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# ORAL CARE FOR PEOPLE WITH HEMOPHILIA OR A HEREDITARY BLEEDING TENDENCY

# Second edition

# **Crispian Scully**

UCL Eastman Dental Institute London, U.K.

# **Pedro Diz Dios**

University of Santiago de Compostela Spain

# Paul Giangrande

Haemophilia Centre, Churchill Hospital Oxford, U.K.



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World Federation of Hemophilia 1425 René Lévesque Boulevard West, Suite 1010 Montréal, Québec H3G 1T7 CANADA

Tel.: (514) 875-7944

Fax: (514) 875-8916

E-mail: wfh@wfh.org

Internet: www.wfh.org

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Treatment of Hemophilia Monographs Series Editor Dr. Sam Schulman

Correspondence to: Professor Crispian Scully CBE, MD, PhD, MDS, MRCS, FDSRCS, FDSRCPS, FFDRCSI, FDSRCSE, FRCPath, FMedSci, FHEA, DSc, DChD, DMed (HC) Director, UCL Eastman Dental Institute; Co-Director, World Health Organization Collaborating Centre for Oral Health, Disability and Culture, 256 Gray's Inn Road, London, WC1X 8LD, U.K.; Tel: 442079151038; Fax: 442079151039

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# Oral Care for People with Hemophilia or a Hereditary Bleeding Tendency

Crispian Scully, Pedro Diz Dios, Paul Giangrande

# Introduction

Oral disease may affect general health and may, in people with a bleeding tendency, cause serious bleeding. Surgery such as tooth extraction and some local anesthetic injections can cause bleeding that persists for days or weeks and that cannot always be controlled by pressure alone.

Since bleeding after dental treatment may cause severe or even fatal complications, people with hemophilia or congenital bleeding tendencies are a priority group for dental and oral preventive health care. Those with HIV infection may develop other mouth problems, such as infections or ulcers.

Maintaining a healthy mouth and preventing dental problems is thus of great importance, not only to quality of life and nutrition but also to avoid the dangers of surgery. Unfortunately, people with bleeding disorders may neglect oral health for fear of bleeding; this has contributed to a lack of good dental care for some.

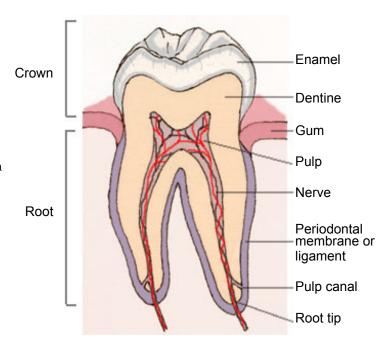
This monograph aims to help people with bleeding tendencies and healthcare professionals achieve good oral health and thus minimize interventions.

# The Healthy Mouth

# The teeth

Teeth are made of a crown of hard enamel surrounding sensitive dentine, and a root which has no enamel covering (Figure 1). Teeth contain a vital pulp (nerve) and are supported by the periodontal membrane, via which the roots are attached into sockets in the alveolar bone of the jaws (maxilla and mandible). The fibers of the periodontal ligament attach through cementum to the dentine surface. The alveolar

bone is covered by the gingivae, or gum. A healthy gum is pink, stippled, tightly bound down, and forms a close fitting cuff around the neck (cervical margin) of each tooth.



**Figure 1.** The first or primary set of teeth (also called deciduous, milk, or baby teeth) includes four incisors, two canines, and four molars in each jaw (total 20 teeth). The normal permanent (adult) set of teeth includes four incisors, two canines, four premolars, and six molars in each jaw (total 32 teeth).

#### **Tooth development**

Tooth development begins in the fetus. At about one month of pregnancy, the tooth crowns begin to form. Indeed, all the deciduous and some of the permanent teeth start developing in the fetus.

The primary teeth have all started to form by birth. Formation of the permanent incisor and first molar teeth begins at, or close to, the time of birth, with formation of other permanent teeth starting later.

