Cystic fibrosis (CF) is a common autosomal recessive condition which typically causes sinopulmonary sepsis, and in most cases, pancreatic insufficiency with a resultant failure to thrive. Management includes treatment of bronchial inflammation and infection with antibiotics and physiotherapy, pancreatic enzyme replacement therapy, and fat-soluble vitamin supplementation. A high-calorie, high-fat diet is recommended for these patients, and high sugar foods are often eaten to maintain the increased calorific intake needed. Cystic fibrosis patients have been reported to have abnormal dentitions, with dental defects related either to the disease itself or as a consequence of treatment.1,2,3

Dental caries

Previous studies on patients with cystic fibrosis have reported variable caries prevalence. Jagels and Sweeney1 reported a reduced caries prevalence in 63 CF patients who were on long-term oral antibiotics, compared with 56 of their siblings. This confirmed the findings of Littleton and White in 1964,4 who looked at children receiving long-term antibiotic therapy and showed lower caries experience in children receiving oral penicillin for an extended period. They concluded that this finding was a result of the antibiotic, rather than a consequence of a specific disease process and could be extrapolated to any group of children on long-term antibiotic treatment. Primosch and co-workers6 reported significantly reduced dental caries prevalence in 86 CF patients compared with an age and gender matched control group, with a greater reduction in caries prevalence in the primary dentition compared with the permanent dentition.

Kininons1 reported a similar reduction in dental caries in 131 patients with cystic fibrosis, compared with a similar number of control individuals; the reduction here was also only significant in the primary dentition. In addition, a more recent study by Martiens et al of 37 CF patients aged 6–38 years, age and sex matched with a similar number of control individuals, showed no difference in caries prevalence between groups.7 Other work by Kininons1 concluded that the reduction in dental caries prevalence seen in the CF patients surveyed, was associated with a significantly higher mean pH and buffering capacity of their stimulated saliva. This researcher suggested that there was a link between altered saliva properties and low caries experience, but found no relation between the changes in saliva and the severity of the disease process.

Periodontal health

Poor oral hygiene with concomitant accumulations of dental plaque and calculus is directly associated with inflammation of the periodontal tissues.8 The prevalence of calculus in children varies widely depending on patient demographics, social class, and general health. Patients with cystic fibrosis have altered amounts of calcium and phosphate in their saliva which can affect calculus formation. Wotman and co-workers9,10 described increased amounts of calculus in children with cystic fibrosis and asthma compared with “healthy” children, particularly in the older age groups, which they felt reflected raised calcium and phosphorus levels in the saliva of these patients. Kininons1 reported similar trends towards a higher prevalence of calculus in CF patients that were positively and significantly related to age. In contrast, another study of patients with cystic fibrosis has shown conflicting results with regard to plaque scores, calculus levels, and periodontal health.11 Jagels and Sweeney,1 in a study designed to measure dental calculus levels over a study period of eight weeks, reported values for the debris index since the prevalence of calculus was extremely low in all groups. They reported no significant difference between CF subjects compared with healthy controls except that those who chewed pancreatic enzyme supplement had a slightly lower debris index.

The dental effects of pancreatin have also been reported in animal experiments on rats by Baumhammers and colleagues.12 In this study, the level of calculus was correlated...
with the use of pancretin, and extrapolating from the results it was postulated that CF patients taking pancretin would show lowered plaque and calculus deposits. This view was also supported by Ennever and Sturzenberger, who studied the benefits of pancretin supplemented chewing gum on oral health in human adults and found a significant reduction ($p < 0.01$) in the occurrence of dental calculus compared with a control group using a placebo gum.

Long term antibiotic therapy has been shown to lower periodontal index scores. Littleton and White showed a slight lowering of mean periodontal index scores in patients on long term antibiotics; however, the changes were generally limited to inflammatory gingival changes only and the results were not statistically significant.

**Enamel defects**

Enamel defects are the result of impaired development of dental enamel. The term has been used to describe a range of appearances of enamel, the structure of which is disrupted during its formation or maturation phases; it represents defective mineralisation.

Clinically, hypomineralisation affects the translucency of dental enamel and can also lead to chipping of the enamel, rough surfaces, and, in some cases, as a consequence, also potential for more rapid caries development. Many investigators have believed that the integrity of the tooth enamel is an important factor in determining caries immunity or susceptibility. In 1943, Brucker investigated the dental health of 931 public school pupils. Only 0.7% of the 28,934 teeth examined were found to have enamel defects, yet nearly 11 times more permanent teeth were found to be decayed. The author reported that there was therefore no link between enamel defects and dental caries, but did comment on a preponderance of hypoplastic first permanent molars in boys, with these teeth showing a higher prevalence of dental caries. Brucker concluded that the two disorders did not have a common origin and that no relation exists between caries and developmental defects of enamel. The fact that the teeth most affected by enamel defects were upper incisors, yet first permanent molars are the teeth most affected by caries, supports this conclusion.

