Clinical Cases in Pediatric Dentistry

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Medically Compromised Patients

Paddy Fleming

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Case 1

Congenital Heart Disease

Figure 1.1.1 Facial photo

A. Presenting Patient

- 2-year-, 8-month-old Hispanic female
- New patient

B. Chief Complaint

- Referred from hospital pediatric cardiac unit for dental assessment and management of asymptomatic dental caries

C. Social History

- Single mother is primary caregiver and receives welfare
- No siblings

D. Medical History

Congenital Heart Defects

- Triscuspid atresia
- Hypoplastic right ventricle
- Restrictive ventricular septal defect (VSD)
- Sub-pulmonary narrowing

No Known Food or Drug Allergies

Current Medications

- Warfarin po 3 mg daily
- Furosemide po 20 mg daily

Several Past Surgeries and Hospitalizations

E. Medical Consult

For This Patient (see Fundamental Point 1):

- Baseline cardiac function
- Baseline INR range: 2.0 to 3.0
- Baseline hemoglobin: 11.7 g/dl
- Baseline pulmonary saturations: 97%
- Baseline blood pressure 80/40 mm Hg
- Baseline respiratory rate: 20/minute
- Baseline oxygen: 0.5 l/minute

F. Dental History

- No dental home
- Currently bottle feeding with sweetened liquids
- High caloric supplementation to increase weight
- Toothbrushing once a day with fluoridated toothpaste with little adult supervision
- No systemic fluoride exposure
- No history of dental trauma

G. Extra-oral Exam

- No significant findings

H. Intra-oral Exam

Soft Tissues

- Generalized erythematous mucosa
- Generalized edematous gingivae

Hard Tissues

- Proximal and smooth surface cavitations on maxillary incisors

BACKGROUND INFORMATION 1

Congenital Heart Disease (CHD)

- Incidence rate of approximately 8 to 10 cases/1,000 live births
- Most lesions occur individually; several form major components of syndromes or chromosomal disorders such as Down (trisomy 21) and Turner syndrome (XO chromosome)
- Known risk factors associated with CHD include maternal rubella; diabetes; alcoholism; irradiation; and drugs such as thalidomide, phenytoin sodium (Dilantin) and warfarin sodium (Coumadin)
- Turbulent blood flow is caused by structural abnormalities of the heart anatomy and presents clinically as an audible murmur
- CHD can be classified into acyanotic (shunt or stenotic) and cyanotic lesions depending on clinical presentation
- Acyanotic lesions are characterized by a connection between the systemic and pulmonary circulations or stenosis of either circulation (left to right shunts). The most common anomalies are:
  1. Atrial septal defect (ASD)
  2. Ventricular septal defect (VSD)
  3. Patent ductus arteriosus (PDA) caused by failure of closure of the ductus connecting the pulmonary artery with the aorta (normally closes soon after birth)
  4. Coarctation of the aorta
  5. Aortic stenosis
  6. Pulmonary stenosis
- All cyanotic conditions exhibit right to left shunting of desaturated blood. Infants with mild cyanosis may be pink at rest but become very blue during crying or physical exertion. Children with cyanotic defects are at significant risk for desaturation during general anesthesia
- The most common cyanotic lesions are:
  1. Tetralogy of Fallot which includes a VSD, pulmonary stenosis, overriding aorta and right ventricular hypertrophy
  2. Transposition of great vessels
  3. Tricuspid atresia
- If cardiac failure develops, the infant is digitalized and prescribed diuretics if necessary. Hospitalization, oxygen, nasogastric tube feeding, and antibiotic therapy for chest infection may also be required

FUNDAMENTAL POINT 1

Consult with Cardiologist and Hematologist to Obtain Baseline Information on:

Current Cardiac Status

- Blood pressure
- Respiratory rate
- Oxygen rate
- Pulmonary saturations
- Blood gases

Cardiac Medications

Current INR Range (Normal: Without Anticoagulant Therapy: –1; Target Range With Therapy: –2 to 3)

Previous Surgical Management

Future Surgical Management

Information is Then Used to:

- Assess risk of cardiac complications under general anesthesia (GA)
- Assess risk of infective endocarditis (IE) and need for antibiotic prophylaxis before invasive dental procedures
- Assess risk of hemorrhage during dental surgery
- Develop a perioperative anticoagulant management plan

- Fissure cavitations on molars
- Smooth surface demineralization on molars

Occlusal Evaluation of Primary Dentition

- Flush terminal plane
- Anterior open bite

Generalized Severe Plaque Accumulation

I. Diagnostic Tools

Bacteriology and Saliva Tests

- Not done

Radiographs

- Intra-oral periapical films (taken at time of dental treatment under general anesthesia)

Photographs

- Pre- and post-operative intra-oral photos (taken at time of dental surgery)

High Caries Risk Due to (Caries-risk Assessment Tool [CAT]) (see Fundamental Point 2):

- Special health needs child
- Visible cavitations
- Enamel demineralizations
- Low socioeconomic status
- Visible plaque score (4/6)
- Dietary chart (>3 sugar exposures/day)
- Medications that impair saliva flow
- Use of fluoridated toothpaste but no fluoridated water nor fluoride supplements
- Toothbrushing once a day

J. Differential Diagnosis

Developmental

- Enamel hypoplasia/hypomineralization

Infective

- Bacterial and/or fungal

Odontogenic

- Loss of tooth structure due to dental caries and/or attrition/abrasion/erosion

Inflammatory

Pulpal Pathology

K. Diagnosis and Problem List

Diagnosis

- Active early childhood caries
- Chronic hyperplastic gingivitis
- Periapical pathology
- Enamel hypoplasia

Problem List

- Untreated carious lesions
- Malocclusion
- Poor infant feeding practice

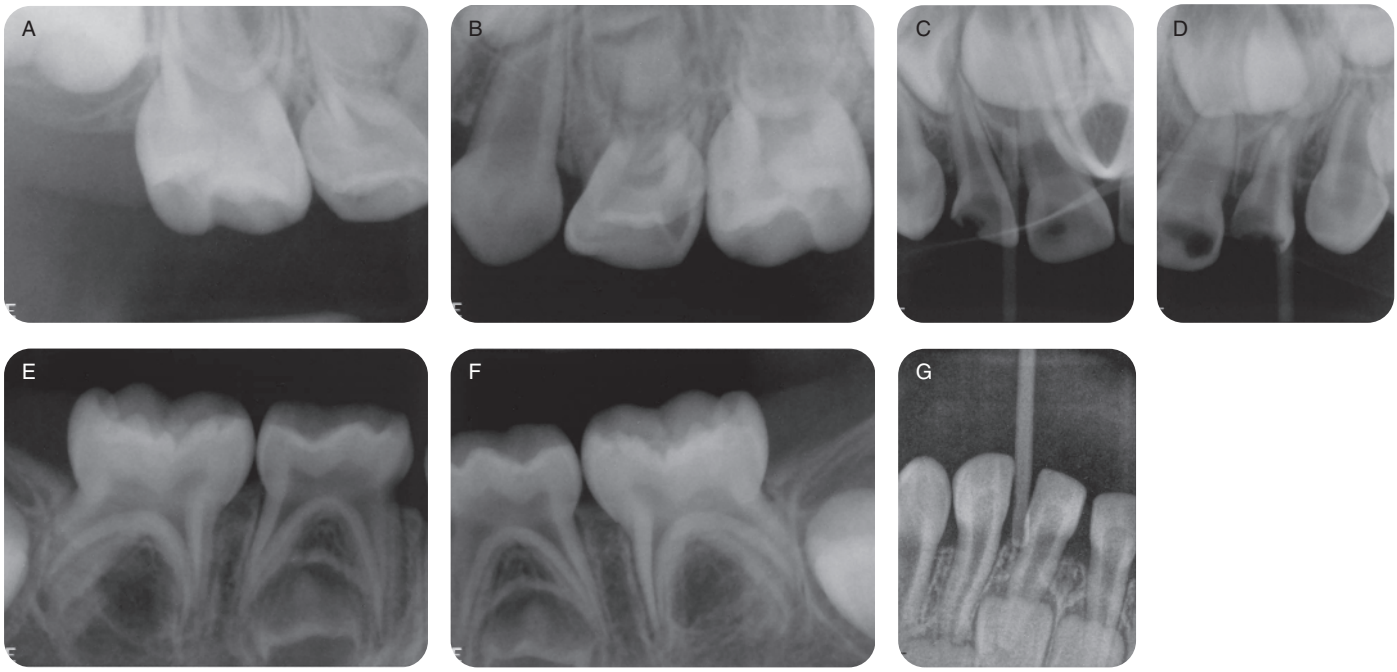

Figure 1.1.2a–g. Pre-op intra-oral radiographs

Figure 1.1.3a–e. Pre-op intra-oral photographs

Figure 1.1.4a–e. Post-op intra-oral photographs

FUNDAMENTAL POINT 2

The Caries-risk Assessment Tool (CAT)

Individuals Using This Tool Should Be Able to:

- Visualize child's teeth
- Access reliable historian for nonclinical elements
- Be familiar with footnotes that clarify elements
- Understand that each child's risk classification is determined by highest risk category where a risk indicator exists

Users Must Understand the Following Caveats:

- Caries risk assessed at one point in time only
- Clinical management remains a professional decision
- CAT does not render a diagnosis
- Advanced technologies (e.g., Diagnodent, microbiological tests) are not essential for using this tool

AAPD (2011–12)

- Parent's limited understanding of potential medical complications
- High risk of infective endocarditis (IE)—see Background Information 2
- High risk of uncontrolled hemorrhage from invasive dental procedure
- Behavior assessment: uncooperative

L. Comprehensive Treatment Plan

Establishment of a Dental Home and Caries Prevention Plan

- Cease bottle feeding
- Improve and increase frequency of oral hygiene
- Limit sugar intake between meals
- Brush after oral medicine intake
- Commence 1 mg fluoride supplement daily

Caries Control Using 0.2% Chlorhexidine Gel or 0.12% Liquid: Apply With Cotton Tip Nightly Two Weeks Prior to Procedure

Remove Any Potentially/Pulpally Involved Teeth to Reduce Future Risk of Chronic Bacteremia

Comprehensive Treatment of Carious Lesions Under General Anesthesia Due to the Complex Medical History, Dental Needs, and Behavior Management Issues

BACKGROUND INFORMATION 2

Infective Endocarditis (IE)

- Both coronary heart disease (CHD) and rheumatic heart disease can predispose the scarred internal lining of the heart to bacterial or fungal infection known as infective endocarditis (IE)
- Bacteremia during or after an invasive dental procedure can lead to the formation of friable vegetations of blood cells and organisms on the scar tissue
- *Streptococcus viridans* is most frequently responsible for chronic IE, while *Staphylococcus aureus* is often implicated in the acute fulminating form (Hallett 2003)
- IE may be prevented by antibiotic prophylaxis but there is little or no evidence to support this practice
- In 2008, the National Institute for Health and Clinical Excellence in the UK recommended against IE prophylaxis for all patients undergoing any type of dental procedures. Despite a 78.6% reduction in the number of prescriptions in the two years after the guideline was introduced, there was no large increase in the incidence of cases of or deaths from IE
- Pre-operative antiseptic mouthwash can reduce oral bacterial load
- Focusing on good oral hygiene practices may be more important than antibiotic prophylaxis

IE Prevention With IV Antibiotic Prophylaxis.

Perioperative Anticoagulant Plan

Follow-up Care

Post-op and Home Care Instructions

- Discharge after recommencement of Warfarin
- Soft cold diet for two days
- Recommence tooth brushing after 24 hours
- Start prevention plan

Recall Plan

- Six weeks post-op, and four- to six-month recall thereafter

M. Prognosis and Discussion

- Prognosis for limiting caries progression and changing the diet is guarded in view of limited understanding of caries risk factors and medical complications by parent. Prognosis could be improved with additional home support.