Table 1. Average age for tooth eruptions

Deciduous (Primary) Teeth				
		Age (in	months)*	
		Upper	Lower	
A.	Central incisors	8-13	6-10	
В.	Lateral incisors	8-13	10-16	
C.	Canines (cuspids)	16-23	16-23	
D.	First molars	13-19	13-19	
E.	Second molars	25-33	23-31	
Permanent Teeth				
		Age (ir	n years)*	
		Upper	Lower	
1.	Central incisors	7-8	6-7	
2.	Lateral incisors	0 0		
۷-	Lateral Incisors	8-9	7-8	
3.		8-9 11-12	7-8 9-10	
3.	Canines (cuspids)	11-12	9-10	
3.	Canines (cuspids) First premolars	11-12	9-10	
3.	Canines (cuspids) First premolars (bicuspids)	11-12 10-11	9-10 10-12	
3.	Canines (cuspids) First premolars (bicuspids) Second premolars	11-12 10-11	9-10 10-12	
<ul><li>3.</li><li>4.</li><li>5.</li></ul>	Canines (cuspids) First premolars (bicuspids) Second premolars (bicuspids) First molars	11-12 10-11 10-12	9-10 10-12 11-12	

<sup>\*</sup> Ages listed are averages; there is a wide range

#### **Teething**

Tooth eruption (teething) occurs after crown formation is largely complete, but before the tooth roots are fully formed.

Just before primary teeth erupt, the gums may be a bluish colour and swollen, usually because of a transient bleed into the gum, which resolves spontaneously. This has nothing to do with the bleeding tendency but may be more obvious in people with a bleeding disorder than in those who do not bleed excessively.

# Key messages:

- Teeth start to develop *in utero*.
- Root formation finalizes after eruption.
- The primary set of teeth has 20 teeth in total.
- The permanent set of teeth has 32 teeth in total.

Teething rarely causes bleeds, but if minor bleeding or oozing from the gums does occur, contact your hemophilia treatment centre (HTC). Due to the moistness of the mouth, blood clots cannot form as easily and might fall out before a cut or tooth socket is healed. If a baby has persistent mouth bleeds, a product may be prescribed to help clots stay in the mouth.

Teething may cause irritability, disturbed sleep, cheek flushing, drooling, and sometimes a small rise in temperature and/or a rash. It does not cause diarrhea or bronchitis, though these may occur coincidentally.

# Problems during tooth eruption

Some mouth bleeding can occur during infancy, when tooth roots are mobile. Antifibrinolytic agents may be useful in these circumstances. Experts say that baby teeth should be allowed to fall out naturally, without pulling, so less bleeding will occur. If bleeding occurs when a tooth falls out or is extracted by a dentist, the child should bite gently on a gauze pad or moist tea bag. If bleeding continues, contact your HTC.

Nearly half of the permanent teeth appear in the mouth by age 10. Children might experience bleeding or oozing when teeth erupt. Dental professionals advise parents to apply firm, gentle pressure to the area and to use an antifibrinolytic agent if necessary. Afterwards, treat your child to soft, cool foods like yogurt, avoid serving hot foods, and avoid using straws. These simple measures can help preserve the blood clot until the area heals.

The teenage years bring many changes, sometimes including the need for orthodontic braces. Children with hemophilia can wear braces just like everyone else, but the orthodontist must be told about the child's bleeding disorder, so that special care can be taken to avoid cutting or irritating the gums when bands and wires are placed on the teeth. Applying dental wax over rough edges protects gum tissue, cheeks, and lips.

Wisdom teeth, or third molars, usually begin to erupt at age 17. Because these teeth cut through the gums, people with bleeding disorders can

experience prolonged bleeding and might need an antifibrinolytic agent or desmospressin (usually administered nasally). If the teeth are not in the right position or the jaw is too small to accommodate them, they may become impacted and need to be extracted. This procedure should be planned out with a dentist or oral surgeon and the HTC.