With respect to the dental effects observed in patients with cystic fibrosis, it remains unclear which relate to the management of the condition and which relate to the metabolic effects of the disease itself. However, Suckling and Pearce, in a study of New Zealand children, reported that the incidence of any one or a combination of a number of common childhood illnesses during the period of tooth formation did not affect the appearance of the enamel.

The aim of this study was to assess the oral health of CF children compared with children with other respiratory disorders by determining the prevalence of dental caries, enamel defects, and dental calculus.

**METHODS**

**Children studied**

The study was approved by the Royal Brompton Hospital Research Ethics Committee. Children with cystic fibrosis or another respiratory disorder diagnosed using standard criteria, who attended the Royal Brompton Hospital as an outpatient during a four week period, comprised the study and control groups, respectively. Those excluded from the study included inpatients who were seen in the outpatient clinic, patients younger than 2 years of age, or those children who were unable to cooperate or where consent was refused. Written informed consent was obtained from the parents or carers of the children. The child’s ethnic group and diagnosis were obtained from their medical notes.

**Clinical dental examination**

A qualified dentist (AN), who had previously undergone a familiarisation exercise with two experienced epidemiologists, undertook an oral examination of the children. No advance notice was given to any of the children and each child was examined under the same conditions, in an outpatient clinic room under artificial light. The patient was seated on a high backrest chair with the examiner positioned behind the patient.

The examination was visual only and a probe used solely for debris removal. No radiographs were taken. The status of the surfaces of all erupted teeth present was recorded. Any missing primary incisors in children aged 5 years or more were considered to have exfoliated, while any missing primary molars in children aged 8 years or younger were regarded as missing because of caries. However, this was only after considering the caries state of the remaining dentition and questioning of the child and/or parent. Dental caries was recorded when there was a visible breach of the enamel surface. From this the DMFS and dmfs indices were calculated for the permanent and primary dentitions respectively.

The Community Periodontal Index (CPI) was recorded on all patients using a CPI probe. Scores were given for each of the six sextants, with the highest score for each sextant recorded. The scoring was 0 for healthy, 1 for bleeding on probing, and 2 for calculus present in that sextant.

The presence or absence of enamel defects was recorded according to the World Health Organisation (WHO) criteria.

The WHO assessment form is based on examination of the upper incisors, canines, and first premolars (or upper first primary molars in the case of the primary dentition) and the first permanent lower molars if present. If these teeth have not occurred or have been extracted, no substitution is made. A score is given for each tooth based on the presence or absence of enamel opacities. No grade or score is given for the extent of the opacity.

Patients attended for outpatient appointments infrequently, therefore all children were examined once only; re-examination to determine intra-examiner reproducibility was not felt to be feasible given the extended catchment area of the department, which is a tertiary referral centre for the specialty.

**Questionnaire**

A questionnaire was used to obtain information about home oral care and other dental aspects of each child’s care that may have influenced their oral health. Parents or carers were asked to complete this at the time of the examination. The information collected included the patient’s name, date of birth, and the parent’s occupation. In addition, the use of fluoride supplements, toothpastes, toothbrushing frequency, and help with toothbrushing, were recorded.

**Statistical analysis**

The significance of the difference between the means of the recorded variables was calculated using Student’s $t$ tests for independent samples. A confidence interval of 95% was selected to determine when the difference was statistically significant.

**RESULTS**

Data were obtained for 180 patients in three age groups: <6 years, 6–9 years, and ≥9 years. There were 74 children with cystic fibrosis; the control group included 106 children (23 with primary ciliary dyskinesia, 53 with asthma, 9 with recurrent chest infections, and 21 with other respiratory problems). The age range of patients was 2.5–16.5 years. During the specific time period only three children refused to participate and six were excluded as they were too young to cooperate.

Since there were no significant differences in the age distribution between children with cystic fibrosis and other respiratory disorders when analysed by age group ($p > 0.4$), the results were analysed further using age group as an independent variable (table 1). For group 1 (<6 years) dental
examination involved the primary dentition only. Group 2 (6–9 years) and group 3 (>9 years) both included children who had mixed dentition.

For group 1 there was no significant difference between CF children and the control children for any parameter (table 2).