N. Common Complications and Alternative Treatment Plans

- Noncompliance with dietary advice
- Post-operative bleeding or infection
- Continued caries progression

Alternative Treatment Plans May Include:

- Use of stainless steel crowns in all teeth presenting with decalfication or white spot lesions
- Alternative pulpal management, e.g., extraction vs. pulp therapy (must consider risks of chronic bacteremia)
- Alternative medications for anticoagulant management (e.g., Heparin)
- Alternative methods for behavior management

BACKGROUND INFORMATION 3

Anticoagulant Therapy

- Anticoagulants are usually prescribed for children with valvular heart disease and prosthetic valves to reduce the risk of embolization
- If dental extractions are required, it may be necessary to decrease the clotting times to facilitate adequate coagulation but not to such an extent so as to cause emboli or clotting around the heart valves
- Commonly used anticoagulant drugs are oral warfarin sodium (Coumadin), which is a vitamin K antagonist depleting factors II, VII, IX and X, or heparin sodium (Heparin), which inhibits factors IX, X, and XII
- Local hemostatic measures include application of topical thrombin, packing of the socket with microfibrillar collagen hemostat, oxidized regenerated cellulose, and suturing of attached gingivae. Splints or stomo-adhesive bandages may also be of benefit. There have been recent reports of the efficacy of a "fibrin sealant" (Tisseel Duo 500) in the management of coagulopathies, but its use on moist oral mucosa is limited
- It is recommended to consult the cardiologist regarding modification of the anticoagulant therapy before oral surgery. Some practitioners cease warfarin three to five days prior to the surgery date, commence enoxaparin sodium once daily via Insulfon, and admit the patient to the hospital on the day of the procedure. In this protocol, recommencement of warfarin and weaning of enoxaparin sodium 24 hours after surgery is required to re-establish correct international normalized ratio (INR), prothrombin time (PT), and activated partial thromboplastin time (APTT)
- However, recent studies on adults who had multiple dental extractions without modification of their anticoagulant therapy showed few or no post-operative complications. The American Dental Association stated in 2003 that the scientific literature does not support routine discontinuation of oral anti-coagulation therapy for dental patients because it can place them at unnecessary medical risk. Their coagulation status, based on the INR, must be evaluated before invasive dental procedures are performed and any changes in their anticoagulation therapy must be discussed with the patient's physician.
- The American College of Chest Physicians recommends that patients who are taking vitamin K antagonists and are about to undergo minor dental procedures continue with the therapy because it does not confer an increase in clinically important major bleeding. The 2008 College guidelines further state that it is reasonable to co-administer an oral pre-hemostatic agent at the time of the procedure until more adequately powered studies are done. In patients receiving aspirin, the College recommends continuing it around the time of the procedure.
- To date, there have been no studies on pediatric patients, only case reports.

Self-study Questions

1. ***What important questions need to be asked when taking a medical history from a patient with congenital heart disease?***
2. ***What are some common medications to improve cardiac function and reduce congestive heart failure in children?***
3. ***Why are children with congenital heart disease more likely to develop dental caries in primary teeth?***
4. ***How would non-compliance with preventive advice alter treatment planning?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. Nature of diagnosis (acyanotic or cyanotic), supportive medications, previous surgical corrections, future surgical corrections, current cardiac function, physical activity limitations, risk of IE
2. Oral elixirs including Digoxin and Furosemide, usually with sucrose or sorbitol base.
3. Enamel is often hypoplastic and susceptible to early childhood caries; high-caloric diet; use of sucrose-rich medications; medications may induce xerostomia; parental indulgence with sweets, juices, sodas, etc.
4. It may be necessary to extract all carious teeth, especially those with pulpal involvement, to reduce the risk of infection and IE.

Case 2

Cystic Fibrosis

Figure 1.2.1a. Facial photograph

A. Presenting Patient

- 5-year-, 1-month-old female
- New patient

B. Chief Complaint

- Referral by general dentist for dental assessment and treatment of asymptomatic carious lesions

C. Social History

- Mother is primary care provider
- No siblings

D. Medical History

Cystic Fibrosis (CF) (see Background Information 1)

- Diagnosed at 3 years and 6 months of age
- Sees pediatrician every six weeks
- Has daily physiotherapy for removal of lung secretions
- Last acute chest infection: three weeks ago

FUNDAMENTAL POINT 1

Questions for the Caretaker of a Cystic Fibrosis Patient Regarding Their Medical History and Health Status

- When was the diagnosis made?
- Who are the lead pediatrician and other pediatricians who are involved in the patient's care and what are their addresses?
- What is the frequency of outpatient appointments?
- What is the frequency of admissions?
- When were the most recent admissions?
- What are the frequency and severity of chest infections?
- What is the microbiology of chest infections? (Infection by pseudomonas is an indication that lung function is severely compromised.)
- What are the current medications?
- Is an indwelling catheter (central line) present?
- What is the patient's experience with general anesthesia?
- Is there a possibility of further radical treatment? (Heart-lung transplant may be considered in patients with terminal respiratory failure.)
- Are there any special dietary requirements/ modifications?

No Known Drug or Food Allergies; Vaccinations Up-to-Date

Medications

- Flucloxacillin 250 mg qid: broad-spectrum antibiotic for prophylaxis against chest infections

- Pancreatin with each meal: pancreatic enzyme replacement
- Salbutamol inhaler bid: β-2 agonist
- Fluticasone Propionate inhaler bid: corticosteroid
- Vitamins A, D, and E supplements
- Ursodeoxycholic acid: dietary supplement to improve flow of bile

E. Medical Consult

If Parent is Not a Good Historian or if Child is Not Cooperative for Dental Care in Office, Contact Child's Pediatrician to:

- Review current medications and history of hospital admissions
- Establish respiratory status
- Discuss other means to treat this patient

F. Dental History

- Patient has dental home and has had restorations done in office
- Cariogenic diet
- Good oral hygiene habits with some parental supervision
- Uses toothpaste containing fluoride
- Has non-fluoridated water supply and no fluoride supplementation
- No history of trauma
- Behavior assessment: ± on the Frankl Palmer Scale

G. Extra-oral Exam

- No significant findings

H. Intra-oral Exam

Soft Tissues

- No significant findings

Hard Tissues

- No significant findings

Occlusal Evaluation of Primary Dentition

- Mesial step molars
- Right anterior crossbite
- Lower midline shift to the right (3 mm)
- Maxillary anterior crowding

Minimal Plaque Seen on Teeth

Dental Exam

- Severe enamel defects on maxillary left second primary molar and mandibular left second primary molar, which have been restored with amalgam. However, there is evidence of secondary caries at margins now

BACKGROUND INFORMATION 1

Cystic Fibrosis

- Autosomal recessive disorder
- Prevalence 1 in 2,500 live births in Caucasians
- Basic defect is a failure to code for cystic fibrosis transmembrane regulator (CFTR) protein which regulates electrolyte and water transport across cell membranes
- The optimal diagnostic test is the measurement of sweat electrolyte levels
- Complex, multisystem disease involving the upper and lower airways, pancreas, bowel, and reproductive tracts. The two main systems affected are respiratory and gastrointestinal.
- Respiratory issues: Viscous secretions accumulate in smaller airways which are prone to infection. Children are on long-term antibiotics to prevent chest infections, which must be treated aggressively. Healing occurs with scarring, further compromising the airway. Regular physiotherapy is required to encourage physical removal of secretions. Many children have reversible airway obstructions treated by salbutamol and steroid inhalers.
- Gastrointestinal issues: Damage to pancreas results in pancreatic enzyme insufficiency, thus oral pancreatic supplements are required with all meals. Fat-soluble vitamin supplements are taken due to the patients' reduced ability to absorb the vitamins. Incidence of celiac disease and Crohn's disease seems to be increased in CF patients.
- Long-term complications of CF include diabetes, liver disease, pneumothorax, sinusitis, nasal polyps, osteopenia, failure to thrive, and infertility
- The cornerstones of management include proactive treatment of airway infection and encouragement of good nutrition, as well as an active lifestyle
- For patients in late respiratory failure, heart-lung transplants are an option but are available in only a minority of cases

FUNDAMENTAL POINT 2

Oral Manifestations and Dental Treatment Considerations in Cystic Fibrosis Patients

- In spite of a high-carbohydrate diet the incidence of dental caries in cystic fibrosis (CF) patients has been found to be consistently lower than in healthy controls (Kinirons 1985, Narang, et al. 2003). This fact has been attributed to the long-term antibiotic therapy of these children (Kinirons 1992), high salivary pH (Kinirons 1985), and raised salivary calcium levels (Blomfield, et al. 1973)
- Patients with CF present increased levels of calculus and lower gingival health (Narang, et al. 2003). These patients have altered amounts of calcium and phosphate in their saliva, which affect calculus formation. However, pancreatin may have a role in decreasing calculus formation and reducing dental caries in these patients (Narang, et al. 2003)
- There is a higher prevalence of enamel defects, which can be attributed to severe systemic upset in infancy, especially in cases which patients are diagnosed late
- General anesthetic is strongly contra-indicated because of the compromised airway and increased incidence of post-operative chest infections
- Decisions on which antibiotic to give for acute dental conditions must be made in conjunction with the pediatrician. The choice of antibiotic should take into account the current regime and the types of antibiotics that may be required for future treatment
- Children with CF are high priority for dental prevention because the condition makes treatment of dental disease more difficult
- Liver disease may be a feature of older children with CF, resulting in complications involving bleeding, infection, and drug metabolism
- Nitrous oxide sedation may be contra-indicated in these children and should only be carried out after consultation with the pediatrician to establish respiratory capacity. The procedure should be carried out in a hospital environment. Careful monitoring of oxygen saturation levels is essential

- Mild enamel defects on mandibular right second primary molar and maxillary right second primary molar; the latter also has occlusal caries
- Evidence of moderate erosion on upper and lower incisors

I. Diagnostic Tools

- Right and left bitewings: no other radiographic lesions noted other than the ones mentioned above.

J. Differential Diagnosis

- N/A

Figure 1.2.1b. Pre-op frontal intra-oral

Figure 1.2.2. Pre-op maxillary intra-oral

Figure 1.2.3. Pre-op mandibular intra-oral

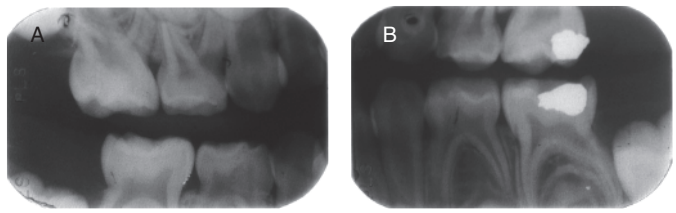

Figure 1.2.4a–b. Pre-op right and left bitewing radiographs

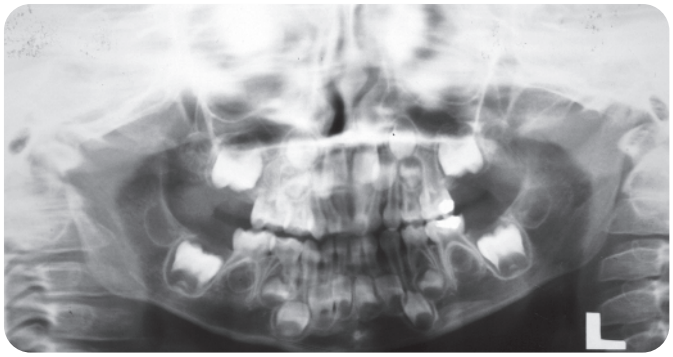

Figure 1.2.5. Pre-op panoramic radiograph

K. Diagnosis and Problem List

Diagnosis

- Early childhood caries with probable pulp involvement
- Occlusion problems
- Enamel hypoplasia
- Early tooth surface loss in incisors

Problem List

- Untreated carious lesions
- Poor dietary habits
- High caries risk (special health care needs, cariogenic diet, caries)
- Possible behavior management issues
- Correction of malocclusion

L. Comprehensive Treatment Plan

- Treatment of all carious lesions
- Establishment of a caries prevention plan

M. Prognosis and Discussion

- This patient was on a typical therapeutic regime. Involvement of the respiratory system meant that she was on ventolin and flixotide inhalers as well as flucloxacillin as prophylaxis against chest infections. Damage to the pancreas had resulted in the requirement of pancreatin, a combination of pancreatic enzymes. In addition, supplementation of fat-soluble vitamins was required as well as a diet high in refined carbohydrate and protein, and a full fat content.

In general, children with cystic fibrosis have low levels of dental caries. It is, therefore, surprising that three cavities were present. A number of factors may have contributed to this. All three teeth had enamel defects and two had been restored with amalgam which had developed secondary caries. Diet analysis revealed a high consumption of sweet drinks between meals, a finding further supported by the degree of erosion present.

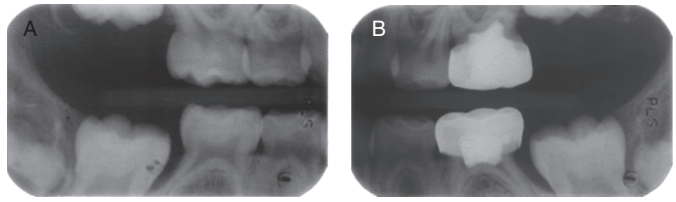

Figure 1.2.6a–b. Post-op right and left bitewing radiographs

Figure 1.2.7. Post-op maxillary intra-oral

Figure 1.2.8. Post-op mandibular intra-oral

The fact that there was some erosion present could be also considered unusual. No studies have specifically examined erosion in children with cystic fibrosis; however, salivary studies have demonstrated higher concentration of salivary bicarbonate and phosphates with consequent increased pH (Kinirons 1985). It would seem that, in this case, the high frequency of acidic drinks negated this protection.

