Correspondence

Clinical outcome of fetal uropathy

Sir,

Madarikan et al, in their article on clinical outcome of fetal uropathy, discussed the antenatal management of fetal uropathy. However, I think the following points deserve mention.

Any fetus found to have a renal dysplasia or an obstructive uropathy should, initially, have a full anomaly ultrasound scan performed to exclude other structural abnormalities. An accurate ultrasound scan can normally be done even in the presence of oligohydramnios by an experienced ultrasonographer. A fetal karyotype should also be considered because of the association between renal tract abnormalities and chromosomal defects. The parents should then be counselled with this information and its significance. If they elect to continue the pregnancy an attempt is made to assess the functional potential of the fetal renal tract by three methods: amniotic fluid volume, kidney size and sonographic appearances, and by the results of biochemical analysis of fetal urine after ultrasound guided needle aspiration. This latter technique is particularly helpful in identifying those fetuses with an obstructive uropathy who could benefit from vesicoamniotic shunting. Appelman and Golbus have shown that a fetal urine osmolality greater than 210 mmol/kg and a urinary sodium and potassium greater than 100 mmol/l and 90 mmol/l respectively, are accurate predictors of renal function. If the kidneys are non-dysplastic, liquor volume is reduced and the obstructive uropathy is infravesical, then vesicoamniotic shunting is generally undertaken. This has been shown to improve the perinatal survival of fetuses with posterior urethral valves, prune-belly syndrome, and urethral atresia. Whether it reduces long term renal and pulmonary morbidity is, however, unknown.

Although no one doubts the value of ultrasound screening, controversies do remain regarding the antenatal management of obstructive uropathy. Further information comparing conservative and surgical treatment of fetal obstructive uropathy is required, not only in relation to perinatal survival, but also long term morbidity and mortality.

References


Brittle or battered?

Sir,

Most paediatricians know very little about osteogenesis imperfecta. From time to time they are called upon to give 'expert' evidence in a case where a child has unexplained fractures. In most cases the differential diagnosis is obvious, but in others not quite so straightforward as Dr H Carty would have us believe.

My concern is that paediatricians faced with the task of providing expert evidence will refer to Dr Carty's authoritative annotation and in consequence may mislead the court. It is in the mild case of osteogenesis imperfecta in which the sclerae are normal colour and the teeth normal that most diagnostic difficulty arises. Paediatricians faced with the onerous task of providing an explanation for fractures should bear in mind the following points:

- No family history is necessary for the diagnosis of brittle bones.
- Wormian bones are not a sine qua non for the diagnosis of osteogenesis imperfecta.
- Metaphyseal fractures are a feature of both abuse and brittle bones.
- Normal bone texture and a normal radiographic appearance of the rest of the skeleton at the time of the first fracture do not exclude brittle bones.
- The fact that no recurrent fractures occur while the child is in care does not exclude the diagnosis of osteogenesis imperfecta. The interval between fractures may be many months or even years.

I believe that in very rare instances at the time of court proceedings it may not be possible to differentiate accurately brittle bones from non-accidental injury on the medical evidence alone.

References


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Dr Carty comments:

The diagnosis of mild osteogenesis imperfecta can be difficult but poses a clinical problem only in older children who are mobile and sustain frequent fractures after minor injury. Medico-legal problems arise, not with these children, but with children, usually under the age of 6 months, who have suffered non-accidental injury and who, it is claimed, have mild osteogenesis imperfecta despite the
absence of a family history and the presence of stigmata classically characteristic of non-accidental injury: complex skull fractures, metaphyseal fractures, rib fractures, and fractures at different stages of healing. Fractures sustained by children with mild osteogenesis imperfecta are the same as accidental fractures sustained by normal children: typically fractures of the shafts of long bones. Astley states that metaphyseal fractures are found only in severe osteogenesis imperfecta and not in mild cases. Rib and skull fractures occur in mild osteogenesis imperfecta with the same frequency as they do in normal children. The only series describing metaphyseal, skull, and rib fractures as a prominent feature of mild osteogenesis imperfecta is the series of Paterson quoted by Dr Blumenthal. I am sure that Dr Blumenthal has read the ensuing correspondence from Taitz who points out that not all courts have accepted Paterson’s evidence. Taitz has calculated, with support of several authoritative references, that the chance of a child having a mild form of osteogenesis imperfecta without a family history is between 1:1 000 000 and 1:3 000 000.