As table 3 shows, group 2 cystic fibrosis children (aged 6–9 years) showed a statistically significant increase in the number of permanent teeth with enamel defects ($p = 0.003$), and a trend towards an increased prevalence of enamel defects in the primary dentition. In addition, there was a non-significant lower caries prevalence in the cystic fibrosis group in both dentitions. The periodontal health of the 6–9 year old CF children was worse than the non-CF children of a similar age; they had a higher mean number of sextants with calculus and a lower mean number of healthy sextants, although the differences were not statistically significant (table 3).

In group 3 (>9 years) the CF children showed a lower caries prevalence in the primary dentition and an increased number of permanent teeth with enamel defects, compared with the control group, although these differences were not statistically significant (table 4).

Table 5 describes the distribution of caries experience between decayed, missing, and filled tooth surfaces for all the children examined. Overall, the proportions of children who were caries-free were quite similar between CF and non-CF children for the youngest age group, while in the older age groups a higher proportion of CF children were caries-free. Fifty per cent of the CF children in the youngest age group had...
active decay, while in the non-CF group this proportion was 80%. Of the 6–9 year old CF children, 78% were caries-free compared with 69% of non-CF children in this age group. Although the overall caries experience was very low in the permanent dentition of the 6–9 year olds, it was represented by filled teeth in the CF children, while in the non-CF children, it was represented by decayed surfaces. The distribution of decayed and filled tooth surfaces in the group 3 children with experience of dental caries was similar for both primary and permanent dentitions.

With respect to the questionnaire, table 6 shows differences between both groups in terms of toothpaste type (fluoride content), frequency of use, and help with toothbrushing. The difference in the use of low fluoride toothpaste (<600 ppm F) between the youngest study and control groups can be explained by the numbers of children in each group (12 in the CF group, 34 in the control group). With respect to toothbrushing, 77% of CF patients were reported to be brushing their teeth more than twice per day compared with 67% of children with other chronic respiratory disorders. In addition, a higher proportion of CF children brushed their teeth unaided (80%), compared with 64% of children in the other respiratory disorders group.

Table 7 shows the distribution of enamel opacities between both primary and permanent teeth, and between the CF group and other respiratory disorders group. Overall this shows an increase in opacities in the permanent dentition compared to the primary dentition, and in the CF group compared to the other respiratory disorders group.
of long term antibiotics in the diet has the effect of reducing lulence can be hypothesised. Firstly, a large proportion of the seen, but some possible reasons for the reduced caries prevale-
mor and other teeth groups in the CF children.
from these data because of the small numbers of patients
more cariogenic diet. It is difficult to draw firm conclusions
similar results even though CF patients potentially have a
in the non CF group.
Only one patient in each group (CF and non-CF) had undergone an extraction of a lower first permanent molar, most likely because of caries according to the history from their parents.

**DISCUSSION**

This observational study showed a difference in the oral health of a group of children with cystic fibrosis compared with children who had other chronic respiratory problems. The difference observed was in the significantly higher (p = 0.003) mean number of opacities in the permanent dentition of the 6–9 year old CF children, while in the older group, the number of enamel defects and, in particular, enamel opacities, in the CF children was higher, but the difference was not statistically significant. The fact that there was very little difference in the mean number of enamel opacities in the primary dentition in all age groups suggests that the metabolic insult causing these enamel defects occurred postnatally. This could be the result of a chronic metabolic disease such as cystic fibrosis; however, the occurrence of opacities may relate to other conditions, for example, prematurity, low birth weight, fluoride ingestion from toothpaste, as well as disorders such as coeliac disease and asthma. Other studies, one in vitro using extracted teeth, and another from medical records of Dutch children, have shown a relation between developmental defects of first molar teeth and respiratory disorders.

Dental caries in the primary dentition was lower in the CF children who, in the youngest age group, had a mean dmfs of 0.17 compared with 0.41 for the non-CF children. This is in broad agreement with other studies, which have shown similar results even though CF patients potentially have a more cariogenic diet. It is difficult to draw firm conclusions from these data because of the small numbers of patients seen, but some possible reasons for the reduced caries prevalence can be hypothesised. Firstly, a large proportion of the cystic fibrosis children are on long term antibiotics as a consequence of their disease. Studies have shown that the addition of long term antibiotics in the diet has the effect of reducing the prevalence of dental caries. Secondly, CF children and their carers tend to be highly motivated and health conscious and may be good and regular dental attenders, although the evidence for this is limited. Certainly, in this study, there was a higher proportion of decayed primary teeth in the younger non-CF children compared with the CF children. Thirdly, the majority of CF patients were taking replacement pancreatic enzymes, which have been shown to be effective in caries reduction. However, as the aetiology of dental caries is multifactorial, other aspects need to be taken into consideration.

