LETTERS TO THE EDITOR

Dental enamel defects and coeliac disease

EDITOR,—The association between coeliac disease and dental enamel defects (DED) is already known. These defects range from discoloration to pitting, grooving, and total loss of enamel, and are considered to be coeliac disease specific when distributed symmetrically and chronologically in all four sections of dentition. The Italian Society of Pediatric Gastroenterology and Hepatology (SIGEP) promoted a multicentre study for evaluating the prevalence of DED in a large group of Italian patients with coeliac disease. A total of 603 children with coeliac disease were studied (327 girls, 276 boys, mean age 17.8 years) in 13 Italian centres for paediatric gastroenterology. All subjects had permanent or mixed (permanent plus primary) dentition. The diagnosis of coeliac disease had been made in all cases using the criteria of the European Society of Paediatric Gastroenterology and Nutrition. The dental enamel inspection was performed in each centre by a paediatric gastroenterologist experienced in identifying enamel defects, with a dentist present. A group of 694 schoolchildren (mean age 12.4 years) served as healthy controls. When a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control, a subject from the control group (mean age 12.4 years) served as healthy control

Table 1 Prevalence of DED in 603 Italian children with coeliac disease (CD) in 13 paediatric gastroenterology departments. The correlation with the age of diagnosis of CD is shown

<table>
<thead>
<tr>
<th>Centre</th>
<th>No with CD</th>
<th>Positive (%) for DED</th>
<th>Mean age of diagnosis of CD (years) Total</th>
<th>With DED (years)</th>
<th>Without DED</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chieti</td>
<td>31</td>
<td>26 (83)</td>
<td>4.3 Total</td>
<td>4.3</td>
<td>2.3</td>
</tr>
<tr>
<td>Milano</td>
<td>40</td>
<td>30 (75)</td>
<td>6.6</td>
<td>8.4</td>
<td>5.0</td>
</tr>
<tr>
<td>Varese</td>
<td>33</td>
<td>17 (55)</td>
<td>6.3</td>
<td>6.3</td>
<td>5.1</td>
</tr>
<tr>
<td>Trieste</td>
<td>90</td>
<td>48 (53)</td>
<td>10.1</td>
<td>13.1</td>
<td>7.1</td>
</tr>
<tr>
<td>Fiesole</td>
<td>40</td>
<td>2 (50)</td>
<td>4.6</td>
<td>6.2</td>
<td>3.5</td>
</tr>
<tr>
<td>Modena</td>
<td>25</td>
<td>12 (48)</td>
<td>8.2</td>
<td>9.5</td>
<td>7.1</td>
</tr>
<tr>
<td>Milano 2</td>
<td>37</td>
<td>15 (40)</td>
<td>6.7</td>
<td>5.1</td>
<td>7.2</td>
</tr>
<tr>
<td>Messina</td>
<td>23</td>
<td>7 (30)</td>
<td>8.4</td>
<td>9.5</td>
<td>7.2</td>
</tr>
<tr>
<td>Cagliari</td>
<td>85</td>
<td>14 (16)</td>
<td>10.4</td>
<td>15.5</td>
<td>5.3</td>
</tr>
<tr>
<td>Bologna</td>
<td>77</td>
<td>12 (15)</td>
<td>6.1</td>
<td>8.4</td>
<td>3.1</td>
</tr>
<tr>
<td>Roma</td>
<td>82</td>
<td>10 (12)</td>
<td>4.5</td>
<td>4.2</td>
<td>5.2</td>
</tr>
<tr>
<td>Cesena</td>
<td>46</td>
<td>2 (4)</td>
<td>7.6</td>
<td>11</td>
<td>4.7</td>
</tr>
<tr>
<td>Palermo</td>
<td>15</td>
<td>5 (33)</td>
<td>4.2</td>
<td>4.9</td>
<td>3.3</td>
</tr>
<tr>
<td>Total</td>
<td>603</td>
<td>195 (32)</td>
<td>6.5</td>
<td>8.1</td>
<td>4.1</td>
</tr>
</tbody>
</table>

DED are therefore present in one third of Italian patients with coeliac disease, with a prevalence lower than that found in the Finnish studies1 but higher than that found in UK.2 This study also suggests that DED may be connected to late diagnosis of coeliac disease. The mechanism of development of DED in coelacis is not clear, but it seems more likely that they are the consequence of immune mediated enamel damage rather than related to malnutrition. In fact, similar lesions that appear to be associated with HLA DR3 haplotypes,3 are common in autoinmune disease (such as polyendocrinopathy), but are rare in nutrition disorders such as rickets. Our study also confirms that coeliac disease can be symptomatic or can present with an atypical clinical picture. We recommend that subjects with symmetrical DED in permanent teeth undergo serological testing for antientomysium antibodies and intestinal biopsy when testing positive.

