Dental Concerns of Children with Cystic Fibrosis – An Overview

Nirmala SVSG* and Rupak Dasaraju
Department of Pedodontics & Preventive Dentistry, Narayana Dental College & Hospital, Andhra Pradesh, India

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*Corresponding author: Nirmala SVSG, Department of Pedodontics & Preventive Dentistry, Narayana Dental College & Hospital, Andhra Pradesh, India
E-mail: nimskrishna2007@gmail.com

Abstract
Cystic fibrosis (CF) is an autosomal recessive disorder occurring in one of every 2000 births. It is the most common genetic disorder affecting whites. The genetically altered protein affects exocrine gland function which leads to micro obstruction of the pancreas, which results in cystic degeneration of the pancreas and ultimately a digestive enzyme deficiency producing malabsorption of nutrients. These children have increased chest diameter, clubbing of fingers and toes, chronic productive cough, decreased exercise tolerance. Cystic fibrosis related diabetes is more common with high incidence of tooth discoloration, low incidence of dental caries, high incidence of mouth breathing and open bite malocclusion associated with chronic nasal and sinus obstruction. It should be preferred to treat in a more upright position to allow them to clear secretions more easily. Extractions can be carried out under local anesthesia. Use of sedative agents that interfere with pulmonary function should be avoided. Along with possible etiological factors, clinical features, diagnosis, tests for CF and treatment options for discolouration of the teeth are discussed.

Keywords: Cystic Fibrosis; Children; Dental Management

Introduction
Cystic fibrosis (CF) is a common autosomal recessive condition which typically causes sinopulmonary sepsis, and in most cases, pancreatic insufficiency with a resultant failure to thrive. It is first recognized in the 1930 [1], CF is one of the most common genetic diseases in worldwide. It affects both males and females and people from all racial and ethnic groups. However, the disease is most common among Caucasians of northern European descent and less common among African Americans, especially the pueblo of Zuni. About 12 million Americans are carriers of a faulty cystic fibrosis gene. It is caused by mutations of the gene situated on the long-arm of the seventh (7q31) chromosome coding. Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) which is responsible for the synthesis of cAMP - dependent membrane chloride channel located on the top surface of epithelial cells of exocrine glands [2]. Accumulation of the secretion in the outlet ducts caused by a dysfunction or lack of CFTR proteins leads to abnormal activity of exocrine glands, especially in the respiratory and alimentary tracts. Carbohydrates, the main dietary component, supply energy to the body, but at the same time are the major cariogenic agent. It is estimated that in Poland (like in most European countries) cystic fibrosis occurs in 1/2500 newborns and every 25th person is a carrier of abnormal CFTR gene, responsible for the disease [3].

Etiology
Although several hundreds of genetic mutations are known, the most common mutation in Western Europe is Delta- F508. The prevalence of the disease is one in every 2000 live new born children [4]. In essence; the disease affects all exocrine glands (including the salivary glands) and renders their respective secretions more viscous than under healthy conditions. CF patients consequently suffer from gastro-intestinal (due to pancreatic insufficiency) and respiratory problems. Because of this, they are compelled to have supplemental pancreas enzymes with every meal, drink and snack, supplemental vitamins (A, D, E and K) if necessary, frequent antibiotics (per os, via aerosols and IV) and daily expectorantia and mucolytica (mostly via aerosols) [5-10].

Signs and Symptoms
Symptoms often appear in infancy and childhood, such as bowel obstruction due to meconium in new born babies [11]. As the child grows, he or she will need to exercise to release mucus in the alveoli. The poor growth and poor weight gain despite a normal food intake in children. There will be accumulation of thick, sticky mucus leads to frequent chest infections, coughing or short breath. Congenital absence of vas deferens in males leads to infertility [12]. CF causes salty tasting skin and sweat will be very salty, as a result body loses large amounts of salt when they sweat. Hence it causes dehydration, increased heart rate, tiredness, weakness, decreased blood pressure, heart stroke and death which is very rare. CF can cause clubbing and low bone density lead to osteoporosis, and in rare cases it can present as a coagulation disorder. Young children are sensitive to vitamin K malabsorptive disorders because only a very small amount of vitamin K passes through placenta, leaving the child with very low reserves. Because factors II, VII, IX, X (clotting factors) are vitamin K dependent, low levels of vitamin K can result in coagulation problems [13].

Other signs and symptoms of CF develops later in life which may include infections that blocks the airways which causes frequent coughing, frequent botus of sinusitis, bronchitis, pneumonia, pneumothorax, Diarrhea, foul smelling, greasy stools, severe constipation pancreatitis, liver disease, diabetes, and Gall stones etc.

Other glands which may be affected are the salivary glands, the submaxillary gland may be enlarged, the secretion of the sublingual gland is thick and tenacious, and there may be destruction as in the pancreas. The parotid gland, which is not a mucus-secreting gland, may have an increased output with high electrolyte content [14].

