National Screening Committee

Child Health Sub-Group Report
Dysplasia of the hip

September 2004

DEVELOPMENTAL DYSPLASIA OF THE HIP (DDH) AND CONGENITAL DISLOCATION OF THE HIP (CDH)

The condition

1. The condition should be an important health problem.

DDH occurs in 1.2 / 1000 births. If untreated, it results in a limp, and later in life leads to pain and early onset of osteoarthropathy.

2. The epidemiology and natural history of the condition, including development from latent to declared disease, should be adequately understood and there should be a detectable risk factor, or disease marker and a latent period or early symptomatic stage.

DDH encompasses DDH and a range of developmental hip abnormalities, whose precise clinical significance is still uncertain. Without screening, some cases would be identified incidentally in the first few months of life and most would present clinically by two years of age. Outcome is thought to be better with early intervention, but not all the late diagnosed cases have adverse outcomes and some poor outcomes occur even with early treatment after screening. It is believed that early treatment reduces the need for surgery and minimises the long term disability due to early degenerative joint disease associated with the disorder.

Screening aims to identify DDH, but ultrasound detects other forms of DDH as well. Screening programmes involve clinical examination, ultrasound (either as a primary screen for all babies or as a secondary screen) and subsequent referral to an orthopaedic surgeon for treatment, which involves splinting or surgery. There are likely to be about 840 cases per year. Thus the number of adverse events prevented is probably much lower than the number of cases.

3. All the cost-effective primary prevention interventions should have been implemented as far as practicable.

N/A

4. If the carriers of a mutation are identified as a result of screening the natural history of people with this status should be understood, including the psychological implications.

N/A

The test

5. There should be a simple, safe, precise and validated screening test. Screening is carried out in the first week of life and again at 6-8 weeks of age. The first screen is a question to identify high risk factors. Babies with risk factors should be referred for ultrasound examination. The second screen is a clinical examination, which should be performed on all babies. The test in the first few weeks of life is observation for visible abnormalities of the lower limbs, and the Ortolani and Barlow tests, which require training and skill.

As cases are rare in the experience of any one screener and true cases should not be repeatedly examined for training purposes, it is very difficult to either train adequately or monitor standards. After 6 weeks of age, the physical signs gradually change and are more difficult to define precisely and to elicit. They are not readily adapted to a screening procedure.

Ultrasound is not currently recommended for primary screening. It has a role in secondary screening. The screen probably misses up to 2/3 of cases in many places but it may be possible to achieve sensitivity of 75-80% or higher with good training and supervision. Specificity is variable but a large number of referrals for second line screening is inevitable.

6. The distribution of test values in the target population should be known and a suitable cut-off level defined and agreed.

Clinical screen – **N/A.** Ultrasound – lack of consensus about protocol and interpretation or reporting. This will be addressed.

7. The test should be acceptable to the population.

YES

8. There should be an agreed policy on the further diagnostic investigation of individuals with a positive test result and on the choices available to those individuals.

Not entirely – there are some variations in methods, but overall agreement about approach to definite cases of CDH. More uncertainty about DDH.

9. If the test is for mutations the criteria used to select the subset of mutations to be covered by screening, if all possible mutations are not being tested, should be clearly set out.

N/A

The treatment

10. There should be an effective treatment or intervention for patients identified through early detection, with evidence of early treatment leading to better outcomes than late treatment.

YES

11. There should be agreed evidence based policies covering which individuals should be offered treatment and the appropriate treatment to be offered.

YES in general terms for CDH, though with many variations, but much less agreement for DDH.

12. Clinical management of the condition and patient outcomes should be optimised by all health care providers prior to participation in a screening programme.

It is not, but is being addressed.

The screening programme

13. There should be evidence from high quality Randomised Controlled Trials that the screening programme is effective in reducing mortality or morbidity.
NO
Evidence shows wide variations in the number of cases detected, but as some series show high sensitivity it is thought that quality of clinical examination may be a key issue. There is no RCT evidence that screening results ultimately in reduced morbidity, though there is observational data to suggest this is the case.

Where screening is aimed solely at providing information to allow the person being screened to make an "informed choice" (eg. Down's syndrome, cystic fibrosis carrier screening), there must be evidence from high quality trials that the test accurately measures risk. The information that is provided about the test and its outcome must be of value and readily understood by the individual being screened.
N/A

14. There should be evidence that the complete screening programme (test, diagnostic procedures, treatment/intervention) is clinically, socially and ethically acceptable to health professionals and the public.
YES in general – but there are problems raised by poor programmes and poor information

15. The benefit from the screening programme should outweigh the physical and psychological harm (caused by the test, diagnostic procedures and treatment).
Probably **YES**

16. The opportunity cost of the screening programme (including testing, diagnosis, treatment, administration, training and quality assurance) should be economically balanced in relation to expenditure on medical care as a whole (i.e. value for money).
Cost per adverse event avoided or improved outcome obtained has been assessed by Dezateux et al. The main finding is the high cost of universal ultrasound. Conclusions

are very sensitive to assumptions about test performance and intervention particularly for universal ultrasound screening.

