Letters to the editor

‘Use of rectal diazepam in the community’

SIR–The paper by Norris et al.1 which concludes that ‘the use of diazepam as first line therapy for children with acute seizures needs to be reviewed’ (p 340), has put community paediatricians in a difficult position.

Norris et al. estimated that the overall incidence of respiratory depression after administration of diazepam was 9%. I am sure that the authors would be the first to agree that, as their study was hospital-based and thus subject to selection bias, their findings cannot easily be extrapolated to community practice.

The study examined children who had seizures from various causes. Secondary analysis of the data suggests that when the condition being treated was epilepsy, the relative risk of respiratory depression was half that of other causes of seizure (4/71 versus 7/59, respectively). Be that as it may, this still implies a 5 to 6% risk when treating people with epilepsy. This figure simply cannot be ignored if true.

I have audited all the children in my trust who have rectal diazepam ‘on site’ in their schools. There were 50 retrospective, documented treatment episodes in eight children, and no evidence of respiratory depression was found in these cases. Two episodes of respiratory difficulties were recorded; in one the rectal diazepam had not been given and the child continued to have seizures, and in the other, the diazepam failed to control the seizure, and the child subsequently developed difficulties as a result. These cases merely illustrate that uncontrolled seizures have serious recognized morbidity.

To find the true risk of treating epilepsy with rectal diazepam in school, I have calculated that 300 treatment episodes in different individuals would have to be analysed. According to my audit, a sample of 100 000 children would have to be surveyed to attain this number of treatment episodes. Clearly such a study is impractical.

These findings prompt two important questions: should the use of rectal diazepam be restricted in our schools? Or should all teachers be trained in advanced life support, and if so, by whom? The first option may be more acceptable to teachers unions (which are currently reticent). However, restricting the drug would undermine the very reasons for using it in the first place and might increase the number of children going into status or having respiratory problems, as occurred in my audit. The second choice would be time-consuming and expensive. The life support coordinator for our trust stipulates that 1–2 hours of tuition in a maximum group of four people are required to become effectively trained in resuscitation techniques. However, measures must be taken if there is a significant (as yet unproven) risk attached to current community practice.

More research is needed in the community before rectal diazepam can be deemed too dangerous to use first line.

Perhaps researchers should think carefully before making sweeping statements about the safety of current practices, which are outside their remit.

Diazepam may carry risks in accident and emergency departments but we must think clearly before stopping its use in our schools.

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References

‘Norris et al. reply’

SIR–We welcome the letter by Dr Mackereth regarding the management of children with acute seizures in the community. We agree it would be inappropriate to extrapolate our findings from a hospital study to the community. We are delighted that Dr Mackereth has been reviewing the use of diazepam as first line therapy for children with acute seizures as we have suggested. We have not stated that diazepam should not be used.

It is essential, however, that research is carried out so that children who are having seizures and require treatment receive the drug that is most effective, with the least toxicity, by the most appropriate route. Research is not restricted to randomized controlled trials1 and more information regarding the efficacy and toxicity of anticonvulsants used for acute seizures is clearly needed2, both in hospital and the community.

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References

‘Transient cytochrome oxidase deficiency with Ohtahara syndrome’

SIR–We reported previously a child with Ohtahara syndrome and cytochrome oxidase deficiency1 and wish to update you with a new and interesting development. At the age of 2 years, this boy was hypotonic with profound developmental delay, normal brain MRI scan, and seizures.

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References
Muscle biopsy at this time showed markedly reduced activity of cytochrome c oxidase ([complex IV] 0.003 µmol/g wet weight/min [reference range 0.1–5.0 µmol/g wet weight/min]) with normal activities of the mitochondrial respiratory chain complexes I, II, and III. Mitochondrial DNA analysis for the common disease causing point mutations and rearrangements revealed no abnormalities.

The quadriceps muscle biopsy has been repeated 3 years later. This showed that cytochrome c oxidase (COX) activity is now 0.299 µmol/g wet weight/min, within our reference range. The ratio of cytochrome c oxidase/succinate to cytochrome c reductase was initially 0.019, well below the reference range of 0.12–2.52 (based on 200 biopsies) and is now within our reference range: 0.192. The activities of complexes I and III were once again normal. Muscle histology paralleled this normalization of muscle biochemical function. The first biopsy showed a fairly uniform staining of all fibres in the COX reaction with no clear checkerboard pattern of darker (type-1) and lighter (type-2) fibres. In the second biopsy, however, the normal checkerboard pattern was clearly demonstrated. There were also residual subtle abnormalities, including a few fibres with a coarse, granular staining pattern, others with patchy staining, and many type-2 fibres had only a few weakly staining mitochondria. Interestingly, the type-1 fibres were of normal size and the type-2 fibres were atrophic; the reverse of the pattern seen in the first biopsy.

