

# Clinician Prize

c1

## **Blepharo-Cheilo-Dontic (BCD) syndrome: presentation and dental management**

**S. ADEBOYE\*, B. O. I. COLE & N. J. JEPSON**

*Newcastle Dental Hospital, Newcastle upon Tyne*

*Presenting problem.* A 12-year-old girl with Blepharo-Cheilo-Dontic (BCD) syndrome presented at the Newcastle Dental Hospital following referral from the multi-disciplinary cleft lip and palate clinic. Blepharo-Cheilo-Dontic (BCD) syndrome is characterized by abnormalities of the eyelid, lip and teeth. Her presenting complaint was missing front teeth. The permanent units missing were 15, 14, 13, 12, 11, 21, 22, 24, in the upper arch and 35, 34, 32, 44 and 45 in the lower arch. The history of her previous extractions was not available. She had Skeletal Class III molar and incisal relationships, left posterior cross bite, occlusal contacts on the second permanent molars and poor oral hygiene.

*Clinical management.* The *initial management* involved preventive and restorative measures including fluoride therapy, fissure sealants, scaling and polishing and tooth brushing instructions. The lower first permanent molars were restored with stainless steel crowns. The *intermediate management* addressed her main concern by the fit of an upper complete over-denture. *Long-term management* would involve bone grafting and implants.

*Discussion.* This patient presented a complex aesthetic problem, which through multi-disciplinary care resulted in a satisfactory aesthetic outcome. She will need continued monitoring into adulthood.

c2

## **Dental injury from air gun pellet**

**G. ABOU AMEIRA\* & M. GELBIER**

*Eastman Dental Institute, UCL*

*Presenting problem.* A 10 year old girl was referred to the Paediatric Unit of the Eastman Dental Hospital (EDH) following surgical removal of an air gun pellet. She had been accidentally shot with an air rifle, at six years of age. This avulsed the upper left primary central incisor and caused arrested root development of the upper left permanent central incisor. In addition she has amelogenesis imperfecta (hypomature type).

*Clinical management.* Although the long term prognosis of UL1 was poor, it was decided to retain the tooth for as long as possible. Calcium hydroxide endodontic therapy was instituted for one month, followed by root filling with Mineral Trioxide Aggregate (ProRoot MTA, Dentsply). Discolouration developed at the cervical margin therefore non vital bleaching was carried out as well as vital bleaching to improve the aesthetics of the hypomature enamel of the incisors. All the treatment was carried out under nitrous oxide sedation because the patient was apprehensive.

*Discussion.* The inappropriate use of air guns can result in severe, potentially fatal injuries in children. This is the first reported case of arrested root development of an upper right permanent central incisor as a result of an accidental air gun pellet injury.

c3

## **Combined speciality management of two dental trauma cases**

**L. BURBRIDGE\*, C. DEERY & K. HARLEY**

*Edinburgh Dental Institute, Edinburgh, UK*

*Presenting problem.* Two patients attended having each suffered a traumatic injury to one of their permanent maxillary central incisors. Patient 1 was aged nine years at the time of injury and patient 2 aged seven. Clinical and radiographic examination revealed that patient 1 had sustained a complicated oblique crown-root fracture to 11 and that patient 2 had sustained a severe intrusion injury to 11 with an associated enamel-dentine fracture.

*Clinical management.* Initial management for both patients was undertaken in conjunction with orthodontic colleagues. The traumatised teeth in both patients were extruded; for patient 1 a removable appliance was utilized to engage a fixed orthodontic attachment and for patient 2 a sectional fixed appliance was used. Extirpation of the pulp of the traumatised tooth for patient 1 was undertaken at the initial appointment and the root canal has subsequently been obturated. For patient 2, following loss of pulpal vitality, endodontic treatment was begun and apexification is currently being awaited. Both traumatised teeth have been restored with directly placed composite resin.

