LETTERS

Osteogenesis imperfecta and congenital dislocation of the hip

EDITOR,—Colin R Paterson and colleagues have drawn attention to an important group of children that paediatricians performing neonatal examinations need to be aware of.[1] I would value some clarification about the content of their paper.

Eight cases of osteogenesis imperfecta were described from a central register of such patients. How many patients were registered in total, and over what period of time were the cases collected? The changes in work practice suggested at the end of the paper have important implications for paediatric, orthopaedic, and radiology department workloads, hence some idea of the incidence of the condition is vital. The figures in their article suggest that 30-60 children a year are born with osteogenesis imperfecta. If all the cases described occurred in one year the incidence of iatrogenic fracture would be 1:75 000 live births compared with 1800 cases of residually unstable hips per year (that is, 1:333 live births). Although paediatricians need to be aware of this rare problem and consider asking all parents if there is a family history of brittle bones, we will no doubt continue to be held responsible for missed congenital dislocation of the hips if we start to take a more lax attitude to routine examination.

Some of the other comments suggest that the authors have not had to do 10-15 neonatal examinations a day on a busy postnatal ward. My own practice has been to warn parents that "most babies cry after this test" even before I have touched the hips, this being the reason it is left to the end of the neonatal examination. Until a suitable long term trial of ultrasound screening of neonatal hips is undertaken, as suggested by N M P Clarke,[2] it will undoubtedly continue to be paediatricians who have to balance the risks of missing congenital dislocation of the hips against causing this rare complication.

ANDREW F MELLON

Department of Child Health,
Royal Victoria Infirmary,
Newcastle upon Tyne NE1 4LP

1 Paterson CR, Beal RJ, Dent JA. Osteogenesis imperfecta: fractures of the femur when testing for congenital dislocation of the hip. *BMJ* 1992;**305**:464-6. (22 August.)
2 Clarke NMP. Diagnosing congenital dislocation of the hip. *BMJ* 1992;**305**:435-6. (22 August.)

EDITOR,—The article by Colin R Paterson and colleagues requires comment and moderation. The straightforward message is that care should be taken examining the hips of a newborn child when there are grounds to suspect osteogenesis imperfecta. That is entirely reasonable, but the case histories do not, in general, give enough information for the reader to verify the diagnosis of osteogenesis imperfecta, and they do not establish that the fractures were caused by examination of the hip.

Osteogenesis imperfecta is a rare condition; congenital dislocation of the hip is common. It is therefore important that the hips be carefully examined. Only the presence of unequivocal signs of osteogenesis imperfecta or an unequivocal family history of osteogenesis imperfecta should prevent this examination. Similarly, the suggestion that the hips should be radiographed if the baby cries after the examination should be resisted. Paterson *et al* discuss the differential diagnosis between osteogenesis imperfecta and non-accidental injury. Previous correspondents in this journal have commented on Dr Paterson's tendency to accept a diagnosis of osteogenesis imperfecta or other bone disorder when others would have diagnosed non-accidental injury.[2,3]

In the context of child abuse hearings, the diagnosis of any brittle bone disorder should be accepted only when there is excellent evidence for that diagnosis. Evidence such as large fontanelle, hyperextensible joints, blue sclerae in infancy, one or two Wormian bones, or a family history of fractures does not constitute such excellent evidence. Hidden non-accidental injury causes many more fractures than hidden brittle bone disease—by at least a thousandfold.[4]

L H P WILLIAMS

Bassetlaw District General Hospital,
Nottinghamshire S81 0BD

1 Paterson CR, Beal RJ, Dent JA. Osteogenesis imperfecta: fractures of the femur when testing for congenital dislocation of the hip. *BMJ* 1992;**305**:464-6. (22 August.)
2 Taitz LS. Child abuse and osteogenesis imperfecta. *BMJ* 1988;**296**:292.
3 Carty H, Shaw DG. Child abuse and osteogenesis imperfecta. *BMJ* 1988;**296**:292.
4 Taitz LS. Child abuse and osteogenesis imperfecta. *BMJ* 1987;**295**:1082-3.

AUTHOR'S REPLY,—We thank Andrew F Mellon for his interest in our paper. The register described has been built up over about 12 years and now includes clinical data on 1100 patients.

Mellon misunderstands the message of our paper. We are not suggesting that our study should influence the choice of screening methods in routine use. We do advise that when osteogenesis imperfecta can be suspected from family history or physical signs the standard Ortolani and Barlow tests should not be used. In these uncommon infants ultrasonic examination is preferable.