Salivary changes, including the pH and buffering capacity, have been linked to altered caries experience, and some studies of patients with cystic fibrosis have equated the reduced caries prevalence observed to anatomical variations of the teeth, which may predispose them to a lower risk.

The prevalence of calculus was not found to be significantly different between the two groups, for all ages studied. However, there was a trend towards a higher number of sextants with calculus in the CF group. No conclusions can be drawn from this because of the small number of patients observed and the fact that calculus deposits are not a common finding in young children. The increased number of sextants with calculus, observed in the cystic fibrosis group in the current study, correlates well with the reduction in number of healthy sextants.

One of the predisposing factors to salivary calculus formation is the higher level of calcium ions in the saliva of CF patients and an increased salivary pH that favours precipitation of calcium ions and thus calculus formation. Conversely, other workers implicate pancreatic replacement enzymes in the inhibition of calculus formation, but based only on evidence from animal studies.

With regard to oral hygiene, the higher frequency of toothbrushing seen in the CF children can be partially attributed to increased motivation of the children and their families, an increased awareness of health problems. In addition, the higher proportion of CF children brushing their teeth unaided probably reflects their increased motivation and awareness, which in turn tends to lead to an increased level of maturity.

**Conclusions**

In this study, while caries prevalence was reduced in the children with CF, the increased prevalence of enamel defects as well as calculus accumulation emphasises the need for a paediatric dentist to be a member of the multiprofessional team involved in the care of children with CF. Where this is not the norm, on diagnosis of CF, early referral should be made to a specialist paediatric dentist for relevant counselling and dental care.

Table 8 Distribution of opacities on incisors, molars, and incisors and molars together, in CF and non-CF children

<table>
<thead>
<tr>
<th>No. of incisors with opacities</th>
<th>No. of pts with incisors affected</th>
<th>No. of molars with opacities</th>
<th>No. of pts with molars affected</th>
<th>No. of pts with first permanent molars extracted</th>
<th>No. of pts with molar and incisor teeth affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>CF group</td>
<td>44 (14.8%)</td>
<td>21 (28.4%)</td>
<td>11 (9.0%)</td>
<td>6 (9.83%)</td>
<td>1</td>
</tr>
<tr>
<td>Non-CF group</td>
<td>30 (7.1%)</td>
<td>17 (16.0%)</td>
<td>6 (4.3%)</td>
<td>4 (5.7%)</td>
<td>1</td>
</tr>
</tbody>
</table>

**REFERENCES**

Mesial temporal sclerosis and temporal lobe epilepsy

Mesial temporal sclerosis is the most common lesion found in adults with temporal lobe epilepsy. For many years it has been held that these lesions may be related to complicated febrile convulsions in childhood but the association is based on retrospective data and a cause-and-effect relationship has not been proved. A magnetic resonance imaging (MRI) follow up study of children after febrile convulsions (R Tarkka and colleagues. Neurology 2003;60:215–8) has shown no evidence of mesial temporal sclerosis.

From a total of 329 children with febrile convulsions 64 were selected for study; 24 who had had a prolonged (30 minutes or more) convulsion, 8 who had had at least one unprovoked seizure on follow up, and 22 controls who had had a single simple febrile convulsion (short (15 minutes or less), non focal, and not repeated within the episode). MRI scans were performed after a mean follow up of 12.3 years. No patient had mesial temporal sclerosis and the volumes of right and left hippocampal formations and amygdala were similar in the three groups. A lower mean difference in volume between right and left hippocampal formations in the prolonged febrile convulsion group was not thought to be clinically significant. These researchers conclude that mesial temporal sclerosis is not a common sequel of even prolonged febrile convulsions and they believe that there is no causal relationship between the two.

On the other hand, mesial temporal sclerosis is common in children with severe temporal lobe epilepsy. In Montreal (C Bocti and colleagues. Neurology 2003;60:191–5) the neuropathology was reviewed of 22 children who had undergone anterior temporal lobectomy for refractory temporal lobe epilepsy during a 20 year period (1979–99). Fourteen of the 22 temporal lobes had cortical dysplasia (CD) of the neocortex and mesial temporal sclerosis was present in 12 of 15 specimens with available hippocampal tissue. Seven children had both kinds of lesion. The importance of dual pathology (mesial temporal sclerosis plus an extra-hippocampal lesion) in patients with severe temporal lobe epilepsy is increasingly recognised as are the clinical differences between paediatric and adult temporal lobe epilepsy. (Children describe fewer auras, have simpler automatisms, and more frequent motor manifestations, mostly tonic.) The relationship between prolonged febrile convulsions and later mesial temporal sclerosis is questionable and temporal lobe epilepsy in children may be a different entity from temporal lobe epilepsy in adults.