*Discussion.* Significantly traumatised teeth in these patients have undergone treatment which was acceptable to them. This involved a joint paediatric-orthodontic approach, in one case to facilitate restoration and in the other to reposition the tooth. It is anticipated that both these traumatised teeth will be maintained for the foreseeable future with the obvious benefits to the developing child.

c4

## **Early temporomandibular joint ankylosis related to mastoiditis**

**C. GARDNER\*, E. M. O'DWYER, H. ZAITOUN &**

**J. C. COOPER**

*Alder Hey Children's Hospital*

*Presenting problem.* A fourteen-year-old boy presented with a 2 year history of progressive limited opening secondary to right-sided temporomandibular joint (TMJ) problems following previous mastoid disease and surgery. He complained of difficulty with eating but no other joint symptoms.

*Clinical management.* Clinical examination demonstrated no evidence of any mandibular deformity or asymmetry though he had considerably reduced mandibular movements and jaw opening (maximum 15–20 mm). Radiographic examination and subsequent CT (computed tomography) scans revealed expansion of the right mandibular condyle with erosive and proliferative changes also affecting the right glenoid and articular fossa. In view of this patient's increasing functional problem, surgical intervention for the release and reconstruction of the right TMJ was planned. This would improve his restricted mandibular movement and opening. The patient was admitted for right mandibular condylectomy and removal of ankylosis via an open preauricular and submandibular approaches. Reconstruction was via a costochondral graft (CCG) placement.

*Discussion.* This case demonstrates a previous temporomandibular joint septic arthritis and subsequent degenerative changes as a result of chronic mastoid disease. Reconstruction of the ankylosed TMJ with a CCG yields a functional condyle with growth potential and a high degree of patient satisfaction.

c5

**Kissing molars and a post permanent dentition****N. GUPTA\*, M. HARRISON & P. AGRAWAL***Guy's, Kings and St Thomas' Dental Institute, London*

**Presenting problem.** Whilst managing the carious dentition of a young boy, there was an incidental finding of 5 supernumerary teeth that developed between the ages of 8 and 9. There was subsequent development of 2 more supernumeraries over the next 5 years. Management of his dentition was further complicated by impaction and direct apposition of molar occlusal surfaces – 'kissing molars' – a rarely reported condition.

**Clinical management.** The development and progression of these anomalies are highlighted in the panoramic views taken periodically through his treatment. Caries management in a crowded dentition plus the development of these dental anomalies over a 5-year period required a multidisciplinary approach. Joint paediatric and orthodontic comprehensive treatment plans were created in an attempt to provide long-term management of this patient.

**Discussion.** The presentation of a non-syndromic post permanent dentition and the additional finding of kissing molars added to this cases complexity and uniqueness. A post permanent dentition is the result of inappropriate dental lamina activity, whereas the molar abnormality is the result of malfunction of the dental follicle. This case discusses whether the conditions are linked and describes their management.

c6

**A different look: three-dimensional imaging of a child with Binder syndrome****E. J. HINGSTON\*, M. L. HUNTER & C. H. KAU***Cardiff and Vale NHS Trust and Cardiff University (Wales College of Medicine)*

Binder syndrome (maxillo-nasal dysplasia) is an uncommon developmental anomaly characterised by an unusually flat, under-developed midface, with an abnormally short nose and flat nasal bridge. A twelve-year-old boy with Binder syndrome is presented. His facial and dental features are described and an overview of his past and future management provided. In addition to standard radiographic examination, a novel technique involving three-dimensional laser scanning has been used to evaluate the patient's facial morphology. The scan produced has subsequently been superimposed on the average face produced by amalgamating the scans of 41 skeletal class I, unaffected twelve-year-old males. This has enabled identification of the degree of deviation of the facial features of the patient from those of the average face. The results highlight the localised midface deformity characteristic of Binder syndrome and identify a deficiency of up to 15 mm in the nasal area.

c7

**Neurofibroma within the mandible of a ten year old child****A. K. HUMPHREYS\* & T. A. GREGG***Dental Department, Royal Belfast Hospital for Sick Children, Grosvenor Road, Belfast, BT12 6BP*

**Presenting problem.** A 10 year old child was referred by a community dental officer for dental extractions. Her medical history included neurofibromatosis type 1 (NF1) of the plexiform type affecting the right side of her face. Clinical examination revealed carious teeth and also delayed eruption of the lower right first permanent molar with thickening of the mucosa in this area. Radiographic examination confirmed displaced lower 46, 47 with associated radiolucency of unknown origin.

