Topic for Debate

AT THE CROSSROADS – NEONATAL DETECTION OF DEVELOPMENTAL DYSPLASIA OF THE HIP

Moderator: David Jones

Contributors: C. A. Dezateux, L. G. Danielsson, R. W. Paton, J. Clegg

The clinical testing for development dysplasia of the hip (DDH) in neonates is falling short because of the unreliability of the Ortolani/Barlow (O/B) manoeuvres and the performance of these examinations by personnel who are relatively inexperienced. The role of ultrasound, either selective or universal, is unclear and the logistics of implementation of a universal programme are a daunting proposition in the British context. Are we clear as to what we may realistically achieve by a screening programme?

Following the response to my Editorial in this journal in November 1998 I asked four experts to consider four questions: What are the goals of screening? What are the current problems? How should these be addressed? Should the current guidelines be altered?

Their views were sought as personal opinions without quotation of supporting references.

Carol Dezateux:

Screening of newborn infants for congenital dislocation of the hip was introduced more than 30 years ago in the belief that a better outcome could be achieved by early detection and abduction splinting rather than by surgical management of cases clinically presenting. The aim was to prevent pain, limitation of function and disability due to dislocated or subluxated hips. With the advent of ultrasound, an additional goal of preventing isolated acetabular dysplasia and

associated osteoarthritis has been proposed. Detection of neonatal instability or dysplasia is the aim of the screening test, but not the goal of screening.

The current policy in the UK is to perform the O/B tests in all newborn infants and again at the age of six to eight weeks. In practice, most infants with positive clinical findings are examined by ultrasound, as is a variable proportion with risk factors. This programme does not identify all affected children and, although it may be possible to improve these results, other information is relevant to policy development. The absolute burden of disease and its potential reduction through screening need to be balanced against the possible disbenefits to those screened or treated. What does the balance sheet look like for the current programme and its alternatives? This can be summarised by posing the following four questions.

What are the long-term consequences for health and quality of life of dislocation/dysplasia presenting clinically? Before screening was introduced, published data suggested that about one child per 1000 developed established dislocation. After surgical management, between a quarter and a half of these may have radiologically abnormal hips at skeletal maturity. Of these, half may develop pain in the hip in their third or fourth decade. This outcome may have improved with advances in surgical management. An additional consideration is the impact on the quality of life for these individuals, yet information on this is limited.

How well does screening identify those at risk and distinguish those who are not? Currently, about half of those affected are not detected by screening and present clinically with dislocation or subluxation, usually as a result of parental concern. There is evidence that the use of expert examiners for screening may reduce this, but is unlikely to eliminate it. Not all late presentations are necessarily preceded by neonatal instability.

Screening will also suggest abnormality in some infants who are normal but may then be treated. This number cannot be measured but, currently, between 3 and 5 infants per 1000 are splinted, suggesting that at least four normal children are treated for each truly affected child treated with a splint. The occurrence of avascular necrosis of the femoral head after abduction splinting in an otherwise normal child constitutes a major hazard of screening and may occur in up to 1% of those treated.

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By how much is the long-term outcome improved by abduction splinting? The evidence that this procedure improves long-term outcome is limited and is mostly derived from single-centre studies. In the long term and despite avoidance of surgery, dysplasia may persist in some, although the frequency of this is unclear. One difficulty in making inferences from observational studies of outcome relates to the proportion of infants with false-positive diagnoses included in these case series, which may bias estimates of the effect of treatment. A further difficulty arises because changes over time in factors which improve outcome among those presenting clinically, due, for example, to advances in surgical management or earlier age at presentation, will tend to reduce the marginal effectiveness of abduction splinting.

