Is our current policy of selective ultrasound screening for developmental dysplasia of the hip justifiable?

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INTRODUCTION

Developmental dysplasia of the hip (DDH) is a congenital condition in which there is an abnormal development of the hip joint, causing a misalignment of the femoral head with the acetabulum. This affects 0.01-0.2% people. It was first depicted by Hippocrates, who suggested that 'uterine pressures and birth trauma' as potential aetiologies.[1] 'Family studies have also provided evidence that there is genetic predisposition to DDH based on polygenic-multifactorial inheritance'.[1,2] DDH can include an array of 'hip abnormalities including dysplastic, subluxated, dislocatable, and dislocated hips'.[3] If left untreated DDH is associated with the development of gait deformities, premature osteoarthritis and chronic pain.[4] The aim of screening is to detect cases of DDH as early as possible to avoid consequential hip malformation, pain and degenerative arthritis. DDH is an issue of controversy across healthcare professionals, although unanimously agreed upon as vital in identifying those requiring further investigation and possible treatment, whether to screen universally or only screen those deemed to have an acceptable risk factor is under question.

Selective ultrasound screening was introduced in the UK in 1969 as part of the diagnostic process in newborn babies to encourage early detection.[5] Early diagnosis is essential for a better outcome of treatment,[6] as often signs do not present until walking age, which at this late stage requires surgery. Screening, in the UK, takes place twice: within 24 hours of birth or on discharge from the hospital; and at six weeks of age.[5] If an abnormality is detected at this early stage, a Frejka splint or Pavlik harness is issued, which works by keeping the hips abducted to reduce the femoral head and keep it in the acetabulum. Screening can be universal (screens all neonates) or selective (targets at-risk neonates).

The risk factors for developing DDH are shown below in table 1, which are taken into account when screening for DDH, although the majority of infants with DDH do not have associated risk factors.[7]

Abnormal findings on clinical examination
– instability
– click
– limited range of movement

Family history of developmental hip dysplasia

Breech presentation at birth
– vaginal delivery
– caesarean section

Postural deformity (moulding)
– torticollis[9,10]
– foot deformity

Other
– sacral dimple
– genetic basis

Table 1 Risk factors for developmental DDH[8]
**Note – although females have a higher prevalence of developmental DDH than males have, sex alone is not considered a risk factor.[8] Other risk factors include a birth weight of over 4kg, and an increased maternal age.[2]*

Epidemiology

The incidence of DDH ranges from 0.7-7 per 1,000.[11] Its prevalence varies 0.1-2 per 1,000 in its moderate to severe forms;[12-15] differences in reported incidences may be explained by the sensitivity of different screening methods (clinical only, selective ultrasound or universal ultrasound), interpretation of results from the screening and treatment chosen.[16] A much higher prevalence, 40 to 60 per 1,000,[15,17-19] exists if the milder forms, Graf type II, detected by ultrasound are included. The Graf method of classification defines DDH according to the size of the alpha and beta angles detected on ultrasonography. Graf type ii is defined in table 2. Absolute risk increases 70-120 per 1,000, when the baby is both female and born by the breech position. There is a lower prevalence in the Afro-Caribbean community due to African traditions where infants are carried on the mother's back or side, at mother's pelvis, thus abducting the hips. By contrast the practice of swaddling, practiced by some cultures, in which hips are adducted by tight binding, is associated with a higher prevalence.[20]

Controversy exists around treatment of DDH detected to be in the milder form. There is abundant evidence that 60-80% of cases detected resolved spontaneously.[21-23] As a result, clinicians and surgeons vary in treatment, some treating all those identified whilst others only offering monitoring.[24]

Overview of normal paediatric anatomy

In the normal adult hip, the acetabulum (socket) is composed of portions of the three pelvic bones (ilium, ischium and pubis) which are ossified together. In the paediatric hip, the three pelvic bones have not fused together, and there is a larger component of non-ossified cartilage. The development of the hips occurs whilst in the uterus, where the head of the femur should sit comfortably within the acetabulum. In DDH, the acetabulum is shallower, and the femoral head may displace out of the hip socket. Figure 1 shows the difference between a normal hip and that affected by DDH.