**Clinical management.** Painful carious primary teeth were extracted and the carious permanent teeth restored. Surgical extraction of the submerged lower right first permanent molar and incisional biopsy of surrounding tissue were undertaken.

**Discussion.** Oral manifestations are common in NF1. A variety of abnormal presentations involving gingivae, bone and displaced teeth has been reported and in a small number of cases malignant neoplasia is described. In this child, due to the displacement of teeth and associated radiolucency in the mandible, surgical investigation and biopsy were required for diagnosis. Pathology confirmed features of a neurofibroma consistent with the history of neurofibromatosis. Dentists should be aware of oral presentation, local complications and the known risk of malignant transformation in this condition.

c8

**Congenital insensitivity to pain: clinical case presentation****A. HUTTON\* & S. MCKAIG***Birmingham Children's Hospital, UK*

**Presenting problem.** A 6 month old female presented with large areas of oral ulceration and bite wounds to the tongue, cheek and fingers. Her brother was also known to the department due to similar problems and had been diagnosed as having congenital insensitivity to pain.

**Clinical management.** This involved identification of the cause, removal of deciduous teeth and encouragement to break the habit of biting her tongue and cheeks. As the adult dentition erupted further traumatic incidents lead to the use of oral mouth guards and advice on behaviour management.

**Discussion.** Congenital insensitivity to pain is a rare condition described within various categories. Differences occur between the mode of inheritance, clinical features and the predominantly affected peripheral neurones. These conditions are difficult to diagnose. They are generally noted from early childhood due to diminished or absent pain sensations. This case report describes the management of a female with congenital insensitivity to pain up to her present age of 6 years. The aim of treatment is to prevent episodes of oro-facial trauma and self-mutilation injuries. The primary teeth were removed on eruption and further management of the permanent dentition has involved the use of soft occlusal guards and behaviour modification techniques.

c9

**The use of multi-directional cross-sectional tomography for localising an odontome****J. JOHNSON\*, E. J. WHAITES & E. C. SHEEHY***Guy's, Kings & St. Thomas' Dental Institute, London*

**Presenting problem.** A healthy male, aged 7 years presented, for the management of his retained upper right primary central and lateral incisors and investigation of a large calcified radio-opaque mass overlying the roots of these primary teeth.

**Clinical management.** Examination revealed the upper right primary central and lateral incisors had a firm hard swelling overlying the buccal aspect of these teeth. Panoramic and occlusal radiographs revealed the presence of an irregular calcified radio-opaque mass apical to the roots of the retained right primary maxillary incisors. They failed to accurately diagnose the extent and location of the lesion in the sagittal plane and its exact relationship to the unerupted right permanent central incisor. Three 2-mm-thick segmental cross-sectional tomographs of the anterior maxilla in the sagittal plane were taken. This revealed a large, but separate, calcified mass, overlying the buccal and palatal aspects of the unerupted upper right permanent central incisor. The patient was scheduled for surgical excision of the lesion.

**Discussion.** This case report illustrates the benefits of supplementing plain film radiographs with multi-directional cross-sectional tomography to localise a large maxillary odontome and accurately establish its relationship with an unerupted upper permanent central incisor.

c10

### Management of double maxillary central incisors: case reports

T. KANDIAH\*, S. J. MCKAIG & C. J. BROWN

Birmingham Dental Hospital, UK

**Presenting problem.** This presentation illustrates the multidisciplinary approach to the management of double maxillary central incisor teeth. Four cases ranging in age from 8–11 years, expressed concern regarding the appearance of their large anterior teeth. In addition one reported bullying. One patient gave a history of partial deafness and asthma, the remaining histories were unremarkable. Clinical examination revealed anterior crowding and double incisor teeth, which were bilateral in 3 cases.

