

**Oral health
and
congenital heart disease**



Photo: taken by Maryann Fløttkjær

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Preface

Congenital heart disease is one of the most common developmental anomalies. Due to advances in pediatric cardiac surgery and modern imaging techniques most of these patients now survive into adulthood. The dental practitioner may now regularly encounter children and adults with congenital cardiac defects and should therefore possess up-to-date knowledge about the consequences of the disease regarding oral health and how to manage this patient population. Previous studies have revealed that many dentists are not confident in treating this group of patients, possibly because of lack of experience and knowledge about this “new” medical group.

This master thesis aims to raise dentists’ awareness of the importance of good oral health among these patients and provide them with up-to-date knowledge about the impact of cardiac medicines and dietary modifications on their oral health status. The latest international guidelines regarding antibiotic prophylaxis will be presented. Other aspects like orthodontic treatment, the use of sedation and local anesthetics, social insurance benefits and syndromes associated with congenital heart disease will also be discussed.

Dentist Audun Rui and cardiologist Per Lunde (UNN) have both contributed with their knowledge and understanding about this group of patients. I would like to give them a special thanks for helping me write about this important subject.

I would also like to thank Marte A. Jystad (FFHB) for inspiring me to write about her “heart patients” and for helping me getting in contact with the right sources.

Last but not least I would like to thank my tutors, Tiril Willumsen and Hilde Nordgarden, for their guidance and for encouraging me to write about this topic.

Historical perspective

Congenital heart disease used to carry a very poor prognosis. Before pediatric cardiac surgery began (in 1938) (Fisher, Hardy and Widmann 2005), most of the children with severe CHD died during their first years of life (Hoffman, Kaplan and Liberthson 2004).

In the last fifty years, innovative cardiac surgery techniques and modern diagnostic tools (e.g., ultrasound) have resulted in the survival of the majority of infants born with congenital heart disease (Warnes 2009). As a result of this, the number of adults with CHD has grown rapidly (Hoffman, Kaplan and Liberthson 2004). Most of these patients have lifelong medical needs.

According to Sunnegårdh (2000), knowledge of the cardiac anatomy was first based on findings from the autopsy of human beings. The first descriptions of congenital heart defects were tetralogy of Fallot (Stensen in 1673), transposition of the great arteries (Baillie in 1797) and ventricular septal defect (Roger in 1879).

The first congenital heart defect corrective surgeries were carried out during the 1940's, mostly on older children and adults. Around 1970, successful cardiac surgery procedures on infants were performed. Since the 1980's surgical repair of most congenital heart defects have reached high standards even though much research remains to further improve the surgical techniques and treatment options (Hoffman, Kaplan and Liberthson 2004).

When conventional surgery was not possible, in the 1980s heart transplantation followed by the use of broad immunosuppressive therapy (cyclosporine) in children and young people become a realistic treatment option (Sunnegårdh 2000).

Method

This master thesis is a literature review based on several studies and scientific literature concerning patients with congenital heart disease.

Most of my research is carried out by using PubMed, a database where one can find several references and abstracts on life sciences and biomedical topics.

At the beginning of my work I used the search query “congenital heart disease and oral health” to find articles related to this topic. I limited my search to articles published the last 10 years. Gradually I started to search for specific themes by using the keywords “congenital heart disease” with “nutrition”, “dental treatment”, “dental hygiene”, “syndromes”, “anticoagulation”, “infective endocarditis”, etc.

As I read through several articles, I checked out their reference lists and studied some of them further, explaining the large list of references used in my thesis.

I have also read several books concerning this issue. At first, I went to the Medical Library at the University Hospital in Oslo to find scientific literature regarding the general aspects of congenital heart disease. After achieving a general medical and historical picture of this congenital condition, I went to the Dentistry Library at the Faculty of Dentistry in Oslo and searched for books related to oral health. Some of these books are published before 2007, the year when major changes in the guidelines of antibiotic prophylaxis were undertaken. This is very important to be aware of and is also a topic of discussion in my thesis.

It should be emphasized that I have tried to use references which are not already outdated, because of the rapid medical and dental “evolution”.

General web searches on the web search engine Google were also performed, but not all the “articles” I found there could be trusted. Therefore, only some of that material is used in my thesis.

I also used some of the dental lectures given to me at the Faculty of Dentistry in Oslo.

I had personal communication with a cardiologist named Per Lunde, who works at the University Hospital of Northern Norway and with Audun Rui, a former dentist with long experience from clinical work in dentistry, now working in the Norwegian seamen's church in Copenhagen. Cardiologist Per Lunde gave me advices on how to perform adequate searches on this topic on PubMed and he also updated me on the different medications used by this patient population. Audun Rui presented me his brochure, which he wrote several years ago in cooperation with the Norwegian Heart Association for children, concerning congenital heart disease and oral health. I compared his brochure to a new pamphlet newly published by this same association.

As well, my personal experience with a congenital heart condition has contributed to my master thesis. It has helped me understand the different aspects of the disease, not only the dental aspects, but also the psychological and medical ones.

Definitions

Oral health

General health: "A state of complete physical, mental and social well being, not merely the absence of disease or infirmity." (WHO 1946)

According to Poulsen and Koch (2009), oral health should be considered a part of general health and therefore the term "oral health" can be defined as:

"A state of sound and well-functioning dental and oral structures as well as absence of dental fear and anxiety" (Poulsen and Koch 2009). Both somatic and non-somatic aspects are included in this definition.

Good oral health implies:

- Absence of pain and discomfort
- Absence of specific dental diseases (e.g., caries, periodontitis)
- The capability to eat, drink and speak
- Acceptable aesthetic appearance

When any of these criteria are affected one may define the condition as an oral disease process (Griffiths and Boyle 2005 a).

Congenital heart disease

“Congenital heart defect” is defined by Mitchell et al. (1971) as **“a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance”** (Mitchell, Korones and Berendes 1971).

The term “congenital” means “present at birth” (con “together”; genitus “born”), but according to Perloff (2003), one should consider congenital heart defect as a dynamic defect that “originate in the early embryo (the heart is the first organ to form in vertebrate embryos), evolve during gestation, and change considerably during the course of extrauterine life.”

ASA Physical Status Classification System (American Society of Anaesthesiologists, 1963)

Patients are classified in different risk categories helping the general practitioner providing safer dental treatment and better meet the patient’s limit of tolerance. The system “estimates” the medical risk presented by a patient undergoing a surgical procedure. This is very important whenever management of pain (e.g., local anesthetic) or anxiety (e.g., CNS depressant) is planned (Malamed 2009 a).

According to Malamed (2009 a) the classification system has been used for all surgical patients regardless of anesthetic technique (e.g., general anesthesia, regional anesthesia, sedation).

The classification system follows:

ASA Physical Status 1 - A normal healthy patient

ASA Physical Status 2 - A patient with mild systemic disease without limitations of daily activities

ASA Physical Status 3 - A patient with severe systemic disease that limits activity but is not incapacitating

ASA Physical Status 4 - A patient with severe systemic disease that is a constant threat to life

ASA Physical Status 5 - A moribund patient who is not expected to survive 24 hours without the operation

ASA Physical Status 6 - A declared brain-dead patient whose organs are being removed for donor purposes

Source: Malamed, 2009 a

(ASA 5 and 6, not for dental use)

Patients with CHD may represent ASA 2, 3, or 4 risks. The dentist can recommend medical consultation to judge the degree of disability present.

Syndromes associated with congenital heart disease

Approximately 20% of congenital heart defects are associated with syndromes and other extracardiac malformations (Meberg et al. 2000, Gatzoulis et al. 2005 a).

Examples of syndromes associated with congenital heart disease are (Sunnegårdh 2000, Meberg et al. 2000, Smith 2001, Gatzoulis 2005 a, Fitzgerald et al. 2010):

- **Trisomy 21:** the association between trisomy 21 and CHD is well acknowledged (Meberg et al. 2000). AVSD and tetralogy of Fallot are the

most common lesions (Gatzoulis 2005 a). Approximately 40% have a congenital heart disease (Sunnegårdh, 2000).

Oral manifestations (Griffiths and Boyle 2005 b):

- Periodontal disease (Lopez-Perez et al. 2002)
- Lack of central incisors in the lower jaw and the lateral incisors in the upper jaw, (Russell and Kjær 1995)
- Missing teeth/supernumerary teeth
- Microdontia
- Abnormal shaped crowns of teeth
- Cleft lip and palate
- Malocclusions
- Abnormal jaw relationships
- Reduced masticatory ability
- Mouth breathing
- Macroglossia
- High arched palate
- Agenesis of the parotid glands (Ferguson and Ponnambalam 2005)
- Gingival hyperplasia from anticonvulsant medication.

According to Griffiths and Boyle (2005 b), they have difficulties in maintaining an adequate oral hygiene due to anatomical factors.

➤ **DiGeorges syndrome:** deletion of chromosome 22 (22q11 deletion).

Around 15% of patients with tetralogy of Fallot have this deletion (Gatzoulis et al. 2005 a, Srivastava 2006).

A majority of the children with this genetic defect have various forms of heart failure. Many of them also have cleft lip and palate and malfunction of the palate-, pharynx- and oral motor function. Speech and language development is often delayed.

- **Williams syndrome:** caused by the deletion of chromosome 7q11.23 and is associated with cardiac, neurodevelopmental and multisystem abnormalities (Gatzoulis et al. 2005 a). Approximately 60 percent have a congenital heart defect (Frambu 2009).

It is characterized by distinctive facial features and varying degrees of mental retardation, abnormal oral function, sucking-, swallowing- and eating difficulties, drooling, deviations in teeth / tooth position, microdontia, agenesis and enamel hypoplasia (Axelsson 2005).