Initial screening of newborns takes place by physical examination, using the Barlow and Ortolani tests, with the infant lying supine. The Barlow test involves adduction of the hips to the midline, whilst the examiner gently presses on the knee, guiding their pressure posteriorly. A positive Barlow test allows the examiner to feel the femoral head dislocate out of the acetabulum. The Ortolani test entails a similar procedure where the examiner flexes the hips and knees to an angle of 90°. Force is then applied on the greater trochanter, in an anterior direction whilst gently abducting the infant's legs.

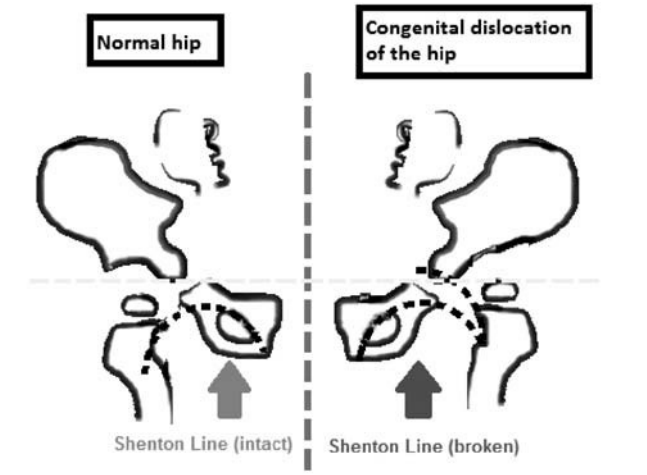


Figure 1 Comparison of hip anatomy

A positive result will reveal a clicking sound, caused by the femoral head moving anteriorly into the acetabulum. Asymmetry in abduction is also checked and full range of movements.

Ultrasonographic screening of the hip is undertaken if there are abnormal findings on physical examination. There are different approaches to the ultrasonagraphic assessment of the paediatric hip, but the standard method used in the UK is Graf's method, which classifies hips into four main groups ranging from normal to severely dysplastic and dislocated hips. This is done by the measurement of the 'acetabular inclination angle (α) and a cartilage roof angle (β)'.[16] Table 2 summarises Graf's classification.

Type I Type Ia Type Ib	Alpha angle >60° (normal) Beta angle <55° Beta angle >55°
Type II Type IIa Type IIb Type IIc	 Alpha angle 50-59°, age up to 12 weeks Alpha angle 50-59°, age greater than 12 weeks Alpha angle 43-49° Beta angle <77°
Type III	Alpha angle <43° Distinguished on the grounds of structural alteration of the cartilaginous roof
Type IV	Alpha angle <43° Dislocated

Table 2 Graf's classification of developmental dysplasia of the hip[25]

During an ultrasound, the infant is placed on their lateral aspect with hips flexed and abducted their legs flexed in the foetal position. Figure 2 gives a simple representation of what is seen on ultrasound of a baby's hip. Although it does not show the beta angle (which measures the cartilaginous coverage), the alpha angle is shown, which passes through the bony roof and middle of the head of the femur).

The criteria for treatment, as assessed by ultrasonography, is an alpha angle of under 55° in a baby of six weeks, or an alpha angle of less than 60° in a baby of 12 weeks. Babies of six weeks of age with an alpha angle of between 56° and 59° have an immature type of hip (Graf type IIa) – these babies do not require treatment at that stage, but are rescanned six weeks subsequently.

When mild DDH is detected a Tübingen hip flexion orthosis device is issued, to be worn for 23 hours of the day (one hour spared for nappy changing and bathing purposes), and a re-scan arranged in six weeks.