The aims of prevention were to halt further development of oral lesions and to maintain the positive attitude that the patient displayed to dentistry. Ultimately, general anesthesia (GA) was strongly contra-indicated given her respiratory status and it was therefore extremely important not to reach a stage when this was necessary.

The patient was vulnerable to chest infections and her mother was naturally keen to protect her when these developed. This resulted in a number of cancelled appointments, especially during the winter and autumn.

The overall response to treatment was very positive. Compliance both with treatment and preventive

messages was excellent and the overall prognosis could be considered very good.

N. Common Complications and Alternative Treatment Plans

- Extensively decayed maxillary left second primary molar and mandibular left second primary molar
- Pulpectomy or extraction if teeth are non-vital with infection. If extracted, space maintenance must be addressed

Self-study Questions

1. ***Why do CF patients have a high rate of calculus?***
2. ***What two main systems are affected in this condition?***
3. ***Studies have shown that caries rates are low in this condition. What reasons have been given for this?***
4. ***What precautions are required for treatment under general anesthesia in CF patients?***
5. ***What are the long-term complications of CF?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. They have altered amounts of calcium and phosphate in their saliva, which affect calculus formation. However, pancreatin may have a role in decreasing calculus formation and reducing dental caries in these patients
2. Respiratory and gastro-intestinal
3. This fact has been attributed to the long-term antibiotic therapy of these children, high salivary pH, and raised salivary calcium levels
4. The precautions for treatment under general anesthesia are:
   - Avoid general anesthesia if possible
   - Make sure that there are no signs of pulmonary infection. May require sputum culture
   - Chest radiograph
   - Blood gases
   - Pulmonary function testing
   - Vigorous course of pre-op and post-op chest physiotherapy to clear as much of the secretions as possible
   - Check for diabetes (blood glucose) and liver disease (LFTs) pre-op
   - Check current antibiotic regimen
   - Peri-operative frequent suctioning and removal of secretions
   - Nasal polyps are a contra-indication to nasal intubation
5. Long-term complications of CF include diabetes, liver disease, pneumothorax, sinusitis, nasal polyps, osteopenia, failure to thrive, and infertility

Case 3

Hemophilia A

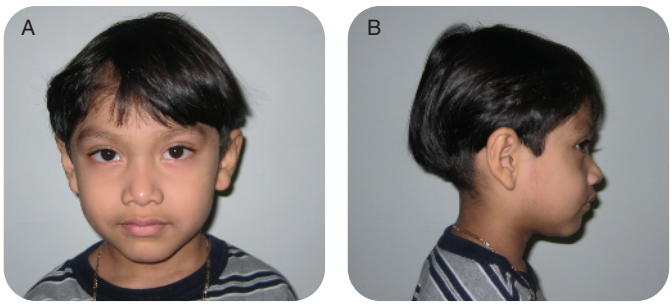

Figure 1.3.1a–b. Facial photographs

A. Presenting Patient

- 4-year-, 2-month-old Hispanic male
- New patient referred by his physician

B. Chief Complaint

- Child states, "My tooth hurts when I eat candy"
- Mother states, "Has only been a problem for one week"

C. Social History

- Patient attends preschool
- Middle class family, married parents, mother is primary caregiver, patient is the only child

D. Medical History

- Severe Hemophilia A
- Two emergency room visits in the past year for injury-associated bleeding (knee and elbow)
- Medications: IV Advate® (recombinant factor VIII) three times a week administered at home by mother
- The patient has no food or drug allergies and his vaccinations are up-to-date

E. Medical Consult

- Hemophilia Team

F. Dental History

- No dental home

FUNDAMENTAL POINT 1

Implications of Bleeding Disorder for Dental Treatment

Question Caretaker Regarding:

- Type and severity of hemophilia
- Hemophilia team contact information
- Compliance with medications
- Frequency and management of bleeding episodes
- Inhibitor status
- History of blood borne diseases such as HIV infection or hepatitis due to blood transfusions
- Limitations or restrictions on activities

- Fair oral hygiene; no adult supervision
- Three meals and two snacks daily. Special snack of candy bar or soda is given as reward after each IV infusion
- Brushes with fluoridated toothpaste once a day
- Lives in optimally fluoridated area
- No history of trauma

G. Extra-oral Examination

- No significant findings

H. Intra-oral Examination

Soft Tissues

- Marginal gingivitis around molars

Hard Tissues

- No significant findings

Occlusal Evaluation of Primary Dentition

- No significant findings

BACKGROUND INFORMATION 1
Hemophilia Management

- Hemophilia occurs in 1 out of every 5,000 males. Hemophilia A (factor VIII deficiency), the most common type (85% of the cases), and hemophilia B (factor IX deficiency) are X-linked recessive traits. Hemophilia C (factor XI deficiency) is an autosomal recessive trait most frequently presenting in Ashkenazi Jews
- Hemophilia presents as impaired secondary hemostasis (stabilization of the platelet plug with fibrin) while primary hemostasis (platelet plug formation) is normal. The disease is classified by level of factor activity (normal: 55% to 100%): severe(<1%), moderate (1% to 5%), and mild (>5%). The activated partial thromboplastin time (aPTT) is usually two to three times the upper limit of normal
- Patients with severe factor deficiencies may bleed spontaneously in muscles, skin, and joints, which may eventually develop arthropy, particularly in the ankles and knees. The incidence of joint pathology is decreased with prophylactic factor administration which is done multiple times a week in cases of moderate and severe hemophilia to increase factor levels to approximately 5%
- For breakthrough bleeds, additional factor replacement is needed together with administration of antifibrinolytic medications such as aminocaproic acid (AMICAR) or tranexamic acid (Cyklokapron). Desmopressin (DDAVP) increases plasma levels of factor VIII and may be useful in mild hemophilia A

Moderate Plaque Accumulation

Enamel Chipping on Incisal Edges of Mandibular Incisors

Intact Dentition on Visual Exam

I. Diagnostic Tools

- Two bitewing radiographs
- Maxillary and mandibular incisor occlusal radiographs
- Interproximal caries noted between molars

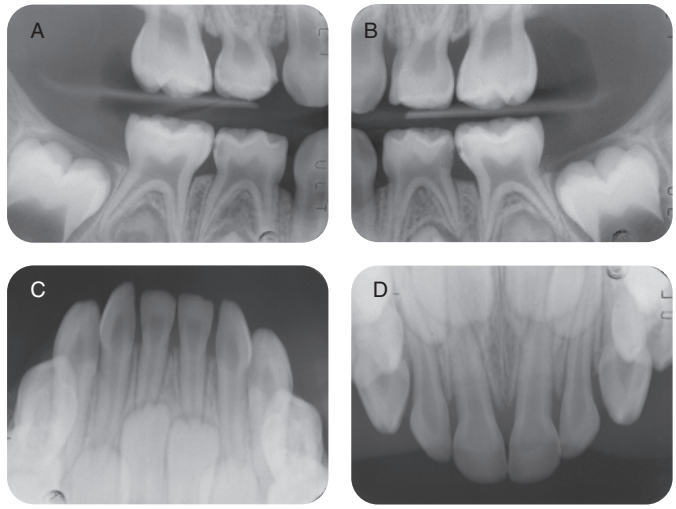

Figure 1.3.2a–d. Pre-op intra-oral radiographs. A. Right bitewing radiograph, B. Left bitewing radiograph, C. mandibular occlusal radiograph, D. Maxillary occlusal radiograph

J. Differential Diagnosis

- N/A

K. Diagnosis and Problem List

Diagnosis

- Severe hemophilia A
- Early childhood caries

Problem List

- High risk of bleeding with invasive dental procedures and nerve blocks
- Untreated caries on the interproximal surfaces of all primary molars and incipient lesions in the maxillary left molars
- High caries risk due to special health care needs, lack of dental home, presence of caries, cariogenic diet, oral hygiene without adult supervision, and visible plaque and radiographic findings

L. Comprehensive Treatment Plan

- Establishment of a dental home and a caries prevention plan
- Prevention of head and oral trauma (helmets and mouth guard)
- Contact hemophilia team for factor management before and after dental care. Plan oral intubation for general anesthesia (GA) to minimize risk of airway trauma, and an aggressive treatment such as restoring all primary molars with stainless steel crowns

FUNDAMENTAL POINT 2

Dental Care for Patients with Hemophilia

- Most patients can receive outpatient dental treatment. Appointments should be planned to minimize the need for factor infusions. Treatment under GA in a hospital should be considered for patients with extensive treatment needs and those with moderate or severe hemophilia. Always consult the hemophilia team before any dental work
- Prevention of dental problems is essential
- Avoid iatrogenic trauma to the oral mucosa by careful use of saliva ejectors, rubber dam, Vaseline lubrication of soft tissues, proper impression technique, use of interproximal wedges, and gentle placement of radiograph film, particularly in the sublingual area
- For local anesthesia, careful technique and aspiration are essential
- In general, buccal, intrapapillary, and periodontal ligament infiltrations can be administered without factor replacement
- Due to the risk of dissecting hematoma formation and potential airway compromise, factor replacement should be raised to 30% to 40% before infiltration into a highly vascular area or loose connective tissue as well as before posterior superior alveolar or inferior alveolar nerve blocks
- Pulp treatment usually presents a low risk of bleeding and is generally preferable to extractions
- Factor replacement is needed for subgingival scaling. Periodontal surgery can be a great challenge to hemostasis
- Fixed or removable orthodontic appliances can be used. Minimize gingival trauma when fitting bands and cementing appliances

For Surgical Procedures

- Carry out treatment as atraumatically as possible
- Observe patients for a prolonged period of time post-surgery; consider hospitalization in moderate/severe cases
- Use local hemostatic agents: pressure, absorbable gelatin product, cellulose materials, thrombin, microfibrillar collagen, fibrin glue, cyanoacrylate, acrylic stents, bone wax, electrocautery, resorbable sutures, periodontal dressings, and epinephrine
- Bleeding can be aggravated by aspirin and other non-steroidal anti-inflammatory drugs. Codeine and acetaminophen are safe alternatives.
- Any swelling, rapid breathing, dysphagia, or hoarseness may be a sign of airway compromise caused by intubation and/or dental treatment. Patient should be sent to the hospital immediately for advanced airway management and the hematologist should be consulted.

- Schedule a follow-up dental appointment in two weeks to check for complications and to reinforce prevention plan
- Three month recall

M. Prognosis and Discussion

- Risk for future caries is high. Prognosis could be improved by establishing a dental home, improving dietary and hygiene habits, and frequent recall visits

N. Complications and Alternative Treatment Plan

- Hemophilia cases with inhibitors present a management challenge because bleeding episodes continue despite appropriate factor replacement. Inhibitors are factor-specific antibodies which develop in 25% to 35% of patients with hemophilia A and 3% to 5% of patients with hemophilia B. In such cases, dental care should be consolidated into one treatment session and may include use of a bypassing agent such as factor VIIa or activated prothrombin complex concentrate.
- The patient should be admitted post-operatively because complications may arise such as a compromised airway and bleeding from trauma to the maxillary midline frenum which may be managed by local and/or systemic measures.

Self-study Questions

1. ***Which laboratory screening test is affected by a reduced level of factor VIII or factor IX?***
2. ***In addition to pre- and post-operative medications, what are special considerations for treatment under general anesthesia for patients with hemophilia?***
3. ***Which analgesic medications are contra-indicated for a patient with a bleeding disorder and why?***
4. ***What is the significance of inhibitors in hemophilia?***
5. ***What local anesthesia techniques should be done only after factor replacement?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. The activated partial thromboplastin time (aPTT) is usually two to three times the upper limit of normal
2. Nasotracheal intubation is contraindicated due to increased risk of airway trauma. Treatment venue is more appropriately in a hospital rather than in-office
3. Aspirin and nonsteroidal anti-inflammatory medications adversely impact hemostasis due to platelet inhibition
4. Bleeding episodes continue despite appropriate factor replacement levels. Care for these patients may include use of a bypassing agent such as factor VIIa or activated prothrombin complex concentrate
5. Infiltrations into a highly vascularized area or into loose connective tissue, and posterior superior alveolar and inferior alveolar nerve blocks.