- **Turner syndrome:** about 30% of patients with this genetic defect have a congenital heart defect (Sunnegårdh 2000). According to Szilágyi et al. (2000), they have significantly less decayed, missing, and filled compared to patients without Turner syndrome, but they have significantly higher plaque and gingival indices. Patients with Turner syndrome have more orthodontic anomalies and smaller tooth crowns. Their alveolar arch of the maxilla is narrower and the mandibular arch is shorter and broader than in healthy patients.

- **Marfan syndrome :** a genetic disorder of the connective tissue that may affect the aorta and the heart valves.

Oral aspects: high arched palate, malocclusion, long small teeth, mandibular prognathism, temporomandibular joint disorders.

According to DeCoster et al. (2002), local hypoplastic enamel spots, root deformity, abnormal pulp shape, and pulpal inclusions are more frequent in patients with Marfan syndrome. DeCoster et al. (2002) also found that calculus and gingival indices were significantly higher in patients with this genetic defect.

- **Noonan's syndrome:** a congenital disorder in which congenital heart disease is one of the principal features. It is characterized by mental retardation, dysmorphic facial features, coagulation defects (Ortega et al. 2008).
Oral manifestations: bleeding disorders, malocclusions, high arched palate and micrognathia (Nunn, 2011).

Patients with syndromes may have social and learning disabilities which can make the delivery and acceptance of dental care more complicated (Foster and Fitzgerald 2005).

Studies have showed that this group of patients has increase levels of caries and untreated caries (Nunn 1987, Urquart and Blinkhorn 1990).

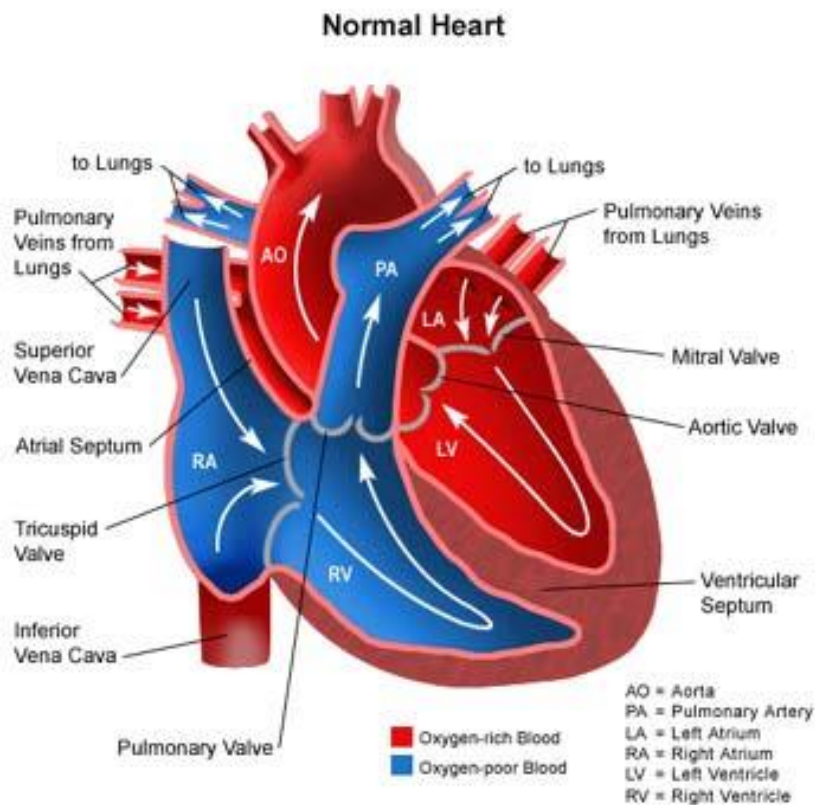
According to Axelsson (2005), an interdisciplinary approach is required, preferably by experienced health- and dental staff with knowledge of the oral characteristics of these syndromes.

Speech therapists, nutritionists and physiotherapists should also be included in this cooperation.

Challenges for the dentist are:

Priorities
Interdisciplinary collaboration
Oral medical knowledge
Communication ability
Empathy
Knowledge of social insurance benefits

Normal heart anatomy



Source: <http://www.lpch.org/DiseaseHealthInfo/HealthLibrary/hrnewborn/0329-pop.html>

The normal heart has two sides, the left and the right, which are separated by a muscular wall called the septum. Each side of the heart also has two parts - an upper chamber called an atrium and a lower chamber called a ventricle.

The cardiovascular system

The cardiovascular system is composed of the heart and blood vessels, including arteries, veins, and capillaries. It includes two circulatory systems: the **pulmonary circulation**, which is a short “loop” from the heart to the lungs and back again where blood is oxygenated, and the **systemic circulation** which sends blood from the heart to the rest of the body to provide oxygenated blood.

Oxygen deprived blood enters the right atrium of the heart from the vena cava and flows through the tricuspid valve into the right ventricle, from which it is pumped through the pulmonary valve into the pulmonary arteries which go to the lungs. Oxygenated blood is then returned to the left atrium of the heart through the pulmonary veins, before flowing through the mitral valve into the left ventricle. There the oxygen-rich blood from is pumped out via the aorta, and on to the rest of the body (Wikipedia 2011).

Congenital heart disease

According to Thaulow (2009), nearly 500 children are born with CHD every year in Norway and 15000-20000 Norwegian adults are living with CHD today. More than 1.8 million adults are living with congenital heart defects around the world (Adult Congenital Heart Association 2010).

The live birth incidence of CHD is around 7-8 per 1000 in Western countries, but this estimate varies between different studies (Mitchell et al. 1971, Dahlgren et al. 1987).

Heart disease in children is usually congenital. About 60% of all CHD is diagnosed in infants, 30% in children and 10% in adults (over 16 years of age) (Gatzoulis et al. 2005 a). Because of increased survival of these children into adulthood (Rosenkratz 1993), there are now more adults than children diagnosed with CHD (Wren and O'Sullivan 2001).

What is a congenital heart defect? A congenital heart defect (CHD) is a deficiency in the heart's structure and great vessels which is present at birth. The defects may only be simple "holes" between the chambers of the heart, but there can also be severe deformations, such as complete absence of chambers or valves. The defects interfere with blood flow in the heart or vessels near it, obstructing it or causing the blood to flow through the heart in an abnormal pattern.

According to Hallett et al. (2008) and Weddell et al. (2011), congenital heart disease is often divided into two types:

- **Cyanotic** (bluish-grey discoloration of the skin caused by a lack of oxygen in the body). Cyanotic conditions are characterized by deoxygenated hemoglobin in the blood stream (oxygen desaturation). Infants with cyanosis may become blue during crying or physical exertion (“blue children”); Examples of such defects include persistent truncus arteriosus, tetralogy of Fallot, transposition of the great vessels, pulmonary atresia, hypoplastic left heart, Eisenmenger’s syndrome.
- **Non-cyanotic.** This group is characterized by a connection between the systemic and pulmonary circulations causing shunting of blood within the heart or stenosis (constriction) of either circulation. These defects include ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), aortic stenosis, pulmonic stenosis, coarctation of the aorta.

The 8 most common structural CHDs account for over 80% of the total (Gatzoulis et al. 2005 a, Dahllöf and Martens 2009).

They are:

VSD 30%
ASD 10%
Patent ductus 10%
Pulmonary stenosis 7%
Coarctation 7%
Aortic stenosis 6%
Tetralogy of Fallot 6%
Transposition of the great arteries 4%
Other 20%

These problems may occur alone or together.

CAUSES: The cause of congenital heart disease is rarely identified and may be either genetic or environmental. In general it is a combination of both (Hoffman 2005).

The known causes of congenital heart disease are frequently sporadic genetic changes with trisomy 21 being the most common genetic cause (Schoen and Mitchell 2010).

It also seems that inheritance may play a role. If a parent or a sibling has a CHD, there is a greater risk of having a child with a heart defect.

Environmental factors during pregnancy include: maternal infections (Rubella), drugs (alcohol, retinoic acid for acne, Warfarin sodium (Coumadin), Thalidomide ingestion, lithium therapy) and maternal illness (diabetes mellitus, phenylketonuria, and systemic lupus erythematosus) (American Heart Association 2011, Wikipedia 2011).

SYMPTOMS: While congenital heart disease is present at birth, the symptoms may not be immediately obvious. Many congenital heart defects have few or no symptoms, but severe defects can cause signs and symptoms, especially in newborns. These signs and symptoms may include: rapid breathing, cyanosis, tiredness, poor blood circulation, heart murmurs (abnormal blood flow through the heart) (National Heart Lung and Blood Institute 2011).

TREATMENT: Some small CHDs do not demand any treatment, but in most of the time CHD is serious and requires surgery and/or medications. Most of these patients require life-long specialized cardiac care and it is estimated that 25% of children born with a CHD will grow up into adulthood with persistent non-operated defects, residual defects or cardiac sequelae after therapeutic procedures (Meberg et al. 2000).

This increase in survival rates leads to “an increased burden of complexity” when managing these patients dental health (FitzGerald et al. 2010).

Nutrition

Malnutrition is common in children with CHD (Moynihan 2006, Vaidyanathan et al. 2008). The causes are multifactorial and may include inadequate energy intake, hypermetabolism, impaired absorption (Smith 2001), difficulty feeding, nausea, vomiting, breathing problems or fatigue (Stecksén-Blicks et al. 2004, Moynihan 2006). Gastroesophageal reflux can also be a problem among CHD patients and may affect growth. However, increased energy requirements and decreased energy are the major issues (Smith 2001). In CHD patients, the

heart needs to pump more to distribute enough oxygen and this requires more energy (Petersen 2011).