Most important effect of the abnormal mucus is in the lungs. Here again there is obstruction by the thick secretion, causing a generalized emphysema which is usually persistent and secondary infection follows, due to Staphylococcus aureus. This chronic pulmonary disease occurs in almost all patients with cystic fibrosis and is often severe and progressive. Further complications of lung condition may be bronchiectasis, collapse of one or more lobes or lung abscess. There is a clinical point to note that the paranasal sinuses are often involved even in very young children and may give rise to symptoms [1].

Dental Caries
Due to its common incidence, dental caries belongs to social diseases. Its main etiological factor is the bacteria most frequently
transmitted by parents or caretakers in the early childhood [15]. Carbohydrates provided to the oral cavity together with food are the medium for cariogenic bacteria. As CF patients receive high carbohydrate diet, it is assumed that the cariogenic process can be more intensified in this group of patients. Various species of bacteria break down saccharides to acids, which leads to enamel demineralization and cavity formation. When oral hygiene is insufficient, dental deposits undergo mineralization and being transformed into dental calculus cause parodontosis [6]. CF patients, burdened with numerous ailments from many organs, receive various medications which affect the quality and quantity of saliva secretion. Moreover, their potential cariogenic effect on teeth is increased by sweeteners added to drugs to improve taste. Inhalers contain steroids which after long-term administration may cause oral mycosis, just like antibiotics taken in great amounts. Due to frequent infections of the upper respiratory tract, CF patients often breathe through the mouth, which promotes malocclusions and predisposes to periodontitis and inflammation of oral mucosa [17].

Literature reported reduced caries prevalence in 63 CF patients who were on long term oral antibiotics, compared with 56 of their siblings [18]. Primosch, et al. concluded that significantly reduced dental caries prevalence in CF patients compared with an age and gender matched control group, with a greater reduction in caries prevalence in the primary dentition compared with the permanent dentition. Previous studies suggested that there was a link between altered saliva properties and low caries experience, but found no relation between the changes in saliva and the severity of the disease process. The prevalence of calculus in children varies widely depending on patient demographics, social class, and general health. Patients with cystic fibrosis have altered amounts of calcium and phosphate in their saliva which can affect calculus formation [19]. Wotman, et al described increased amounts of calculus in children with cystic fibrosis and asthma compared with "healthy" children, particularly in the older age groups, which they felt reflected raised calcium and phosphorus levels in the saliva of these patients [20]. Kinirons reported similar trends towards a higher prevalence of calculus in CF patients that were positively and significantly related to age. Literature published in the 1980s and at the beginning of the 1990s claimed that CF youngsters (mean age seven years) had significantly less caries experience than control subjects This was attributed to their higher salivary buffer capacity, higher salivary calcium concentration and their frequent use of antibiotics [21].

### Enamel Defects

The term has been used to describe a range of appearances of enamel, the structure of which is disrupted during its formation or maturation phases; it represents defective mineralization. Enamel defects are the result of impaired development of dental enamel [22]. Clinically, hypomineralization affects the translucency of dental enamel and can also lead to chipping of the enamel, resulting in unprotected dentine and, as a consequence, the potential for more rapid caries development [23]. The integrity of the tooth enamel is an important factor in determining caries immunity or susceptibility [19]. The teeth most affected by enamel defects were upper incisors, yet first permanent molars are the teeth most affected by caries.

Enamel formation occurs in three stages: matrix formation, during which proteins involved in amelogenesis are produced; calcification, during which mineral content is acquired and the proteins are removed; maturation, during which the enamel is calcified and the remaining proteins are removed. Disorders in the early stages of enamel development evoke enamel hypoplasia, clinically detectable as fissures or enamel loss [24]. In contrast, disorders occurring in the calcification or maturation stage can cause hypomineralization [25].

### Diagnosis

It can be done by three methods as follows:

- **Newborn screening**
- **Sweat testing**
- **Genetic testing**

New born screening measures initially for raised blood concentration of immunoreactive trypsinogen.

Infants with an abnormal newborn screen need a sweat test in order to confirm the diagnosis of CF. In many cases, a parent makes the diagnosis because the infant tastes salty [26].

Sweat testing involves application of a medication that stimulates sweating such as pilocarpine. In order to deliver the medication through skin, iontophoresis is used.

CF can also be diagnosed by identification of mutations in the cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene.

Radiographs and computed tomography scans are used to examine the lungs for signs of damage or infection. Examination of spumut is required to isolate organisms. Pulmonary function tests to measure how well the lungs are functioning, and response to antibiotic therapy. Blood tests can identify liver abnormalities, vitamin in deficiencies and the onset of diabetes.

The most common complications are chronic respiratory infections which include coughing up blood, chronic respiratory failure, diabetes, liver disease, malnutrition, osteoporosis, arthritis, pneumonia, pneumothorax etc [27].

Management of CF continues throughout a patient’s life and is aimed at maximizing organ function; hence the quality of life will be enhanced.