Clinically at 4 years of age the child has increased tone in all four limbs. He responds to light, but his vision remains impaired. His bearing is normal. Seizures continue to be a problem.

It appears that this is a rare case of transient reversible cytochrome oxidase deficiency of which there have been previous case reports. In the other cases reported, the children appeared to have a good clinical outcome, with the disappearance of clinical signs. Unfortunately this does not appear to be the case with our patient. It is possible that in this patient the early COX deficiency altered or impaired brain development, leading to the neonatal epileptic encephalopathy and consequent developmental delay, continued visual, cognitive and motor impairment, and epilepsy.

Dr Cathy Higgins
Dr George Gray
Dr Pramila Raman
Mr Kelvin Poulton
Dr William Whitehouse

References

‘Pseudohypertrophy of the temporalis muscle in Xp21 muscular dystrophy’

SIR–A 17-year-old male with Xp21 muscular dystrophy confirmed by cDNA probe CFS6b, and clinical progression in keeping with a Duchenne phenotype, presented to his neurologist for investigation of episodes of loss of consciousness. Although these episodes were thought to be related to a developing cardiomyopathy, an MRI of the brain was performed. No significant intracranial abnormality was found. However, there was striking symmetrical enlargement of the temporalis muscles with signal characteristics in keeping with the fatty infiltration of pseudohypertrophy (Fig. 1).

Duchenne muscular dystrophy is often referred to as pseudohypertrophic dystrophy because the affected muscles characteristically enlarge as a result of fatty fibrous replacement. Pseudohypertrophy of the calf muscles occurs in 95% of cases. Other muscle groups commonly affected include infraspinatus, deltoïd, wrist extensors, serratus anterior, quadriceps and glutei, and the anterior tibial muscles. Although macroglossia and hypertrophy of the masseters have previously been described, this is the first time, to our knowledge, that pseudohypertrophy of the temporalis muscle has been reported.

Figure 1: MRI showing no intracranial abnormality but striking symmetrical enlargement of temporalis muscle with signal characteristics in keeping with fatty infiltration of pseudohypertrophy.
References


Notices

International Symposium on West’s Syndrome and Related Infantile Epileptic Encephalopathies (ISWS)
Yayoi Memorial Hall, Tokyo Women’s Medical University, Shinjuku-ku, Tokyo, Japan. February 10–11, 2001

This meeting is relevant to those who wish to gain a better understanding of etiopathogenesis and exploration of new treatment strategies in various intractable epileptic encephalopathies, including West’s syndrome, early infantile epileptic encephalopathies with suppression burst (Ohtahara syndrome), and severe myoclonic epilepsy in infancy (Dravet). The official language of the meeting is English. The deadline for Abstracts is October 31, 2000. For further information, please contact Yukio Fukuyama, Secretariat, ISWS, c/o Child Neurology Institute, Samban-cho TV Plaza, 5F1, 24 Samban-cho, Chiyoda-ku, Tokyo, 102-0075, Japan. Tel: +81 3 3238 1580; Fax: +81 3 3238 1502.
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4th Congress of the European Paediatric Neurology Society
Baden-Baden, Germany. September 13–16, 2001

Dr F Hanefeld, President of the European Paediatric Neurology Society cordially invites you to participate in the forthcoming congress in Germany. For further information, please contact, AKM Congress Service GmbH, Obere Schanzstrasse 18, D-79576 Weil am Rhein, Germany. Tel: (+49) 7621 98 330. Fax: (+49) 7621 78 714. E-mail: akmweil@akmcongress.com. Web site: www.akm.ch/epns2001.

6th International Conference of the Baltic Child Neurology Association
Kaunas, Lithuania. June 13–16, 2001

For further information, please contact, Dr Nerija Vaiciene/Dr Milda Endziniene, Department of Neurology, Kaunas University of Medicine, Eiveniu 2, LT–3007 Kaunas, Lithuania. Tel/Fax: (370–7) 330 423. E-mail nerija.vaiciene@takas.lt.
Web site: www.info.kma.lt/BCNA.

Submissions to Developmental Medicine & Child Neurology

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