**Clinical management.** Treatment planning involved a multidisciplinary approach with restorative and orthodontic colleagues. Clinical management of the double teeth varied from, no active intervention, extraction with and without subsequent orthodontic alignment, to surgical division and orthodontic alignment.

**Discussion.** Double teeth can influence oral development affecting aesthetics, tooth alignment and plaque control. Treatment planning is complex and a multidisciplinary approach is essential.

c11

### Cysts in the mixed dentition: interesting cases and their management

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Radicular cysts arising from deciduous teeth are reported to be rare, less than 0.5% of radicular cysts reported being related to the primary dentition. Although this appears to be a rare condition we report on 5 cases presenting to the Department of Paediatric Dentistry at The Royal London Hospital in the last 2 years. This presentation investigates the differential diagnosis of cysts associated with primary molar teeth. We review some interesting case studies and their management, illustrating the good potential for healing following either enucleation or marsupialisation. Research regarding these lesions will also be discussed.

c12

### Maxillary mucormycosis in a 14-year-old boy undergoing oncologic therapy

E. MCGOVERN<sup>1</sup>\*, P. FLEMING<sup>1</sup>, E. COTTER<sup>2</sup>, J. RUSSELL<sup>1</sup>, A. O'MARCAIGH<sup>1</sup>

<sup>1</sup>Our Lady's Hospital for Sick Children, <sup>2</sup>Practice limited to Prosthodontics, Dublin

**Presenting problem.** A fourteen-year-old boy undergoing chemotherapy for management of acute lymphoblastic leukaemia was referred for dental management of a suspected mucormycosis infection prior to haemopoietic stem cell transplant (HSCT). Intra-oral examination revealed denuded alveolar bone labial and palatal to the central maxillary incisors. Radiological examination revealed extensive bony destruction of the anterior palate, nasal septum and maxillary anterior alveolar ridge. Treatment of the infection including eradication of infected and/or necrotic tissues was required.

**Clinical management.** The necrotic area of the anterior maxilla and incisive alveolar region was surgically excised. This also included removal of maxillary incisor and canine teeth. An obturator was fitted to replace the surgically excised teeth and bone. High dose antifungal drugs were also required to treat the now confirmed mucormycosis. Recurrent infection warranted further surgery and dental extractions twenty-seven days post transplant.

**Discussion.** Mucormycosis is a rare invasive fungal infection that occurs in an immunocompromised host. This case describes the man-

agement of such infection prior to HSCT and its recurrence in the immediate post transplantation period. It describes the immediate dental management, dental reconstruction and the role of paediatric dentistry in the multidisciplinary care of child with a life threatening illness.

c13

### Hypomelanosis of Ito

M. MOFFAT\*, L. BARRETO & S. MCKAIG

Birmingham Dental Hospital and School

**Presenting problem.** A 9 year old boy was referred to the paediatric dental department for a consultant opinion. Clinical examination showed a pattern of light patches on his skin, midface hypoplasia, enamel hypoplasia, microdontia affecting maxillary teeth with a marked contrast in tooth size in the opposing arches and a marked class III malocclusion. Radiographic examination demonstrated multiple missing permanent teeth and microdontia in the maxillary arch. Medical history revealed he had previously been diagnosed with a genetic condition.

**Clinical management.** Comprehensive management included; liaison with medical colleagues and dental colleagues; intensive preventive regime and acclimatisation; restoration of carious upper second deciduous molars; restoration of hypoplastic lower incisors.