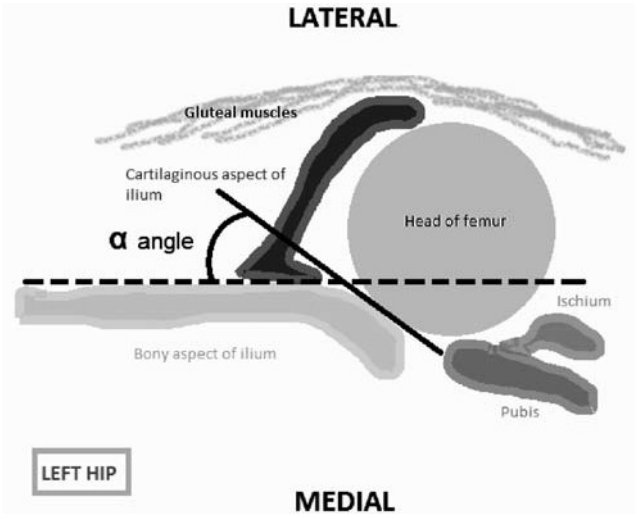


Figure 2 Ultrasound anatomy

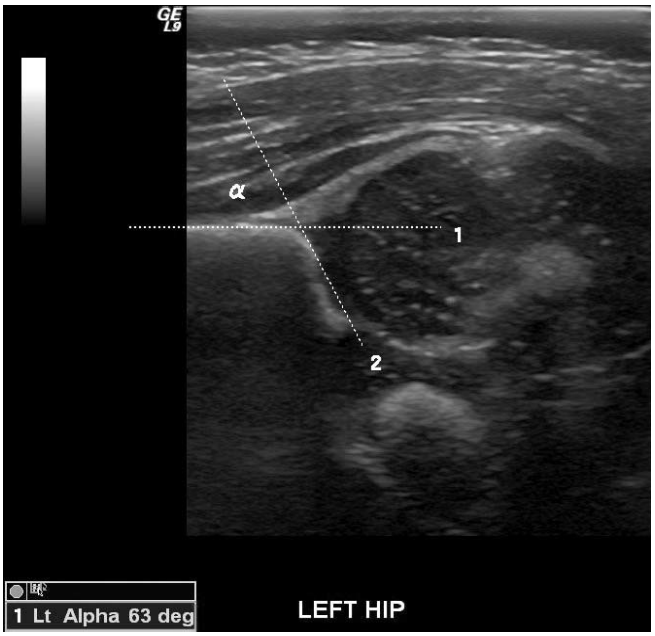
Graf suggested treatment when the alpha angle fell below 45° and monitoring of an infant eight weeks or younger with an alpha angle below 55°.[26] In Lancaster, the standard for measurement is an alpha angle greater than 60% at 12 weeks. Treatment for DDH can be through monitoring, a harness (eg a Pavlik harness), use of a Frejka pillow or Hip spica, or surgery.

DISCUSSION

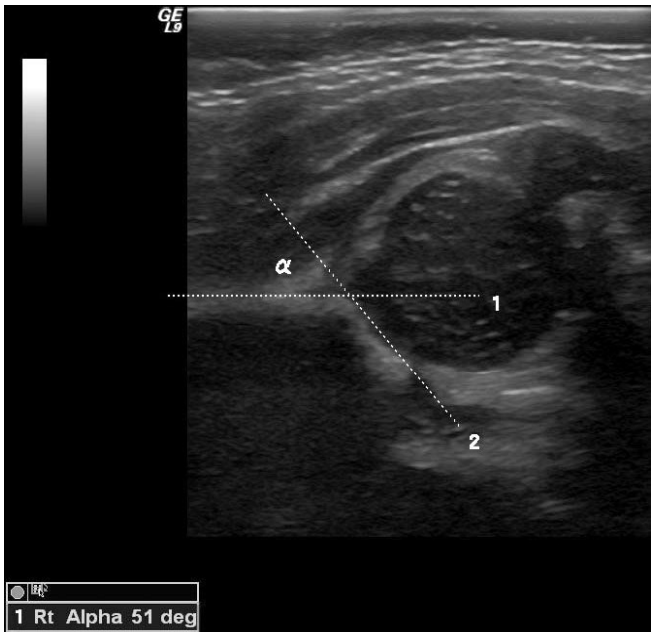
Although it is agreed upon that screening for DDH is imperative, controversy and disagreement lies around who and when to screen, as well as when to treat.

