sought. A total of 161 patients (male/female ratio 1 : 1·3) were identified from the general anaesthetic day-case records. These were 8–11 years old at time of extraction and had one or more first permanent molars removed at the Royal London Dental Institute 1999–2001. Each patient was reviewed aged 12–16 years. A pilot study of 10 patients was carried out. Pre-extraction data included chronological and radiographic age determination and axial inclination of the second permanent premolar and molar measured from radiographs. The postextraction data collected included age and axial inclination of the second permanent premolar and molar measured from radiographs and intra/interarch occlusal relations measured from study models. This study will provide evidence-based data to assist in treatment planning for the early loss of first permanent molars.

4

A clinical evaluation of mineral trioxide aggregate for root-end closure of nonvital immature permanent incisors in children – a pilot study

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Aim of the research. To evaluate the clinical efficacy of MTA as an apexification material when used in nonvital immature permanent incisors in children.
Methods. The investigation was completed in two phases: an *in vitro* evaluation of the handling properties of MTA on five extracted human premolars and an *in vivo* determination of the clinical procedures on four subjects. The main clinical study consisted of 16 children and 18 nonvital permanent incisors. An apical plug of 3–4 mm was created using MTA and final obturation was completed using thermoplastisized gutta-percha. Clinical and radiographic follow-up was performed 3-monthly for 1 year.
Results. The 1-year follow-up will be completed end of April 2004. From preliminary data on 16 teeth MTA placement was considered to be adequate in 12 teeth (75%). Only 11 cases were reviewed clinically and among these, clinical success was 81·3%. Radiographic success was found to be 68.8% and in 5 cases (31·2%) the outcome was considered to be uncertain.
Conclusions. MTA appears to produce good clinical results when used as an apexification material. However, more controlled clinical studies are required to evaluate the long-term success of this procedure.

2004 BSPD Clinical Case Presentation Prize Entrants, sponsored by RA Medical

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1

Papillon–LeFevre Syndrome in a young boy: a clinical case presentation

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Presenting problem. Papillon Le–Fevre Syndrome (PLS) is characterised by Palmo plantar keratosis and a rapidly progressing periodontitis, which leads to early loss of both deciduous and permanent dentitions. Inheritance is though to be autosomal recessive and the incidence has been estimated at 1 in 4 million. PLS patients have a mutation in the Cathepsin C gene. This case focuses on the genetic, clinical, immunological and radiographic characteristics and their Dental Management.
Clinical Case Presentation. A 3-year-old boy, originally seen by a Specialist Practitioner for receding gums was referred to the Department of Periodontology of Birmingham Dental Hospital where a diagnosis of PLS was made from the clinical signs alone. Later he was seen in the

Paediatric Department for his long-term management. Genetic analysis showed a mutation in Cathepsin C gene by both the patient and his father which confirmed the diagnosis of PLS. His initial treatment consisted of mechanical debridement and the use of topical antimicrobials every 10–12 weeks with intensive oral hygiene instructions issued. This was supplemented by a combined course of Amoxicilin and Metronidazole for 10 days were signs of disease progression ever noticed.

Discussion. The patient is now $4^{1}/_{2}$ years, has retained all his deciduous dentition and there has been very little change in bone levels surrounding his teeth since the initial presentation. This case suggests that controlling the pathogenic microbiota by appropriate antibiotic and conventional periodontal therapy can slow down or arrest the progress of periodontitis in PLS patients as shown by other reports.

2

Advances in medical management: a 21st century dental dilemma

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Presenting problem. Two cases are presented; the first describes the dental management of a patient following chemo- and radio-therapy used in the treatment of rhabdomyosarcoma of the right mastoid at age two. Problems include hemifacial hypoplasia, hypodontia, microdontia, crowding and 'stacking' of the molars. The second describes a patient with Hurlers syndrome (mucopolysaccharidosis I) who underwent a bone marrow transplant. Problems include severe hypodontia and poor root formation of all adult teeth.

Clinical management. The management of both cases centred on simplifying all dental treatment and maintaining the dentition.

Discussion. The challenge faced in the dental management of the increasing number of survivors of severe childhood disease is discussed.

3

Goldblatt's syndrome in a 4-year-old boy: clinical case presentation

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Presenting problem. Spondylo-epimetaphyseal dysplasias (SMD) are an apparently heterogeneous group of disorders involving the epiphyses of joints. Their classification is based on predominant rather than exclusive anatomical localization. A peculiar type of chondroplasia related to these SMD's was reported by Jack Goldblatt in 1991. In addition to the typical features of SMD, in Goldblatt's syndrome there is also dentinogenesis imperfecta (DI) and ligamentous hyperextensibility (LH) present. We report the genetic, clinical, radiographic and dental manifestations of a 4-year-old who was diagnosed with Goldblatt's Syndrome and the dental management of this child.