**Discussion.** This case illustrates an unusual pattern of microdontia and hypodontia in a young patient diagnosed with Hypomelanosis of Ito. Comprehensive management included consultation and liaison with medical and dental specialties. Treatment included preventive advice and restorative treatment. This presentation stresses the importance of a multidisciplinary approach in the management of paediatric dental patient.

c14

### Management of a traumatised tooth – the holistic approach

S. NORTH & G. C. MOONEY\*

Department of Paediatric Dentistry, Charles Clifford Dental Hospital, Sheffield, UK

**Presenting problem.** An 11-year-old boy in the mixed dentition was referred by his General Dental Practitioner for management of traumatised 21. Previous dental treatment comprised fissure sealants on first permanent molars and composite bandage to 21. Examination revealed an enamel dentine fracture affecting 21 which was discoloured. All lower deciduous molars were infraoccluded, there was no evidence of 65 and 26 was tilted mesially. An OPT confirmed formation of all permanent teeth together with 65, which was completely submerged. The developing 25 was displaced mesially overlapping 24. **Clinical management.** 21 responded positively to sensibility testing and was restored with composite. 26 was distalised with an URA to create space prior to surgical removal of 65. At surgery 53 and 63 were removed to address incorrect path of eruption of 13 and balance respectively. A transpalatal arch with Nance button was subsequently fitted to maintain 26 position allowing adequate space for eruption of the upper left premolars. Vitality of the upper incisors is being monitored along with exfoliation of the infraoccluded molars.

**Discussion.** Diagnosis of the submerged deciduous molar was an incidental finding elicited from the history and clinical findings. Under normal circumstances treatment of infraoccluded deciduous molar teeth is not warranted when the permanent successor is present since approximately 97% will exfoliate naturally. The path of eruption of 13 has been addressed though future orthodontic treatment may be required to align 25 which is currently palatally placed.

c15

### **C.I.N.C.A. (chronic infantile, neurological, cutaneous, articular) syndrome: rare cause of periodontitis**

**E. M. O'DWYER\*, H. ZAITOUN & D. R. LLEWELYN**

*Alder Hey Children's Hospital*

**Presenting problem.** An 11 year old female with C.I.N.C.A. syndrome presented with pain from her mobile permanent lower canine. There was a history of previous multiple extractions of permanent teeth under general anaesthesia. Furthermore she was dental phobic and an irregular attender.

**Clinical management.** On examination, the patient appeared to be extremely short in stature. She had dysmorphic features, including, frontal bossing, angelic appearance and a saddle nose. Dental examination revealed severe generalised aggressive periodontitis with localised aggressive periodontitis in the lower anterior quadrants. Specific tests including DNA analysis for periodontopathogenic bacteria and a genetic predisposition test to identify interleukin 1 (II-1) polymorphisms associated with periodontal disease were carried out. The patient was II-1 genotype negative although she had an increased bacteria load of periodontopathogens. The teeth of poor prognosis were extracted. Following this prevention, acclimatisation, full mouth one-stage disinfection and new upper and lower partial prostheses were constructed.

**Discussion.** C.I.N.C.A. syndrome, is a rare, chronic, systemic inflammatory condition with no reported dental features. Currently trials are underway to assess the effectiveness tumour necrosis factor-alpha (TNF-alpha) and IL-1 blockers for treating patients with C.I.N.C.A. As much of the periodontal tissue destruction in periodontal disease is attributed to IL-1 and T.N.F-alpha, it would be interesting to research if the above treatment of C.I.N.C.A. improves their periodontal condition.

c16

### **Provision of a partial denture after mandibular resection**

**S. SHAH\*, L. K. MCCAUL & C. DEERY**

*Edinburgh Dental Institute*

**Presenting Problem.** A 12-year-old girl was referred by her General Dental Practitioner to the Paediatric Department of the Edinburgh Dental Institute with regards to an asymptomatic bony swelling to the right side of the mandible. Radiographic examination revealed a radiolucency causing displacement of the lower right canine and first premolar. The biopsy results confirmed that this was consistent with a chondrosarcoma and/or chondroblastic osteosarcoma.