We can reassure L H P Williams that there is ample evidence of osteogenesis imperfecta in each of the patients reported in our paper. In the two patients in whom non-accidental injury was initially seriously considered we now have follow up data for nine and 10 years, with a typical history of fractures in each case.

There are some patients in whom the diagnosis of osteogenesis imperfecta cannot readily be made at the time of the first fracture (about 10% of all cases). This difficult area of diagnosis is outside the scope of the present correspondence, but a previous report may be helpful.[1]

C R PATERSON

Department of Biomedical Medicine,
Ninewells Hospital and Medical School,
Dundee DD1 9SY

ROBIN J BEAL
JOHN A DENT

Department of Orthopaedic and Trauma Surgery,
Dundee Royal Infirmary,
Dundee DD1 9ND

1 Paterson CR, McAllion SJ. Child abuse and osteogenesis imperfecta. *BMJ* 1987;**295**:1561.

Advice to authors

Priority will be given to letters that are less than 400 words long and are typed with double spacing. All authors should sign the letter. Please enclose a stamped addressed envelope for acknowledgment.

Diagnosing congenital dislocation of the hip

EDITOR,—N P M Clarke states that clinical neonatal screening has failed to reduce and may actually have increased the incidence of congenital dislocation of the hip.[1] This statement is inaccurate and misleading. It is true that unsupervised and poorly organised screening programmes have often been ineffective. Many reports from around the world, however, have shown that late diagnoses may be greatly reduced (>90%) by neonatal screening.[2] Indeed, the Malmö study, cited in support of Clarke's contention, itself provides striking evidence of the success of neonatal screening.[3]

The Malmö study, of all 98 891 infants born in Malmö, Sweden, between 1956 and 1987, was initiated by Von Rosen, a pioneer in neonatal screening. Under his direction the incidence of late diagnosis fell to the exceptionally low figure of 0·07/1000 births during a 17 year period. While the incidence has risen in recent years, it still remains low compared with the incidence before screening.[4] Furthermore, over the whole 31 years no cases were diagnosed late among infants who presented in the breech position, though such infants accounted for a third of all neonatal diagnoses. That experience mirrors experience in Bristol.[5] The most probable explanation in both cases is that infants delivered in the breech position, being known to be at high risk, were checked particularly carefully at birth.

Clarke also repeats the unsupported speculation that clinical examination may actually provoke hip instability or, alternatively, may cause harm. The editorial draws attention to Paterson *et al*'s report on femoral fractures after hip examination in infants with osteogenesis imperfecta.[6] Unfortunate as these cases may be, they are exceptionally rare. The eight reported cases were culled from the Brittle Bones Society's files for the United Kingdom and Republic of Ireland since 1972. During the same 20 years in these countries it can be estimated that more than 100 000 infants would have been born with hip instability, a readily treatable but potentially crippling deformity.

PETER M DUNN

Bristol University,
Department of Child Health,
Southmead Hospital,
Bristol BS10 5NB

1 Clarke NMP. Diagnosing congenital dislocation of the hip. *BMJ* 1992;**305**:435-6. (22 August.)
2 Dunn PM. Screening for congenital dislocation of the hip. In: MacFarlane A, ed. *Progress in child health*. Vol III. London: Churchill Livingstone, 1987:1-13.
3 Sanfridson J, Redlund-Johnell I, Uden A. Why is congenital dislocation of the hip still missed? *Acta Orthop Scand* 1991;**62**: 87-91.
4 Palmen K. Prevention of congenital dislocation of the hip. The Swedish experience of neonatal treatment of hip joint instability. *Acta Orthop Scand* 1984;suppl 208.
5 Dunn PM, Evans RE, Thearle MJ, Griffiths HE, Witherow PJ. Congenital dislocation of the hip: early and late diagnosis compared. *Arch Dis Child* 1985;**60**:407-14.
6 Paterson CR, Beal RJ, Dent JA. Osteogenesis imperfecta: fractures of the femur when testing for congenital dislocation of the hip. *BMJ* 1992;**305**:464-6. (22 August.)

EDITOR,—I agree with N M P Clarke that ultrasonography is a reliable method of examining infants' hips and that it quickly and clearly distinguishes immature hips from those with frank dysplasia in a way that clinical examination may not be able to do.[1] But Clarke's other conclusion—