Many have decreased appetite and food intake due to pain, surgery, fever, infection, psychological distress, medications, treatment (Roth-Isigkeit et al. 2005, Turkel and Pao 2007, Moursi et al. 2010) and frequent hospitalizations (Rocha et al. 2006). Congenital heart disease may result in a hypermetabolic state, resulting in high energy demands and weight loss (Moursi et al. 2010). Higher energy demands require frequent meals, more snacks and energy-dense supplements (Rocha et al. 2006).

Medical treatment

The most common congenital heart defects do not require any kind of medication after they've been repaired by surgery, but the more complicated the defects are, the higher the risk of rest defects and thereby symptoms that require medications. These patients usually have two major problems: heart failure and arrhythmias.

Some will also have high blood pressure (coarctation of the aorta) that requires medications, and others need anticoagulation / antithrombotic therapy.

The most common medications used by patients with CHD (American Heart Association 2007, Hægermark 2010, Felleskatalogen 2011)

Diuretics: if the heart's pumping ability is impaired, too much water will be held back and blood volume increases. This increases the heart load further. Diuretics counteract this effect and therefore relieve the heart. Today these patients usually apply furosemide (Diural®, Lasix®, Furix®), often supplemented with spironolactone (Spirix®, Aldactone®) (Hægermark 2010)

Oral effects: Have been reported to increase the risk of caries (Rosén and Stecksén-Blicks 2007) and have also been associated with hyposalivation.

ACE inhibitors (angiotensin-converting-enzyme inhibitors): the treatment of choice for heart failure (according to the American Heart Association). ACE inhibitors cause your blood

vessels to expand and reduce peripheral vascular resistance. In addition, ACE inhibitors will stop your body from making angiotensin, a protein that causes blood vessels to constrict.

Captopril (Capoten®) and Enalapril (Renitec®) are two similar drugs belonging to this family and they both relieve the heart (Hægermark 2010). Some of the side effects of ACE inhibitors are changes in your ability to taste, dry cough and generalized swelling.

Angiotensin II receptors antagonists (antihypertensive medications): Angiotensin II causes blood vessels to constrict and cause high blood pressure. This group of drugs blocks the action of angiotensin II, permitting the blood vessels to relax and dilate, which lowers the blood pressure. They have effects similar to those of ACE inhibitors and are often used when ACE inhibitors cannot be tolerated by patients.

Antiarrhythmics: drugs which regulate heart rate in different ways. These drugs include: digoxin, beta-blockers (inhibit stimulating effects of adrenaline to the heart and the sympathetic nervous system), amiodarone (Amiodaron®, Cordarone®) and flecainid (Tambocor®).

The development of arrhythmias is one of the most common consequences of CHD and is a common cause of sudden cardiac death in patients with known CHD (Polderman et al. 2004).

Digitalis: Digoxin is a widely used drug which increases the cardiac muscle work capacity and regulates certain types of heart rhythm disturbances.

Oral side effects: Association between digoxin and dental caries has been found because this medicine is available in a sucrose-based suspension (Lanoxin®) (Stecksen-Blicks et al. 2004, Moursi et al. 2010).

Beta Blockers: they keep your heart rate down and lower your blood pressure. Examples of these include propranolol (Pranolol®) (Felleskatalogen 2011), atenolol (Atenolol®), sotalol (Sotalol®) (Hægermark 2010).

Oral side effects of beta blockers include xerostomia and lichenoid reactions (Griffiths and Boyle 2005 d).

Anticoagulant/antiplatelet therapy: prevents blood coagulation and inhibits the blood platelets' tendency to clump together and form a blood clot, which may result in strokes or heart attacks. The most common used medicines are warfarin (Marevan®), acetylsalicylic acid (Albyl-E®), heparin (Heparin®) (Felleskatalogen 2011).

Side effects include: bleeding gums, bruising, nosebleeds, waterbrash, heartburn, pyrosis and gastric catarrh (Hægermark 2010).

Calcium channels blockers (only used in some cases of CHD): used for treating high blood pressure and abnormal heart rhythms. They disrupt the movement of calcium (Ca^{2+}) through calcium channels. They cause vasodilatation and reduced heart rate.

Oral side effects: gingival hyperplasia caused by nifedipine (Adalat ®) (Seymour 1991, Hægermark 2010).

Antibiotics: used to treat bacterial infections which unfortunately are common in children with CHD and to prevent or treat endocarditis. (see section “Infective endocarditis”).

These drugs have enabled patients to live normal lives despite their heart condition.

What is infective endocarditis?

Infective endocarditis is one of the serious and fatal complications in children and adults with congenital heart disease (Child and Perloff 1991, Dodo et al. 1996). Infective endocarditis is more likely to occur in the presence of damaged or traumatized endothelium which is often the case in these patients.

Infective endocarditis (IE) is a rare and potentially life-threatening disease with a reported incidence of approximately 1.7 to 6.2 cases per 100,000 person-years in Western countries (Berlin et al. 1995, Hogevik et al. 1995). Despite improvements in diagnosis and treatment, IE still has a high rate of mortality, up to 20%, and is associated with significant morbidity (Bayer et al.1998).

Patients with congenital heart defects are at increased risk for developing IE and account for up to 20-35% of cases (Gatzoulis et al. 2005 b).

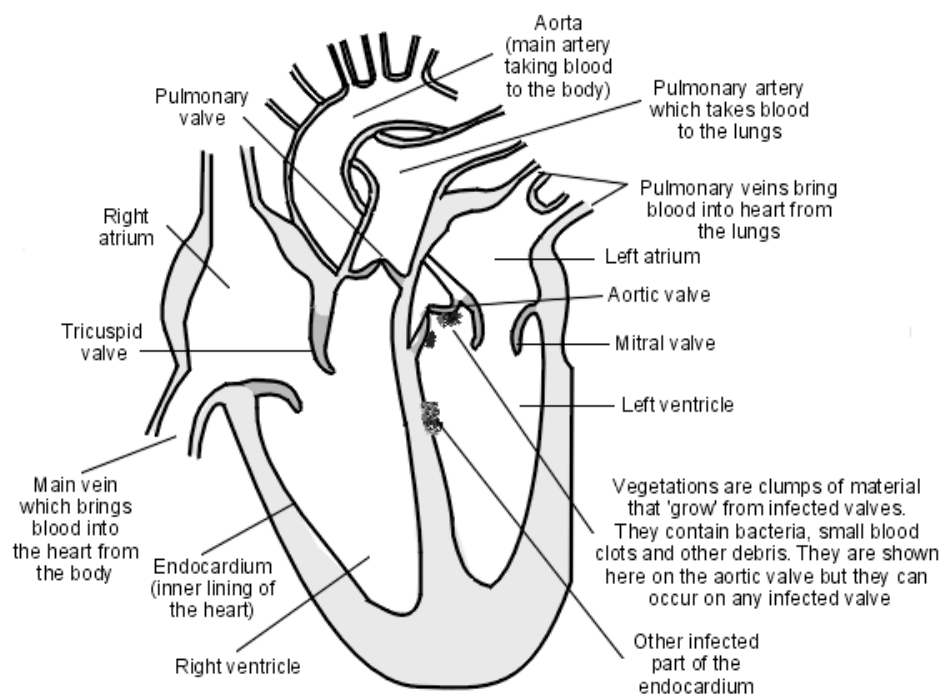
Infective endocarditis is an infection caused by microorganisms that affects a damaged part of the endocardium. The endocardium is the tissue that lines the inside of the heart chambers. The infection usually involves one or more heart valves (that have been previously damaged due to CHD) which are part of the endocardium (Tong and Rothwell 2000), but may also involve nonvalvular areas or mechanical devices that are implanted in the heart, such as artificial heart valves, pacemakers, or implantable defibrillators (Cabell et al. 2003).

Etiology: small clumps of material called vegetations develop on infected valves. The vegetations contain bacteria or fungi, small blood clots, and other 'debris' from the infection (Tong and Rothwell 2000). Vegetations may embolize and cause renal, pulmonary or myocardial infarcts or cerebrovascular accidents.

Invasive dental procedures can introduce bacteria into the bloodstream (bacteraemia), where colonization can occur on the damaged areas of the endocardium (Rhodus and Miller 2008, Fiske et al. 2007). *Viridans* streptococci, a large group of commensal streptococcal bacteria commonly found in the oral cavity, are most frequently responsible for infective endocarditis (Roberts et al. 2006), whereas *Staphylococcus aureus* is often implicated in the acute fulminating form of infective endocarditis (Cabell et al. 2003, Naber 2008, Farbod et al. 2009).

For IE to occur, three conditions are necessary:

- A damaged endocardium.
- Bacteraemia (presence of bacteria in the bloodstream).
- Bacteria of sufficient virulence to evade the body's innate defenses, and able to attach, colonize, invade and cause infection of the damaged endocardium.



Heart With Infective Endocarditis

Source: <http://www.patient.co.uk/health/Endocarditis-Infective.htm>

Diagnosis: IE is usually suspected based upon the patient's history, symptoms, and findings such as a new murmur. It may be confirmed by positive blood cultures for organisms typical of the disease and echocardiographic evidence of cardiac involvement: vegetations, valvular damage, and regurgitation.

Symptoms: The clinical symptoms of IE include fever, malaise, anorexia, chronic renal failure, weight loss and arthralgia. Inflammation of the endocardium increases cardiac destruction, and murmurs subsequently develop. Painful fingers and toes and skin lesions are also important symptoms.

Treatment: Patients with IE are hospitalized and given antibiotic therapy to minimize cardiac damage. Sometimes surgical intervention is necessary.

Prognosis: The outlook is good if the infection is diagnosed and treated early. Many patients are cured with a course of high dose intravenous antibiotics. Complications develop if the infection is left untreated, or if treatment is delayed. Some people die from the complications.

(Prendergast 2002, European Society of Cardiology 2004).

