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Screening in Developmental Dysplasia of the Hip (DDH)[☆]

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ABSTRACT

Screening for Developmental Dysplasia of the Hip (DDH) is a controversial subject. Screening may be by universal neonatal clinical examination (Ortolani or Barlow manoeuvres) with the addition of sonographic imaging of the hip (selective 'at risk' hips or universal screening in the neonate). In the UK, the NIPE guidelines recommend universal neonatal clinical assessment of the hip joints, a General Practitioner 6–8 week clinical 'hip check' and assessment clinically with sonographic imaging at 4–6 weeks for certain 'at risk' hips for pathological DDH.

The effectiveness and difficulties arising from the UK current screening policy (clinical and sonographic) are highlighted. The purpose of the review was to assess the risk factors and efficacy of diagnostic methods in DDH, based on longitudinal cohort studies of 10 years or more.

Conclusion: Hip screening in DDH does not meet most of the World Health Organisation's criteria for an effective screening programme and should only be considered as surveillance due to its low sensitivity and positive predictive value (PPV). There is a significant risk of over diagnosis and over treatment. There is no International consensus on screening in DDH. Pathological DDH is mainly a female condition and 'at risk'/General Practitioner screening identifies few pathological cases in male subjects. The General Practitioner 6–8 week 'hip check' has a very low PPV for pathological DDH and is of doubtful value in screening and diagnosis. Unilateral limitation of hip abduction is a time dependent and useful clinical sign in the diagnosis of pathological DDH. The majority of the previously considered 'at risk' factors are not true risk factors with little or no association with pathological DDH.

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Introduction

The traditional guidance for screening for Developmental Dysplasia of the Hip (DDH)/Congenital Dislocation of the Hip (CDH) in the UK was produced by the Standing Medical Advisory Committee (SMAC) in 'a little red book' and was active between 1969 and 2004.¹ In this document it was stated that 60% of CDH was associated with risk factors. These risk factors included:

Caesarean section
Foot deformities (including postural)
Intra uterine growth restriction (IUGR)
Family history,
Breech presentation
Oligohydramnios.

Unfortunately, there were no references from the literature published at the time, confirming or validating this statement. The guidance was flawed as it appeared to be based on expert opinion (level 5 evidence) without a robust or identifiable evidence base. The purpose of this review was to evaluate the current evidence base on the association of 'at risk' factors and the effectiveness of the current screening guidelines in the diagnosis of pathological DDH.

'An effective screening programme must identify the cases of DDH earlier than would have been identified in the usual course of care and must lead to better functional outcomes than late treatment. Any benefit should outweigh the harms of screening'.² DDH does not meet this aspiration or most of the criteria for an ideal screening programme⁴⁰ (Figure 1).

There are too many variables in this condition. These include:

The natural history of the condition is unknown
Currently there is no scientifically proven effective and accepted early treatment
Opinions on who should be treated are not agreed (National and International)

The condition should be important *
The examination & or treatment are/is acceptable to the patient *
Continuously rolled out and repeated *
Treatment and diagnostic facilities should be available *

There should be an effective & accepted treatment
Recognisable latent & early symptomatic stage should be present
Opinions on who should be treated are agreed
The natural history of the condition should be known

Guaranteed safety, sensitivity & specificity of the test (ideally > 90%)
Tests are inexpensive & simple
Cost effective programme

Those marked with a * are WHO criteria fulfilled in the DDH screening programme.

No recognisable latent and early symptomatic stage
Sensitivity and Positive Predictive Value (PPV) of the clinical screening tests are poor.

The term Congenital Dislocation of the Hip (CDH) was superseded by the new name of Developmental Dislocation of the Hip (DDH) in 1989.³ This was in recognition of the fact that not all cases of pathological hip conditions associated with DDH were present at birth. This opinion has important legal ramifications. If some hip joint conditions that are stable at birth deteriorate and are diagnosed at a later date as an irreducible hip dislocation, they cannot be considered to be 'missed' cases following negative neonatal clinical hip screening by a competent screener. DDH is a dynamic condition in which the hip abnormality may improve or deteriorate with growth.³ The spectrum of presentation varies from hip dysplasia, to reducible subluxation/dislocation and eventually irreducible hip joint dislocation.³ Neurological, neuromuscular, syndromes and skeletal dysplasias are excluded, as the hip abnormality is secondary to a primary pathology and is not idiopathic.⁴ The traditional outcome measure is that of irreducible hip dislocation.⁵

The diagnosis of hip pathology in DDH screening may be clinical and or sonographic. Clinically positive Ortolani and or Barlow⁶ hip instability manoeuvres will spontaneously resolve in 70–90% of cases within 2–4 weeks post-natally.^{6,7} The problem with clinical hip screening tests are that the Ortolani manoeuvre is only 60% sensitive^{14,15} and the Barlow manoeuvre has a PPV of only 22%.¹⁶ The Barlow and Ortolani manoeuvres failed to identify 66.7% of those hip joints that subsequently required surgical intervention.¹⁷ Sonographic screening of the hip joints may be universal in type in the neonate or be undertaken selectively as 'at risk' screening at 6 weeks of age. An unresolved issue is that sonographic diagnosis of DDH has a higher prevalence of abnormality than clinical diagnosis, raising the possibility of an over diagnosis of the condition which may lead to over treatment.² Sonographically 90% of Graf Type II hip dysplasias, <25% of Graf Type III hip dysplasias and <90% of Graf Type IV dysplasias may resolve spontaneously.^{8–12} There are numerous sonographic classification systems with a lack of validation in the diagnosis of DDH. The technique is operative dependant with variable Kappa scores (intra and inter observer error). The natural history of sonographic hip joint instability and dysplasia has not been accurately defined (no controlled clinical trials with and without splintage of the hip joints).^{18,30,31,33} Sonographic abnormalities could be considered to be a driver of over diagnosis in DDH: 'ability to detect smaller abnormalities axiomatically tends to increase the prevalence of any given disease'.³²

The effectiveness of the clinical hip screening programmes in the UK and North America have been disputed in the published literature.^{14–17} The 'late' or overall irreducible dislocation rates vary internationally from between 0.07 and 0.5 per 1000 live births^{18–22} and may be affected by various genetic and local environmental factors. A general review of the literature on screening in DDH shows that the national/international published levels of evidence are generally of an imperfect standard and the studies published are mainly uncontrolled and observational.²⁴ There are some systematic

Fig. 1 – 'Ideal' screening criteria, after Wilson & Jungner⁴⁰ (as applied to screening in DDH).

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