What are the other options? One is to use ultrasound in infants with clinical instability or with specific risk factors. This may reduce the numbers treated with abduction splinting, but information on the longer-term outcome is needed and is the subject of the current MRC multicentre 'Hip Trial'. Using ultrasound to assess infants with risk factors but without instability, will identify only a small number of those who escape detection by the O/B tests. Furthermore, the value of treatment in this group, especially those with 'borderline' ultrasound appearances, has not been established. An alternative is to screen all infants by ultrasound. This has been introduced in some European countries, where 21% of all term infants are reported to have abnormal ultrasound appearances requiring treatment or follow-up, and 40 to 50 per 1000 are splinted. Lower yields and rates of treatment have been reported from the only UK centre to operate such a policy, but even assuming that this experience is generalisable, questions remain as to the rates of detection and the effectiveness of treatment. At present, there is only limited evidence to suggest that universal ultrasound screening is associated with more complete identification of affected children, and none to show that it leads to a better long-term outcome. Furthermore, it is unclear whether those at risk of isolated acetabular dysplasia as adults can be identified in infancy, and whether early treatment by abduction splinting can prevent the development or persistence of dysplasia.

How may these problems be addressed? The effectiveness of the current programme in the UK was challenged almost as soon as it was introduced. Since then, much has been published on rates of detection, but little on outcome. This needs to be addressed. Existing data should be used to quantify the prevention and cost-effectiveness of current and alternative screening policies within the UK setting, and to prioritise research relevant to the screening policy. The clinical paediatric orthopaedic community is central to ensuring that the necessary primary research is carried out. Some individual centres have carried out small quasi-randomised trials and the current MRC Hip Trial represents the first multicentre exercise in this field. Other collaborative studies should be encouraged, since these conditions

are relatively rare, and large, adequately powered studies are needed. Although most controversy has centred on how well the screening test works, the effectiveness of treatment with abduction splinting is central to the rationale for screening, irrespective of the test used. Can we continue to defend the lack of trials of treatment in this very important area of paediatric orthopaedic practice?

In the UK, all existing and proposed screening programmes are subjected to systematic evaluation under the auspices of the National Screening Committee. Given that the last policy review was in 1986, it is timely that current policy be updated to reflect the best available evidence as well as the criteria currently established for screening programmes. In developing a new policy, the guiding principle should be to maximise benefit while causing least harm.

Lars Danielsson:

In Malmö all living neonates are clinically examined for instability of the hip by a paediatrician once or twice within the first 48 hours. The infants are examined on weekdays, including holidays, but not on Sundays. In about 80% of the cases the procedure is limited to two neonatologists; the remainder are examined by other paediatricians with experience of the technique.

Children with clinically unquestionably dislocated or dislocatable hips are immediately referred to the Orthopaedic Outpatient Clinic, examined by a paediatric orthopaedic surgeon and treated in a von Rosen splint for three months. Those with clinically unstable but not dislocatable hips are assessed using a dynamic anterior ultrasonic technique, introduced in 1990. The radiologist and the orthopaedic surgeon measure the instability while the latter is performing the dislocation-reduction manoeuvre. If the head is not dislocatable but the displacement at sonography exceeds 1/4 of its diameter (> 3 mm), the patient is treated for six weeks in the von Rosen splint. If the displacement of the femoral head at sonography is 1/4 of the diameter the patient is not treated. All patients are clinically examined at least one week after the first orthopaedic examination, at the age of three months and at one year. Radiological examination is included at the age of three months and one year even in those who have not been treated.

The goals of screening are to detect DDH as early as possible and before the hip has become stabilised in a dislocated position. Some abnormal hips are still not detected, often without an obvious explanation. At one time too many paediatricians were involved in the diagnostic procedure in Malmö. This may explain why out of 21 cases diagnosed late between 1956 and 1998, 12 were missed in 1980 to 1986 when the screening routines in the Paediatric Department were changed so that all physicians were involved. In one year 22 took part!

It has been suggested that repetition of a Barlow test, performed roughly, may increase instability of the hip and cause dislocation. In 1963 a more extensive Barlow test

with stabilisation of the pelvis with one hand when examining the hip with the other, was introduced. The same method is still being used and cannot explain the increased number of missed cases in 1980 to 1986.

We examined whether children born on Saturdays, Sundays, days before holidays or holidays, had an increased risk of being missed. No such tendency was noted.

Plain radiographs performed by inexperienced radiologists not familiar with the provocation-dislocation test may be misleading. We therefore excluded this assessment from the screening routine in 1983.

We have found that hips reported to be unstable or even dislocatable in the morning, stabilised during the day. Could the opposite happen? Is this an explanation for the fact that there is no series without one single failure? Nevertheless, failures in detection should be considered as such and not regarded as developmental.