ALESSANDRO VENTURA
STETANO MARTELLOSI
(COORDINATORS)
SIGEP Study Group on Dental Enamel Defects in Coeliac Disease
Istituto di Clinica Pediatrica, Universita' di Pisa and *Istituto per l'Infanzia e IRCCS (Burlo Garofolo), Trieste, Italy

recovery has been gradual but sustained, and is not yet complete. Pupillary involvement in GBS appears to be rare as it is not cited in at least one reference text;1 furthermore, it has been stated that oculomotor abnormalities do not occur in GBS. However, pupillary abnormalities have been reported in seven adults with GBS; all except one required ventilatory support and two (the non-ventilated, and one of the ventilated patients) died of a cardiac arrhythmia.1,2 Although there may be a reporting bias, pupillary involvement would appear to be associated with severe disease and an increased risk of dying. The pathophysiology is unclear but almost certainly represents a manifestation of autonomic dys-function. Pharmacological testing previously has suggested either simultaneous postganglonic involvement of sympathetic and parasympathetic nerves or isolated parasympathetic involvement.1 In our patient, pupillary pares is developed at the height of autonomic instability. It appeared to affect both sympathetic and parasympathetic nerves, although this was not confirmed pharmacologically. Bilateral ptosis and failure of full pupillary dilatation in the dark indicated sympathetic involvement, while absence of light and consensual responses suggested parasympathetic involvement. The ‘mid-point’ dilatation of the pupils was probably due to the presence of intrinsic muscle tone within the pupillary muscles. We believe that the recognition of iridoplegia as an associated feature of GBS is important and its presence warrants intensive monitoring.

JEREMY C. HUNG
RICHARD E. APPLETON
The Roald Dahl EDG Unit, Royal Liverpool Children’s Hospital, Alder Hey, Eaton Road, Liverpool L12 2AP

Predictive value of preschool surveillance in detecting learning difficulties

EDITOR.—In trying to predict learning difficulties using preschool data, Corrigan et al have overlooked some basic principles of health surveillance and screening for developmental problems.1 Detection of learning difficulties is a reasonable goal of health surveillance. However, health surveillance does not include using data from one point in time to predict problems seven to eight years later. In fact, the efficacy of preschool developmental screening as a global phenomenon is still not clear and certainly has not been shown to be useful in detecting mild to moderate learning difficulties.2 There is also generally a poor correlation between perinatal events and subsequent learning outcomes except in extreme cases. Predictive learning difficulties using data from the preschool or even neonatal period also seems a paradox given that learning difficulties are by definition related to educational problems that occur in schools. The authors have supported this theory by specifically choosing children who had been at school a minimum of two years, thereby allowing teachers time to detect learning problems.

The outcome measures of developmental delay or learning difficulties in the preschool period that are used in this study are vague and both require further definition. The diagnosis of developmental delay using referral to a psychologist or documentation in the child health record would fit best with surveillance methodology rather than an outcome measure. Preschool learning difficulties, once again seem to be an incongruent concept given the previous definition. The results do not seem to answer any hypotheses of clinical relevance and this is evident by the fact that being a single mother in the neonatal period appeared to be protective against learning difficulties, when in fact we know the opposite is true.

In conclusion, the authors have reiterated the supposition that has been part of the literature for a number of years; learning difficulties cannot be assessed until the child is in an educational facility. Mild learning difficulties may be a reflection of maturational variability and the importance of detecting this in the preschool period is still being debated. There is no doubt that it is important to detect learning problems early in a child’s schooling and services should be in place to support these children.3 There seems little point in mounting extensive surveillance programs in an attempt to predict difficulties years later. Outcomes of health surveillance should be rather directed to interventions which can be implemented in the present.