Congenital heart disease and oral health

Congenital heart is a systemic disease, and systemic diseases are often connected to oral diseases. They may be manifested in the oral cavity or their treatment may have impact on dental and oral structures; the consequences of oral diseases in patients with systemic diseases can be life-threatening. Systemic diseases may also affect dental treatment (or vice versa). Therefore, it is of great importance for the general dentists to understand and be aware of the patients' medical status before treating them in practice.

Although dental problems are common in patients with CHD (Stecksén-Blicks 2004, Tasioula et al. 2008, da Fonseca et al. 2009, Rai et al. 2009), their oral health is frequently overlooked by the medical profession (Balmer and Bu'Lock 2003, Tasioula et al. 2008). Rosén and Stecksén-Blicks (2007) reported in their study that many of the dentists noticed the increased cariological problems in patients with CHD, but less than one half of them gave them more caries prevention than to the healthy group.

Children with CHD often have more untreated caries (in primary dentition) than other healthy children (Franco et al. 1996). Of major significance is the fact that untreated caries can be a contra-indication for heart surgery (Hayes and Fasules 2001).

The development of the dentition may be affected by the systemic effects of CHD and/or its treatment. Patients with CHD (especially those with cyanotic conditions) have more enamel anomalies (Franco et al. 1996) in the primary dentition and thereby higher risk of early childhood caries (Hallett et al. 1992). Cyanotic children suffer from chronic hypoxia which may disturb the enamel formation and cause enamel hypomineralization (Jalevik and Noren 2000).

The use of sugar based liquid medications and dietary prescription with high-caloric supplements further increase the caries risk.

Poor oral health in these patients also leads to a lower quality of life (da Fonseca et al. 2009).

Meticulous oral hygiene and preventive dental care is essential to reduce the risk of dental disease in these patients which can severely compromise their medical management and overall prognosis. Children with CHD handle stressful treatment procedures poorly compare to healthy children, which accentuate the importance of disease prevention (Rosén and Stecksén-Blicks 2007).

Infective endocarditis (IE)

Revisions of the guidelines

The requirement for antibiotic cover is one of the more confusing areas in dentistry with different regimes adopted in different countries. Rosén and Stecksén-Blicks (2007) concluded in their study that one-third of the dentists thought the information about antibiotic prophylaxis was unclear.

In theory, the provision of antibiotic cover for dental treatment is linked to the prevention of IE. However, no published data demonstrate convincingly that the administration of prophylactic antibiotics prevents IE. The lack of any supporting evidence has “forced” the major heart associations to simplify and restrict the previous recommendations given (Norsk Cardiologisk selskap 2008)

In 2007, the American Heart Association published new, revised guidelines for the prevention of infective endocarditis (Wilson et al. 2007). Similarly, new guidelines were adopted by Norwegian cardiologists in 2008. These new recommendations represent a fundamental change of attitude regarding the use of antibiotics. The practice of giving antibiotics cover to all patients with cardiac abnormalities and for almost all kinds of dental procedures is stopped. Only patients with underlying cardiac conditions, who are associated with the most serious outcome if IE develops, should receive prophylaxis prior to dental

procedures that is likely to cause bleeding (Norsk Cardiologisk selskap 2008, Lockhart et al. 2009).

The new guidelines emphasize that maintaining optimal oral health and good oral hygiene are more important in reducing the risk of IE than taking preventive antibiotics before dental procedures (Lockhart et al.2009).

The practitioner should be aware that guidelines change and they have a professional responsibility to keep up to date.

Traditionally, guidelines were based on studies that demonstrated that almost all dentogingival manipulative procedures could induce bacteraemia and cause IE in susceptible patients (Elliott 1939) and the use of antibiotic prophylaxis could reduce this bacteraemia and the risk of IE (Durack 1994, Farbod et al. 2009).

However, studies show that very few IE cases are preventable with antibiotic therapy prior to dental procedures. According to Van Der Meer et al. (1992), dental procedures may cause a small fraction of IE cases. On the other hand, prophylaxis would only prevent some of them, even if the therapy was effective.

IE is more likely to result from daily activities, such as brushing, flossing teeth, using wooden toothpicks or chewing food than from bacteraemia caused by dental procedures (Roberts 1999, Wilson et al. 2007, Lockhart et al. 2008, Farbod et al. 2009, Delahaye et al. 2009).

According to Roberts et al. (1999), “it is far more likely that such everyday procedures are the cause of bacterial endocarditis because the cumulative exposure is often hundreds, thousands, or even millions of times greater than that occurring following surgical procedures such as extraction of teeth.”

Several studies show that a majority of patients who develop IE have not had a dental treatment within two weeks prior to the onset of IE (Durack 1994, Durack 1998, Strom et al. 1998).

Table 2: Prevalence of bacteraemia arising after various types of dental procedures and oral cavity.

Procedure	Prevalence of bacteraemia
Extractions Single Multiple	51% 68-100%
Periodontal surgery Flap procedure Gingivectomy	36-88% 83%
Scaling and rootplanning	8-80%
Periodontal prophylaxis	0-40%
Endodontics Intracanal instrumentation Extracanal instrumentation	0-31% 0-54%
Endodontic surgery Flap refelction Periapical curettage	83% 33%
Tooth brushing	0-26%
Dental flossing	20-58%
Interproximal cleaning with toothpicks	20-40%
Irrigation devices	7-5%
Chewing	17-51%

Source: Seymour et al. Infective endocarditis, dentistry and antibiotic prophylaxis; time for a rethink? Brit Dent J 2000;189:610-6.

Adverse effects and resistance

The administration of prophylactic antibiotics is not risk free (Wilson et al. 2007). It has been reported adverse drug reactions due to amoxicillin, which is the primary antibiotic used for IE prophylaxis. Skin reactions gastrointestinal effects, hematologic complications and liver reactions are some of the adverse effects seen with the administration of amoxicillin (Salvo et al. 2007).

Finally, indiscriminate use of antibiotics may lead to increase resistance (Farbod et al. 2009). The frequency of multidrug-resistant viridans group streptococci has increased dramatically during the past 2 decades (Wilson et al. 2007). The shift in microorganisms implicated in the development of IE, namely from streptococci to staphylococci is very disturbing (Farbod et al. 2009).

This compromises those who may benefit from antibiotic therapy, reducing the efficacy and number of antibiotics available.

Maintaining good oral health and hygiene may present a more direct method to prevent and reduce bacteraemia. The presence of dental disease may increase the risk of bacteraemia associated with these routine activities.

Even if dental procedures have not been confirmed as risk factors for IE, they should not be excluded as causative factors. Several known cases of IE are often infected with microorganisms frequently encountered in the oral microflora (Strom et al. 1998).

There may be some cases of IE that could be prevented by antibiotic cover prior to dental procedures, but this should be restricted to those patients at highest risk of adverse outcome if IE should occur (Wilson et al. 2007, Farbod et al. 2009).

Nutrition

Nutritional deficiency during the first years of life of these children, in the period when teeth are formed, is one of the causes of defects in their structure (e.g., enamel hypoplasia) (Hallett et al. 1992). Studies have shown that children with CHD have increased levels of caries and

enamel hypoplasia (Hallett et al. 1992, Stecksén-Blicks et al. 2004). However, in 2008, Tasioula et al. (2008) found that children with congenital heart disease had similar levels of dental disease than children without this congenital anomaly. This may be the result of early intervention and successful management of CHD which has reduced the degree of systemic disturbances (cyanotic patients) during enamel formation.

Caloric needs in infants with CHD vary, but may be around 120 to 170 kcal/kg/day (Lewis and Hsich 2005). Nutritional intervention should include increasing the caloric density of formula by increasing the concentration and adding carbohydrate or fat (Smith 2001, Moynihan 2006).

Unfortunately, frequent consumption of carbohydrates increases the risk of developing caries (Gustafsson et al. 1954, Winter 1990). Of particular importance is the frequency of intake. Each exposure can drop the pH (Donly 2009). Dentists should therefore recommend larger, nutrient-dense meals instead of small, frequent meals. Products with fermentable carbohydrates and other sugars should be consumed with meals, instead of between meals (Touger-Decker and van Loveren 2003).

Xylitol-sweetened candies and healthy snacks should replace frequently ingested cariogenic snacks (Donly 2009).

Studies have shown that parents of chronically sick children tend to overprotect and overindulge their child (Van Dongen-Melman et al. 1986). Because of the reduced appetite of these children, parents often let their child eat and drink what they prefer (mostly sugar-containing beverage and cariogenic snacks), whenever they want (between main meals, at night) making oral health a low priority (Ievers et al. 1994, Moursi et al. 2010).

Unfortunately, the well meaning of these parents contributes to their child's development of oral diseases (Winter 1990). These children also receive sweet treats given by relatives who feel sorry for them (Foster and Fitzgerald 2005).

Frequent intake of soft drinks increases the risk of erosion (irreversible loss of tooth structure) (Bartlett 2009). This risk is even higher when these highly acids liquids are given in nursing bottles.

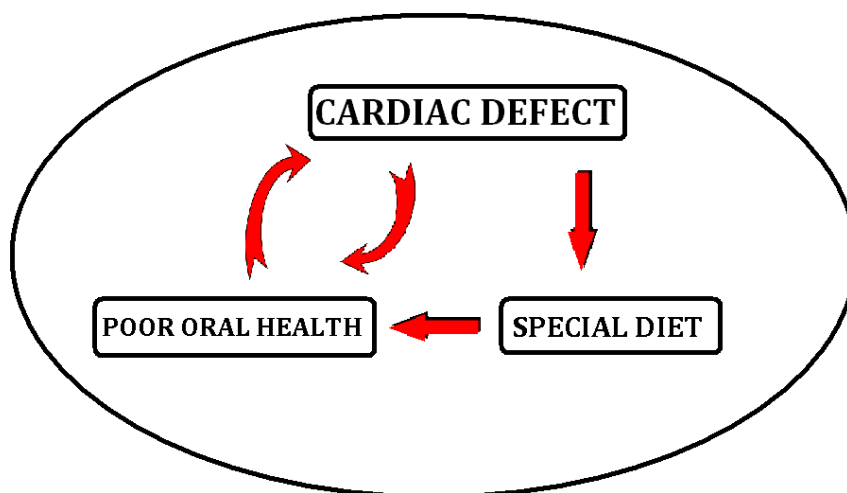
Table 1: Nutrition and oral health status in patients with congenital heart disease.