The third statement (about metaphyseal fractures) in Dr Blumenthal’s letter is quoted directly from Paterson who in turn claims authority from Astley but Astley’s full quotation is: ‘There is evidence of obvious generalised bone disease of osteogenesis imperfecta so that confusion with non-accidental injury does not occur.’

For these reasons I cannot accept Dr Blumenthal’s claim that courts will be misled by what I have written.

References

Non-accidental injury and copper deficiency

Sir,

Recent excellent articles by Dr Helen Carty1 and Dr J C L Shaw2 on non-accidental injury and copper deficiency correctly and clearly state that the court is not the right place to decide on the child’s welfare. I tend to agree with them. It appears that on both prosecution and defence sides winning a case if often of more importance than the truth. In achieving this each side tries to exploit the subtle nature of the medical evidence presented by the so called expert witnesses; this evidence has to be grasped and decided upon by the non-medical judges and juries—an almost impossible task. In my opinion the medical experts are as much to blame as the non-medical law profession in compromising the child’s welfare. Surely society and the medical profession, as part of society, should not accept this unsatisfactory situation and should campaign for a system, perhaps away from the courts, which seeks the real truth and ultimately does justice to the child.

Good tolerance of pyrazinamide in children with pulmonary tuberculosis

Sir,

By adding pyrazinamide to the usual triple treatment consisting of rifampicin, isoniazid, and ethambutol, it is possible to reduce the duration of antituberculous treatment with equal efficacy.1 Despite recommendations2 paediatricians seem reluctant to prescribe pyrazinamide, probably because of fear of hepatotoxicity and effects on uric acid metabolism.

We have assessed the tolerance of pyrazinamide in 43 children with primary tuberculosis and abnormalities on chest radiography (especially hilar lymphadenopathy with or without parenchymal infiltration). The diagnosis of tuberculosis was confirmed by a positive Mantoux test (>10 mm) and the finding of Mycobacterium tuberculosis in the children or their parents, or both. Mean age at diagnosis was 6 years (range 2 months to 15 years). Nineteen children were less than 3 years old.

Each morning, one hour before breakfast, during the initial phase all the children received rifampicin, isoniazid, and ethambutol at recommended paediatric doses3 and pyrazinamide (mean dose 23.3 mg/kg, range 20 to 37 mg/kg). This regimen of four drugs was prescribed for two months in 38 children, three months in two, and four months in three children (because of initial resistance to isoniazid). The second phase of treatment consisted of daily rifampicin and isoniazid.

Plasma uric acid and plasma glutamic oxaloacetic and glutamic pyruvate transaminases were measured before initiation of treatment, one month later, and at the end of treatment with pyrazinamide. Plasma uric acid concentrations were normal before treatment (150–360 μmol/l). They increased slightly (360–450 μmol/l) in nine of the children (21%) and returned to normal when pyrazinamide was stopped. There were no signs of gout nor arthralgias.

Liver enzymes were normal before treatment in 42 children and remained normal in 40. In two children transaminases increased (to three times normal values) at one month but returned to normal without modification of treatment. In one child the four antituberculous drugs were started just after viral A hepatitis; this did not prevent the normalisation of liver functions.

This paediatric study is in agreement with those carried out in adults and shows that pyrazinamide is well tolerated when used in doses <35 mg/kg/day even in infants. Furthermore, except in those five children with initial resistance to isoniazid, the duration of treatment was reduced to six months, which compares favourably with