SHARON GOLDFIELD
THERESA LAZZARO
FRANK OBERKLAD
Centre for Community Child Health and Ambulatory Paediatrics, Royal Children’s Hospital, Flemington Road, Parkville, Victoria 3052, Australia

Predicative value of preschool surveillance in detecting learning difficulties


Bisphosphonates in osteogenesis imperfecta

EDITOR.—I was interested to read the recent articles by Allgrove1 and Williams et al on the use of bisphosphonates in children and would like to report my experience of their use in a child with osteogenesis imperfecta. The patient was referred to me age 9 years with a history of recurrent fractures since infancy and a family history consistent with osteogenesis imperfecta. She had a nine month history of low back pain, evidence of a thoracolumbar kyphosis, and tendon oedema of the lumbar spine. She had become wheelchair bound following a fractured femur three months previously and a recent hospital admission with pyrexia and a painful back. Osteoporosis was associated with upper motor neurone signs in her legs. Radiography of her spine showed collapse of numerous vertebrae in the thoracic and lumbar spine with marked osteoporosis. Routine biochemistry showed no abnormality of serum calcium, phosphate, alkaline phosphatase, parathyroid hormone, or 25-hydroxyvitamin D and a normal urine calcium/creatinine ratio. Bone density of the lumbar spine (L-1, L-2, L-3) using a Lunar DPX-L DXA scanner with the paediatric software, was 0.395 g/cm2 with a Z score for age of –4.0. After discussion with her parents it was decided to treat her with bisphosphonates. This was initially with pamidronate 35 mg/kg given intravenously every three months for six months which was then increased to 1 mg/kg/day for two consecutive days for a further six months. Because of difficulties with venous access this was then changed to etidronate given orally for a period of two weeks in every three month period in a dose of 600 mg/day (9 mg/kg) which has continued for the past nine months. No adverse effects were seen during the period of treatment.

The change in bone density during this period is indicated in table 1. There was a 44% increase in the bone density of the lumbar spine with no further fractures occurring during this time. This has been accompanied by a progressive improvement in her mobility such that she is now walking with the aid of a rollator and uses the wheelchair only for long distances. She now has no evidence of spinal deformity. Although it could be argued that some of this improvement was due to the onset of puberty, the improving Z score indicates an effect independent of changes in body size. Thus this case supports other reports indicating the potential benefits of bisphosphonates in osteogenesis imperfecta, although it is important that there is careful selection of cases, and as indicated by Allgrove that monitoring of bone biochemistry and density is undertaken. The need for appropriate informed consent is also essential.

White matter attenuation and megalencephaly

EDITOR—In 1985 one of us (ROR) published a case of unusual self resolving leukodystrophy.1 We now describe a second case. As with the first, neuroimaging was prompted by a large head size. At 14 months, she had an occipitofrontal circumference of 53.5 cm and her head circumference was similarly increased being 63 cm. His father’s head was also said to have been large. Unfortunately the patient’s head circumference at birth was not available. Apart from a slight delay in acquisition of head control and sitting attributable to the mechanical disadvantage of her large head, her development up to that time was age appropriate. Computed tomography showed pronounced low attenuation throughout the cerebral white matter (see fig 1A). Investigations for progressive leukodystrophy including relevant lysosomal enzyme studies, very long chain fatty acids, peripheral neurophysiology, and urine for N-acetyl aspartate were all normal. A second computed tomogram at 2.2 years showed partial resolution of the white matter changes in all areas, but to a lesser extent in the frontal lobes (see fig 1B).

Her head growth continued parallel with the normal growth trajectory at 3.5 to 4 SDs above the mean. When seen aged 8.6 years, her occipitofrontal circumference was 57 cm. She was otherwise normal neurologically and academically functioning around the mean. Magnetic resonance imaging of her brain at this stage showed slight increase in signal in the terminal myelination areas and a small discreet high signal area in the anterior frontal lobe of uncertain aetiology and unlikely to be of clinical significance.

This second case confirms that occasionally children with familial macrocephaly initially have pronounced white matter changes which appear alarming but are nevertheless benign.

Salmonella meningitis acquired from pet snakes

EDITOR—Salmonellosis associated with reptiles is well documented but a proved link between salmonella meningitis and reptilian carriage has not been reported.

