now established through screening programs in many countries. In the UK routine clinical examination of the hips was introduced in 1969 with both Ortolani and Barlow manoeuvres performed within the first 24hrs of birth and repeated periodically in the first year. Despite these tests, late established dislocation of the hip persists at similar rates⁵. It is not clear if these cases represent "missed" diagnosis or late dislocations. There are several published reports of infants with normal neonatal clinical examination who went on to develop DDH, consistent with the hypothesis that the disorder may develop post-natally^{9,10}.

Using ultrasound as a screening tool for DDH has been under study for some time but its role has still not been clearly delineated. The majority of maternity units have access to ultrasound imaging.¹¹ It can be applied universally to all new born infants or targeted to high risk groups. One of the problems that has been identified with ultrasound screening is an increased rate of diagnosis. The "sonographic" incidence of DDH has been quoted as high as 55.1/1000.¹² This has led to concerns of over treatment. Classification systems have been devised on the sonographic appearance of the neonatal hip and when these are applied, clinically relevant DDH incidence drops dramatically (13% -to 2.3% in one series)¹³

Clinical trials of ultrasound in DDH

There is a paucity of good quality evidence available.

1. What type of follow up after neonatal hip instability?

The UK hip trial randomized infants with clinical hip instability to ultrasonographic follow-up or follow-up with clinical examination alone. There was no significant difference in radiographic incidence of DDH (hip dysplasia, subluxation or dislocation) between the two groups.¹⁴

2. What type of ultrasound screening is most effective ?

- **Rosendahl et al¹** studied a cohort of 11,925 newborn infants allocating them randomly to receive either general, selective, or no ultrasound screening in addition to the standard clinical examination. All examinations were carried out at < 48hrs of age.
 - The treatment rate was greatest in the general ultrasound group (Splintage rates: general US = 3.4%, selective US = 2%, no US = 1.8%; P < 0.001)
 - The follow up rate was highest for non-treated infants in the ultrasound screening group because of inconclusive early findings.
 - Late DDH was less prevalent in the general ultrasound screening group, but did not reach significance.
- **Holen et al¹⁵** randomised 15,529 babies to universal clinical and ultrasound screening or clinical screening with selective ultrasound used in those at risk (instability or uncertain clinical findings, family history, breech position and foot deformities).
 - 11% of the selective ultrasound group had ultrasounds.
 - There was one case of late diagnosed DDH in the universal group vs 5 cases in the selective group, not a significant difference (p=0.22)
- **Roovers et al¹⁶** describe DDH detection before and after introducing a program of post-neonatal hip screening. 5170 babies were screened at 1,2 and 3 months of age and this was compared with 2066 babies that were screened clinically in child health centres.
 - The sensitivity of the US screening was higher but so was the treatment rate.
 - Detection was at an earlier age in the ultrasound screened group.
 - The rate of late diagnosed DDH was lower, but not significantly so, in the ultrasound screened group
 - The authors raise concerns that universal US screening may be leading to over treatment without clear benefits in terms of reducing cases of late diagnosis.

3. What are the cost implications of ultrasound screening ?

Rosendahl¹⁷ then followed up the above study with an analysis of the cost effectiveness of the three methods used and showed that total costs for each varied by less than 5%. When broken down by proportion of total cost contributed by late DDH treatment: General US = 22%, selective US = 65%, no US = 65%.

The UK Hip Trial looked at costs of all infants with hip instability randomised to either ultrasound assessment or clinical examination alone. There was a trend towards lower overall costs in the ultrasound group but this did not reach significance¹⁴.

4. How effective can clinical examination be?

Reports on the literature on the effectiveness of clinical screening vary in their findings. Some authors such as Hadlow¹⁸ and Goss¹⁹ suggest that with experienced examiners, neonatal clinical screening will miss very few cases of DDH. Hadlow examined 20,657 infants and reports 2 missed cases of CDH¹⁸. Goss¹⁹ studied 5166 babies in order to determine the incidence of DDH in Australia and as to whether all late DDH cases could be eliminated by an effective clinical screening program. They found 100 babies with hip instability were tested, (77%% female, 26% breech, 25% family history) = 19.4/1000. There was one late diagnosis of DDH diagnosed at 4 months, which they put down to failure of the screening process, not the test itself.

This experience is not consistent with other observers. Bialik et al¹⁰ reported on regional data where all babies were examined shortly after birth by both an orthopedic surgeon

and a paediatrician. Between them, they detected 76 cases of neonatal hip instability from about 18,000 births. However there were a further 36 babies on whom the neonatal examination was normal who required later treatment for DDH. Fiddian et al²⁰ reported their 10 year experience of screening by specifically trained physiotherapists. They detected 255 cases of neonatal hip instability in 42,000 babies screened however 13 cases of DDH presented late that had had normal newborn examination.

Other reports have showed that the incidence of late diagnosis has not changed since the introduction of clinical screening programmes^{21,22}

5. Is splinting safe ?

Gardiner has examined radiologically the effects of early splinting on hip development. 76 infants with dislocatable hips were randomised to further ultrasound surveillance or immediate splinting.^{23,24} Results showed:

- Dislocatable hips can be safely assessed with a repeat ultrasound examination at 2 weeks of age without an adverse outcome.
- Splinted hips showed poorer epiphysial maturation but these changes were less marked at the age of 1 year.
- The specificity of hip ultrasound in the first week of life (approx. 70%) makes it unsuitable as a tool for primary screening.

