Current Concepts Review

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Universal or selective ultrasound screening for developmental dysplasia of the hip? A discussion of the key issues

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Abstract

Purpose To summarize recent developments and provide recommendations as to whether universal or selective programmes are advisable.

Methods A literature review was performed and preference given to studies with higher levels of evidence. All programmes reviewed included clinical screening.

Results Recent studies underline the need for high quality screening programmes to promote the early detection of developmental dysplasia of the hip (DDH). A small number of cases may be missed clinically but with universal ultrasound screening programmes the late presentation rates appear to be virtually zero. Contemporary studies show treatment rates with universal screening programmes which are now lower than those with selective ultrasound. There is little agreement over the criteria used for selective programmes. Alternative outcome measures, such as the first operation rate or the percentage undergoing major (open) surgery are both lowest with universal ultrasound screening programmes. Furthermore, a significant reduction in the rate of surgery for DDH later in life was seen after the introduction of universal ultrasound screening, whereas the defined criteria for selective screening may not detect the majority of patients who require late surgery. Abduction bracing with modern orthoses is associated with a zero rate of avascular necrosis (AVN), whereas closed reduction techniques have an overall risk of 10%.

Conclusion On clinical grounds, if future studies confirm that hip abduction in flexible orthoses is not associated with AVN, it may be time for a paradigm shift of screening for DDH towards a universal ultrasound protocol. The costs associated both with each type of screening programme and with the management of late presenting cases are also important but may be secondary to clinical benefit.

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Introduction

Developmental dysplasia of the hip (DDH) is a term that encompasses a wide spectrum of pathology ranging from mild acetabular dysplasia with or without instability to a complete dislocation at birth which may or may not be reducible. Many aspects of the condition ranging from definition through diagnosis and management remain controversial, and perhaps the only certainties are that early diagnosis is better than late and that avascular necrosis (AVN) of the femoral head represents a poor outcome. It is well recognized that clinical examination is not infallible at detecting neonatal cases and this article discusses the ways in which the use of ultrasound screening can aid diagnosis. The paper summarizes recent developments and provides recommendations as to whether universal or selective programmes are advisable. Preference has been given to studies with higher levels of evidence.

The definition of DDH and its natural history

The exact definitions and the methods of diagnosis associated with the well-recognized range of morphological and clinical disorders that constitute DDH are variable and the natural history is surprisingly poorly understood. This relates primarily to the lack of good long-term studies that follow all categories of infantile hip instability to skeletal maturity and the scarcity of well-designed prospective studies.[1]

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DDH is diagnosed by physical examination supplemented by the use of static and/or dynamic ultrasound assessment.[2] The prevalence of clinical instability is known to be age-dependent and does diminish in the first week of life as a consequence of increasing muscle tone[3] and the changing hormonal environment. Therefore, the vast majority of clinically unstable hips at birth stabilize within three months. Persistent acetabular dysplasia reportedly leads to degeneration of the hip joint and premature osteoarthritis.[3,4] Mild dysplasia may not become clinically apparent until adult life (or may indeed never be manifested clinically), whereas severe dysplasia will most likely become symptomatic earlier (in adolescence).[5,6] The incidence of both is largely unknown. Whilst more than 90% of immature hips will have improved by the age of six weeks, it is also true that hips regarded as normal in the neonatal period have a 0.2% risk of deteriorating over time.[7] Surprisingly, controversy still exists about the influence of treatment on the natural history both in terms of improving early dysplasia and avoiding later degenerative change.

Persistent childhood dysplasia and neonatal hip instability predispose to adult hip disease. The Norwegian Medical Birth Registry was correlated with the Arthroplasty Registry;[8] and when adjusting for gender and year of birth, there was a 2.6-times increased risk (95% confidence interval 30 to 105) for children with neonatal hip instability to undergo total hip arthroplasty. Of the 442 patients undergoing hip arthroplasty, 95 had the surgery due to degenerative joint disease from residual hip dysplasia, yet only eight had neonatal hip instability. It was suggested that there is a significant amount of hip dysplasia, with no physical findings in childhood, that becomes symptomatic in adult life.[2,6,9]

Clinical screening for DDH is considered effective in the presence of structural or functional abnormalities but it is not always able to detect abnormalities in stable hips or indeed identify all unstable hips. Thus, all clinical screening programmes have a late presentation rate.[10-14]

Ultrasound imaging methods are able to detect the different stages of DDH from a mildly dysplastic, concentrically located, stable hip to a severely dysplastic, unstable or even dislocated hip. Several classification systems are used to define categories of dysplasia and instability.