Medical disorder	Dietary factors	Special diet	Oral health status
Congenital heart defect	Decreased appetite Difficulty feeding Impaired absorption Inadequate energy intake Increased energy requirements Hypermetabolism Gastroesophageal reflux Nausea Vomiting Early satiety	Frequent meals Frequent consumption of carbohydrates Energy-dense supplements Cariogenic snacks Soft drinks	Increased caries Enamel hypoplasia Erosion Periodontal disease

Source: Inspired from Moursi et al. 2010 (table 1).

Children with CHD often need dietary modifications which may have damaging effects on their oral health and poor oral health will itself have negative effects on their general health status, creating a vicious cycle.

Fig. 1 Cycle of interactions between cardiac defects, dietary modifications and oral health.



Source: inspired from Moursi 2010 (fig. 1).

Nutrition is an essential component of oral health (American Dietetic Association 2003). Healthy food choices contribute to healthy teeth and healthy teeth play an important role in the chewing and eating process which influence the nutritional status. A close collaboration between nutritionists, parents and dentists is recommended for oral health promotion and disease prevention in these patients (Glick 2005, American Academy of Pediatric Dentistry 2008).

Dentists should promote healthy food policies and thereby contribute to their patient's well being.

Oral side effects of medications

The principles of good pharmacotherapy involve balancing the benefits of medication against the potential risks and adverse effects. The oral soft tissues exhibit rapid cell growth and cell turnover and are therefore more likely to be affected by side effects of medication.

Medical treatment can cause an upset in the equilibrium between the micro-organisms (bacteria, fungi, viruses) which usually populate the oral cavity and cause infections (Griffiths and Boyle 2005 c).

Sugar-based cariogenic medicines (Griffiths and Boyle 2005 c, Matias Freire 2006)

Caries in chronically sick children may be life-threatening because of the consequences of the disease or the dental treatment. A serious decayed tooth can develop an infection in the root canal where bacteria can spread to other parts of the body, including a susceptible heart. An attempt to treat a decayed tooth can also cause bleeding which will increase the risk of infective endocarditis in susceptible patients.

An area of particular concern is the use of sugar-based liquid medication for children, aiming to make them more palatable (Kenny and Somaya 1989, Roberts and Roberts 1989). A high concentration of sucrose has been found in several kinds of children's medicines (Novais et al. 1998). This significantly elevates the cariogenic potential of these medications (Hobson

1984). Studies show that children who use liquid medicines containing sugar for a long time have higher levels of caries lesions compared to control groups (Feigal et al. 1984, Roberts and Roberts 1989). Children on digoxin therapy, administered with a sucrose-containing syrup, have increased caries prevalence (Stecksén-Blicks et al. 2004). According to Maguire et al. (1996), children with chronic illness taking liquid medication for more than 1 year have significantly more dental caries of their primary anterior teeth than their siblings. Health practitioners should be aware of sugared medicines in the form of liquids (Feigal et al. 1981) and a full medical history is therefore essential.

Today, sucrose is avoided as a sweetener in most medicines and in many countries the pharmaceutical industry has developed alternative formulas without sugar or with non-cariogenic sweetener substances. The use of pills instead of liquids has also been well accepted (Berlin et al. 1995). The use of other methods of administering medicines, such as the rectal, intramuscular, inhalation, and transdermal ways should be considered.

According to Rosén and Stecksén-Blicks (2007), “seventy-two per cent reported that their CHD-patient(s) had caries but only 34 % knew that some medicines used by CHD-patients could increase the risk for caries. Younger dentist reported statistically significantly more often than older that medicines used by this group of patients could increase the caries risk”.

Patients should take their medicines in tablet form when possible, avoid medications containing sugar in collaboration with their doctor, take sugar-based medicines with meals unless otherwise instructed (Moursi et al. 2010), avoid liquid medications just before bedtime, brush with fluoride toothpaste or chew sugarless gum after ingesting their medication (Feigal et al. 1981). Digoxin (Lanoxin®) is a sucrose-based liquid medication, but it can also be found in tablet form (containing lactose) (Felleskatalogen 2011).

Xerostomia/Hyposalivation

Heart medications such as antiarrhythmics, diuretics, ACE inhibitors, calcium channel blockers can cause salivary dysfunction (Griffiths and Boyle 2005 c, Nissim 2005, Dahllöf and Martens 2009).

Xerostomia is defined as the subjective sensation of dry mouth (Greenspan 1996). It is uncomfortable for the patients and can cause difficulties in speaking, tasting and eating.

Hyposalivation is a term based on objective measures of the saliva secretion, when the flow rates are significantly lower than the accepted “normal value”. Flow rates of unstimulated saliva less than 0,1ml/min, and those of chewing-stimulated saliva less than 0,7ml/min, fulfil the criteria for hyposalivation (Axelsson 2005, Bradley 2010).

Saliva is very important in providing remineralization, clearing food debris from the oral cavity and has an important role in the buffering of acids produced by the bacterial biofilm from sugars (Featherstone 2000, Stookey 2008). Furthermore, salivary proteins (histatins, lactoferrin, peroxidase, and lysozyme) aid in antimicrobial activity by inhibiting bacterial growth (Tabak 2006).

Some medications can cause a decrease in saliva secretion; therefore, it is important to note all medications taken when completing a medical history and find out whether salivary flow has been compromised due to medication side effects by taking saliva tests. The salivary flow rate affects saliva composition. A decrease in salivary flow increases the susceptibility to caries, dental erosion and other oral infections (Stookey 2008).

The use of artificial saliva, chewing gum and lozenges can stimulate salivary flow (Griffiths and Boyle 2005 c). Greater consumption of water, the use of fluoride products and good oral hygiene is also recommended when salivary flow rate is low.

Dental treatment of patients with CHD

Anticoagulation therapy as a complication for dental treatment procedures

In patients with CHD, anticoagulation and antiplatelet therapy may be necessary to prevent thrombosis or embolism. Those on chronic anticoagulation therapy may require adjustment before undergoing oral surgery.

Patients taking anticoagulants (e.g., warfarin), have their blood-clotting level regularly monitored by a doctor. A value called the INR (International Normalized Ratio) is used to determine the clotting tendency of blood. In healthy people the INR is about 1. The INR value of patients taking anticoagulants is normally higher (2-3) (Neppelberg and Brokstad Herlofson 2008), meaning that the blood clots more slowly (the bleeding time is two to three times longer than normal). The dosage of the anticoagulant is adjusted to maintain an appropriate INR. When undergoing oral surgery, their INR values may be set below the therapeutic range, to avoid excessive post-operative bleeding (Sacco et al. 2006).

Interruption or reduction of anticoagulation and antiplatelet therapy prior to oral surgery has been discussed for several years (Neppelberg and Brokstad Herlofson 2008).

According to Sacco et al. (2006), stopping or reducing the anticoagulation therapy before oral surgery is not necessary if simple measures for local hemostasis are implemented (haemostatic gauze, sponges, and sutures, e.g., acid. tranexamic (Cyklokapron®), Surgicel®).

Michael J. Wahl reported in his article “Myths of dental surgery in patients” (2000) that “serious embolic complications, including death, were three times more likely to occur in patients whose anticoagulant therapy was interrupted than were bleeding complications in patients whose anticoagulant therapy was continued (and whose anticoagulation levels were within or below therapeutic levels)”. He therefore recommends that patients continue their anticoagulant therapy when undergoing surgical dental procedures, if the patient’s anticoagulation level is within the recommended therapeutic range.

At the Department of Oral Surgery at the University in Oslo, patients treated with warfarin must present an INR obtained within 24 hours prior to the surgical procedure with an INR value lower than 2.0-2,5. Minor procedures can be performed with $INR < 3$, if local haemostasis and post-operative information is given (Neppelberg and Brokstad Herlofson 2008). The INR value should also have been stable over time. If reduction/interruption of the anticoagulant therapy is needed, this must always be done in cooperation with the patient’s doctor. Interruption of treatment with antiplatelet therapy (Albyl-E®) prior to surgical procedures is normally not necessary (Neppelberg and Brokstad Herlofson 2008).

Cyanotic patients have both a bleeding and a thrombotic diathesis. Cyanosis is associated with an increased risk of bleeding (abnormal platelet function and coagulating factors, increased tissue vascularity, and collaterals). Mucosal bleeding and easy bruising are the most frequent problems.

Occasionally there is a thrombotic tendency.

The use of anticoagulants in patients with cyanosis is complicated by the presence of bleeding diathesis, and achieving the desired INR can be difficult due to reduced plasma volume (Gatzoulis et al. 2005 a).

Not many dentists are aware of the associated haemorrhagic tendencies in such patients. Haemostatic abnormalities associated with CCHD are an important aspect that is often overlooked by the dentists (Auluck and Manohar 2006).

Immunosuppressive therapy and its influence on oral health

The use of immunosuppressive drugs (e.g., cyclosporine) is essential for heart transplant patients to avoid the possibility of rejection (Lindenfeld et al. 2004). The number of pediatric heart transplantations for complex congenital heart disease has increased over the last years.