# Delays in eruption

A delay in eruption of up to 12 months is rarely alarming and is often caused by local factors, such as the tooth impacting against another tooth. This occurs most often in the third molar (wisdom tooth), premolar, and canine regions, because these are normally the last teeth to erupt. An X-ray is recommended if tooth eruption is delayed for more than one year.

# Key messages:

- Teething may cause irritability, drooling, and a very small rise in body temperature.
- Failed eruption of a single tooth is usually caused by impaction.

# **Problems Affecting Teeth and Gums**

## Tooth damage

Teeth may be damaged by

- decay (dental caries);
- erosion by carbonated drinks (such as cola), fruit juices, or gastric acid regurgitation;
- wearing down by grinding (attrition), or hard brushing (abrasion);
- trauma (such as falls, sports, road accidents, or fights).

# Early tooth loss

Tooth extraction as a result of dental caries or, in adults, periodontal (gum) disease, is the most common cause of early tooth loss. Teeth, particularly incisors, may also be lost through injuries, such as in sports, assaults, falls, or road traffic injuries.

If a permanent tooth is knocked out, pick it up by the crown (avoid touching the roots), rinse it in clean cold water, and, if possible, place it in milk. Hurry to a dentist or emergency room; they might be able to reinsert the tooth. On the way, apply firm pressure to the bleeding site with a piece of clean gauze.

Go directly to the emergency room if bleeding on the tongue, cheek, or floor of the mouth doesn't stop; if the tongue, throat, or neck is swollen or bruised; or if breathing or swallowing is impaired.

# Dental plaque

Caries and inflammatory periodontal (gum) disease, both a result of dental bacterial plaque, are the most prevalent mouth diseases. Plaque is a biofilm containing various germs (microorganisms). Plaque forms on teeth, particularly between them, along the gum margin, and in fissures and pits, and can only be removed by brushing and other oral hygiene aids. Plaque formation can be reduced by some toothpastes and mouthwashes. If plaque is not regularly removed at least each day, it irritates the gums, causing gingivitis, and may become hard, forming tartar (calculus). This cannot be removed by brushing – only by a dentist or hygienist.

#### Key messages:

- Caries and periodontal disease are the most common oral diseases.
- Dental plaque is the underlying cause of both these problems.
- Daily removal of plaque is important.

#### **Dental caries**

Dental caries are caused by the destruction of the enamel and the dentine of the teeth. Sugars, particularly non-milk sugars found in food and drink are the major causes of caries (i.e. they are cariogenic) (see Table 2). Sucrose (cane sugar) and other non-milk sugars like glucose and maltose are the main sugars that cause caries. Concentrated fruit juices and dried fruits also have a high concentration of sugars and are cariogenic, whereas fresh fruits and vegetables are not. Milk sugar (lactose) is less cariogenic than the other sugars.

Dietary starch is changed slowly by saliva enzymes to glucose and maltose. Sugars are changed by plaque bacteria to acids, which cause dental caries. The main plaque bacteria are *Streptococcus mutans*.

Saliva protects against caries: stimulation of saliva, by chewing, for example, reduces caries, while a dry mouth can increase decay. Untreated, caries will cause tooth damage, pain, and eventually, a dental abscess.

The number of caries has been declining for some years in many parts of the world, mainly because of the protective effect of fluoride (see page 7).

# Key messages:

- Caries destroy the enamel and dentine of the teeth.
- Caries are the result of fermentation of sugars by plaque bacteria.

# Gum (gingival) bleeding

Most gum bleeding is due to gingivitis, an inflammation caused by plaque accumulation due to poor oral hygiene. A low platelet count (thrombocytopenia) and von Willebrand disease (VWD) aggravate any bleeding. Since plaque is the main cause, anti-plaque agents and increased tooth brushing are important to minimize gingivitis.

# Key messages:

- Gum bleeding is usually caused by gingivitis, a result of plaque accumulation.
- Increased brushing is needed.
- Gum bleeding is aggravated by von Willebrand disease and thrombocytopenia.