To date, there is no agreement about the best screening protocols for the neonatal hip: should it be clinical alone, ultrasound alone or a combination of both techniques?

Late presenting DDH

There is the potential to miss DDH in an infant particularly if the clinical examination is performed by untrained or inexperienced examiners and especially in the absence of risk factors.[15] There is no universally accepted definition of late presentation but late presentation and late diagnosis do lead to the need for more treatment, a higher complication rate and a poorer outcome.[16] A 12-fold increase in relative risk of requiring open reduction following late presentation was reported by Price et al.[17]. Children presenting before six weeks of age were treated successfully with abduction bracing in 84%, whereas 86% of children presenting after ten months eventually required open reduction surgery.[17]

Late diagnosis is more prevalent in children without risk factors implying that those with risk factors are assessed more closely at birth and identified more promptly.[18]

Screening strategies

The aim of all screening strategies is to identify all cases of hip instability and dysplasia promptly so that observation and/or early appropriate treatment leads to normal hip development. The clinical challenge is to separate the neonatal hip instability which resolves spontaneously from that which may persist and lead to symptoms and/or early degenerative change.[2] Overtreatment should be avoided particularly if there is a risk that such treatment might harm.

Clinical screening with or without selective ultrasound seems to be widely accepted whilst universal ultrasound screening remains controversial.[1,3,16]

Good universal clinical screening programmes confirm that experience and training are associated with less missed cases and lower treatment rates and that the examiner can be either a paediatrician, surgeon or physiotherapist.[12-14] In their clinical practice guideline summary for the American Academy of Orthopaedic Surgeons in 2014, Mulpuri and co-workers recommended clinical screening for DDH of children up to six months of age. Universal ultrasonography screening of newborn infants was not recommended; however, performing an imaging study before six months of age in selected infants with significant risk factors was suggested.[16] The problem is deciding what constitutes a significant risk factor: currently the only agreed risk factors are clinical instability and/or physical signs suggestive of a dislocated hip and a positive family history.

In contrast, universal ultrasound screening has been an integral part of the national surveillance programmes in Austria since 1992 and in Germany since 1996 (programmes which also include a clinical assessment).[19,20] There is evidence that these programmes are associated with a reduction in the number and the severity of surgical interventions related to DDH and that they are cost effective.[20,21] Opponents of universal screening emphasize the high rate of spontaneous correction and false positive screening results leading to over treatment and

a potentially higher rate of AVN of the femoral head.[1,16] Several review articles in the past scrutinized the most favourable screening methods,[1,16] but often excluded studies with lower levels of evidence or a focus on single outcome measures. The rate of late presenting DDH (false negative rate) is commonly used as an outcome measure in the evaluation of a screening programme.[11,22] This, however, requires large patient numbers to achieve significant results. Valid survey data have been suggested as a feasible alternative to assess the rate of first operative procedures on newborn hips during the first five years of life.[20] More recently,[7,23] the rate of major (open) surgery was used to evaluate the effectiveness of screening for DDH. Finally, the effect of hip screening on reducing the late surgery rate for DDH such as corrective osteotomies is a crucial parameter to define the value of this investigation. In recent years a series of articles have been published adding new aspects to this much-debated topic.[7,9,21,23-25]

For those who do not support a universal programme, some degree of selection takes place although there is no universally accepted definition of what constitutes a risk factor for DDH: we know that DDH is common in girls and firstborns but neither of these patient demographics constitute a risk factor. Overall, the greater the percentage of the population that is screened with ultrasound, the lower the late presentation rate is.[26]

There have been two prospective randomized controlled trials looking at selective versus universal ultrasound screening strategies. Both trials also included expert clinical examination as part of the protocol. Holen et al[11] included 15 529 infants in their study of screening strategies, either good clinical screening and universal ultrasound examination or good clinical screening of all hips and selective ultrasound examination. The rate of late cases in Holen et al's[11] study was 0.13/1000 with universal ultrasound screening and 0.65/1000 with selective screening. The difference in late detection was not statistically significant. One could argue that it may be of clinical significance and certainly of clinical benefit had the children been identified earlier in the selective group by having universal screening. It is of note that the late presenting cases in the universal ultrasound screening cohort had actually not undergone a scan, thus these cases represent a process failure rather than a technique failure.