If too many examiners are involved in diagnosis there is a tendency to treat for safety rather than by conviction. The possibility of the development of avascular necrosis must be considered. The incidence of this in our patients is 1:163 treated. Since ultrasonic examination has become routine in all unstable cases the number of such patients treated has decreased. Only 34 unstable but not dislocatable hips out of the last 178 consecutive cases have been treated.

Further improvements in detection can be achieved by adherence to an established routine. Central registration of failures in diagnosis should be established in order to allow proper assessment to be made of the reasons for the failure in diagnosis.

Robin Paton:

The aim of screening is the early detection and treatment of DDH, thereby preventing or limiting long-term disability. The screening programme should employ a regime which has good sensitivity and specificity and does not lead to overtreatment. The cost of the programme should neither be prohibitive nor more than if the conditions were left untreated. Current methods do not meet these criteria. The low prevalence combined with relatively poor sensitivity of clinical tests makes false-negative results more likely. The O/B tests are 100% specific but only 60% sensitive in expert hands. Hips may become clinically stable but remain abnormal on ultrasound examination.

The O/B tests are more sensitive in the hands of expert examiners. In the UK most neonatal examinations are carried out by inexperienced staff, thereby further reducing sensitivity. Other signs such as limited abduction, shortening and asymmetrical skin creases are of no value in diagnosis in the neonate. 'At-risk' factors have a poor predictive value if used as a screening test. In my experience, only 31% of neonatal dislocations have 'at-risk' factors without positive O/B tests.

Although ultrasound is highly sensitive (>90%) it also has deficiencies. Treatment of unstable and dysplastic hips was advocated before the natural outcome of abnormalities

was assessed by serial testing. Most clinically unstable hips settle spontaneously and serial scanning of such hips confirms this, although acetabular dysplasia may persist. Most of the abnormalities apparent on ultrasound are signs of immaturity rather than pathological abnormality.

Because of the poor relationship between abnormalities observed by ultrasound at birth and the ultimate development of the hip, universal ultrasound screening should not be advocated. Thousands of normal infants would have to be examined often serially, with few positive results. Over-treatment would occur. True universal screening is also impracticable because an appreciable minority would not be able to attend for examination. Even if ultrasound screening was implemented there would still be patients needing surgery, such as those with irreducible dislocations and persistent dysplasia without dislocation. 'At-risk' ultrasound screening is not as valuable as previously thought. It is of little benefit in significantly reducing the rate of late dislocation, but there is a higher rate of detection in clinically unstable hips or those with 'at-risk' factors.

Clinical screening programmes should continue but the examination must be undertaken by the most experienced clinician available. Unstable hips should then be referred for ultrasound examination to confirm the diagnosis. All clinically unstable hips should be assessed by ultrasound scans within one to two weeks. They should be followed by weekly scans until the hip stabilises. If this fails to occur within two to three weeks, or if major dysplasia persists after nine weeks, the hip should be treated by dynamic splintage. Hips which are clinically stable but 'at risk' because of family history or breech presentation should be examined by ultrasound at nine weeks, by which time immature hips will have usually developed. An additional clinical assessment at three to four months is advisable in order to detect unilateral limited abduction. Such hips should be assessed by ultrasound to determine if there is major dysplasia as ultrasound is more sensitive at this age than radiographs. After four to six months radiography is more reliable than sonography and is the preferred investigation.

The term 'missed' dislocation should be discouraged. 'Late' dislocation is a better title since stable but dysplastic hips at birth may progress to irreducible dislocation. Late dislocation will never be abolished by screening because of the nature of the condition.

John Clegg:

DDH occurs with sufficient frequency and poses considerable problems of management if diagnosed late to justify determined efforts towards early diagnosis, when simpler, more conservative management has a greater chance of success. The subtle changes of acetabular dysplasia can be responsible for pain and premature degenerative changes and are rarely identified by clinical examination in infancy. The few long-term reviews available confirm the incidence of failure of early diagnosis. The combination of residual

subluxation of the hip together with avascular changes in the femoral head, inevitably result in premature degenerative changes, with increasing morbidity in the more active years between the third and fourth decade. Considerable effort has been put into the use of clinical screening by the methods of Ortolani and Barlow since the introduction of the SMAC guidelines in 1986. Sadly, this effort has not been generally rewarded by a dramatic improvement in rates of early detection. A depressingly large number of cases of dislocation still present late and tax the skill and expertise of the paediatric orthopaedic surgeon.