Salmonella azaramo was isolated from the cerebrospinal fluid, blood, and stool cultures of a 5 month old white boy, admitted with symptoms, signs, and typical findings of bacterial meningitis. Treatment with cefotaxime 200 mg/kg daily for three weeks appeared to provide a rapid clinical response but the child was readmitted three days after discharge with vomiting and irritability. S azaramo was reisolated from the cerebrospinal fluid.

Ciprofloxacin was added to cefotaxime and this time the infection was successfully eradicated, the child being discharged after a further three weeks with normal cerebrospinal fluid and no apparent neurological deficit.

The family owned three snakes: an Indian python and two Royal pythons, which lived in two separate tanks in the dining room. The Indian python was unusual in that it roamed freely around the house. Faeces samples from the Indian python grew S azaramo; faeces from the Royal pythons grew S azaramo, S arizonae, S lome and an unnamed salmonella species. None of the family members had suffered significant diarrhoeal episodes and stool samples were negative.

Import regulations on reptiles have been significantly eased in recent years and a large market in reptile purchase and exchange exists in the UK. Most reptile owners are probably unaware of the likelihood of salmonella carriage in their pets, even though carriage of salmonella species in reptiles is almost universal (as high as 94%) and information is not provided to the potential purchaser on possible health risks and the importance of hand washing; young children are at particular risk of serious infection, the majority of salmonella meningitis cases occurring in patients under 1 year. Investigators of cases of salmonella infection should be aware of the possible significance of reptiles or exotic pets and liaise with local microbiology departments; faeces samples can be obtained relatively easily from lizards and snakes.
random' or some variant in the title or abstract/summary section.

These results were compared with the simple (and easily remembered) Medline search strategy of 'random*'-where * is the truncation character.

The simple search strategy found two thirds (59/90) of the handsearch defined RCTs, of which 78% (46/59) had the word 'random' or variants in the title or abstract. However, of the 31 RCTs which failed to be retrieved by this strategy, none (0/31) had 'random' or variants in the title or abstract (p<0.001).

We propose that the Archives adopts some of the CONSORT recommendations for the reporting of RCTs, by encouraging authors to include the word 'randomised' in the title and abstract of all these papers, enabling easier retrieval of useful clinical studies.

ROBERT PHILLIPS
TAMAR ROSENBERG
Oxford Medical School,
John Radcliffe Hospital,
Oxford OX3 9DX


The editors comment-
We agree and we are currently adopting this proposal.

Acquired rectovaginal fistulae in South Africa

EDITOR,—Acquired rectovaginal fistula associated with HIV positive children is a new and rather curious entity. 1 The underlying pathology has been reported to be chronic diarrhoea with abscess formation in the anterior rectal wall; after breakdown of the abscess, a persistent fistula appears. 2 We have seen seven such infants with fistulae in the past three years. We treated all these cases with diversion sigmoid colostomy. Biopsy of the fistula confirmed the presence of chronic inflammatory cells only. Our result with colostomy has been disappointing, with no spontaneous healing so far.

We would like to point out that in all these cases the fistula tract was from the upper anal canal to the vestibule just posterior to the hymen. Both of these children suffered from acute onset of diarrhoea one week before the fistula appearance. No causative organisms were grown from their stool cultures. If one is to take account of their clinical histories, rectovesicu- lar fistula may be caused by a specific but as yet undescribed organism, which is more prevalent in HIV positive infants.

B BANIEGHBAL
J PONSECA
Department of Paediatric Surgery,
Baragwanath Hospital,
Soweto,
Witwatersrand University,
Johannesburg,
Republic of South Africa


About 177,000 legal abortions are performed annually in Great Britain. (30 Jan 97, Col 333-334; 18.2.97)

The mean number of decayed, missing, or filled teeth in 5 year olds in England in 1987–8 was 1.73 and in 1993–4 it was 1.74. The corresponding figure for 12 year olds was 1.49 in 1988–9 and 1.15 in 1992–3. (28 Jan 97, Col 161-162; 18.2.97)

The government’s estimate of the proportion of UK gross domestic product spent on health is 4.7% for 1979–80, 5.2% for 1990–1, and 5.8% for 1995–6. (6 Feb 97, Col 722; 18.2.97)