The major long-term risk of splinting is avascular necrosis of the femoral head. This was studied by Bradley who found a 3.8% rate of avascular necrosis in children splinted with either a Pavlik harness or Von Rosen splint.²⁵

What we do at RPA Hospital

There are not the resources to institute a universal ultrasound screening program notwithstanding the equivocal evidence of benefit. The mainstay of screening therefore will continue to be clinical examination (as described above) in the routine newborn examination. We will however use ultrasound screening in cases that have significant risk factors. Residents should ask the duty registrar, fellow to review any case where they are uncertain about the physical findings of the hips. or consultant So:

1. *If either hip is dislocated*, in other words, cannot be abducted:
 - arrange hip ultrasound and if diagnosis confirmed refer to Dr Angus Gray (Sydney Childrens Hospital. 9650 4983)
2. *If either hip is dislocatable* on routine newborn examination:
 - book into Dr Jeffery's Friday clinic for a repeat examination at about one week of age.
 - arrange hip ultrasound and if either hip is still dislocatable at that clinic, the baby will be referred to Dr Angus Gray (Sydney Childrens Hospital. 9650 4983) for bracing.
 - If neither hip is dislocatable, then the baby will be reviewed by Dr Jeffery again at 6 weeks after a hip ultrasound.
3. *If either hip has movement detectable* but is not dislocatable:
 - arrange hip ultrasound at 6 weeks of age and make appointment after ultrasound for RPA hip follow up service.
4. *If a baby has a family history of DDH* in a 1st degree relative, or is a female breech

- arrange hip ultrasound at 6 weeks of age and make appointment after the ultrasound for RPA Hip follow up service(Dr Jeffery's clinic).
5. *If a baby has a 'clicky' hip*, this is not an indication for further screening or follow up as long as there is no movement in the hip joint. If in doubt, get the baby reviewed by a senior member of the medical staff.

Action on basis of ultrasound findings at 6 weeks:

There is a lack of normal data for percent bony coverage (PBC) at 6 weeks (the normative data is all at birth). However considering the normal range at birth and the natural growth of percent bony coverage, we should use a cut off for normal of 50%. So:

1. *If PBC is 50% or over at 6 weeks* and clinical examination is normal, no further investigation is indicated.
2. *If PBC is between 40 and 50%* and clinical examination is normal, then repeat ultrasound at 4 months.
3. *If PBC is less than 40%* but the clinical examination is normal, repeat in a further month (i.e. 10 weeks of age) and refer to Dr Angus Gray (Sydney Childrens Hospital. 9650 4983) if the coverage is still less than 40%.

RPA Hospital Hip Screening Follow up:

All babies requiring follow up for hip examination or screening will be seen in Prof Heather Jeffery's clinic on a Friday morning. When booking the baby into the clinic please inform the secretary that this is a hip follow up. The hip screening ultrasounds are usually performed on a Wednesday or Thursday, it is VERY important that you book the baby into the hip clinic for the second Friday after the date of the ultrasound scan not the first Friday.

The only exceptions to the above are babies who are being discharged from HDU or SCBU and need a screening hip ultrasound. If the baby is being followed up anyway by one of the neonatal consultants then the hip screening follow up can be done as part of that appointment. If the baby would not have otherwise been followed by one of the neonatal consultants then they should be booked into the hip clinic as outlined above.

Treatment

- Subluxable hips with a positive Ortolani sign are treated with an abduction splint (after consultation with Dr Gray), either with a Von Rosen splint or a Pavlik harness. The latter (used by this hospital) is a soft splint that holds the hips in flexion and abduction. Follow up ultrasounds can be obtained with the harness in place.
- Once a reduced hip is shown to be stable and of a mature configuration the harness can be removed. Surgery is therefore reserved for those patients having failed conservative therapy.

Summary

- DDH represents a spectrum of dynamic abnormalities of the hip joint.

- Clinical screening will detect neonatal hip instability at a certain degree of severity however even with experienced examiners there will be cases of late presentation of DDH (with dislocation) in whom the newborn clinical examination was normal.
- Universal ultrasound screening at birth result in a statistically insignificant trend to reduced rate of late presentation DDH but at the cost of a high false positive rate with potential for over-treatment and consequent clinical harms (such as avascular necrosis).
- The AAP clinical practice guideline does not recommend universal ultrasound screening.
- The large majority of hips that are sonographically abnormal at birth become normal over the first 4 to 8 weeks of life.

Key points

Screening with ultrasound increases treatment rates because it has a higher sensitivity than clinical examination, but with only a trend to reduction of late DDH cases.	★★★★★ ¹
Ultrasound at 2 weeks of age is optimal for high risk group screening (classically breech presentation and first degree relatives, possibly all girls and boys with risk factors). Ultrasound at 6 weeks of high risk infants with normal examination does not affect outcome	★★★★ ^{1,4}
Late DDH is very rare if both ultrasound and clinical examination are normal.	★★★★ ^{1,10,11}
The great majority (> 90%) of dysplastic/unstable hips improve spontaneously without intervention.	★★★★ ^{3,10}
There is no difference in total program costs between general ultrasound screening, high risk group screening or clinical examination alone.	★★★★ ^{17,19}
Screening by clinical examination has not reduced surgery for late DDH	★★★ ^{12,24}

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