Case 4

T Cell Lymphoblastic Leukemia/Chemotherapy

Figure 1.4.1. Facial photograph

A. Presenting Patient

- 3-year-, 4-month-old Caucasian male

B. Chief Complaint

- Referred by hospital oncology unit for dental assessment and management of oro-facial cellulitis

C. Social History

- Two-parent, middle class family unit, one sibling, mother is primary caregiver

D. Medical History

- Cell lymphoblastic leukemia
- Influenza A positive
- Norovirus positive
- No known food or drug allergies
- Vaccinations up to date

Current Medications

- Chemotherapy and supportive therapy (on Children's Oncology Group study): Five-week induction, five-week consolidation, eight-week

BACKGROUND INFORMATION 1

Acute Lymphoblastic Leukemia (ALL)

- Accounts for 80% to 85% of acute childhood leukemias with a peak incidence at four years of age
- Defined by the presence of more than 25% lymphoblasts in the bone marrow
- The most common signs and symptoms are anorexia, irritability, lethargy, anemia, bleeding (including oral), petechiae, fever, lymphadenopathy, splenomegaly, hepatomegaly, and bone pain
- The most common head, neck, and intraoral manifestations of leukemias are sore throat, lymphadenopathy, laryngeal pain, gingival bleeding, and oral ulceration
- Therapy is tailored to the risk of relapse dependent on cytogenetic markers, and includes a combination of induction chemotherapy, central nervous system prophylaxis, and maintenance chemotherapy for 2.5 to 3.5 years
- Intrathecal therapy (commonly methotrexate) has been used to replace cranial irradiation
- Cure rates for standard risk ALL are now over 90% on current protocols. If relapse occurs, 40% to 50% can be cured with chemotherapy and/or hematopoietic stem cell transplantation, which is reserved for very-high-risk or relapse patients
- Prognosis depends on age of onset, initial white cell count, cytogenetic abnormalities, and other features

interim maintenance, seven-week delayed intensification, reinduction if required, 12-week maintenance

- Multiple medications including Vincristine, Methotrexate, Prednisone

E. Medical Consult

Oncology Team

- Gather information about the underlying disease, time of diagnosis, modalities of treatment the patient has received since the diagnosis, planned treatment, surgeries, complications, prognosis, current hematological status, allergies, and medications
- Baseline complete blood count (CBC)
  - Platelets 81,000/mm3 (normal: 150,00 to 400,000)
  - White blood cells 4,100/mm3 (5,000 to 15,000)
  - Absolute neutrophil count (ANC) 2,600/mm3 (1,500 to 8,000)
  - Red blood cells 4.16×10^{12}/L (4 to 5.2)
  - Hematocrit 34% (34% to 40%)
  - Hemoglobin: 11.2 g/dL (11.5 to 13.5)

F. Dental History

- No dental home
- Good oral hygiene with supervision
- Cariogenic diet with high caloric supplementation to increase body weight
- Brushes once daily with fluoridated toothpaste
- Lives in an area without fluoridation
- No previous trauma history
- Behavior assessment: uncooperative for routine dental treatment

G. Extra-oral Exam

- No significant findings

H. Intra-oral Exam

Soft Tissues

- Localized marginal gingivitis with bleeding

Hard Tissues

- No significant findings

Occlusal Evaluation of the Primary Dentition

- Flush terminal plane
- Canines class I
- Anterior tooth contact on closure

Other

Multiple carious lesions on visual exam: Maxillary left first molar, maxillary right first molar, maxillary anterior teeth

Figure 1.4.2a–e. Pre-op intra-oral photos

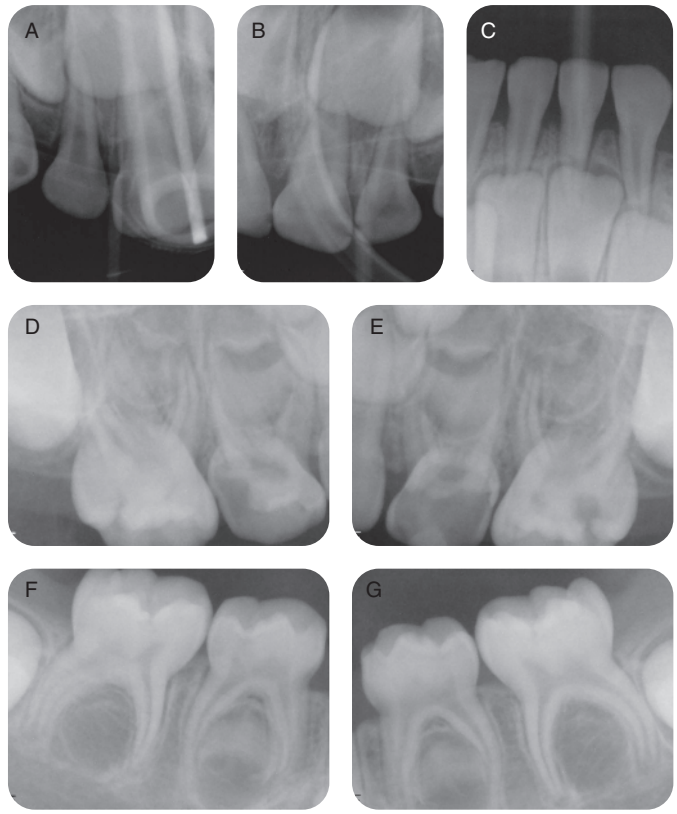

Figure 1.4.3a–g. Pre-op Intra-oral radiographs. A. Right maxillary occlusal radiograph, B. left maxillary occlusal radiograph, C. mandibular occlusal radiograph, D. right maxillary periapical radiograph, E. left maxillary periapical radiograph, F. right mandibular periapical radiograph, G. left mandibular periapical radiograph

- Fissure discolorations on molars
- Smooth surface demineralization on maxillary molars

I. Diagnostic Tools

- Intra-oral periapical films taken at time of dental care under general anesthesia

J. Differential Diagnosis

Developmental

- Enamel hypoplasia
- Enamel hypomineralization

Infective

- Bacterial
- Fungal

Odontogenic

- Loss of tooth structure
- Dental caries
- Attrition/abrasion/erosion

Inflammatory

- Pulpal pathology
- Oro-facial cellulitis

K. Diagnosis and Problem List

Diagnosis

- Active early childhood caries
- Acute apical periodontitis
- Facial cellulitis due to odontogenic infection

Problem List

- High caries risk due to:
  - Special health needs
  - Visible cavitations
  - Enamel demineralizations
  - Visible plaque
  - Cariogenic diet (>3 sugar exposures/day)
  - Usage of medications that impair salivary flow
  - Lack of fluoridated water and fluoride supplements
- Limited understanding of potential medical complications by parent
- High risk of oral sepsis and subsequent medical complications

L. Comprehensive Treatment Plan

- Explain importance of maintaining optimal oral health for children with acute lymphoblastic leukemia (ALL) being treated by chemotherapy (see Fundamental Point 1).
- Explain oral health care protocol (mucositis care), the risk of oral sepsis complicating medical management, and long-term oro-dental effects from therapy
- Caries control with antimicrobial therapy prior to surgical management
  - Prescribe 0.2% chlorhexidine gel application four times daily prior to procedure but avoid using alcohol-containing products when mucositis is present (it will sting and burn the mucosal tissues)
- Remove any potentially/pulpally involved teeth
- Remove any sources of tissue irritation
- Establish a caries prevention plan as well as a dental home
- Restore multi-surface caries lesions with stainless steel crowns due to their longevity and ability to prevent further caries progression
- Apply sealant on discolored molar fissures
- Apply fluoride varnish on demineralized areas
- Administer IV antibiotic prophylaxis at time of dental treatment to reduce risk of bacteremia if the patient is immunosuppressed and treatment cannot be delayed
- Address behavior management considerations
  - Provide comprehensive dental treatment under GA to facilitate all necessary procedures at the one time and to avoid delaying the cancer therapy. This can be done concurrently with other medical procedures such as lumbar puncture and bone marrow aspiration, if required

Post-op and Home Care Instructions

- Discharge after full recovery
- Soft, cold diet for 48 hours
- Recommence toothbrushing in 24 hours

Prevention Plan

- Recall plan
  - Six weeks post-op
  - Four-month review during chemotherapy
- Apply fluoride varnish to demineralized areas at both visits

M. Prognosis and Discussion

- Prognosis for limiting caries progression is fair in view of understanding of caries risk factors and potential medical complications by parent. However, the burden of a cancer diagnosis and all

FUNDAMENTAL POINT 1

Oral Care During Cancer Treatment

- Patient and caretaker education is a must
- Aggressive oral hygiene with a soft toothbrush or an electric brush should be done throughout treatment, regardless of the child's hematological status
- Ultrasonic brushes and flossing can be used if the patient is properly trained
- Avoid using toothpicks and water irrigation devices during periods of immunosuppression
- Patients who do intensive oral care have a reduced risk of developing moderate/severe mucositis without increasing the risk of septicemia in the oral cavity. For oral mucositis care, review the guidelines at www.mascc.org (under "Study Group and Resource Center")
- Nystatin is not effective for prevention and treatment of fungal infections in immunocompromised hosts
- Patients should brush or at least rinse the mouth with water after taking oral medications or nutritional supplements
- Children at high risk for xerostomia or caries should be prescribed supplemental fluoride

of its complex treatment may push oral/dental care to a low priority status. Indulgence of the sick child with sweet candies and liquids can also be a problem.

- Prognosis for changing current dietary habits is uncertain in view of the difficulty in maintaining adequate nutrition during chemotherapy.

N. Common Complications and Alternative Treatment Plans

- Non-compliance with dietary advice
- Post-operative bleeding or infection

Figure 1.4.4a–e. Post-op intra-oral photos

- Continued caries progression
- Acute and long-term oral complications related to chemotherapy (see Background Information 2)

Alternative Treatment Plans May Include:

- Alternative restorative materials, e.g., composite resin, amalgam, glass ionomers
- Alternative pulpal management, e.g., extraction vs. pulp therapy in posterior teeth (must consider risks of chronic bacteremia)
- Alternative methods for behavior management
- Nitrous oxide vs. outpatient sedation vs. GA vs. nothing

FUNDAMENTAL POINT 2

Dental Treatment During Cancer Treatment

- Ideally, all dental care should be completed before cancer therapy starts. When that is not possible, prioritize procedures and place temporary restorations until the patient is stable
- Infections, extractions, scaling, and sources of tissue irritation should be taken care of first, followed by carious teeth, root canal therapy, and replacement of faulty restorations. The risk of pulpal infection and pain determines which carious lesions should be treated first
- A complete blood count (CBC), including the absolute neutrophil count (ANC), is necessary before dental procedures
- Elective dental treatment should be delayed when the child is immunocompromised, i.e., the ANC is <1,000, although some oncology teams allow for invasive dental care to be done at ANC >500. In emergency cases, discuss antibiotic prophylaxis with the patient's physician before proceeding
- Because many acute lymphoblastic leukemia (ALL) patients have also been receiving systemic corticosteroids, the possibility of adrenocortical suppression should be considered and additional steroid cover provided as appropriate
- Primary teeth in need of pulpal therapy during the induction and consolidation phase of chemotherapy should be considered for extraction due to possibility of failure of the pulp treatment, which will lead to an odontogenic infection
- When pulpal therapy of permanent teeth is needed, the risk of bacteremia and potential septicemia must be weighed against the potential benefits of tooth conservation
- Braces should be removed if the patient has poor oral hygiene or is at risk for the development of moderate/severe mucositis. Smooth, well-fitting appliances (e.g., band and loops) can be kept if the patient has good oral hygiene
- Root tips, teeth with periodontal pockets >6 mm, teeth with acute infection, significant bone loss, involvement of furcation, symptomatic impacted teeth, and non-restorable teeth should be removed at least seven to 10 days prior to initiation of therapy.
- A platelet count >75,000/mm^3 does not require additional support, but be prepared to treat prolonged bleeding with local measures after oral surgical procedures
- If the platelet count is <75,000/mm^3 and invasive procedures need to be carried out, consult with the physician before providing care
- Coagulation tests may be in order for individual patients, especially those with liver disease or coagulation problems
- Consult an experienced oral surgeon before oral surgical procedures in patients who have had or are currently on head and neck irradiation and/or bisphosphonates, given the risk of jaw necrosis
- Children in full remission can be treated routinely, although a CBC is prudent if an invasive procedure is planned
- There is no evidence to support the administration of antibiotic prophylaxis to prevent catheter-related infections associated with an invasive oral procedure in patients with chronic indwelling central venous catheters

BACKGROUND INFORMATION 2

Acute and Long-Term Effects of Chemotherapy on the Craniofacial Complex

- The cytotoxic drugs used during chemotherapy can cause damage to several body organs, including the craniofacial complex
- Neutropenia is defined as <1,500 neutrophils/mm^3 and can predispose the child to oral sepsis by commensal organisms
- Direct stomato-toxicity is caused by the cytotoxic action of the chemotherapeutic agents on oral mucosal cells leading to inflammation, thinning, and ulceration of the mucosa (mucositis)
- Recent case reports suggest that the incidence and severity of mucositis, the most common and painful side effect of chemotherapy, may be reduced with the concomitant administration of granulocyte colony stimulating factor (G-CSF) during chemotherapy
- Salivary function may also be diminished, although this response has not been reported as common in children
- Other acute oral side effects of chemotherapy include secondary infections, bleeding, and neurotoxicity
- These problems are commonly encountered in the induction and consolidation phases of chemotherapy when relative high doses of multi-agent therapy are employed
- Children younger than 10 years of age who receive chemotherapy and/or radiotherapy (total body irradiation or localized radiotherapy to the head and neck) may present dental developmental defects such as tooth agenesis; short, tapered roots; early apical closure; crown disturbances in size and shape; microdontia; enlarged pulp chambers; and dentin and enamel opacities and defects