Clinical Management. A 4-year-old boy was referred to the Department of Paediatric Dentistry, Birmingham Dental Hospital, suffering with toothache. His initial management consisted of extraction of his deciduous upper right second molar and both his deciduous lower first molars under general anaesthetics for relief of his symptoms of acute pain. These teeth were sent for histopathological examination. His long-term management consists of close liaison with other medical specialities, preventive treatment, orthodontic monitoring and restorative treatment to maintain occlusal height and restore aesthetics.

Discussion. The histopathological findings confirmed the clinical diagnoses of DI, which is a feature of Goldblatt's syndrome. Extra orally he displayed signs of SMD and LH which are the other two features of this syndrome. The genetic, clinical, radiographic characteristics and histopathological report of his extracted teeth of this boy confirmed the typical features of Goldblatt's syndrome.

4

Dental findings and treatment in an extended family with amelogenesis imperfecta

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Amelogenesis imperfecta (AI) is a genetic condition of defective enamel. It is inherited as a single gene defect occurring in approximately 1 : 40 000. This paper presents the dental findings and management of 5 boys from the same extended family (3 brothers and their 2 cousins) with hypocalcified amelogenesis imperfecta. All children showed a maternal inheritance pattern. The hypomineralised enamel was yellow/brown in colour and showed rapid breakdown. All children complained of sensitivity and poor appearance. Additional features associated with the AI included taurodontism and anterior open bite. Stainless

steel crowns were placed on the posterior teeth as soon as breakdown or sensitivity occurred and acid etch composite on the anterior teeth when indicated. All children received their treatment under local anaesthesia. SS crowns were placed on primary molars from as young as 3 years. This paper illustrates the dominant inheritance pattern and variations in clinical manifestation of this genetic condition and discusses the importance of early management

2004 BSPD Poster Prize Entrants, sponsored by Dentsply

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1

The frequency of repeat general anaesthesia for paediatric dentistry at Liverpool University Dental Hospital

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Aim. To investigate retrospectively the frequency of repeat extractions under general anaesthesia (GA), the pattern of disease and treatment for children who had received GA.
Methods. Records of patients attended GA chairs at Liverpool University Dental Hospital in January and February 2003 for extractions were studied. A data collection form was designed to record the relevant information.
Results. A total of 174 patients were seen for GA extractions with a mean age of 6·5 years. Of those, 22 patients (12·6%) had a history of previous GA extractions with a mean age of 5 years at first GA. The mean interval at second GA was 2·5 years. The main diagnosis for all patients was caries in the primary dentition (80%) and the main reason for the use of GA was lack of patient cooperation (88%), however, preventing long-term phobia in young patient was only found in 12% of the cases. In general, radiographs were taken in 38%, however, children who needed repeat GA, radiographs were only available in 13% at first GA. The mean number of teeth extracted generally was 4·4 compared to 3 teeth at first GA for those patients needed a repeat GA.
Conclusion. Although the frequency of repeat GA is relatively low it still indicate the need of more radical treatment planning, regular use of radiographs when possible and preventive dentistry to avoid the use of unnecessary dental general anaesthesia.

2

Has a preassessment clinic reduced repeat general anaesthetics?

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Aim. A previous audit showed 17% of children were having a second or third general anaesthetic for dental extractions. A preassessment clinic prior to general anaesthesia was set up at this time. The aim of this audit was to determine whether the clinic has reduced the number of repeat general anaesthetics. The objectives were: (i) to investigate the number of children having a repeat general anaesthetic for extractions within 3 years; and (ii) to establish if decayed or heavily restored teeth remained postoperatively.
Method. Patient records of 200 consecutive children undergoing general anaesthetic for dental extractions at the Bristol Dental Hospital from June 2000 were selected. Data collection included identifying which teeth were removed, whether radiographs were taken preoperatively, the presence of decayed or heavily restored teeth post-operatively and whether a repeat general anaesthetic was carried out within 3 years.
Results. Eight patients (4%) had a repeat general anaesthetic within the 3-year period and 50 patients (25%) had heavily restored or carious teeth remaining postoperatively.
Conclusion. The preassessment clinic has significantly reduced the number of children having a repeat general anaesthetic. This is in comparison to the initial audit carried out in 1997 in which 14% of patients had a second