**Clinical management.** The patient underwent several courses of chemotherapy and surgical resection of the right mandible. The area was surgically reconstructed with a fibula graft. After a period of healing, she was seen back in the Paediatric Department for provision of a temporary lower partial acrylic denture.

**Discussion.** The lower partial denture was made over several visits and proved to be a difficult clinical problem due to contraction and scarring of the soft tissues as well as the overeruption of her opposing upper right dentition. Her long-term treatment plan is to place implants in the reconstructed area.

c17

### **Multidisciplinary treatment of a boy with crown root fractured teeth**

**S. SOOD\*<sup>1</sup>, A. O'CONNELL<sup>1</sup>, B. O'CONNELL<sup>2</sup> & M. O'SULLIVAN<sup>2</sup>**

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**Presenting problem.** A 14 year old boy attended the Dental Hospital after traumatizing his anterior teeth. He had sustained complex crowns

fractures of teeth 21 & 22 with luxation injuries. Both were treated conservatively with calcium hydroxide and composite restorations. The teeth were splinted and monitored over the next week and continued to respond positively to vitality testing. He then sustained a second trauma to his teeth which resulted in a spiral root fracture of tooth 21 and a complex crown root fracture of tooth 22.

**Clinical management.** The initial management involved root canal therapy on tooth 22 and stabilisation of tooth 21. During this course of treatment, tooth 21 lost vitality and a buccal sinus developed. A multidisciplinary approach was taken and the following treatment plan was developed: Tooth 21 is to be extracted and Bioss is to be placed in the extracted socket with a resin retained bridge to replace this tooth with the future possibility of an implant. Following successful root canal therapy Tooth 22 will be restored with a post crown.

**Discussion.** This case highlights the complexity of treatment planning following trauma in a young patient. The double trauma sustained and the patient's age adds to the difficulty in determining the best long-term treatment to maximize the aesthetic and functional outcome for the patient. The importance of the multidisciplinary approach is reinforced.

c18

### **The management of hypodontia: a comprehensive training exercise**

**V. SRINIVASAN\*, R. S. HOBSON, B. O. I. COLE & A. J. SHAW**

*Newcastle Dental Hospital and School*

**Presenting problem.** A 13-year old girl with missing upper lateral and lower central incisors was concerned with irregularity and spacing of her teeth, diminutive lower front teeth and 'pointy' upper canines. She was in the permanent dentition and teeth 71 and 81 were retained. She presented with a Class 1 skeletal pattern and Class I incisor relationship. She was molar Class 1 on the right and 1/2-unit Class II on the left. Her overbite was incomplete, over-jet was less than 1 mm and there was generalised spacing.

**Clinical management.** *Short term:* Preoperative study models and Kesling set-ups were cast to consider treatment options. Diagnostic composite additions to the upper permanent canines to demonstrate adequate camouflage, was carried out. *Mid term:* Extraction of teeth 71 and 81, followed by upper and lower fixed orthodontic appliance treatment to close all spaces and allow for a 6 mm space in the lower anterior region for a single pontic. *Long term:* Camouflage of teeth 13, 23 as 12, 22 after debond. Retention and fabrication of a resin retained bridge to replace one lower central incisor.

**Discussion.** There is much to be gained by the patient and clinician when, all aspects of hypodontia treatment is carried out by the paediatric dentist. Such treatment can be very successful with regular interdisciplinary reviews and contributes to a comprehensive training exercise.

c19

### **The use of overdentures in children with cleft-lip and palate**

**Z. TEJANI\*, E. C. KOK, C. MASON & B. GRIFFITHS**

*Great Ormond Street Hospital*

**Presenting problem.** Case 1: A 9-year-old girl with bilateral cleft-lip and palate, presented with nasal food escape (due to a residual palatal fistula) and poor aesthetics resulting from retroclined upper incisors, hypodontia (15 missing permanent teeth) and a severe Class III malocclusion. Case 2: An 11-year-old boy with Rapp-Hodgkin Syndrome and bilateral cleft-lip and palate was being teased at school. He presented with missing upper incisors, abnormally erupting upper canines, hypoplastic amelogenesis imperfecta and a severe class III malocclusion.

