The early diagnosis of congenital dislocation of the hip

The saga concerning the early diagnosis of congenital dislocation of the hip (CDH) continues to expand, and this issue of the Journal adds three more papers to the literary store. Two of them report experience with clinical and ultrasound scanning for early diagnosis and the third discusses, in a controversial way, the value of these diagnostic methods.

For orthopaedic surgeons, early diagnosis enables successful treatment to result in normal development of the hip. However, despite scientific advances, numerous problems remain. The legal profession is interested, since some parents seek compensation for the poor results of treatment after what they regard as late or missed diagnosis. What progress has been made in early diagnosis and management?

It has for long been known that early diagnosis is possible (Ortolani 1937) and it is more than 30 years since the papers by Barlow (1962) and von Rosen (1962) caused an enthusiastic explosion of interest. Longer-term results, however, from other centres were not as good as had been hoped, and even with early diagnosis the outcome was not always completely successful. The incidence of hip instability detected in the newborn was much greater than the true incidence of congenital dislocation, yet infants still presented for late treatment at a rate of approximately 0.4 to 0.6 per thousand live births (MacKenzie and Wilson 1981). This represents about one-third of the true incidence of CDH in unscreened populations. Either the tests were inadequate or they were badly performed; or perhaps some infants who were apparently normal at birth developed late instability and subluxation. It was shown that dedicated, thorough clinical examination could reduce but not eliminate the rate of late diagnosis (Moore 1989; Macnicol 1990; Krikler and Dwyer 1992).

It is now believed that there is a spectrum of conditions which may be diagnosed during the first few months of life (Catterall 1984). These range from an established dislocation irreducible at birth (which is often missed) to simple neonatal instability, which usually settles completely without treatment. An important group are those with an eccentrically placed hip; this may go on to acetabular dysplasia with subluxation, although some represent no more than normal variants. Ortolani (1937) suggested the term congenital dysplasia, since it is clear that not all abnormal hips result from congenital dislocation. Klisic (1989) suggested the term Developmental Dysplasia of the Hip (DDH) and this term has now been adopted by the American Academy of Orthopaedic Surgeons. This implies a spectrum of conditions which may evolve over time, and is preferred to 'CDH' for future reports.

One approach to the continuing incidence of late presentation has been to pay increased attention to infants known to be at increased risk. Risk factors include breech position, a family history of DDH, neonatal hip instability, clicking hips, and the presence of other deformities such as abnormal feet, torticollis or plagiocephaly (Cunningham et al 1984; Jones and Powell 1990). In this issue of the Journal Boeree and Clarke (p. 525) report the combination of clinical and ultrasound methods of diagnosis for the 'at-risk' group. They found that 7% of all infants required screening at a special clinic and that 6.23% of these needed treatment (4.4 per thousand live births). Nevertheless, they report late diagnosis in 0.2 per thousand. Their risk categories did not include infants born by caesarean section or those who had been cared for in special care units, although Hansson, Nachemson and Palmén (1983) reported a high incidence of hip anomalies in these groups. Ultrasound screening of the high-risk groups did reduce the incidence of late referral, but only to the level reported after the dedicated clinical examination of all cases (Moore 1989; Macnicol 1990). The unhappy conclusion must be that whatever tests are used at birth or later, they need to be applied to all infants and not just to the 'at-risk' population.

Ultrasound examination was introduced in the early 1980s (Graf 1984; Clarke et al 1985; Berman and Klenerman 1986). Its great advantages over radiography are that there is no irradiation and that the cartilaginous femoral head is visualised directly. For enthusiasts, it is 'the solution', but for others it provides a blur of black and white dots and smudges whose interpretation may be a matter of conjecture. Protagonists claim to be able to monitor acetabular development and to separate the signs of immaturity (Graf 2a) from those of serious dysplasia. Examination involves finding the cartilaginous femoral head and observing the acetabulum in relation to it. The orientation of the acetabulum is rarely considered.
although it is known to be forward-facing at birth and may subsequently be associated with the benign condition of rotational acetabular dysplasia (Dyson, Lyskey and Catterall 1987). Dynamic tests of instability increase the usefulness of ultrasound examination; they may be the most essential part of it (Clarke et al 1985; Harcke and Grissom 1990). It is not difficult to accept that an entirely stable joint will probably develop normally, but an unstable and eccentrically placed femoral head may become dysplastic with time. Adding ultrasound imaging to clinical examination at birth dramatically increases the number of abnormal hips detected, but most of these ‘abnormalities’ get better without treatment. Ultrasound may detect more of the benign abnormalities, without diagnosing more of the true cases of DDH.

The paper by Marks et al (p. 534) reports the results of a policy of ultrasound examination at birth for all cases. Using the grading of Terjesen, Bredland and Berg (1989), they found that 6% of infants had abnormal scans but that 90% of these had become normal by the age of nine weeks. The percentages are similar to those reported by Boeree and Clarke (p. 525) from their selective approach. Treatment was required for 59 hips (2.42 per thousand), 37 (1.5 per thousand) for persisting clinical instability and 22 (0.85 per thousand) for persisting ultrasound abnormality. The last figure is very similar to that for late referral reported in other studies. Ultrasound imaging at birth is therefore capable of detecting nearly all abnormal hips. No hip which was normal at birth had become abnormal with time, although there is still the possibility of the late development of dysplasia in adolescence; Tucci et al (1991) reported late dysplasia in 17% of children after apparently successful treatment by a Pavlik harness from birth. It is therefore essential that the outcome study of Marks et al is continued in the longer term.

Jones and Powell (1990) used clinical examination of all infants, supplemented by ultrasonic examination of the ‘at-risk’ population and reported no late cases. However, Clarke, Clegg and Al-Chalabi (1989) and Boeree and Clarke (p. 525), using a similar protocol, report late incidences of 0.6 and 0.2 per thousand, no better than the best results of clinical examination alone. The ultrasound patterns of the Terjesen groups (1989) describe only eccentric position of the femoral head. This implies instability, which is what ultrasound should really be assessing. If progress is to be made in early diagnosis, ultrasound must diagnose the degree of instability present as well as the apparent dysplasia.

In the third paper on DDH in this issue Hernandez et al (p. 539) introduce us to the complexities of Decision Analysis Methodology, a type of statistical analysis which may be a bridge too far for the average orthopaedic surgeon. They analyse historical data and assess the ‘value’ of various diagnostic strategies. Their conclusion is that routine ultrasound scanning, of whole populations or of ‘at-risk’ groups, has limited value, confirming the view of many clinicians that careful clinical examination at any age and plain radiography at three to four months of age is a better option.

At present, the cornerstone of diagnosis must be the clinical examination of all infants by experts on at least two occasions in the first three months of life. Ultrasound examination may be justifiable for the ‘at-risk’ population, to which may be added infants born by caesarean section and those treated in special care units. It is probable that the important contribution of ultrasonic imaging is its ability to recognise instability. A new diagnostic classification is required which measures both instability and dysplasia. Until ultrasound examination can distinguish between true dysplasia and the normal variations of anatomy, radiography at three to four months of age is still the best method of establishing the true diagnosis in a doubtful hip.

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REFERENCES


Cunningham KT, Moulton A, Benningfield SA, Maddock CR. A clicking hip in a newborn baby should never be ignored. Lancet 1984; i: 668-70.