Self-study Questions

1. ***What important questions need to be asked when taking a medical history from a patient with ALL?***
2. ***What are some common side effects of chemotherapy?***
3. ***Why are children with neutropenia at risk of oral sepsis?***
4. ***How would non-compliance with preventive advice alter treatment planning?***
5. ***How would management differ if the child had a poor medical prognosis?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. Questions regarding the underlying disease, time of diagnosis, modalities of treatment the patient has received since the diagnosis, planned treatment, surgeries, complications, prognosis, current hematological status, allergies and medications
2. Mucositis, opportunistic infections, oral bleeding, salivary dysfunction, and neurotoxicity
3. They do not have enough neutrophils to defend them from an infection
4. Non-compliance with preventive measures such as daily oral hygiene would indicate a poor prognosis for minimizing adverse long-term oro-dental effects such as enamel demineralization and high caries rates
5. Management of children with a poor prognosis is generally palliative or symptomatic relief of pain and oral discomfort

Case 5

Liver Transplant

Figure. 1.5.1. Facial photo

A. Presenting Patient

- 4-year-, 11-month-old Caucasian male

B. Chief Complaint

- New patient referred by pediatric gastroenterology for dental assessment and treatment
- Mother noted decayed teeth but no symptoms
- Mother states that teeth have always been somewhat discolored but she is not concerned

C. Social History

- Lives at home with 6-year-old sibling and parents
- Middle class family

D. Medical History

- Biliary atresia, liver transplant at 13 months of age
- Medications: Tacrolimus 0.8 mg twice daily and Prednisolone 1 mg daily
- Frequent hospitalizations and hospital visits since birth for management of biliary atresia and liver transplant
- Allergic to eggs and nuts, no known drug allergies
- Vaccinations are up to date, including measles, mumps, and rubella, which is a live vaccine that was administered before the transplant when the patient was immunocompetent

E. Medical Consult

Pediatric Gastroenterologist

- All tests (liver function, urea and electrolytes, blood pressure) were within normal range for this patient

FUNDAMENTAL POINT 1

Medical History of a Liver Transplant Patient

- The dental professional needs to know if the child's liver transplant is functioning well prior to commencing a course of dental treatment. The liver is a complex organ with many important functions, including the synthesis of blood clotting proteins. The prothrombin time (PT) and partial thromboplastin time (PTT) are blood tests that may be used to assess the ability of the liver to synthesize clotting factors. Each laboratory has its own set of reference ranges for PT and PTT for children of different ages. The results of such a coagulation profile should be discussed with the child's physician before commencing a course of dental treatment. It is also important to remember that live vaccines are contraindicated in patients with solid organ transplants because of concerns of disseminated infection associated with immunosuppressive therapy.

BACKGROUND INFORMATION 1

Liver Function and Disorders

Some Important Functions of the Liver

- Metabolism of carbohydrates, lipids, and proteins
- Metabolism of drugs (detoxification) prior to excretion
- Synthesis of plasma protein (albumin) and clotting factors
- Storage organ for glycogen, vitamin B_{12}, and iron
- Breaks down hemoglobin; bilirubin and biliverdin are added to bile as pigment

Most Common Indications for Pediatric Liver Transplantation

- Chronic liver disease
  - Biliary atresia (most common reason for pediatric liver transplantation)
  - Alpha-1 antitrypsin deficiency
  - Autoimmune hepatitis
- Metabolic liver disease with extra-hepatic complications
  - Crigler-Najjar syndrome
  - Urea cycle defects
- Acute liver failure
- Hepatic tumors

Figure 1.5.2a–c. Pre-op intra-oral photos. A. Anterior view, B. maxillary view, C. mandibular view

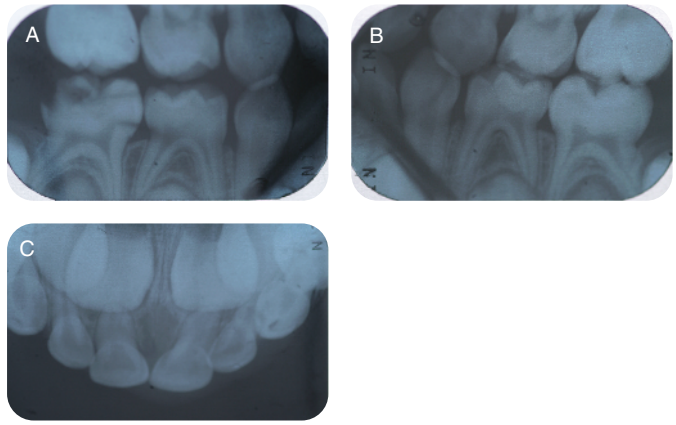

Figure 1.5.3a–c. Pre-op radiographs. A. Right bitewing radiograph, B. Left bitewing radiograph, C. Maxillary occlusal radiograph

F. Dental History

- No dental home
- Drank juices from a bottle until 4 years old
- Fair oral hygiene with parental supervision
- Uses toothpaste containing fluoride
- Optimal water fluoridation levels
- No history of dental trauma
- Asymptomatic dental decay

G. Extra-oral Exam

- No significant findings

H. Intra-oral Exam

- Soft and hard tissues: no significant findings
- Occlusal evaluation of primary dentition: class I canines and mesial step molars
- No visible plaque
- Carious lesions on primary molars
- Greenish discoloration of teeth consistent with history of biliary atresia
- Possible enamel hypomineralization and/or hypoplasia of mandibular right primary second molars with post-eruptive breakdown and caries
- Erosion of palatal surfaces of maxillary anterior teeth
- Cooperative for examination and radiographs

I. Diagnostic Tools

- Radiographs confirm extensive and deep decay involving mandibular right primary second molar

J. Differential Diagnosis

- Hemolytic disease of newborn
- Amelogenesis imperfecta
- Dentinogenesis imperfecta

- Coronal dentin dysplasia (Shields type II)
- Congenital porphyria

K. Diagnosis and Problem List

Diagnosis

- Early childhood caries
- Liver transplant
- Possible enamel hypomineralization and/or hypoplasia of mandibular right primary second molars with post-eruptive breakdown and caries
- Erosion of palatal surfaces of maxillary anterior teeth
- Intrinsic staining of teeth due to incorporation of unconjugated bile pigments in calcifying dental tissues

Problem List

- High caries risk due to:
  - Use of juice in a bottle until 4 years of age
  - Hospitalization as infant and young child due to biliary atresia
  - Need for daily ingestion of medications
  - Possible enamel hypomineralization/hypoplasia of mandibular primary second molars
  - Special health care needs
  - No dental home
  - Untreated caries
- Immunosuppressant therapy to prevent rejection of liver transplant
- Significant dental treatment required in a 4-year-old with history of extensive medical intervention since birth

L. Comprehensive Treatment Plan

Establishment of a Caries Prevention Plan as well as a Dental Home

Comprehensive Dental Treatment Under General Anesthesia (GA)

- Intravenous administration of hydrocortisone 25 mg at time of induction because of long-term steroid use

FUNDAMENTAL POINT 2

Dental Treatment Considerations in a Child with Liver Disease

- Decreased metabolism (detoxification) of many drugs including lidocaine: administer less than maximum recommended dose of local anesthetic (lidocaine <4.4 mg/kg)
- Decreased metabolism (detoxification) of general anesthetic and sedative agents: except for nitrous oxide/oxygen analgesia/anxiolysis, the use of sedation or general anesthesia should be restricted to specialist hospital units
- Decreased production of vitamin-K-dependent blood clotting factors (II, VII, IX, X): Increased potential for post extraction bleeding
- Enlargement of spleen may cause platelet sequestration with associated low platelet count: Increased potential for post extraction bleeding
- Local measures to control post-extraction bleeding include the use of oxidized regenerated cellulose (Surgicel or Gelfoam) and the use of resorbable sutures
- Hematology consult if platelet count <75,000/mm^3: Consider platelet transfusion to prevent prolonged bleeding
- Varices (enlarged blood vessels) at base of esophagus due to obstruction of blood flow to diseased liver: Chronic gastrointestinal hemorrhage may result in anemia
- Greenish discoloration of teeth due to incorporation of unconjugated bile pigments, especially biliverdin, during period of calcification of teeth
- Yellow discoloration of skin and mucosa (jaundice) due to presence of unconjugated bilirubin in the tissues
- Do not use nonsteroidal anti-inflammatory drugs (NSAIDS) for analgesia because they increase the risk of gastrointestinal bleeding
- Use acetaminophen in reduced dosage for analgesia because high doses are hepatotoxic

- Stainless steel crown (SSC) restoration of mandibular right primary second molars following caries removal
- High-strength glass ionomer or composite resin restorations of occlusal lesions in both maxillary primary second molars and mandibular primary first molars
- Post-operative pain relief with acetaminophen 500 mg as a suppository and intra-oral buccal infiltration of 0.5 ml 2% lidocaine with epinephrine

Follow-up Care

- Post-operative and home care instructions
- Establishment of a frequent recall schedule

M. Prognosis and Discussion

Good Prognosis if Aggressive Preventive Plan is Established and Consistent Home and Professional Dental Care is Provided

- Three-month dental recall visits
- Fluoride varnish application biannually
- Fissure sealant application to the permanent first molars when erupted sufficiently to allow isolation for moisture control
- Promotion of twice daily toothbrushing with fluoride toothpaste and restriction in the frequency of ingestion of sugary drinks/juices and sugary snack foods

N. Common Complications and Alternative Treatment Plans

Extensively Decayed Mandibular Right Second Primary Molar

- Vital pulpotomy and SSC if pulp exposure
- Extraction of tooth if non-vital with infection and subsequent space management/maintenance when permanent first molar erupts

Distal Shoe Appliance Relatively Contraindicated in View of Immunosuppression

Long-term Complications of Steroid and Immunosuppression Agents with Dental Implications: Gingival Overgrowth, High Blood Pressure, Osteoporosis, etc.

FUNDAMENTAL POINT 3

Dental Management of Child Before Liver Transplantation

- Prevention: Educate child and caretakers on the importance of optimal oral care at time of transplantation and afterwards
- Treat/stabilize active dental decay so that the teeth will not be a potential source of infection at the time of transplantation and for the following three to six months
- Extract extensively decayed teeth and teeth with pulp or potential pulp pathology
- Consult the pediatric gastroenterology team: seek advice on coagulation status and ensure a platelet count of >75,000/ml^3 before extracting teeth

Dental Management Following Liver Transplantation

- Regular dental visits for preventive and ongoing care from six months following transplant
- Immunosuppression issues
- Scrupulous cross-infection control measures
- Decreased immunosurveillance
- Increased risk of lymphomas (non-Hodgkins lymphomas or post-transplant lymphoproliferative disease)
- Increased risk of skin cancer: Reinforce safety in sun and the use of sunscreen with high sun protection factor
- Cyclosporine with/without the antihypertensive drug nifedipine may cause gingival hyperplasia and delayed eruption of teeth
- No gingival problems with tacrolimus, which is an alternative to cyclosporine
- Caution with use of NSAIDs for analgesia because these may increase the nephrotoxicity of cyclosporine and tacrolimus
- Glucocorticoids may be used in low dosage as immunosuppressant: Additional dosage not usually required tor dental treatment unless treatment provided under general anesthesia
- Azathioprine and mycophenolate mofetil are other immunosuppressant drugs that may be used following solid organ transplantation. They have no specific oral or dental side effects

Self-study Questions

1. ***What is the most common liver disease in children that requires transplantation?***
2. ***What is the most likely cause of greenish staining of teeth in children with liver disease?***
3. ***What is the minimum platelet count that is recommended for dental extractions?***
4. ***What analgesic should be prescribed with caution in children with liver disease and why?***
5. ***What immunosuppressant medication may induce gingival hyperplasia?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. Biliary atresia
2. The incorporation of unconjugated bile pigments, especially biliverdin, during the period of calcification of teeth
3. 75,000/mm^3
4. Acetaminophen should be prescribed in reduced dosages for analgesia in children with liver disease because high doses may be hepatotoxic
5. Cyclosporin

Case 6

Chronic Benign Neutropenia

A. Presenting Patient

- 10-year-, 11-month-old Caucasian male

B. Chief Complaint

- New patient referred by pediatric hematologist with concerns about gingival inflammation.
- Mother states that "his gums are red and bleed during brushing."