#### Periodontal disease

There are two types of periodontal disease, gingivitis and chronic periodontitis. Gingivitis is an inflammation of the gums caused by the bacteria found in plaque. Without proper oral care, this may progress and damage the attachment of the teeth to the jawbone (periodontal membrane), causing inflammation and damage to this membrane (chronic periodontitis), which can lead to tooth loosening and eventually, tooth loss. Tartar (calculus) may form from hardening plaque (calcification) above and/or below the gum-line, and the plaque that collects on tartar makes the inflammation worse.

Lifestyle factors such as cigarette smoking, and diseases such as HIV infection and diabetes lead to more rapidly progressive gum disease.

An exaggerated inflammatory reaction to plaque during pregnancy can also lead to gingivitis.

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Cariogenic		Non-cariogenic			
Sugars	Mixtures	Fresh fruit or vegetables	Bulk sweeteners	Intense sweeteners	
Dextrose	Brown sugar	Fructose	Hydrogenated glucose	Acesulfame	
Fructose (except in fresh fruits and vegetables)	Golden syrup		Isomalt	Aspartame	
Glucose	Honey		Lactitol	Cyclamate	
Hydrolyzed starch	Maple syrup		Maltitol	Saccharin	
Invert sugar	Treacle		Mannitol	Thaumatin	
Maltose			Sorbitol		
Sucrose			Xylitol		

Pregnancy-associated gingivitis usually develops around the second month and reaches a peak in the eighth month.

# Key messages:

- Plaque can cause inflammation of the gums (gingivitis).
- Involvement of the underlying tissues causes periodontitis.
- Periodontitis can lead to tooth loss.

Gingivitis is painless but may lead to bleeding of the gums, particularly during brushing. Chronic periodontitis (pyorrhoea) is typically seen in adults. It is painless but may be associated with bleeding, bad breath (halitosis), and a foul taste in the mouth, and there may be increasing tooth looseness. Gum disease may also be linked to a range of disease conditions including atherosclerosis, hypertension, coronary heart disease, cerebrovascular disease, diabetes, and low birth weight.

# **Tooth staining**

Superficial tooth discolouration is usually caused by poor oral cleaning, or habits such as smoking, betel, foods, beverages (e.g. tea), or medicines such as iron, chlorhexidine, and long-term oral antimicrobials.

# **Tooth hypersensitivity**

Tooth hypersensitivity is often a result of abrasion from over-enthusiastic brushing. Exposing the tooth to cold air, water, or fruit drinks can cause pain. A dentist should be consulted to ensure there are no cavities and, eventually, for treatment. Use of a good toothbrush coupled with an effective cleaning method minimizes the risk of tooth hypersensitivity.

The treatment of tooth hypersensitivity includes:

- Modification of brushing technique to ensure the gums are not damaged
- Application of desensitizing agents
- Daily use of a fluoride mouthwash
- Regular use of a desensitizing toothpaste

#### **Halitosis**

A degree of halitosis (bad breath) is common in healthy individuals, particularly after sleep (morning breath). Halitosis usually comes from the tongue coating or gum crevice. The bad odour is caused by volatile sulfide compounds released by plaque bacteria.

Individuals who have bad oral hygiene soon develop halitosis, but it is made worse by any form of oral infection, including:

- gingivitis
- periodontitis
- dental abscess
- dry (infected) extraction socket
- sinusitis
- tonsillitis

Many foods and drinks can also cause halitosis, most obviously garlic, onions, curries, the tropical fruit durian, etc. Smoking and drugs including alcohol, isosorbide dinitrate, disulphiram, and others may also be implicated.

Rare causes include other problems such as:

- sinusitis, nasal or tonsillar infections
- diabetes
- lung problems
- · kidney disease
- liver disease
- psychiatric disease

# Key messages:

- Most halitosis arises from the mouth.
- Poor oral hygiene and infections are the main causes.
- Improved oral hygiene is the most effective way to control it.

## **Prevention of Dental Problems**

Families dealing with hemophilia have so much on their plate that dental care can often fall to the bottom of the list. However, it needs to be given a much higher priority.