There were 53 UK deaths from abuse of butane gas lighters and refills in 1990, 37 in 1991, 39 in 1992, 36 in 1993, and 28 in 1994. Most of the deaths were concentrated in the 15–19 age group for which the corresponding figures were 39, 29, 20, 21, and 14. (5 Feb 97, Col 641; 18.2.97)

The Adoptive Mothers (Maternity Leave) Bill which had its first reading in February seeks to give adoptive mothers the same maternity benefits and employment rights as biological mothers. (11 Feb 97, Col 143-145; 4.3.97)

Published perinatal mortality rates in European Union countries in 1993 varied from 5.1 per 1000 births in Finland to 9.1 in the UK but the figures are not comparable because the minimum gestation defining stillbirth varies from 22 to 28 weeks in different countries. (10 Feb 97, Col 13; 4.3.97)

Just under 2000 children in the UK were treated with human pituitary growth hormone between 1959 and 1985. A judge has found the Department of Health to have been negligent ‘in certain aspects’ beginning on 1 July 1977. Appeal against the judge’s decision is possible for those who had treatment before that date. (24 Feb 97, Col 123-130; 4.3.97)

At the Institute of Child Health, London 20 former patients are known to have died of Creutzfeldt-Jakob disease and two have symptoms of the disease. (25 Feb 97; Col 183, 234; 4.3.97)

Around 2% of the abortions performed in England and Wales are on girls under 16. The number of girls having an abortion fell from 4075 in 1987 to 3167 in 1992 but rose each year after that to reach 3401 in 1995. (18 Mar 97, Col 543-544; 1.4.97)

In 1995 about one in three births in England, Wales, and Scotland was to an unmarried mother. In Northern Ireland it was less than one in four. (20 Mar 97, Col 793-794; 1.4.97)

In January 1996 the proportion of pupils in each local education authority in England who were statemented for special educational needs varied from 1.97 to 4.59%. (10 Mar 97, Col 2-4; 1.4.97)

The number of tonsil and adenoid removals in children in each health authority in England in 1994–95 varied from 9.4 to 156.8 per 10,000 children under 16. The health authority with a rate of 9.4 was the only one less than 34 per 10,000. The mean rate was 71.6 with 71717 operations performed nationally. (20 Mar 97, Col 798-800; 1.4.97)

MORE MEETINGS IN 1997

ESPGAN Summer School: Paediatric Gastroenterology and Nutrition
30 August–6 September, Stockholm, Sweden
Further details: Dr Yigael Finkel, Paediatric Gastroenterology, St Göran’s Children’s Hospital, S-112 81 Stockholm, Sweden

Global Strategies for the Prevention of HIV Transmission of Mothers to Infants
3–6 September, Washington, DC, USA
Further details: Conference on Global Strategies, 7101 Wisconsin Avenue, Suite 1300, Bethesda, MD 20814, USA

British Society of Endocrinology and Diabetes
23–24 October, London
Further details: Dr R Stanhope, Department of Endocrinology, Great Ormond Street Hospital for Children NHS Trust, Great Ormond Street, London WC1N 3EH

Commonwealth Congress on Diarrhoea and Malnutrition
21–24 November, Karachi, Pakistan
Further details: Dr Z A Bhutta, Department of Paediatrics, Aga Khan University Hospital, PO Box 3500, Karachi 74800, Pakistan

National Symposium on Angelman Syndrome
29 November 1997, Brussels, Belgium
Further details: Dr B Can, 147 Avenue du Parc, 1190 Brussels, Belgium

Hot Topics ’97 in Neonatology
7–9 December, Washington, DC, USA
Further details: A Lynn Stillman, Registrar, 52 Overlake Park, Burlington, VT 05401, USA

Eighth Annual Course in Paediatric Gastroenterology
8–10 December, London
Further details: Professor J A Walker-Smith, University Department of Paediatric Gastroenterology, Royal Free Hospital, London NW3 2QG

Correction

Notice
There are a limited number of indexes for the Fetal and Neonatal Edition available. If you would like a copy please write to: Ms Sue Heels, Archives of Disease in Childhood, BMA House, Tavistock Square, London WC1 9JR.