C. Social History

- Lives at home with his parents
- Middle class family
- No siblings

D. Medical History

- Chronic benign neutropenia (absolute neutrophil count [ANC] of 300 cells/mm^3 at time of diagnosis)
- Chronic lower respiratory tract infection
- Medications: Azithromycin 250 mg oral prophylaxis on alternate days
- Granulocyte–colony stimulating factor (G-CSF) subcutaneous administration twice weekly
- Hospitalizations: Parenteral antibiotics and physiotherapy for management of recurrent pneumonia. Recent surgical resection of chronically infected lower lobe of left lung
- ANC today: 2,100 cells/mm^3 (see Fundamental Point 1)
- No known allergies to medications or foods
- Vaccinations up to date

E. Medical Consult

- Pediatric hematologist to confirm medical history and advise about any special precautions when providing dental treatment

F. Dental History

- Regular visits to a general dentist for routine care, including management of hypomineralized permanent first molars
- Brushes teeth twice each day with parental supervision
- Healthy low-cariogenic diet
- Uses toothpaste containing fluoride
- Optimal water fluoridation

G. Extra-oral Examination

- No significant findings

BACKGROUND INFORMATION 1

Immunological Defects and Susceptibility to Infection

- An immunocompromised child has one or more defects of the immune system that are sufficiently severe to predispose him to life-threatening infection. A deficiency in the number or quality of phagocytic cells, humoral immunity (immunoglobulin and complement), or cell-mediated immunity is associated with an increased risk of infection by certain pathogens, depending on the type of defect. It is important to remember that immunological defects rarely exist in isolation and frequently there is more than one defect present

Infections in Children With Neutropenia or With Disorders of Neutrophil Function

- Patients are particularly susceptible to recurrent and severe bacterial or fungal infections of the lungs, paranasal sinuses, oropharynx and skin
- They are also susceptible to overwhelming infection, including those of dental origin

See Table 1.6.1: Pathogens and immunological defects

Table 1.6.1. Pathogens and immunological defects

Abnormality	Bacterial	Fungal	Viral	Protozoal
Neutropenia or qualitative defects of phagocytes	Gram-positive Staphylococci (coagulase positive and negative) Streptococci (enterococcus and α-hemolytic) Gram-negative *E.coli K.pneumoniae* *P. aeroginosa* Other Enterobacteriaceae	Candida spp Aspergillus spp		
Defective cell-mediated immunity	*Legionella* *Salmonella* Mycobacteria	Histoplasma capsulatum Cryptococcus neoformans Candida Aspergillus	Cytomegalovirus Varicella-zoster Herpes simplex Epstein-Barr Live viral vaccines (measles, mumps, rubella, polio)	*P. carinii* *T. gondii* Cryptosporidia
Immunoglobulin deficiency	*S. pneumoniae* *H. influenzae*		Enteroviruses	Giardia
Complement deficiency	*S. pneumoniae* *H. influenzae* Neisseria species			
Splenectomy	*S. pneumoniae* *H. influenzae* Neisseria species			Babesia

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H. Intra-oral Examination

Soft Tissues

- Gingivitis, especially around mandibular and maxillary incisors

Bone Tissues

- No significant findings

Occlusal Evaluation of Mixed Dentition

- Class 1 molar relationship with crowding in the canine and premolar regions, more severe in the maxillary arch; overjet: 3 mm, overbite: 80%. There is a mandibular midline shift to the left associated with premature exfoliation of the mandibular left primary first molar (see Figure 1.6.1)

Dental Exam

- Minimal visible plaque with significant gingivitis
- Enamel hypomineralization affecting the permanent first molars with opacities on the labial surface of two permanent incisors
- Defective restoration of mandibular right permanent first molar and mandibular left primary second molar

I. Diagnostic Tools

Panoramic Radiograph (Exposed Six Months Before This Examination) (Figure 1.6.2)

- Unerupted premolars and permanent cuspids show insufficient space to erupt in good alignment. All permanent teeth, including permanent third molars, are present.
- The decayed and hypomineralized maxillary right permanent first molar has been restored since the radiograph was exposed. The restorations of the mandibular right permanent first molar and mandibular left primary second molar are defective and are scheduled for restoration by the local dentist

J. Differential Diagnosis

Gingivitis Due to:

- Poor plaque control
- Incompetent lips/mouth breathing causing drying of gingiva
- Crohn's disease (inflammatory bowel disease)

FUNDAMENTAL POINT 1

Neutropenia

- Neutrophils and monocytes are phagocytic cells that function within the body as the chief defense against infection by bacteria and fungi
- The neutrophil is the predominant phagocytic cell in the peripheral circulation
- Neutrophils have a major role in protecting those surfaces of the body that are in direct contact with the external environment
- The absolute neutrophil count (ANC) is a measure of the number of neutrophils in the blood.
- ANC values can determine what procedures can safely be performed
  - 1,500 to 8,000 cells/mm^3 is normal
  - 500 to 1,500 is safe (no restrictions)
  - 500 or less is low (restrictions apply)
- The normal lower limit for circulating neutrophils is 1,500 to 2,000 cells/mm^3. Severe neutropenia occurs with an ANC below 500 cells/mm^3, which puts the patient at risk of serious infection, including those of oral origin. With an ANC of 100 cells/mm^3 there is a dramatic increase in the incidence of severe infection.

Causes of Neutropenia

Different Diseases May Result in Low Numbers or Absence of Neutrophils. The Causes of Neutropenia In Children Include:

Neutropenias Present at Birth

- Severe congenital neutropenia (Kostmann syndrome) with associated severe periodontitis
- Cyclic neutropenia. Primary cyclic (every 21 days) decrease in maturation of precursor cells in bone marrow with recurring fever, skin infections, oropharyngeal disease, and periodontitis
- Neutropenia in association with a primary immunodeficiency
- Neutropenia in association with a metabolic disorder (Shwachmann-Diamond syndrome and Glycogen–storage disease type 1b)
- Neutropenia as part of certain syndromes (pancytopenia in Fanconi's anemia; dyskeratosis congenita with progressive marrow failure)

Neutropenias Acquired During Life

- Idiopathic neutropenia
- Autoimmune neutropenia
- Chronic benign neutropenia
- Hematological dysplasias and malignancies
- Solid tumor invasion of bone marrow
- Cancer chemotherapy and radiotherapy
- Drug toxicity
- Viral infections

Qualitative Defects of Phagocytes With a Normal Number of Neutrophils Occur in the Following Inherited Conditions:

- Chronic granulomatous disease (CGD)
- Leucocyte adhesion deficiency (LAD)

BACKGROUND INFORMATION 2

Medical Management of Neutropenia

Medical Management Depends on its Cause and Severity. The Primary Goal is to Prevent Infection With:

- Prophylactic antibiotics
- Granulocyte–Colony Stimulating Factor (G-CSF)
- Interferon gamma (IFN-γ) for management of chronic granulomatous disease (which presents a normal number of neutrophils but with a functional defect)
- Hematopoietic stem cell transplant (e.g., for Kostmann and Shwachmann-Diamond syndromes because of high risk of developing acute myeloid leukaemia; for chronic granulomatous disease, transplant is done to prevent life-threatening infection)
- Immunoglobulin and steroid therapy

Figure 1.6.1a–c. Pre-op intra-oral photos. A. Anterior, B. maxillary, C. mandibular

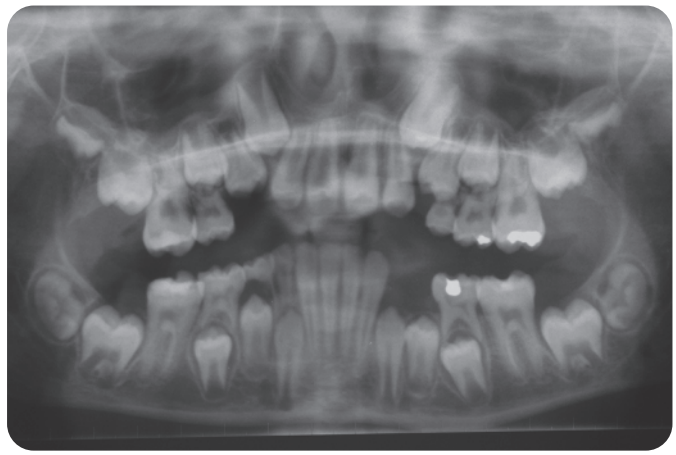

Figure 1.6.2. Panoramic radiograph

FUNDAMENTAL POINT 2
Oral Manifestations of Neutropenia or Disorders of Neutrophil Function

- Gingivitis
- Periodontitis
- Oral ulceration

BACKGROUND INFORMATION 3
Molar-incisor-hypomineralization

- Molar-incisor–hypomineralization or idiopathic enamel hypomineralization usually manifests as hypomineralization or hypoplasia of one or more permanent first molars with opacities of one or more permanent incisors
- It may be associated with a history of illness during infancy or early childhood such as pneumonia, otitis media, and fever, or there may not be any relevant past medical history

- Prepubertal periodontitis
- Primary immunodeficiency disease
- HIV/AIDS
- Hematological dysplasia or malignancy
- Chemotherapy
- Poorly controlled diabetes mellitus

Hypomineralized Permanent First Molars

- Caries
- Chronological enamel hypoplasia
- Amelogenesis imperfecta
- Fluorosis (severe)

K. Diagnosis and Problem List

Diagnosis

- Gingivitis
- Hypomineralized permanent first molars
- Dental crowding and mandibular midline shift to the left

L. Comprehensive Treatment Plan

Treatment of Gingivitis

- Toothbrushing and flossing instruction
- Chlorhexidine mouthwash (0.12%) or gel (0.2%) for initial period of two to three weeks to help resolve gingivitis
- Use of G-CSF leads to increase in ANC with a consequent reduction of the gingivitis

Treatment of Hypomineralized Permanent First Molars

- Interim restorations using resin or resin-modified glass ionomers
- Avoid use of stainless steel crown restorations because of anticipated exaggerated gingival response in association with medical history of neutropenia and associated gingivitis
- Prosthodontic referral when older for long-term restorations of permanent first molars

FUNDAMENTAL POINT 3

Dental Treatment of Children with Neutropenia or Disorders of Neutrophil Function

- Contact medical specialist for consult
- Practice preventive dentistry from age of diagnosis to prevent the need for invasive dental treatment
- Advise use of sugar-free medications
- Extract pulpally involved teeth
- Extract teeth with severe periodontal disease
- Use antibiotic prophylaxis for surgical dental treatment (consult with patient's physician)
- Follow-up visit after dental treatment to check that healing is satisfactory and any infection is resolved

Caries Prevention

- Reinforce advice about balanced healthy diet
- Advise use of sugar-free preparations of liquid medications (antibiotics)
- Reinforce twice daily use of fluoride toothpaste
- Fissure sealant application on eruption of premolars and permanent second molars

Orthodontic Assessment

- Referral for orthodontic assessment of dental crowding and evaluation of hypomineralized permanent first molars
- Extraction of severely hypomineralized permanent first molars may be considered in some cases as part of the orthodontic treatment plan

M. Prognosis and Discussion

- Ongoing liaison and communication with hematologist who will continue to review and manage patient's neutropenia
- Regular review of gingival condition. Consult with periodontist for best time to refer for regular periodontal review into adulthood
- Referral for orthodontic management of crowding
- More definitive restorations of the hypomineralized permanent first molars may be provided in late teenage years or in young adulthood, preferably by a prosthodontist because of the history of neutropenia and gingivitis

N. Common Complications and Alternative Treatment Plans

- An exacerbation of gingivitis may be anticipated during fixed orthodontic appliance treatment and the orthodontist should be informed of this
- An alternative treatment is to monitor the development of occlusion, without orthodontic treatment, and accept some minor crowding and lower midline shift to the left
- Another option, following consultation with an orthodontist, is to extract the most severely hypomineralized permanent first molars to facilitate mesial movement of the unerupted and unaffected permanent second molars. Spontaneous space closure should occur in the majority of cases but there may be mesial inclination of the second permanent molars (Jalevik and Moller 2007).