The most important objective is the prevention of complications. Patients and their relatives must be informed about the importance of good oral hygiene in order to avoid the need for invasive dental care and reduce the number of visits to the dentist. That being said, regular visits mean you will deal with small issues rather than large ones (emergency root canals or extractions).

Caries, periodontal disease, and halitosis are largely preventable, and this is very important for people with bleeding tendencies. As outlined in Table 3, dental problems can be prevented by:

- Reducing the frequency and amount of sugars in the diet
- Avoiding smoking
- Using fluoride
- Practicing regular oral hygiene (at least twice daily)

#### Diet

Refined carbohydrates and sugars, particularly non-milk sugars in items other than fresh fruits and vegetables, are the major causes of caries. The frequency with which you consume these sugars is more important than the amount. Frequent intake of sticky, sugary foods between meals leads to increased dental decay. Thus, to

lessen the development of dental plaque and caries, it is important to limit the consumption of sugary foods and to reduce the frequency, such as by restricting them to meal times.

Drinks other than milk and water should not be given to children in feeding bottles and should be given only at main meals. Children should be introduced to a cup at about six months and should have stopped using bottles by the age of one year.

Foods used to wean children should be free of or very low in sugars other than those in fresh milk and raw fruits or vegetables.

For older children and adults, snack foods and drinks especially should be free of sugars. Because of the risk of erosion as well as of caries, frequent consumption of carbonated and cola type drinks should be discouraged. Fruit juices such as grapefruit, apple, or orange can also cause tooth erosion. For children, water and milk are the best options.

# Table 3. The four main ways to maintain oral health

# Diet: reduce the consumption and especially the frequency of intake of sugar-containing food and drink.

- Sugar-containing food and drink should be consumed as part of a meal.
- Snacks and drinks should be free of sugars.
- Avoid the frequent consumption of acidic drinks.

# Tooth cleansing: brush and floss the teeth thoroughly twice every day with a fluoride toothpaste.

- Effective plaque removal is essential to prevent periodontal disease.
- Brushing alone cannot prevent dental decay, but fluoride toothpastes offer major benefits.
- Other aids to remove plaque should be discussed with your dentist.

#### Fluoridation:

- Request the local water company to supply water with the optimum fluoride level. Fluoridation is a safe, equitable, and highly effective public health measure.
- Consider use of fluoride supplements for children at high risk and living in areas without water fluoridation.

# Regular visits to the dentist: have an oral examination every year.

• Children, people at special risk of oral disease (such as people with a lack of saliva), and those for whom oral disease can be a particular risk to their health (such as people with bleeding problems) may need to be examined more frequently.

Adapted from: The Scientific Basis of Dental Health Education, Health Education Authority, 1996

Chewing sugar-free gum or cheese after meals may help, since they increase the amount of saliva in the mouth, which may protect against plaque acids.

Sugar-free medications should always be used.

# Key messages:

- Sucrose and refined carbohydrates are the main causes of caries.
- The frequency of exposure to these is more important than the total amount consumed.

#### Fluoride

Fluoride protects teeth against caries.

#### Water fluoridation

Water fluoridation has consistently been shown to be the most effective, safe, and fair means of preventing caries, resulting in a decrease of approximately 50%.

# Fluoride supplements

Where the water supply contains less than 300 micrograms/litre ( $\mu g/L$ ), or 0.3 parts per million (ppm) of fluoride, infants and children over the age of six months who are at high risk from caries may be given fluoride supplements daily. Supplements are available as drops or tablets, and are given according to the regimen outlined in Table 4. No fluoride supplements are required if the water supply contains more than 700  $\mu g$  /L (0.7 ppm) of fluoride. Fluoride

supplements should be stored out of reach of children. While it is safe for pregnant women to take fluoride supplements, they have little impact on the future dental health of the child.