17. There should be a plan for managing and monitoring the screening programme and an agreed set of quality assurance standards.

NO

Few districts monitor coverage, training or performance.

18. Adequate staffing and facilities for testing, diagnosis, treatment and programme management should be made available prior to the commencement of the screening programme.

There are not enough orthopaedic surgeons with an interest in paediatrics to provide an optimum service and ultrasound facilities are already hard pressed. This is without all appropriate infants with risk factors having an ultrasound examination.

19. All other options for managing the condition should have been considered (e.g. improving treatment, providing other services), to ensure that no more cost effective intervention could be introduced or current interventions increased within the resources available.

There is probably no alternative to screening.

20. Evidence based information, explaining the consequences of testing, investigation and treatment, should be made available to potential participants to assist them in making an informed choice.

New information has been developed for the Personal Child Health Record.

21. Public pressure for widening the eligibility criteria for reducing the screening interval, and for increasing the sensitivity of the testing process, should be anticipated. Decisions about these parameters should be scientifically justifiable to the public.

N/A

22 If screening is for a mutation the programme should be acceptable to people identified as carriers and to other family members.

N/A

Summary – pragmatic considerations, state of the art, recommendations

If proposed now as a new programme CDH/DDH screening would probably not be accepted. However, it is so ingrained in the clinical practice of so many people that it would be almost impossible to stop it unless overwhelming evidence of ineffectiveness could be obtained. This is unlikely. Therefore, we recommend as an interim measure:

Every baby to be reviewed within the first week of life for risk factors, examined by the clinical screening procedure described above, and referred for ultrasound if risk factors or clinical signs present. Although ideally the examination would not be done in the first two days because there are more false positives at that time, in practice this is often unavoidable.

A second examination within the first ten days has not been shown conclusively to increase identification of DDH. However, given the variability in age of discharge from maternity units and the uncertain coverage of the newborn examination in hospital, a practical case can be made for a second examination some time within the first week if this could be associated with other health care contacts at that time.

The hips should be examined again before 8 weeks of age at the latest and the evidence although weak suggests that since treatment before six weeks is preferable, the examination should be done before that time. See also overview of programme.

Ultrasound should not be introduced as a primary screening measure at present. Although excellent results have been reported (e.g. Clegg), very high levels of intervention are being reported and can probably only be avoided by a high level of senior level involvement by imaging and orthopaedic specialists.

Ultrasound for further examination of referred cases is useful but more work is urgently needed to decide on reporting criteria and management of the less severe abnormalities.

After 6-8 weeks, case-finding continues but does not merit the status of a screening procedure. Parental concern is important and should be taken seriously. Inspection for physical signs suggestive of hip abnormalities should be regarded as good clinical practice, not as screening. Recall specifically for this assessment is not warranted and an opportunistic approach should be adopted.

Information about hip problems should be included in the PCHR.

There is an urgent need to establish proper training where it is not provided and to decide which professional group will do this screen. Though there is no hard evidence, most people think that the Hippy plastic model facilitates training. Records should be kept of training as a risk management procedure.

Sources of information

Research programme and literature reviews by Dezateux et al (MRC programme).
Other literature. Expert seminar.

Status of the recommendation

Discussed and agreed by the Children Health Sub-Group of the National Screening Committee-to be kept under review. Awaiting further to clarify precise guidelines on ultrasound..

Quality of evidence

Mainly II-2 and II-3.

Strength of recommendation

Some evidence to continue clinical and risk factor screening – strengthened by pragmatic considerations.

The evidence for rejection of universal ultrasound at present is considered good, but is related to high intervention rates and high cost for few additional adverse events prevented. The data on which this is based vary widely in findings and interpretation. Further studies may therefore change this view in due course.

Research agenda

1. A trial of screening versus no screening has been debated at length but is not generally thought to be a practical proposition.

2. The issue of universal ultrasound screening will need to be re-visited in due course, but more work is needed first on the natural history and management of DDH.
3. Short term, the issues are to define the risk factors more clearly and determine whether and how staff use them; establish a working protocol for reporting and managing DDH detected during secondary US screening; agree on the key physical signs and physical examination procedures for case-finding after 8 weeks of age. These may each give rise to further research questions.
4. There are few data on how best to teach the clinical examination or how to monitor and maintain quality.