Immunosuppressive drugs can have adverse oral effects (Rhodus and Miller 2008):

1. Increased potential for infection
2. Xerostomia
3. Candidosis
4. Increased risk for activation for the latent herpes virus → intraoral herpetic infection
5. Cyclosporine: may cause gingival overgrowth (Felleskatalogen 2011)

Pharmacological management of pain and anxiety

Patients with CHD, especially children, have difficulties to deal with the “usual” degree of stress associated with dental therapy (reduced tolerance to bradycardia, tachycardia, fall in blood pressure, hypoxia, etc) (Den Norske Legeforening 2010). When these patients are undergoing dental procedures (associated with increased stress), there is a risk of exacerbation of their underlying disease (exacerbation of heart failure and cardiac dysrhythmias). Reduction of stress is the primary goal in the successful dental management of these patients. Awareness and knowledge of the underlying disease process is essential (Malamed 2009 b).

Effective pain management is also a challenge. The different methods of pain control vary from simple behaviour management to general anesthesia (Roberts and Hosey 2005).

General anesthesia and other sedative agents impair these patients’ cardiovascular reserves (positive pressure ventilation, drugs with cardio-depressive effect). Planned procedures require enhanced monitoring and the use of specially trained staff (Den Norske Legeforening 2010).

Sedation and pain control are of much greater importance in these patients than in ASA 1 patients (Malamed 2009 b).

Local anesthesia

Pain control through the use of local anesthetics* plays a major role in reducing stress in patients with CHD (Malamed 2009 b).

*Local anesthetics provide a reversible regional loss of sensation without affecting consciousness.

Cardiovascular effects

In general, there is no contraindication to the use of vasoconstrictors (e.g., adrenaline) in local anesthetics administered in these patients (Jastak et al. 1995). However, adrenaline-containing local anesthetics affect the cardiovascular system (Meechan 2005). An increase in the concentration of adrenaline in the blood may cause an increase in cardiac output and

stroke volume. Blood pressure and heart rate can also be affected (Kaneko et al. 1989). Cardiac arrhythmias are also among the potential adverse effects (Malamed 2009 b).

Accidental intra-vascular injection or rapid systemic absorption can induce cardiovascular problems (Jowett and Cabot 2000, Malamed 2009). In these patients, the use of aspiration to prevent intravascular injection is essential.

Nevertheless, the addition of adrenaline is a protective measure against the toxicity of the anesthetic: it delays the absorption of the anesthetic into the cardiovascular system (which removes it from the injection site and results in higher plasma levels of the local anesthetic) and therefore avoids the need for repeating the injections during the procedure (Malamed 2004).

The side effects of absorbed adrenaline must be weighed against those of elevated local anesthetic blood levels.

Contraindication: Intraligamentary anesthesia (intraosseous injection) may introduce a substantial numbers of bacteria into the bloodstream. In patients susceptible to infective endocarditis this method should be avoided, unless antibiotic prophylaxis is already indicated (Meechan 2005, Brand and Fennis 2009).

Drug interactions

Local anesthetics and a considerable number of drugs bind to plasma proteins thereby competing with each other. This leads to fewer binding sites available for local anesthetics, thus increasing their toxicity.

In addition, beta-blockers decrease hepatic blood flow and interact with liver enzymes that metabolise amide-type anesthetics decreasing their metabolism, leading to an increased plasma concentration of the anesthetic (Brand and Fennis 2009, Malamed 2009 b).

In patients with cardiac anomalies treated with non-selective beta-blockers (propranolol, sotalol, nadolol. etc), adrenaline may cause hypertension (the blockade of the beta 2-receptors prevents the vasodilatation in skeletal muscles that compensates the alfa-adrenergic

vasoconstriction). This can lead to cardiovascular disturbances (Goulet et al. 1993, Brand and Fennis 2009, Malamed 2009 b).

Patients on digoxin therapy should have a medical consultation before administering an adrenaline-containing anesthetic solution, because of an increased risk of cardiac dysrhythmias (Malamed 2009 b).

In patients taking several heart medications, it can be recommended to use a local anesthetic without adrenaline to avoid any potential cardiac damage.

Sedation and general anesthesia

Sedation: reduction of anxiety, stress, irritability or excitement by administration of a sedative agent (nitrous oxide) or drug (Midazolam, Diazepam, Flunitrazepam).

General anesthesia: “a drug-induced reversible modification of signal handling in the central nervous system which results in a decrease of the responses to and subjective perception of external stimuli”. Its purpose is to decrease the patients’ sensitivity and subjective reaction to pain. Cardiovascular function may be impaired (Hallett et al. 2008).

There are no set rules for deciding whether dental treatment should be provided under general anesthesia or by using other sedative drugs and patients need to be assessed individually.

General anesthesia may be considered when children with CHD require multiple dental procedures to limit the number of visits, thereby minimizing stress and the need for repeated courses of antibiotics if necessary. If general anesthesia is indicated, the treatment should be completed in a hospital environment, where adequate supportive care is available if needed and where the patient can receive a more in-depth medical evaluation (Weddell et al. 2011).

In some children with complex cardiac defects, sedation has to be performed in cooperation with the child’s cardiologist (Stecksén-Blicks et al. 2004).

Children with cyanotic defects (decreased oxygen saturation in the blood) are at significant risk for desaturation during general anesthesia and preoperative consultation with the pediatric cardiologist and anesthesiologist is very important (Hallett et al. 2008).

During nitrous oxide sedation (inhalation) the medical practitioner must monitor both the patient and the machinery. Vital signs and oxygen saturation must always be monitored (Hallett et al. 2008). In healthy patients with ASA I or II, the radial pulse is checked during the procedure. For patients with ASA III or IV (children with severe CHD with high blood pressure or cyanosis), treatment within a hospital environment, the use of blood pressure cuff, and/or electrocardiograph monitoring is also recommended (Roberts and Hosey 2005). It is important that appropriate monitoring equipment is available.

It is essential to provide adequate sedation without inducing hypoxia. The myocardium of patients with CHD may be more vulnerable to hypoxic episodes. Inhalation sedation (e.g., conscious sedation with nitrous oxide) is therefore an excellent technique for this group of patients, because of the additional levels of O₂ supplied during the procedure. Minimal to moderate intravenous sedation can also be recommended, however supplemental O₂ should be administered to reduce the risk of hypoxia (Malamed 2009 b, Den Norske Lægeforening 2010).

Intramuscular sedation technique is considered a last-choice technique if other sedative procedures are unavailable or ineffective (Wilson et al. 2007).

Sedation and general anesthesia can carry greater risks for patients with CHD than for healthy patients, but may be necessary to achieve a non-stressful environment and adequate dental care. Consultation with the patient's cardiologist and the anesthetic team before the procedure is essential.

Managing patients with CHD in oral care services

Managing patients who require antibiotic cover (Fiske et al.

2007, Rhodus and Miller 2008, Hallet et al. 2008):

- The medical and allergy history must always be updated before giving antibiotics and the dental team should be trained in the recognition and handling of allergic reactions, including anaphylaxis. Consultation with the patient's doctor or cardiologist is recommended.
- Optimal oral health should be encouraged. Patients must be aware of that their day-to-day oral hygiene measures are more important than professional treatment and taking antibiotics.
- Regular recalls to prevent dental disease.
- 'Group' together a number of invasive dental procedures to be performed in the minimal number of appointments (e.g. consider the use of general anesthesia) to reduce the need for repeated courses of antibiotics. This approach has the twofold benefit of reducing the likelihood of development of antibiotic resistant strains and of allergy. The same antibiotic should not be prescribed within 14 days.
- Edentulous patients may develop bacteraemia from ulcers caused by ill-fitting dentures. New dentures should be checked regularly to correct any problems that may cause mucosal ulceration.

The dentist is responsible for identifying patients at risk for IE. According to Rhodus and Miller (2008) the dentist must:

- Obtain the patient's medical status and history
- Have up-to-date knowledge about the new published guidelines
- Identify dental procedures likely to cause bacteraemia in susceptible patients

- Select the appropriate antibiotic regimen
- Eliminate/remove all sources of infection that could serve as a nidus for cardiac infection

Special care (Fiske et al. 2007, Rhodus and Miller 2008)

- Care must be taken when prescribing antibiotics for patients who are taking other drugs; for example, the anticoagulant warfarin (the effect of which can be intensified) and oral contraceptives (the effect of which can be diminished). It is recommended that potential drug interactions are routinely checked before prescribing.
- Intraligamentary anesthesia carries the risk of a high load bacteraemia and should be avoided.
- Prior to cardiac surgery, oral hygiene should be optimised and any dental interventions should be carried out at least 14 days prior to surgery to allow for mucosal healing. Elective dental procedures should be delayed until at least three months after cardiac surgery (Weddell et al. 2011).

For all patients requiring the provision of antibiotic prophylaxis the following should be recorded in their notes (Fiske et al. 2007):

- Cardiac risk for IE
- The type and dose of antibiotic chosen
- The absence of allergy to the chosen drug
- Time of administration of drug
- The dental procedure performed
- Time dental procedure started
- Post-operative instructions given to the patient
- Preventive advice given.

Pediatric dosing (Hallett et al. 2008):

- The dose for any child should be calculated up to, but not exceeding the maximum adult dose.
- Dosage should always be prescribed according to weight (dose per kg).

Other considerations:

- If antibiotic cover has not been given prior to a dental procedure, antibiotics prescribed up to 6 hours after a procedure may give effective cover (Hallett et al. 2008).
- Extraction of primary teeth which have a poor prognosis is preferred prior to pulp therapy because of the high incidence of associated chronic infection (Weddell et al. 2011).
- Endodontic therapy in the permanent dentition can usually be performed.
- If a general practitioner feels insecure in treating patients who are susceptible to IE he must refer them to a dentist who have the appropriate knowledge about this group of patients (Weddell et al. 2008).