Self-study Questions

1. ***What is the absolute neutrophil count (ANC)?***
2. ***What is the normal range of ANC values in a healthy child?***
3. ***What is one of the primary aims of medical management of a child with neutropenia?***
4. ***What are the predominant types of pathogens likely to cause infection in a child with neutropenia?***
5. ***What oral conditions may be typically seen in a patient with neutropenia?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. The absolute neutrophil count (ANC) is a measure of the number of neutrophils in the blood expressed as the number cells/mm^3 of blood
2. The normal range of ANC in healthy children is 1,500 to 8,000 cells/mm^3
3. Prevention of infections and their sequelae
4. Bacteria and fungi
5. Gingivitis, periodontitis, and/or oral ulceration

Case 7

Asthma

A. Presenting Patient
- 5-year-old Caucasian female
- New patient referral from community dental service

B. Chief Complaint and History of Present Illness
- Intermittent toothache over the past two weeks keeping child awake at night

C. Social History
- Lives at home with parents and two older siblings
- Low socio-economic status

D. Medical History
- Moderate persistent asthma: generally well-controlled with infrequent acute exacerbations
- Allergic rhinitis
- Allergies: dust mite, pollen
- Current medications: Beclomethasone twice daily, Albuterol as required, nasal corticosteroid spray
- Admitted with an acute asthmatic episode eight months ago
- No known food or drug allergies, vaccinations up to date

E. Medical Consult
- No need to contact pediatrician because asthma is generally well controlled

F. Dental History
- Has a dental home but despite numerous previous dental visits to manage early childhood caries (ECC), no treatment has been accomplished
- Diet: frequent consumption of fruit juices
- Poor oral hygiene
- Brushes once daily, unsupervised
- Only occasionally uses fluoridated toothpaste
- Lives in an area with water fluoridation of 0.8 ppm
- Very anxious concerning dental treatment

G. Extra-oral Exam
- Open mouth breathing

H. Intra-oral Exam
Soft Tissues
- White plaque-like lesions overlying hard palate
- Generalized gingivitis

Occlusal Evaluation of Primary Dentition
- Class I canines and molars

Dental Exam
- Heavy plaque
- Multiple teeth with carious lesions
- Surface wear consistent with bruxism is evident on some teeth

I. Diagnostic Tools
- Microbiology: swab of palatal mucosa
- Periapical radiographs (taken under general anesthesia)

J. Diagnosis and Problem List
Diagnosis
- Dental anxiety
- Severe ECC
- Oral candidiasis; pseudomembranous candidiasis of hard palate
- Tooth wear
- Pulpal pathology

Problem List
- Dental pain
- Risk of developing dental infection
- Very anxious child
- Impact of asthma medication on oral health

High Caries Risk Due to:
- Special health needs
- Use of medication that can impair salivary flow
- Presence of dental caries

BACKGROUND INFORMATION 1

Asthma

Definition

- Asthma is a common chronic disorder of the airways, characterized by variable and recurring symptoms, airflow obstruction, bronchial hyper responsiveness and an underlying inflammation.

Prevalence

- Estimated 300 million people suffer from asthma worldwide
- One of the most common chronic diseases among children

Etiology

- The etiology is not fully understood. Precipitating factors include pollens, mold spores, house dust, viral infections, cigarette smoke, cold air, extreme emotional arousal, exercise, and anti-inflammatory medication

Pathophysiology

- Airflow limitation is due to a number of changes influenced by airway inflammation
- Bronchoconstriction following irritant exposure
- Airway hyper responsiveness
- Airway edema and mucous hypersecretion

Signs

- Wheeze
- Tachypnea

Symptoms

- Wheeze
- Shortness of breath
- Chest tightness
- Cough

Classification

Based on Etiology

- Extrinsic: Allergic
- Intrinsic (specific triggers, e.g., exercise)

Based on severity

- Intermittent
- Persistent: Mild, moderate, severe

Management

Goals

- Reduce impairment and maintain (near) normal lung function and normal activity levels
- Reduce risk: Prevent exacerbations, minimize need for emergency care, prevent reduced lung growth, have no adverse effects of therapy

How

- Assessment and monitoring
- Patient education
- Control of environmental factors and co-morbid conditions
- Medications

FUNDAMENTAL POINT 1

Assessment of Asthma

History

- Type and severity of asthma
- Frequency of asthmatic attacks
- Precipitating factors
- Last acute episode and hospital admission
- Symptoms associated with sports/exercise
- Type of medication used regularly and during an acute episode

Consult With Pediatrician in Uncontrolled or Severe Cases. Consultation May Include Assessment of:

- Shortness of breath
- Coughing
- Wheezing
- Rate and depth of respiration
- Use of accessory muscles of respiration
- Auscultation of the lungs
- Oxygen saturation
- Pulmonary Function Test
- Peak Flow Test
- Spirometry

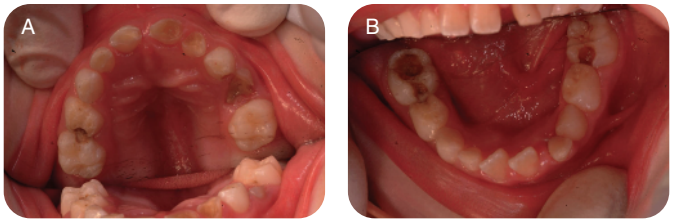

Figure1.7.1a–b. Pre-op intra-oral photos. A. Maxillary, B. Mandibular

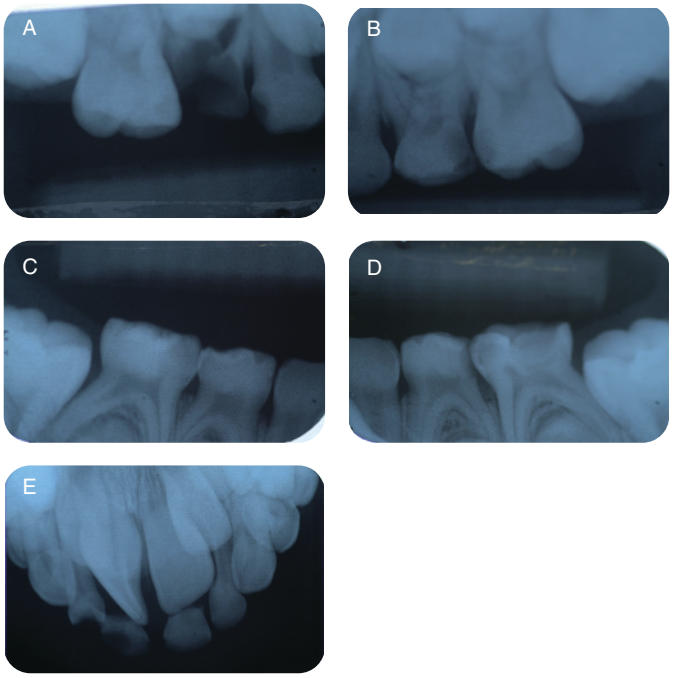

Figure 1.7.2a–e. Pre-op radiographs. A. maxillary right periapical radiograph, B. maxillary left periapical radiograph, C. mandibular right periapical radiograph, D. mandibular left periapical radiograph, E. Maxillary occlusal radiograph

- Low socio-economic status
- Frequent between-meal sugar exposure (fruit juice)
- Unsupervised toothbrushing only once a day
- Visible plaque

K. Comprehensive Treatment Plan

Preventive Plan

- Diet
  - Stop frequent juice intake
  - Limit between-meal snacks
- Oral hygiene
  - Brush twice daily (morning and night)
- Fluoride exposure
  - Consistently brush with fluoridated toothpaste
  - Continue to consume fluoridated water
- Visit dentist for regular review and biannual fluoride varnish application

Management of Oral Candidiasis

- Prevention
  - Consider use of aerosol holding chamber
  - Rinse with water following exposure to inhaled corticosteroids
- Treatment
  - Rinse with chlorhexidine mouthwash daily while candidiasis is present
  - Consider topical antifungal drug therapy in more persistent cases

Comprehensive Dental Treatment Under General Anesthesia (GA)

- Medical
  - Preventive asthma medication as normal
  - Many anesthetic gases are bronchodilators
- Dental
  - Removal of extremely carious, mobile (e.g., maxillary central incisors), and non-vital teeth
  - Restoration of remaining carious teeth: Stainless steel crown (SSC) restoration of two or more surface carious lesions, ferric sulphate vital pulpotomy and SSC if pulp exposed
  - Restoration of single surface carious lesions in molar teeth with high-viscosity glass ionomer cement
  - Space maintenance
- Behavioral management considerations
  - Comprehensive dental treatment under GA to facilitate all necessary procedures at one time
- Follow-up care
  - Post-op and home care instructions: Asthma medication as normal, resume toothbrushing
  - Recall plan: Two weeks post-op, establish regular recall schedule

L. Prognosis and Discussion

- Prognosis for limiting caries progression is guarded
- Prognosis for changing current dietary habits is guarded
- Prognosis for prevention of asthmatic attack is good because of regular use of asthma medication and bronchodilator effect of many anesthetic agents

BACKGROUND INFORMATION 2

Pharmacotheraputic Management of Asthma

- Therapy is initiated based on asthma severity and adjusted as necessary based on asthma control. Stepwise approach to asthma management
- Every patient with persistent asthma, regardless of disease severity, should use a daily controller medication, i.e., an inhaled corticosteroid (ICS)

Quick-relief Medication

- To treat acute symptoms and exacerbations
  - Inhaled short-acting beta agonists, e.g., Albuterol

Long-term Control Medication

- To maintain and achieve control of persistent asthma
  - First choice: ICS (e.g., Beclomethasone)
  - Inhaled long-acting beta$_2$-agonist
  - Leukotriene receptor antagonist
  - Systemic corticosteroids
  - Immunomodulators
- Long-term use of ICS within labeled doses is safe for children in terms of growth, bone mineral density, and adrenal function
- Low- to medium-dose ICS are not associated with the development of cataracts or glaucoma in children

Oral Implications of Asthma and/or its Management

Oral Mucosal Changes

- Gingivitis (associated with mouth breathing)
- Oral candidiasis (associated with use of inhaled corticosteroids)
- Dryness of mouth (associated with use of inhaled corticosteroids)

Dental Caries

- There is insufficient evidence to confirm the increased risk of dental caries and/or erosion in patients with asthma
- Lactose is the carrier for many devices; it gives taste so patient knows that a dose was dispensed
- Inhalers do not taste good; thus, patients increase consumption of flavored beverages

Asthma Medication

- Beta$_2$-agonist medications are associated with:
  - Decrease in salivary flow
  - Decrease in plaque pH
  - Muscle relaxation with subsequent gastroesophageal reflux and associated acid reflux

- Prognosis for prevention of further candidiasis is guarded because of regular use of inhaled corticosteroids and difficulty in rinsing mouth

M. Common Complications and Alternative Treatment Plans

- Non-compliance with dietary advice
- Continued caries progression
- Acute asthmatic episode (see management in Chapter 7)
- Alternative treatment plans may include:
  - Control behavior with conscious sedation
  - Alternative restorative materials, e.g., amalgam or composite for small single or two-surface restorations
  - Alternative pulpal management (e.g., pulpectomy) in non-vital teeth

BACKGROUND INFORMATION 3

Dental Management of Patients with Asthma

- Optimal asthma control is desired prior to dental treatment
- If wheezing, severe, or poorly controlled asthma: reappoint ± medical consult ± consider a hospital setting
- Asthma medication should be taken as normal on day of dental treatment. Bronchodilator should be brought along to the dental appointments
- Behavior management is essential to alleviate anxiety and reduce risk of an acute episode
- Non-steroidal anti-inflammatory drugs (NSAIDS) should be used with caution in all children with asthma: 4% of asthmatics are allergic to aspirin and other NSAIDS. Use acetaminophen instead
- Avoid use of opiates
- For patients taking long-term oral corticosteroids, steroid supplementation is necessary in anticipation of a stressful situation such as dental extractions or during general anesthesia
- Nitrous oxide is contraindicated in severe asthmatics: consult physician
- Unhumidified nitrous oxide may dry out secretions
- Use rubber dam whenever possible
- Certain materials used in the dental office can trigger attacks: sealants, enamel dust, cotton rolls, sulfites, dentrifices, methyl metacrylate
- Avoid long appointments

Conscious Sedation

- Oral: benzodiazepines (e.g., midazolam 0.5 mg/kg) in patients with mild/moderate asthma
- Inhalation sedation (e.g., nitrous oxide) in patients with mild/moderate asthma
- Intravenous: Use extreme caution in patients with asthma (consult pediatrician and anesthesiologist)

General Anesthesia

- Pre-anesthetic review for children with severe or uncontrolled asthma. Non-urgent dental treatment should be postponed until asthma is controlled

Self-study Questions

1. ***What important questions need to be asked when taking a medical history from a patient with asthma?***
2. ***List some potential stimuli/triggers of an episode of asthma.***
3. ***What are the four main areas of asthma management?***
4. ***Name one inhaled medication used to prevent asthma and one inhaled medication used to provide quick relief from an acute episode of asthma.***
5. ***Name a common oral manifestation of inhaled corticosteroid therapy.***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. Type and severity of asthma, level of asthma control, precipitating factors, frequency of acute asthmatic episodes, last acute episode and whether admitted to the hospital, and type of medication used regularly and during an acute episode
2. Pollens, mold spores, house dust, viral infections, cigarette smoke, cold air, extreme emotional arousal, exercise, and certain anti-inflammatory medications
3. Assessment and monitoring, patient education, control of environmental factors and other asthma triggers, and pharmacotherapy
4. Beclomethasone and Albuterol
5. Oral candidiasis