Late detection of DDH was also assessed in the second study by Rosendahl et al,[10] where there were three matched study groups: general ultrasound screening, risk factor screening and only clinical screening. The ultrasound groups also underwent clinical screening. Late cases identified by group were 0.3/1000, 0.7/1000 and 1.3/1000, respectively and again although showing a trend towards less late cases where more ultrasound screening was performed, these differences were also not statistically significant.

Clarke et al[27] also demonstrated a decrease in late DDH presentation from 1.28/1000 to 0.74/1000 by using selective hip ultrasonography in a prospective cohort of patients over a 20-year period; a rate that remains higher than those using universal ultrasound screening programmes.[27] The selection criteria included clinical instability, breech presentation, family history and foot deformity.

In a recent prospective, longitudinal study[28] of a cohort of 64 670 live births in the United Kingdom, 31 infants were detected with an irreducible dislocation of the hip, representing an incidence of 0.48/1000 live births. Of these, 18 (0.28/1000 live births; 58%) presented late, despite universal clinical and selective ultrasound screening. Selection criteria included clinical instability, family history and breech position. Infants with torticollis, foot deformity (including metatarsus adductus) and oligohydramnios were also accepted, if referred. All the late presenting infants had a documented normal newborn clinical examination and no abnormality reported in the six to eight weeks check: 13/18 (72%) late presenting cases had no risk factors.

The most commonly accepted risk factors are clinical instability, family history and breech presentation but many studies also include foot deformity[11,27] which may even be postural.[29] Cephalic presentation and swaddling were recently identified as a risk factor for late-presenting DDH.[24] In Japan, tight swaddling in extension was strongly associated with DDH and the incidence dropped from 52.9/1000 to 5.6/1000 live births in Kyoto after an educational campaign.[30] Theoretically, modern methods of caring for infants in developed countries could also affect hip development[1] and late presenting DDH might also be a secondary phenomenon of an initially normal hip, in which case it may not be possible to detect in the neonatal period as it is not present at that stage. Late dislocation of the hip following normal neonatal clinical and ultrasound examination has been reported previously.[31]

Another aspect to consider is that reportedly, the relationship between the results of clinical and ultrasound examinations is low.[32] A total of 93% of clinically subluxable hips in Kyung et al's[32] study were normal or immature based on static ultrasound examination, and only 74% of dislocating hips and 67% of limited abduction hips presented with morphology below Graf IIa.

Treatment rate following screening programmes

Rosendahl et al's[10] study from 1994 found that general ultrasound screening resulted in a higher treatment rate (3.4%) than either selective ultrasound screening (2.0%) or clinical screening (1.8%). The higher rate with universal screening was statistically significant. However, observational studies

from countries with long-standing screening traditions have shown a decrease in treatment rates with increasing experience of a screening programme.[33] In a recent study from the first author's (R.B.) centre analyzing the hips of 28 092 newborns, a treatment rate of 1% was reported excluding cases with Graf type IIa hips. Surgical interventions (closed and open reductions) were necessary in approximately 10% of patients with DDH but the rate of major (open) surgery was only 0.07/1000 live births and substantially lower than in previous studies.[7]

Godward et al[34] reporting on the value of United Kingdom DDH screening initiatives in 1998 showed that the ascertainment-adjusted incidence of a first operative procedure for congenital dislocation of the hip (CDH) within the first five years of life was 0.78/1000 live births, which was similar to the prevalence recorded in previous studies of the condition in the United Kingdom. They also found that CDH had not been detected by routine screening before three months of age in 70% of children reported to the national orthopaedic surveillance scheme. Suggestions were that the clinical screening programme introduced in the United Kingdom in 1969 had been ineffective in lowering the incidence of surgery for the condition and that reassessment of current and alternative screening policies was required.