Current guidelines (Wilson et al. 2007, Norsk Cardiologisk selskap 2008)

The new guidelines are aimed at patients who would have the greatest danger of a bad outcome if they developed a heart infection.

Preventive antibiotics prior to a dental procedure are advised for patients with:

- prosthetic heart valves
- a previous history of infective endocarditis
- certain congenital heart defects such as
 - unrepaired or incompletely repaired cyanotic congenital heart, including those with palliative shunts and conduits
 - completely repaired congenital heart disease with prosthetic

material or device, whether placed by surgery or by catheter intervention, during the first six months after the procedure*

- repaired congenital heart disease with residual defects at the site or adjacent to the site of a prosthetic patch or a prosthetic device (which inhibit endothelialisation)
- a cardiac transplant with cardiac valvular disease

*Endothelialization of prosthetic materials occurs up to 6 months after the procedure.

At-risk dental procedures (Wilson et al. 2007, Norsk Cardiologisk selskap 2008)

Antibiotic prophylaxis is now indicated for all dental procedures that involve manipulation of the gingival, mucosal or periapical tissues that is likely to cause bleeding.

- Prophylaxis recommended: tooth extractions, periodontal procedures, dental implants, endodontic procedures, local anesthesia injection (intraalveolar), scaling, oral surgery.
- Prophylaxis not recommended: local anesthesia injections (non-intraalveolar), post-procedure suture removal, placement and adjustment of orthodontic appliances, shedding of deciduous teeth, taking dental radiographs, fluoride treatment, bleeding from trauma to the lips or oral mucosa.

Recommended regimens

- Antibiotic prophylaxis given orally is sufficient. Parenteral antibiotic prophylaxis can be given when peroral administration is not possible.
- Peroral antibiotic prophylaxis is given 1 hour before the procedure starts. Parenteral prophylaxis is given 30 min. before the procedure starts.
- One single dose is enough.

- If a patient uses an antibiotic (therapeutic) before a procedure, prophylaxis should be administered with another agent.

Recommended antibiotic prophylaxis

Orally:

Adults: Amoxicillin 2 g single dose orally, 60 minutes before procedure.

Children < 12 years of age: Amoxicillin 50 mg/kg body single dose orally (max. 2 g), 60 minutes before procedure.

Parenterally:

Adults: Ampicillin 2 g i.v. (intravenous) or i.m.(intramuscular) 30 minutes before procedure.

Children < 12 years of age: Ampicillin 50 mg/kg body (max 2 g) i.v. or i.m. 30 minutes before procedure.

If allergic to penicillin:

Orally:

Adults: Clindamycin 600 mg single dose orally, 60 minutes before procedure.

Children <12 years of age: Clindamycin 20 mg/kg body weight (max 600 mg), single dose orally, 60 minutes before procedure.

Parenterally:

Adults: Clindamycin 600 mg i.v. 30 minutes before procedure.

Children <12 years of age: Clindamycin 20 mg/kg body weight (max 600 mg) i.v. 30 minutes before procedure.

Alternative option: Ceftriaxone 1 g (children 50 mg/kg) iv.

AWARE: Ceftriaxone must not be administered if known immediate allergy to penicillin.

If a surgical procedure is planned on infected tissue with a known microorganism, antibiotic prophylaxis should be selected after resistance determination.

Oral health promotion and disease prevention

All dentists have an important role in promoting oral health and prevent oral diseases, but this often has a lower priority in comparison to treatment and care. However there have been recent changes in global and government policies that now place the promotion of health in a more important position (Helsedirektoratet 2011).

Methods of preventing oral diseases:

- **Supervised tooth brushing:** Daily supervised tooth brushing has been shown to reduce dental caries in children (Jackson et al. 2005). It is recommended to brush twice a day.
- **Practice effective oral hygiene from the first tooth inclusive:** The establishment of cariogenic bacteria in the oral cavity occurs when the first deciduous tooth erupt (Berkowitz 1975). Recently erupted teeth have immature enamel and post-eruptive mineralization occurs during the initial two years. During these 2 years, the tooth is more susceptible to develop caries (Matias Freire 2006).
- **Use fluoride products:** Fluoridated toothpastes play a major role in caries reduction (Marinho et al. 2004). Professionally applied topical fluoride also contributes to a decrease in caries prevalence (Attin et al. 1995).
- **Use xylitol products** (American Academy of Pediatric Dentistry 2008).
- **Use antimicrobials products (e.g. Chlorexidine)** (Anderson 2003): must be used with caution and in collaboration with your dentist. Chlorexidine is not meant as a long-term treatment. It can cause staining of teeth, tongue and white fillings.
- **Maintain regular dental attendance** (Prendergast 2001).

- **Limit sugar consumption:** High sugar intake leads to high caries prevalence (Winter 1990, König and Navia 1995, Touger-Decker and van Loveren 2003).
- **Recommend large, nutrient-dense meals at fixed hours, instead of small, frequent meals.**
- **Restrict sugary food and drink consumption to meals only** (Touger-Decker and van Loveren 2003).
- **Avoid the consumption of milk or other sugar-containing beverage at night:** the saliva flow rate is very low at night, increasing the risk for developing caries by drinking sugar-based drinks (Touger-Decker and van Loveren 2003).
- **Encourage the use of sugarless gum and non-cariogenic snacks between meals to increase salivary flow** (Stookey 2008).
- **Promote consumption of water to avoid dehydration in sick children!**
- **One should recommend the doctors to prescribe sugarless medicines whenever they are available** (Matias Freire 2006).
- **Avoid smoking** (Malhotra et al. 2010).

Oral health promotion in primary health care and hospitals (Griffiths and Boyle 2005 a, Matias Freire 2006):

- Teaching effective oral hygiene routines to health workers.
- Health education about the risks of sugared medicines.
- Administration of sugar-free medicines (children who used sugary medications for a long time had higher levels of caries lesions compared to control groups). (Roberts and Roberts 1979, Foster and Fitzgerald 2005). Sugar-free medicines do not contain sucrose, fructose or glucose.
- Avoid oral administration of sugar based medicines (if possible).
- Feeding guidelines for special care groups and closer collaboration between nutritionists and dental professionals (Touger-Decker and Mobley 2007).
- Nutritional policies for hospital catering.

Orthodontic treatment

Few studies have compared craniofacial growth in children with CHD compare to healthy children, even though these children usually experience delayed growth (often because of malnutrition). In 2007, de Andrade Goldner et al. (2007), conducted a study to evaluate the craniofacial characteristics of patients with heart disease. They found that boys with heart disease had sharper maxillary protrusion and the girls without heart disease had increase ANB angles and protrusion of the lower lip. However, these findings are not significantly relevant when it comes to orthodontic treatment.

There has been suggested that children with CHD have more mesial occlusions and crowding of teeth than other healthy children (Franco et al. 1996).

Nevertheless, the most important aspect of orthodontic treatment is to keep the braces clean to reduce the risk of gingival disease which is very unfortunate for this patient population.

Use of electromagnetic dental devices

Knowledge of the safe use of electromagnetic dental devices in patients with pacemakers or implantable cardio-converter defibrillators (ICDs) is poor (Thompson et al. 2007).

There is a potential for current generated by dental devices to interfere with function of pacemakers or ICDs. One should avoid the use of electrosurgery, ultrasonic scalers and electronic pulp testers in case of potential interference. The use of sonic scaler, endodontic ultrasonic instrument, radiographic unit or electric toothbrush is for the moment considered as safe (Rhodus and Miller 2008, Hallett et al. 2008).

Social insurance benefits (Helse- og Omsorgsdepartementet 2011)

In Norway, dental treatment is not covered by the public sector after one has reached 18 years of age*. But every rule has exceptions.

The Social Insurance in Norway gives benefits for some well defined groups of patients, including patients with cardiac defects and patients with rare medical conditions (e.g., Williams syndrome, Noonan's syndrome, Marfan syndrome, etc).

If a patient is undergoing heart surgery, dental treatment is covered to prevent the spread of infection from the oral cavity that may involve a serious and life-threatening risk for the patient.

The Social Insurance also gives benefits if hyposalivation caused by medications or medical disorders is diagnosed and resulted in an increase in caries activity. The diagnosis must be documented by clinical findings for at least 1 year.

Patients with rare medical conditions get covered all necessary dental treatment.

* Patients aged 19 to 20 years old get 75% of their dental treatment covered.

Norwegian heart associations for congenital heart disease

In several countries different heart associations are established to give patients information about their medical disorder and help them dealing with everyday issues.

In Norway "Foreningen for hjertesyke barn (FFHB)" is the major organ for children with congenital heart disease.



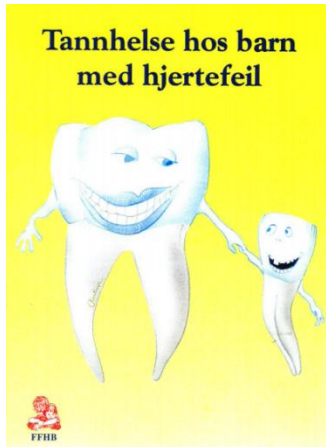
Due to the increased survival of children with CHD into adulthood, leading to a significant increase in the population of adults with congenital heart disease (219), a new heart association was established in Norway in April 2010, “Foreningen voksne med medfødt hjertefeil (VMH)” (Association for adults with congenital heart disease).



The heart association for children with congenital heart disease in Norway (FFHB) has recently published a new and revised booklet about oral health and congenital heart disease. The first booklet concerning this issue was written by dentist Audun Rui and published in the late 1980s.

Both dental professionals and parents of children with congenital heart disease are recommended to read the new brochure, even though it is addressed specifically to parents of children with cardiac anomalies.