#### Fluoride toothpastes

Since the 1970s, fluoride has been added to toothpastes, which is largely responsible for the decline in caries. Brushing at least twice a day using a small-headed, medium hardness toothbrush and a fluoride toothpaste will help prevent caries. Fluoride toothpastes are usually available in one of three strengths (Table 5). Children under about six years of age may swallow toothpaste, so only a pea-sized amount of a toothpaste with less than 1000 ppm of fluoride should be used, and brushing should be supervised.

#### Fluoride mouthwashes or gels

Fluoride mouthwashes and gels are particularly recommended for people who have a dry mouth (xerostomia), as they may be particularly at risk of dental decay. Fluoride mouthwashes can be used on a daily or weekly basis and may be used in addition to fluoride-containing toothpastes.

#### Key messages:

- Fluoride helps prevent caries.
- Most toothpastes contain fluoride.
- Fluoride mouthwashes help protect erupted teeth.

Table 4. Fluoride supplement doses to reduce caries in children at high risk
(in relation to water fluoride content)

Fluoride in water supply* (ppm–parts per million)	Up to 6 months	6 months to 3 years	3 to 6 years	Over 6 years
less than 0.3	-	250 μg daily**	500 μg daily**	1 gram (g) daily**
0.3-0.7	-	-	250 μg daily**	500 μg daily**
over 0.7	-	-	-	-

<sup>\*</sup> The local District Dental Officer or equivalent or the water company should be able to help with this information.

<sup>\*\*</sup> Amount of fluoride found in twice the dose of sodium fluoride.

Content of fluoride in toothpaste	Fluoride content ppm (approx.)	Comments
Low	<600	Useful mainly in children with low risk of caries who live in areas with fluoridated water or receive fluoride supplements
Medium	1000	Useful for all over 6 years of age
High	1500	Useful for people at high risk of caries over 6 years of age

Table 5. Fluoride toothpastes

# **Fissure sealants**

Plastic resin coatings placed by a dentist or dental hygienist in the pits and fissures of permanent teeth can also help reduce caries.

# Oral hygiene

Oral hygiene measures that remove plaque can prevent gingivitis, periodontitis, and halitosis. Teeth should be brushed at least twice each day. Oral hygiene is particularly important in view of the current opinion that periodontitis may affect general health.

Some people with hemophilia fear that brushing and flossing will result in mouth bleeds. However, brushing and flossing are important for everyone, and neglecting these basics has a more profound impact on people with hemophilia than on anyone else. Healthy gums do not generally bleed during brushing and flossing, even in the person with hemophilia. The only time some bleeding might occur is if you are overly aggressive with the toothbrush when brushing.

Bleeding gums are a sign of dental disease. If bleeding continues for more than 20 minutes, or stops and then starts again, contact your HTC. Treating with factor concentrate or other therapies may temporarily stop the bleeding, but not the disease.

Toothbrushes only remove plaque from smooth surfaces of teeth and not from the depths of pits and fissures; removal of plaque from between teeth requires regular flossing (some dental floss also contains fluoride).

#### *Toothbrushes*

The most important oral hygiene device is a toothbrush. Hard brushes are not advisable as they can wear down the teeth and gums and may lead to tooth hypersensitivity. Brushes that are too soft will not effectively remove plaque and debris, and are only recommended when there is extreme tooth hypersensitivity.

The ideal toothbrush should have synthetic bristles of an even length and of medium hardness (about 0.15-0.2 mm diameter bristles). It should be small enough to be easily placed in the mouth and yet suitably designed to effectively remove all the dental plaque – a toothbrush head of about 2-3 cm length and 1 cm width is usually sufficient for adults. For children, a brush of 2 cm by 1 cm is suitable.