The effect of ultrasonography on reducing the overall number of surgical interventions for DDH is disputable. The sensitivity of ultrasound as well as of physical examination will never be 100%, so there will always be unidentified newborns. Although this does not imply its uselessness. The principal problem with sonographic screening is its abililty to detect 'minor abnormalities of both stability and acetabular development that will resolve by a later stage without treatment'.[8] A 2011 study,[27] in west Middlesex, on selective ultrasound screening, suggested that screening of an infant before the age of six weeks made it hard to distinguish between immature hips and dislocated hips. Therefore, ultrasound screening should be performed after the period of six weeks, to reduce the number of infants wrongly diagnosed with DDH.[27]

There is also a real danger that overtreatment can result in complications such as avascular necrosis. These false-positive results also reduce the cost-effectiveness of the screening programme.[8] In Germany, it was found that the use of universal screening with ultrasonography 'reduced the rate of surgical intervention for DDH by 80%',[28] suggesting that there was a huge benefit in the use of ultrasonography. A compromise approach is to limit the ultrasound examination to the high-risk infants as defined in table 1 – so-called selective screening, rather than embarking on population-based screening.



(a) Shows normal hip ultrasound with Graf alpha angle of greater than 60° (Graf type I)



(b) Shows hip with mild shallowness of acetabulum, with Graf alpha angle 51° (Graf type IIa)

Selective screening, can potentially miss detection of true-positive results. Furthermore selective screening with ultrasonography 'does not significantly lower the point prevalence of late diagnosis or late presentation'.[27] Worldwide, the point prevalence can vary from 1.5 to 20 per 1,000 live births.[11]

The effectiveness of the current screening protocol in the UK is undecided. Some reports suggest there has been a decrease in the overall number of surgeries performed on infants,[30-33] whilst others have seen no difference.[34-36]

A study performed in 1998 showed that 'despite screening, the incidence of surgery was high and in most children congenital dislocation of the hip had not been detected by screening or surveillance before the age of three months'.[5] Conversely, a study performed in Germany showed a 75% reduction of operative procedures once the national screening programme was started, scanning all high-risk newborns and everyone else after four to six weeks.[28] A randomised trial in which 15,529 newborns were subjected to either universal or selective screening in Norway found that only one subject was detected late with DDH in the universal group compared to five in the selective group, giving a 'rate of 0.13 and 0.65 per 1,000, respectively'.[14] This suggests that a significant proportion of infants with DDH are being missed when selective screening is chosen.

Interpretation of ultrasound requires a subjective evaluation. There is a variation in definition of true-positive results from those performing it. Also, possible harmful risks could include 'operator-induced hip pathology' through rigorous testing. It has also been suggested that there is increased risk of particular cancers from the elevated exposure to radiation during follow-up radiographic tests,[37] and an element of 'parental psychosocial stress' from true-positive results and unfamiliar treatment regimes, as well as false-positive results that can lead to unnecessary and potentially harmful follow-ups and intervention.[37]

The biggest issue surrounding the use of universal or selective ultrasound screening is the cost-effectiveness of the procedure, which is measured by looking at how many cases are diagnosed per 1,000 in relation to its funding. Currently, this is very low.

Every DDH case is different, so it is difficult to use selective screening when there are no regular trends in the group, and anomalies can occur. Therefore, universal screening is a more effective and equal way of screening. Also, although each surgeon has their own interpretation of when to perform surgery or when to screen, and there is no unified approach to dealing with DDH, in general there is a low incidence of pathologies induced by early/unnecessary treatment.

LIMITATIONS AND FUTURE APPLICATIONS

DDH is an issue of controversy with many different professional opinions as for the screening effectiveness. Rather than handling patients on a case-by-case basis a more unified approach is needed. A main limitation was that diagnosis of DDH was completely based on the orthopaedic surgeons judgement, there was a distinctive lack of a standardised diagnostic criterion for DDH.[24] There is no consensus on when to treat either.

Steps to improvement of the diagnosis of DDH would be to have a 'gold standard' acceptable diagnostic test/criteria used universally by all professionals. This would help standardise the identification of DDH. A universal criterion would also allow audit and developments for improvement to be made.

CONCLUSION

The low point prevalence in the UK could warrant reasonable justification for selective screening of DDH. But arguments of missed diagnoses also arise and questions of whether the screening programme is fair for all infants with or without risk factors surface.

There are positive and negative aspects about selective and universal screening, but a balance has to be made to prevent unnecessary radiation exposure of infants and to ensure the maximum benefit arises from the most minimal cost. The current selective screening programme in the UK is sufficient enough to detect the most vulnerable infants to develop DDH. It can be concluded that those without any risk factors, which could potentially be missed due to the selective screening, are most likely to have a mild form of DDH which is likely to resolve as the infant gets older.

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