The electrolyte level in the sweat may be raised several times above normal level and virtually diagnostic of this condition in children with chronic respiratory disease. Its major importance is that in an environment with a high temperature there is excessive loss of sweat with dehydration and depletion of sodium and chloride ions. The patient may vomit and further aggravate the dehydration. In extreme cases, without treatment, this can result in cardiovascular collapse, coma and death [28].

Treatment of patient with cystic fibrosis is directed primarily at control of the pulmonary conditions. Many of these children are on regular or frequent antibiotic therapy, often the broad spectrum antibiotics like tetracycline. This may be supplemented by aerosols to produce further concentration of the therapeutic agent in the lungs and to reduce the dryness of the atmosphere. Many bacteria are resistant to multiple antibiotics and require weeks of treatment with intravenous antibiotics such as vancomycin, tobramycin, meropenem, ciprofloxacin and piperalcillin. This prolonged therapy often require hospitalization and insertion of a more permanent IV such as a peripherally inserted central catheter (PICC line) or Port-a-Cath. Lung transplantation is considered when lung function declines to the point where assistance from mechanical devices is required or patient survival is threatened [29].

Physiotherapy is an important part of the treatment to promote drainage of the affected parts of the chest. The nutrition of the patient is supervised, as well as the pancreatic extracts he may need, he may be given vitamin supplements. Instruction is given
in the avoidance of conditions which produce sweating especially in summer, and the patient is expected to take additional salt with his meal in hot weather. If severe salt depletion occurs, this is an emergency requiring intravenous saline, and the patient should be admitted in the hospital.

Discoloration of dentitions by continuous or frequent tetracycline therapy since infancy is one of the problems which will need to be faced in these children. The discoloration is usually, though not always, severe and may be yellow at first turning grey brownish yellow later. Enamel hyperplasia is not a feature of this disease. When a patient with CF presents with a history of pain, and no obvious dental cause is discovered, the possibility of sinusitis should be investigated.

Management

Management includes treatment of bronchial inflammation and infection with antibiotics and physiotherapy, pancreatic enzyme replacement therapy, and fat soluble vitamin supplementation. A high calorie, high fat diet is recommended for these patients, and high sugar foods are often eaten to maintain the increased caloric intake needed. Cystic fibrosis patients have been reported to have abnormal dentitions, with dental effects related either to the disease itself or as a consequence of treatment [30].

Dental Treatment

When treating these patients in the dental surgery, excessive temperature of the environment should be avoided because of the problem associated with sweating. It is important that these children are kept free of chronic dental sepsis as part of their general care. Even if such a patient is on prophylactic antibiotics, sources of dental infection should be eliminated to keep him as possible and not add to the burden of coping with other fields of actual or potential infection.

1. Patients should be treated in a more upright position to allow them to clear secretions more easily.
2. Use of sedative agents that interfere with pulmonary function should be avoided.
3. Routine dental treatment offers few problems and there is no contraindication to local anesthesia.
4. Extractions can be carried out under local anesthesia normally, but
5. General anesthesia is essentially a matter for in-patient care, and should never be attempted in the dental surgery except where the pulmonary involvement is negligible and the patient’s physician has been consulted.
6. Children with cystic fibrosis have dry airways, and administration of inhalation sedation can be dangerous when the gases are not humidified. General anesthesia is also problematic because concurrent administration of anticholinergic drugs further aggravates airway dryness. If anesthesia is required for dental care, the patient should be hospitalized and managed by an anesthesiologist.
7. General anesthetics for adult patients with pulmonary disease must also be used with caution. Mild COPD or restrictive lung disease is generally not problematic.
8. Moderate to severe pulmonary disease can be aggravated and degenerate to severe respiratory distress when inhalation anesthetics are used. This is particularly so in conjunction with intravenous drugs that depresses the respiratory center of the central nervous system, and with anticholinergic drugs that may be administered during intubation.
9. When general anesthesia is necessary for patients in this category, they should be hospitalized and managed by an anesthesiologist. In addition, the dentist should consult with the patient’s physician prior to rendering treatment.
10. In a patient with severe pancreatic involvement, it would be wise to check the coagulation time of the blood before surgery, in view of the possibility of the lack of vitamin K.
11. Discoloration of the teeth by tetracycline is the most serious problem to be faced in the dental care of these children.
12. Most of them need aesthetic improvement as the appearance is quite unacceptable.
13. Acrylic caps appear to present the best improvement until the patient is old enough for full jacket crowns [31].

Conclusion

Even though Cystic Fibrosis is not very common in children but whenever pediatric dentist come across the case, he should be able to manage them with precautions to improve the quality of life of the children. Before advances in antibiotic therapy, physical therapy and nutritional supplementation is important and the median life expectancy of the children will be increased.

References


*Corresponding Author: Nirmala SVSG, Department of Pedodontics & Preventive Dentistry, Narayana Dental College & Hospital, Andhra Pradesh, India, E-mail: nimskrishna2007@gmail.com

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