**Clinical management.** Case 1: A maxillary overdenture with full palatal coverage was constructed in a Class II division 2 relationship.



This successfully improved aesthetics and prevented nasal regurgitation while avoiding surgery. Case 2: A transitional horse-shoe shaped denture was constructed, replacing the upper incisors and masking the poor appearance of the erupting upper canines. Both cases are on regular review for oral hygiene monitoring and overdenture adjustments.

*Discussion.* Overdentures in children with cleft-lip and palate are not a conventional form of treatment due to advances in cleft surgery. However, in cases where alveolar bone grafting has not resulted in the desired outcome, such as the cases presented, an alternative in the form of overdentures can be used to improve aesthetics and function in a developing child/adolescent until the patient has fully grown, when a more definitive treatment can be employed, if suitable.

c20

### **Heimler Syndrome in 11-year-old Monozygotic twin girls: clinical case presentation**

**S. VISRAM\* & S. McKAIG**

*Birmingham Dental Hospital, UK*

*Presenting problem.* Heimler Syndrome first documented by Heimler *et al.* in 1991. Features are: 1) Bi-lateral sensorineural deafness 2) Beau's lines and Leuconychia of nails 3) unaffected primary dentition but enamel abnormalities affecting the secondary dentition particularly the posterior teeth resembling Amelogenesis Imperfecta. Twin girls presented at 3 years of age with bi-lateral sensorineural hearing loss. Later presented at age 10 with hypoplasia of the secondary dentition affecting first permanent molars but not the incisor teeth. Radiographs suggest other permanent molars have also been affected. No history of abnormalities to primary dentition. Also noted were Beau's lines and Leuconychia of nails. Clinical findings remarkably similar between both girls. We report genetic, clinical, radiographic and dental manifestations of 11-year-old twin girls diagnosed with Heimler Syndrome and dental management of these girls.

*Clinical management.* 10-year-old girls referred to Paediatric department at Birmingham Dental Hospital suspecting Amelogenesis Imper-

fecta. Initial management included preventative advice, orthodontic assessment, removal of all four grossly hypoplastic first molars with local anaesthetic. Long-term management requires preventative advice, monitoring eruption of permanent teeth, restorative treatment of erupting permanent teeth and orthodontic treatment.

*Discussion.* Clinical findings suggest an unusual form of Amelogenesis Imperfecta affecting the secondary dentition but not the primary dentition, co-incident with other cases of Heimler Syndrome reported previously. The twins also show other features of Heimler Syndrome. The genetic, clinical, dental and radiographic findings seem to suggest that twins have Heimler Syndrome.

c21

### **An unusual case of cemento-ossifying fibroma**

**H. ZAITOUN\* & J. C. COOPER**

*Alder Hey Children's Hospital*

*Presenting problem.* A seven-year-old girl presented with a history, for eight months, of an asymptomatic bony swelling arising from the left maxillary primary molar region with considerable expansion. She reported that the swelling was causing difficulty when eating and tooth brushing.

*Clinical management.* Clinical and radiological examination including CT (computed tomography) scanning confirmed a large lesion, arising from the left maxilla, causing bony expansion and tooth displacement. A bone biopsy together with the appropriate haematological and biochemical studies were undertaken yielding an initial diagnosis of cemento-ossifying fibroma. Reduction of the bony swelling and cystic enucleation was undertaken via a standard intra-oral approach. Further histological examination revealed that there was an aneurysmal bone cyst (ABC) within the cemento-ossifying fibroma.

*Discussion.* This case demonstrates the rare presentation of an ABC that secondarily complicates cemento-ossifying fibroma for which review of clinical progress is essential as rapid expansion due to the presence of the cyst can occur.

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