Assessing the rate of first operation on the newborn hip during the first five years of life across Germany, von Kries and co-authors[20] found an adjusted rate of 0.26/1000 live births, much lower than before the introduction of an universal ultrasound screening programme in Germany. In a study from South Australia the incidence of surgery for CDH in the first five years of life was 0.46/1000 live births, but only 0.19/1000 live births for those diagnosed three months or older, which made the authors suggest that the local clinical screening programme was successful.[18]

The incidence of a first operative procedure may be only of limited value as a method of assessing the effectiveness of a screening programme as the number of first operations for DDH may remain similar, regardless of early or late diagnosis.[7] Severe forms of primary DDH or secondary dislocations on the basis of severe hip dysplasia that may be reducible in a closed manner after early diagnosis, may require open reduction when diagnosed late.[7] Recommendations for ultrasound screening have considerable regional variation.[9] In Taiwan, 1.2/1000 live births were reported to have dislocated hips at birth or were late diagnosed DDH. In all, 40% of these children underwent surgery, 85% of which had major interventions.[23] This led the authors to conclude that their screening programme was inaccurate. The incidence of major surgery (or percentage of open reduction with or without osteotomy of all surgical cases) with clinical screening in other studies was 0.38 (47%) in Northern Ireland,[35] 0.3 (47%) in the United Kingdom[34] and 0.15 (29%) in South Australia[18]. Using universal ultrasound screening, the reported incidence of primary major surgery in Germany was 0.09 (33%)[20] and 0.04 (4.2%) in a recent study from Austria.[7] Some authors suggested that open reduction is associated with late diagnosis rather than failure of primary management.[15] Therefore, the number of open interventions seems a more reliable indicator and was reportedly zero after universal screening.[7,15]

In a retrospective study by Thaler et al,[21] which compared two five-year time periods before and after the introduction of universal ultrasound screening, a decrease of 76% in DDH-related surgery in children and adolescents was demonstrated in the second time period.

AVN of the femoral head

AVN is reported to be the most common and potentially harmful complication of both non-surgical and surgical treatment for DDH.[36] In a recent meta-analysis a mean rate of 10% AVN was identified five years after closed reduction.[37] This risk has been related to both the radiological severity of the dislocation, the amount of abduction used to obtain/maintain the reduction and the method of reduction.[38] Williams et al[39] reported the risk of AVN to be less than 1% with screening, early detection and the use of the Pavlik harness. In a long-term follow-up study of a randomized controlled trial from Norway, the authors did not find higher rates of AVN with the higher treatment rates, associated with the universal screening strategy.[40] Also the type and rigidity of the orthosis as well as the position during abduction reportedly play a role for AVN.[25,38] Recently, a zero rate of AVN was observed using the Tübinger abduction brace.[25]

Conclusion

Late detection of DDH is reduced by all screening modalities be they clinical, selective or universal ultrasound programmes, with the latter having the lowest late presentation rates and now low treatment rates too. The rate of first operation on the infant hip or the percentage of major (open) surgery have both been used as alternative outcome measures and both are lowest when universal ultrasound screening has been used.[7,10,20,25] A significant reduction in the rate of surgery for DDH later in life was shown after the introduction of universal ultrasound screening.[21] Iatrogenic AVN is a potential risk associated with abduction splinting, but recent studies using modern orthoses have a zero rate of AVN compared with an overall risk of 10% following a closed reduction.[1,25]

In 2007, Dezateux and Rosendahl[1] suggested that the extension of clinical screening to include universal ultrasound was not justified scientifically or ethically and they pointed out the need for randomized controlled trials to

assess the effectiveness and safety of neonatal screening and early treatment as well as high quality studies of the adult outcomes of DDH and the childhood origins of early degenerative hip disease. Such studies, however, require an observation period over many decades and furthermore, several tens of thousands of patients would have to be included to achieve significant results. Ambitious studies that planned to collect information on 80 000 British babies throughout their lives and to trace 100 000 children in the United States from birth have both ended early because of recruitment difficulties.[41]

In conclusion, it may be time for a paradigm shift of screening for DDH towards a universal ultrasound protocol.

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