Case 8

Crohn's Disease

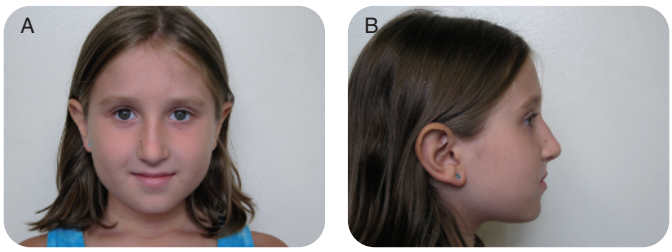

Figure 1.8.1a–b. Facial photographs at 6 month follow up visit

A. Presenting Patient

- 7-year-, 4-month-old Caucasian female

B. Chief Complaint and History

- New patient referred from pediatric gastroenterology for assessment of swollen lower lip and angular cheilitis as part of a diagnostic work-up of inflammatory bowel disease
- Mother noted progressive swelling of lower lip for the past three to four months concurrently with abdominal cramps and diarrhea

C. Social History

- Lives at home with both parents
- Sister (5 years old) and brother (2 years old)
- Middle class family
- Maternal aunt has Crohn's disease

D. Medical History

- Hospitalized because of weight loss, abdominal cramps, and diarrhea. Gastroenterologists undertaking investigations to determine if child has Crohn's disease (CD)
- Three-month history of abdominal cramps, diarrhea, anorexia, and weight loss
- No medications present during medical work-up
- No known allergies to foods or medications
- Vaccinations are up to date

Specific Examinations

- Weight: 21.5 kg (Below the 25th Percentile), Hemoglobin: 9.7 g/dl (Normal Range: 11 to 14), Platelets: 507 × 10^9/l (Normal Range: 140 to 400), C Reactive Protein: 13 mg/l (Normal Range: <10), Albumin: 26 g/l (Normal Range: 35 to 50)
- Barium meal and follow-through showed irregular stricture involving the distal portion of the terminal ileum
- Upper endoscopy and colonoscopy reveal multiple regions with inflammation but mainly involvement of terminal ileum with large ulcers

Histopathology

- Biopsies from stomach, terminal ileum, and colon show focal areas of acute and chronic inflammation
- Gingival biopsy (mandibular incisor labial gingiva) shows marked chronic inflammation and non-necrotizing granulomas, which, in combination with acute and chronic inflammation in the other biopsies, supports the clinical diagnosis of CD with involvement of multiple regions of the GI tract

E. Medical Consult

- Pediatric gastroenterologist

F. Dental History

- One previous dental visit at 6 years of age and dentist noted decay-free primary dentition
- Poor oral hygiene during last two weeks due to illness; supervised by parents. Difficult to brush teeth because of swollen lower lip
- Reduced appetite during last three months but good diet before
- Uses toothpaste containing fluoride
- Lives in optimally fluoridated area
- No history of dental trauma

BACKGROUND INFORMATION 1

- Inflammatory bowel disease (IBD) can be sub-classified as ulcerative colitis (UC) or Crohn's disease (CD). Indeterminate colitis is the term used when unable to clearly discriminate between UC and CD.

Crohn's Disease

- Chronic granulomatous inflammatory disorder of unknown etiology, likely the result of an inappropriate inflammatory response in a genetically susceptible individual to an environmental stimulus
- May affect any part of the gastrointestinal (GI) tract from the mouth to the anus
- Typically involves the terminal segment of the small intestine (ileum) and first segment of the large intestine (colon)
- Peak incidence is in second and third decades of life with up to one-third of cases occurring before 20 years of age
- There is familial clustering with a history of IBD in approximately 15% of cases of CD
- There are probably several genes involved in conferring susceptibility to IBD and there may be genetic heterogeneity, with different genes having similar phenotypic expressions
- NOD2 is the best characterized susceptibility gene, mutations of which confer increased risk for CD. The gene encodes for a protein that is involved in recognizing pathogens and intracellular signaling of the innate immune response

Treatment of Crohn's Disease

- Medical induction and maintenance of remission of intestinal inflammation
- Nutritional support may require nasogastric infusion of formulated food in severe cases with malnourishment and growth retardation, to increase caloric intake
- Medical treatment of relapses/acute exacerbations
- Surgical intervention if there are intractable symptoms despite medical therapy or if there are intestinal complications such as obstruction, infection, fistula, perforation, or hemorrhage

BACKGROUND INFORMATION 2

Presentation of Crohn's Disease in Children

- Abdominal pain
- Diarrhea ± blood in stools
- Poor appetite
- Weight loss
- Impaired growth and pubertal delay
- Anemia due to malabsorbtion and blood loss
- Erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) and high platelet count are indicative of an inflammatory process
- Low albumin due to protein losing enteropathy oral soft tissue manifestations

G. Extra-oral Examination

- Bilateral angular cheilitis
- Gross swelling of lower lip with vertical fissures
- Dry lips

H. Intra-oral Examination

- Soft tissues: Swelling of lower labial mucosa, soft tissue tags and ulcers in lower labial sulcus, and swelling of marginal and attached gingiva around mandibular incisors
- Early mixed dentition with class I relationship of permanent first molars. Mandibular permanent lateral incisors erupting lingual to permanent central incisors with retention of both mandibular primary lateral incisors and the mobile mandibular left primary central incisor
- Generalized plaque accumulation, particularly on labial aspect of incisors
- Caries-free dentition

Figure 1.8.2. Swelling and angular cheilitis of lower lip

I. Diagnostic Tools

- Gingival biopsy when under general anesthesia for endoscopy and colonoscopy

Figure 1.8.3. Intra-oral soft tissue inflammation and ulcerations

FUNDAMENTAL POINT 1

Oral Manifestations of Crohn's Disease

Up to 40% of Children May Have One or More of the Following Oral Manifestations of CD at Initial Presentation:

- Lip and/or cheek swelling
- Angular cheilitis
- Mucogingivitis (inflammation of marginal and attached gingiva), most commonly in the anterior region
- Irregular nodular swelling or "cobblestoning" of buccal mucosa
- Long, deep ulcers in mandibular buccal sulcus
- Mucosal tags
- Multiple aphthous ulcers

J. Differential Diagnosis

- Orofacial granulomatosis (OFG) that may be associated with hypersensitivity to foodstuffs or additives
- Ulcerative colitis (UC): Inflammatory bowel disease affecting the colon in which there may be associated oral aphthous ulceration. Pyostomatitis vegetans with tiny yellow pustules in the oral mucosa has been reported in individuals with UC and CD
- Hereditary angioedema
- Allergic angioedema

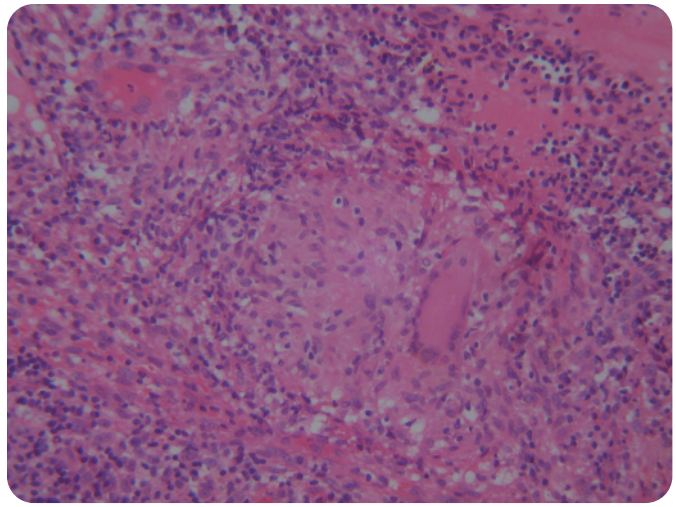

Figure 1.8.4. Histological image of gingival granuloma, formed by an aggregate of epithelioid macrophages. To the right of the center of the image is a multinucleated giant cell

FUNDAMENTAL POINT 2

Biopsy of Oral Lesion

- A non-necrotizing granuloma is a key histopathological finding in Crohn's disease. It consists of an aggregrate of epithelioid macrophages ("epithelioid": lots of pink cytoplasm similar to squamous epithelial cells) and multinucleate giant cells
- Serial sections of a specimen must be examined by a pathologist to find loosely formed non-necrotizing granulomas that are scattered in affected tissue
- Biopsy of a gingival or mucosal tag lesion, if present, is recommended because non-necrotizing granulomas should be found in a biopsy from these sites
- Biopsy of the lower lip is not recommended because there is a risk of damage to the labial branches of the mental nerve. Furthermore, granulomas in an extensively edematous lip will be sparse and scattered and may not be captured in a biopsy specimen

K. Diagnosis and Problem List

- CD with oral lesions
- Retained primary incisors

L. Comprehensive Treatment Plan

- Gingival biopsy to obtain histological confirmation of granulomatous inflammation and diagnosis of CD

- Extraction of retained mandibular primary incisors
- Oral hygiene instruction
- Monitor developing occlusion

BACKGROUND INFORMATION 3
Orofacial Granulomatosis

- Orofacial granulomatosis (OFG) is a condition characterized by orofacial swelling with biopsy-positive non-necrotizing granulomas in a patient without Crohn's disease (CD) or other systemic disease
- May be associated with a hypersensitivity to certain food and drink additives such as benzoates, cinnamon, and tartrazine
- OFG that is associated with hypersensitivity to food additives may improve or resolve with an exclusion diet; however, compliance with such a diet may be very difficult
- Intra-lesional steroids may be required to treat lip swelling in OFG
- A child with OFG may not have overt symptoms or signs of CD but may subsequently develop intestinal CD
- A child with OFG should be referred to a specialist in oral medicine. Review by a pediatric gastroenterologist may also be required because OFG may precede the development of CD.

- Fissure sealant application to permanent first molars when erupted sufficiently to isolate for moisture control

M. Prognosis and Discussion

- Regression of oral lesions anticipated with treatment of CD
- Oral lesions may reappear if there is relapse of systemic CD

Initial Medical Treatment Following Diagnosis of CD

- 6 Mercaptopurine 25 mg daily
- Prednisolone 25 mg daily for four weeks. Then reduce dosage by 5 mg each week until tapered course is completed
- Iron supplements for treatment of anemia: 6 mg/kg daily of elemental iron

Dental Treatment

- Delay treatment in the dental office until medical improvement in CD
- Monitor retained mandibular primary incisors and anticipate spontaneous exfoliation
- Frequent preventive dental visits to encourage improved oral hygiene; professional application of fluoride varnish and fissure sealant application to permanent first molars
- If general anesthesia is planned, Prednisolone therapy may result in suppression of the normal adrenocortical response, with a risk of developing hypotension. Hydrocortisone should therefore be

BACKGROUND INFORMATION 4
Categories of Drugs Used in the Management of Crohn's Disease

Drug category	Drug	Comments
Anti-inflammatory	Glucocorticoids Prednisolone	Conventional corticosteroid, initial high dose to control disease
	Budesonide	Controlled ileal release corticosteroid
	Enteral nutrition	Typically exclusive use of elemental or semi-elemental diet for six to eight weeks (mechanism of action unknown)
	Aminosalicylates (ASA) Sulfasalazine Mesalamine	Active component, 5-ASA, acts locally in intestinal mucosa to inhibit inflammation
Antibiotic	Metronidazole Ciprofloxacillin	Antibacterial effect on intestinal flora
Immunomodulator	6-Mercaptopurine Azathioprine	Immunosuppressant Steroid-sparing
Biologic	Infliximab/Remicade	Monoclonal antibody that neutralizes bioactivity of the key inflammatory cytokine, tumor necrosis factor-alpha (TNF-α)

administered IV at induction of anesthesia by an anesthesiologist if general anesthesia is administered within three to six months of glucocorticoid therapy. There is no need for IV hydrocortisone or glucocorticoid medication prior to providing dental treatment under local analgesia in a dental office.

Self-study Questions

1. ***What parts of the gastrointestinal tract may be affected in CD?***
2. ***Name four possible orofacial manifestations of CD.***
3. ***What is one of the key histopathological findings in CD?***
4. ***What categories of drugs may be used in the management of CD in children?***
5. ***Orofacial granulomatosis in some cases may be associated with a hypersensitivity to certain agents. What agents may be implicated?***

Answers are located at the end of the case.

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SELF-STUDY ANSWERS

1. Any part of the tract, including the mouth
2. Lip swelling and/or cheek swelling, angular cheilits, mucogingivitis, cobblestoning of buccal mucosa, mucosal tags, aphthous ulceration, and long, deep ulcers in the mandibular buccal sulcus are some of the possible orofacial manifestations of CD
3. Presence of non-necrotizing granuloma(s)
4. Anti-inflammatories (glucocorticoids, enteral nutrition, aminosalicylates), immunomodulators, biologics, and antibiotics may be used in management of CD in children
5. Certain food and drink additives such as benzoates, cinnamon and tartrazine