These two pamphlets reflect the “mental evolution” concerning antibiotic prophylaxis, which is mentioned in the article. In the old brochure, a liberal approach is recommended concerning the use of antibiotics while in the new brochure, where the new revised guidelines are introduced, optimal oral hygiene and good oral health are presented as the two major factors to prevent infective endocarditis. The last brochure (written in Norwegian) can be downloaded from their website: www.ffhb.no



Old brochure



New brochure

Discussion

Patients with chronic health conditions have an increased risk of oral diseases due to consequences of the disease or/and the medication given. The general dentist must have up-to-date knowledge of any changes in health condition and medication that may affect the dental treatment, and initiate the necessary preventive measures if necessary. For example a thorough preoperative assessment of the patient's regular medication (including anticoagulants, antiarrhythmics and antihypertensives) is essential to avoid any potential drug interactions during treatment. But many general practitioners took their dental degree many years ago and do not possess up-to-date knowledge about these new medicines and this "new" group of patients. According to Jowett and Cabot (2000), many dentists are not confident in treating children with CHD. Further education about new medical groups in dentistry is needed. A close collaboration with the medical treatment team is an important part of the management of oral health care in patients with CHD.

Health-promoting behaviour

Several studies have examined the health-promoting behaviour among children/adults with congenital heart disease, their parents and their dentists.

Among adults with CHD, knowledge about dental health practices is satisfactory, even though many of these patients are unaware of the risk factors of endocarditis (Moons et al. 2001). According to Chen et al. (2007), adolescents with CHD practice health-promoting behaviour similar to that of their counterparts without CHD.

In a study conducted in 2009 (da Fonseca et al. 2009), one-fifth of the parents of children with CHD did not know that oral health was important for their children's heart. According to Rai et al. (2009), parents' awareness on the importance of maintaining good oral hygiene is very poor. There are studies showing that many children with CHD have never seen a dentist (Saunders and Roberts 1997, Balmer and Bu'Lock 2003).

Balmer and Bu'Lock (2003) reported that among children considered to be at risk from infective endocarditis and registered with a dentist (79% of them) only 29% had received instruction in oral hygiene, 42% had received dietary advice and 13% had received advice regarding fluoride supplementation or had had fluoride professionally applied.

It seems that both dentists and parents of children with CHD lack knowledge about the importance of maintaining a good dental health in these patients. Dentists have a responsibility to give the appropriate advices to both patients and parents of children with CHD. They actually play a very important role in preventing oral diseases in this patient population.

Knowledge about IE among dentists and parents

Parents of patients with cardiac risks lack knowledge about IE (Saunders and Roberts 1997, Balmer and Bu'Lock 2003). According to Balmer and Bu'Lock (2003) only 64% of parents were aware of the link between the oral health of their children and infective endocarditis.

Hayes and Fasules (2001) also reported a deficiency of knowledge among dentists regarding the indications for prophylaxis and the antibiotic regimen required to prevent IE.

Among dentists there is a lot of confusion as to which patient groups and cardiac conditions require prophylaxis and for which particular dental procedure. Thompson et al. (2007) conducted a study that identified potential for under- and over-prescription of antibiotics.

Antibiotic prophylaxis has been a debated topic for many years. In 1965, the American Heart Association (AHA) published their first document related to antibiotic cover prior to dental procedures. Since then their “guidelines” have been revised several times and in 1997 they classified patients with CHD into high, moderate and low risk groups with the low risk group not requiring any antibiotic cover. The groups in these guidelines included a larger number of heart conditions and more specific cardiac defects than in the new guidelines which were published in 2007 and are now adopted by several countries, including Norway (Farbod et al. 2009). The new recommendations are clearer regarding to which patient should receive antibiotic cover in comparison with previous ones and are expected to reduce antibiotic resistance that has increased during the past years (Sanchez-Rodriguez et al. 2008).

Bacteraemia: daily routine activities versus dental procedures

Some studies emphasize the fact that daily routine activities such as tooth brushing present a greater risk of bacteraemia than dental procedures such as a single tooth extraction.

According to research reported in *Circulation: Journal of the American Heart Association* (2008) (Lockhart et al. 2008) tooth brushing poses a risk of bacteraemia similar to that of dental extraction. Although amoxicillin significantly decreased the incidence of bacteraemia resulting from a single-tooth extraction, tooth brushing may be a greater threat for susceptible patients, simply due to frequency. Roberts estimated that brushing teeth two times a day for 1 year had a 154,000 times greater risk of bacteraemia than that following a single tooth extraction (Roberts 1999).

The incidence and magnitude of bacteraemias of oral origin have been found to be directly proportional to the degree of oral inflammation and infection (Bender et al. 1984, Pallasch and Slots 1996) and occur more frequently in individuals with high dental plaque scores and gingivitis than in persons with good oral health (Sconyers et al. 1979).

Lockhart et al. (2009) reported that bacteraemia after tooth brushing is associated with poor oral hygiene and gingival bleeding after tooth brushing. They concluded that improvements in oral hygiene may reduce the risk of developing IE.

Cooperation between dentists and nutritionists

Even though dentists should recommend large, nutrient-dense meals at fixed hours instead of small and frequent meals, this is not always feasible when it comes to children with CHD. These children have decreased appetite and higher energy demands than healthy children. To ensure these energy requirements, parents often have no other choice than feeding them whenever they are hungry (also at night) with food containing a large amount of carbohydrates and fat, which may increase the risk of developing caries. Frequent meals are also recommended by nutritionists to meet these children's energy demands. It is important for the dentists to be aware of this nutritional issue and not change these patients' dietary habits without knowing their underlying disease process. A closer cooperation between dentists and nutritionists is desirable. Dentists must focus on prevention of oral disease when children with CHD are recommended to eat frequently by their nutritionists. The dental check-up intervals should be individualized and these patients must receive regularly oral hygiene instruction and fluoride supplementation. It is also of great importance to inform children and their parents about the oral consequences of frequent ingested cariogenic meals and advice them to seek a dentist regularly to avoid any dental diseases.

New project

An interesting research project between the Faculty of Dentistry in Bergen, the university hospital of Haukeland and the public dental services is planned here in Norway (2010). Oral health status among Norwegian children with congenital heart disease has never been studied before and this new study aims to examine oral health in these children and find out if early intervention and close follow-up will improve their oral health.

This study is still ongoing and we will have to wait for the results to come.

Conclusion

Studies have shown that patients with congenital heart disease have poorer oral health than other healthy patients (Hallett et al. 1992, Stecksén-Blicks et al. 2004).

They are at increased risk of developing oral diseases and the consequences of these are much greater than in healthy patients. Many of these patients have a high caries activity and

untreated gingivitis (Franco et al. 1996). Unfortunately, dental care is often a low priority in this group.

Dentists play an important role in raising awareness of the importance of good oral health among these patients and their families and providing them adequate oral healthcare.

General practitioners should focus on caries prevention and give these patients dietary advice, oral hygiene instruction, and fluoride supplements if necessary.

Several children with CHD experience long hospital stays because of surgery or illness. Many of them have never been to a dentist until a caries problem is evident (Stecksén-Blicks et al. 2004).

They also have an increased vulnerability to stressful dental procedures making the use of sedation techniques an important aspect in the dental treatment of these patients.

Poor oral hygiene has been shown to be a risk factor for infective endocarditis (Lockhart et al. 2009).

Optimal oral hygiene may reduce the incidence of bacteraemia from daily activities and maintenance of good oral health probably plays a more important role than prophylactic antibiotics prior to dental procedures (Delahaye et al. 2009).

Avoidance of dental disease in patients susceptible to endocarditis should be given more attention (Lockhart et al. 2008).

The revised guidelines related to antibiotic prophylaxis emphasize the importance of establishing good dental hygiene for this group.

A closer collaboration between all medical professionals is needed (Stecksén-Blicks et al. 2004, Rosén and Stecksén-Blicks 2007).

Cooperation between cardiologists, nutritionists, dentists with special training and general dentists could help improve dental care for these patients and improve general dentists' confidence in treating this group of patients. They must receive maximal preventive dental

care, to minimize the need for dental procedures and thereby antibiotic cover (Oliver et al. 2008). Parents of children with CHD should also be given information in how to prevent oral diseases in their child.

Children with CHD should have their first dental check-up when they are one to two years old and the check-up intervals should not exceed 6 months (Rasmussen 1999).

Several evidence-based guidelines for the prevention and management of dental caries in “at-risk” children are available and should be followed (Helsetilsynet 1999, Department of health 2007).

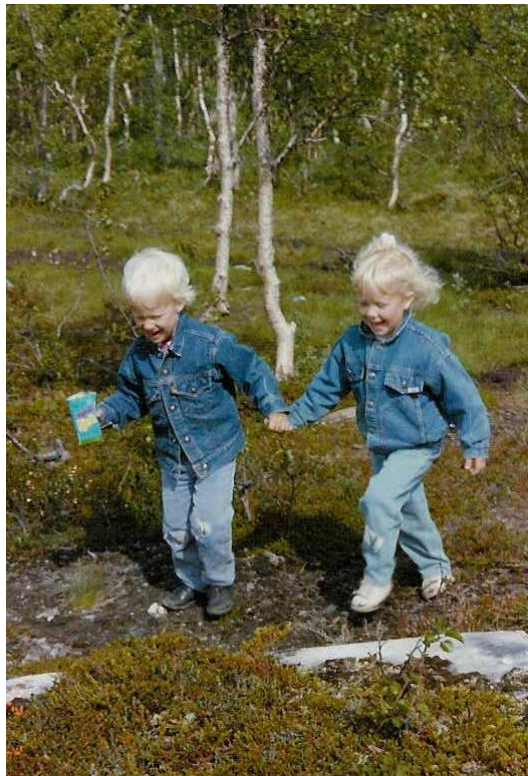


Photo: taken by Maryann Fløttkjær

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