A variety of toothbrushes are available, including:

- Angled brushes to help access areas of the mouth that are difficult to reach.
- Altered bristle length brushes the middle row of bristles is shorter than the outer rows. These brushes clean above and below the gum without causing overbrushing and are excellent for patients with generally healthy mouths.
- Easy-grip brushes are particularly useful for people who are too weak to grip closely or firmly (e.g. individuals with arthritis).
   The toothbrush can be enlarged by fixing a ball of sponge rubber, a nail brush, or bicycle handlebar grip to the handle.
- Extended-handle brushes are particularly effective for patients who cannot lift their arms.
- Electric toothbrushes are increasingly popular and often more effective at removing plaque. They are often light and easy to hold and are ideal for people with limited manual dexterity.
- Interdental brushes help clean between teeth and underneath bridges.

# Brushing techniques

The ideal brushing technique should remove plaque without damaging the teeth or gums. The most effective techniques are:

#### • The roll technique:

Particularly useful for people with healthy gums. The brush is placed with the bristles on the gum, then pressed onto the gums to make the bristles spread. Maintaining the same pressure, the bristles are moved across the gums onto the tooth surface. Behind the front teeth, the brush is held vertically and pulled upwards or downwards.

# • The Bass technique:

Particularly useful for patients with gum disease, the bristles are placed on the gum such that they point away from the crown of the tooth at a 45-degree angle. The brush is vibrated backwards and forwards with a horizontal scrubbing movement. This method can be time consuming, difficult to master, and may cause mild trauma if not done properly with the correct brush.

Few children develop sufficient manual dexterity to clean teeth effectively until about six years of age. Parents should therefore brush their teeth. This is best achieved by standing behind the child, and tilting the child's head back so that the teeth can be brushed.

#### *Toothpastes (dentifrices)*

Toothpastes typically help remove and prevent the formation of dental plaque, and provide a pleasant-tasting mouth and fresh breath. They can deliver fluoride, antiseptics, and desensitizing agents. Toothpastes should not be swallowed.

- Fluorides contained in most toothpastes cause a hardening of the enamel and protect against caries. As fluoride has no notable harmful effects, people with bleeding tendencies are advised to use a fluoridecontaining toothpaste.
- Toothpastes containing triclosan and chlorhexidine have anti-plaque activity and have been proven to protect against periodontitis without harmful reactions.
   Products containing phosphates and

pyrophosphates may help prevent tartar, but some have produced unfavourable gum reactions.

• Many "luxury" toothpastes claim to whiten teeth, but few have supporting evidence.

#### Mouthwashes

Mouthwashes are a controversial issue. Most effective anti-plaque mouthwashes can be safely used for up to one month. They include:

- Chlorhexidine helps control plaque and periodontal disease, but it can affect taste briefly. Also, it binds to tannins found in coffee, tea or red wine, and can therefore contribute to dental staining. These stains can be cleaned off by dental professionals.
- Listerine® contains essential oils which help reduce plaque. It does not stain teeth, but it contains alcohol.
- Triclosan also significantly reduces plaque.

#### Other aids to tooth cleaning

Everyone except people with severe bleeding disorders should use interdental cleaning devices (dental floss and dental tape) to remove plaque that remains between the teeth after brushing. The floss or tape should be threaded between the teeth and gently curled around the side of the tooth, slid down to the gums, and gently brought back up to the top of the tooth. It can be difficult to use and can damage the gums, so it may be best to use a dental floss holder, such as a Flossette.

Disclosing tablets or rinses, which are used after tooth cleaning, stain dental plaque, thus showing where plaque has been left behind.

#### Key messages:

- Good oral hygiene is essential to prevent gingival and periodontal disease.
- Brushing AT LEAST twice daily is required for plaque control.
- Toothpaste containing fluoride should be used.
- Mouthwashes containing triclosan or chlorhexidine can also help reduce plaque.
- Dental floss and interdental brushes help reduce plaque.

# Mouth protection

People with hemophilia should always wear mouth guards when they play sports, especially contact sports. Soft plastic mouth guards or splints, may be needed to prevent damage from trauma, tooth grinding (bruxism), acid erosion, or radiation during radiotherapy.

#### Risk factors for oral disease

Lifestyle may significantly affect oral health. Smoking and tobacco use may contribute to periodontal disease and oral cancer, and some chewing tobacco, candies and even medications containing sugars may increase the risk of caries.