Some abnormal hips cannot be detected by clinical means in the first few months of life. This has been the case for a small number of our patients in the course of a longitudinal study of the performance of a routine ultrasound screening programme over the past ten years. Routine ultrasound examination offers a window through which hip instability and potential developmental dysplasia may be identified. In our experience it has been shown to improve the detection of hip abnormality to the extent of reducing the incidence of late presentation to zero, a dramatic improvement from the days when we relied on clinical examination alone. Such a screening programme requires adherence to strict standards of performance of the investigation and interpretation of the scan.

Ultrasonic screening of patients with 'at-risk' factors alone cannot identify all of the patients with abnormality of the hip. In our experience, almost half of the patients who have been found to have a clinical dislocation do not have such 'at-risk' factors and would never qualify for examination.

If ultrasound screening were to be introduced nationally, there would be a requirement for the development of a Training Centre with regular audit and reassessment of the examiners, as has been established in Austria following the pioneering work of Reinhard Graf. Piecemeal implementation, without structure, supervision and assessment, will fail to introduce and maintain the necessary standards and will discredit the method. Our experience over the past ten years has shown that early detection has resulted in better results than treatment in a harness, with a significant overall reduction in the number of operations required. The need for major surgical intervention has been all but eliminated. Time will tell if this will be reflected in a reduced long-term morbidity and a decrease in the frequency of joint replacement. Audit of the financial aspects of this routine ultrasound screening programme suggests that it need not be more expensive than the costs of surgical treatment in the first few years of life. If the costs of later surgery, and any litigation following upon a missed diagnosis, are also added then routine ultrasound screening offers much better value for money.

Centres which report only a low incidence of missed dislocation after clinical examination are perhaps understandably reluctant to consider any change in screening methods on the grounds that financial benefits may be small

but an increase in the use of medical manpower will be needed, thus making the introduction of a routine ultrasound screening programme less worthwhile. A few patients and their families who are subjected to the trauma and uncertainties of surgery would no doubt have preferred to avoid this. The psychological effects on the whole family of the child in whom there is a late diagnosis, should not be underestimated.

Criticism has been levelled at the use of a routine ultrasound screening programme since it may result in a considerable increase in treatment. In our experience this has not been the case. With a proper protocol for management, overtreatment can be avoided. Serial screening has reduced the need for routine treatment in all cases of subluxation, a condition identified by Barlow as often showing spontaneous resolution.

Of equal importance is the detection of those patients with clinically normal hips, but who show signs of persisting abnormality on ultrasonic examination. It is tempting to suggest that treatment in this group will reduce the incidence of later dysplasia of the hip.

Published work concerning the diagnosis and management of dislocation of the hip rarely includes a consideration of the dysplastic hip, a condition which otherwise may only be identified fortuitously on a chance radiological examination, or if it produces pain in adolescence or early adult life. Ultrasound examination offers the opportunity to identify and treat this dysplasia early in infancy, with the possibility of modifying its development. So far, this idea has featured little in discussions on the diagnosis and management of abnormality of the hip in childhood, but may well assume greater importance in time to come.

Clinical examination has been with us since the beginning of medical practice, but it has failed to diagnose at birth all patients with a significant abnormality of the hip and can never identify those with incipient acetabular dysplasia. At present, ultrasonic examination is the optimum method of diagnosis, but it will, no doubt, eventually be overtaken by other developments such as MRI. Our experience with routine ultrasound screening for the diagnosis of DDH suggests that serious consideration should be given to its general introduction in the UK. The excellent results obtained in Austria could, with appropriate training and supervision, be reproduced here. We should not be deflected by financial or practical arguments based on a narrow perspective aimed simply at diagnosis. The improved opportunities for treatment of both established dislocation and dysplasia will lead to a reduction in morbidity and offer a better quality of life for the child and the family. Only the orthopaedic surgeon is in a position to comprehend truly the difficulties in management after late diagnosis, as well as the long-term complications and disability which may follow. It is appropriate that we should take the lead in supporting the more widespread introduction of a routine ultrasound screening programme. This should result in a reduction in major surgery which

would be confined to a few specialist centres, with better outcomes and improved quality of life for the patient.