# Dental Treatment for People with Bleeding Disorders

Informing your dentist that you have hemophilia before beginning any procedure is the first step. In order to help your dentist plan the correct course of treatment, you should also provide the following information:

- The type and severity of your hemophilia
- The medications you take
- Whether pre-treatment with factor concentrate, nasal desmopressin or an antifibrinolytic agent (tranexamic acid or epsilon amino caproic acid) is required
- Your HTC contact information
- Whether you have:
  - An inhibitor
  - An infectious disease, such as hepatitis
  - o A joint replacement
  - A venous access device (port)
- People with bleeding disorders need close cooperation between their physician and their dentist to receive safe, comprehensive dental care.
- Dental appointments of children with bleeding disorders, as well as education in preventive dentistry for children and caregivers, should be started when the baby teeth begin to erupt.
- Deep injections, surgical procedures particularly those involving bone (extractions, dental implants) – and regional

- local anesthetic blocks should be avoided, where possible, as they may start a bleeding crisis
- Comprehensive dental assessment is needed at the age of about 12 or 13, to plan for the future and to decide how best to forestall difficulties resulting from overcrowding or misplaced third molars or other teeth.
- For people with mild or moderate hemophilia, non-surgical dental treatment can be carried out under antifibrinolytic cover, but a hematologist must be consulted before other procedures are done.
- For people with mild hemophilia A (factor VIII >10%) and most people with VWD (type 1), scaling and some minor surgery may be possible under desmopressin (DDAVP) cover. However, DDAVP is not effective in hemophilia B (even mild cases) as it does not boost factor IX levels.
- For those with severe hemophilia, factor replacement is necessary before scaling, surgery or regional block injections. For example, a dose of 50 international units per kilogram of body weight (IU/kg) of factor VIII is desirable before a tooth extraction in a person with hemophilia A; in hemophilia B, a dose of 100 IU/kg of factor IX is required.
- Local use of fibrin glue and swish-andswallow rinses of tranexamic acid before and after dental extractions are safe and cost-effective methods to help control bleeding.
- Tranexamic acid used topically significantly reduces bleeding following dental treatment. A mouthrinse of 10 mL of a 5% solution used for two minutes, four times daily for seven days, is recommended. It may be used in combination with oral tranexamic acid tablets for up to five days.
- Bleeding can be aggravated by painkillers (analgesics) such as Aspirin® and other nonsteroidal anti-inflammatory drugs such

as indometacin. Codeine and paracetamol (acetominophen) are safer alternative analgesics.

- After tooth extraction, a diet of cool liquid and minced solids should be followed for five to ten days.
- Any swelling, difficulty swallowing (dysphagia), or hoarseness must always be reported to the doctor immediately.
- People with hemophilia with injuries to the head and neck are at risk from bleeding into the brain or neck and should, therefore, be given factor replacement.
- Your doctor may recommend that you take antibiotics before any invasive dental procedures if you have had a joint replaced or venous access device.
- Screening for factor VIII inhibitors is necessary before any invasive procedure, including dental work. For patients with inhibitors, treatment with recombinant factor VIIa or FEIBA® may be required.
- Blood-borne infections should not influence access to dental care.
- HIV has been an issue in the hemophilia community. It can cause mouth problems, particularly infections such as candidosis and ulcers. Treatment can be complicated by thrombocytopenia, which can worsen the bleeding tendency.
- Hepatitis C is extremely common in people with hemophilia and can be associated with a prolonged prothrombin time or INR (International Normalized Ratio) and thrombocytopenia. In such cases, bleeding cannot be prevented with factor VIII (or IX); fresh frozen plasma may be required.

# Conclusion

People with hemophilia or congenital bleeding tendencies are a priority group for dental and oral health care, since bleeding after dental treatment may cause severe or even fatal complications. Maintenance of a healthy mouth and prevention of dental problems is of great importance, not only to quality of life and nutrition but also to avoid the dangers of surgery.

# **Further Reading**

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