COMMENTS

David Jones:

Divergence of opinion on this issue is to be expected but each participant has made important contributions to the debate.

Carol Dezateux gives us a searching and constructive analysis of our shortcomings. Such an epidemiological review is essential, especially as many disciplines are involved in the detection of DDH. She clearly indicates the need for trials to assess the effectiveness of splintage and when it is needed. In my experience there is little harmony in the use of the Pavlik harness in this country. Some centres are diligent and experienced in its application, while others seem to fit it as an adornment rather than a splint. There are wide variations in how the infants are monitored and whether the device is worn constantly or intermittently.

Lars Danielsson reinforces the message of a strong clinical screening programme backed up by ultrasound and the use of the Von Rosen splint. He reports no long-term complications with this device. From my own experience I agree with him. While recognising that some cases will always be missed, the Malmö group sees its future with a clinically-based programme which involves maintaining vigilance and awareness with central reporting of cases which are diagnosed late.

He feels that primary examiners are able to differentiate between subluxation and dislocation. In the former, orthopaedic surgeons and radiologists decide on the basis of dynamic ultrasound whether the infant should be splinted. I doubt if such skills and co-operation would be universally implemented in the UK.

Robin Paton's contribution highlights the practical difficulties of implementing a reliable screening programme in the UK. Even with his enthusiasm and ultrasonographic skills he accepts that there will be cases which are diagnosed late. This is an important clinical and medicolegal message.

I do not think that his ultrasonographic policies could be applied throughout the country. Weekly ultrasonographic examination of a clinically unstable hip would not be widely accepted. Most orthopaedic surgeons would splint an unstable hip after one to two weeks. There would be no quibble about overtreatment.

I am also unsure as to why he does not make more use of radiographs which would be more acceptable to most

clinicians when 'at-risk' infants are being reviewed at nine to 12 weeks. We agree that at eight months, radiographs are the preferred option in clinically suspicious cases. I do not think that a separate orthopaedic examination of normal infants is indicated at four months.

John Clegg has shown great organisational and ultrasound skills in implementing a programme based on universal ultrasound. It is right that he should consider the technique as the optimum. I think it unlikely, however, that his achievement would be reproduced throughout the UK. Not only would it be difficult to agree protocols and train the number of ultrasonographers needed, but there is not widespread enthusiasm among paediatricians and orthopaedic surgeons for universal ultrasound. By his admission, John Clegg's project has important questions yet to be answered. In particular, we need a long-term review to confirm that universal ultrasound does indeed lead to reduced long-term morbidity and decreased frequency of hip replacement. A review of those stable hips with persisting ultrasound abnormality is necessary to decide whether treatment of this group will eliminate later dysplasia of the hip.

CONCLUSIONS AND RECOMMENDATIONS

- 1) Clinical surveillance programmes should continue. The skills of examiners and awareness of the spectrum of presentation of DDH should be improved. Orthopaedic surgeons must be active in these processes.
- 2) When splintage is indicated, it should be effective. If a Pavlik harness cannot be applied properly and monitored reliably then a splint such as the Von Rosen should be used.
- 3) Since ultrasonographic services and skills vary widely, radiographs at three months for infants significantly 'at risk' and any time thereafter for suspected cases, are a better option.
- 4) At present, the introduction of the universal use of ultrasound for DDH in the UK is impracticable. Those centres, however, which undertake it should confirm the long-term effectiveness of the method and, in particular, whether treatment of clinically stable but ultrasonographically abnormal hips will eliminate late-presenting dysplasia.
- 5) Clinical trials are needed to assess the effectiveness of splintage and to confirm the superiority of experienced examiners.
- 6) Guidelines for the screening of DDH should be reviewed. A clear definition of good practice is needed. In the British context the 1986 guidelines need updating.