Orthodontic treatment considerations in Down syndrome patients

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ABSTRACT

Background: Down syndrome is an easily recognized congenital disease anomaly, a common autosomal chromosomal anomaly with high prevalence of malocclusion. Orthodontic treatment demand should be high but it seems difficult to be done because of specific condition of disability. Purpose: The purpose of this literature review was to describe the orthodontic problems found in Down syndrome patients and several consideration that should be done to treat them. Reviews: Many studies report the high prevalence of malocclusion among people with Down syndrome. There is a greater frequency of clas III relationship, crossbite, crowding and also open bite. Several problems might appear in the treatment because of dental, medical, mental, and behavioural factor. Conclusion: It is concluded that orthodontic treatment can be performed in Down syndrome patient, although several difficulties may appear. Good consideration in mental, behavior, medical and also dental condition will influence whether the treatment will success or not. Special care and facilities will support the orthodontic treatment.

Key words: Down syndrome, orthodontic, malocclusion

INTRODUCTION

Down syndrome is the most common clinically recognizable syndrome characterized by generalized physical and mental deficiencies. It was firstly reported in 1866 by John L. Down and the prevalence is variative, in between 1 in 750\(^1\) and 1 in 1250 live births.\(^2\) Statistic in United States showed an increasing prevalence, currently observed at 11.8 per 10,000 births.\(^3\) The prevalence increases with the age of the mother.\(^4\)
nowadays there are approximately 300,000 people with Down syndrome, whereas in Jawa Timur every year there are 689 Down syndrome patients. In 1959 LeJeune, Gautier dan Turpin found the association between Down syndrome and chromosome 21.5 Normal person has 46 chromosomes, but 95% Down syndrome patients have extra chromosome #21. Nondisjunction problem makes total chromosome become 47; therefore it is also known as trisomy #21.6-8 There is a typical mongoloid appearance and virtually all patients have learning disabilities.7

Forty percents of Down syndrome patients can be accompanied with congenital heart problem such as mitral valve prolapse (MVP). Other medical conditions are upper respiratory tract infections, hepatitis, leukemia and even that 100% of person with Down syndrome over 35 years old develop neurological signs of Alzheimer disorder. Besides, there are still problem of speech and hearing, eye problem, and epilepsy.5

The prevalence and the variety of malocclusion on Down syndrome patients have already been studied. Study on 112 subjects with Down syndrome was found 92% of the subjects had malocclusion. Class III malocclusion was most frequently observed (43.8%). Crowding and unilateral cross bite were found in 15% of the subjects respectively. Bilateral cross bite was present in 5.4% of the subjects.9

Study on 57 children with Down syndrome in Mexico also found that the prevalence of anterior open bite was 31.6%.10

Down syndrome patients are like other normal children, who need orthodontic treatment. Parents of Down syndrome children in France often have difficulties to access oral health care and the patients are less likely to receive dental services.11 On the other hand, Down syndrome is categorized to a person with disabilities, so they need special management. Down syndrome often accompanied with systemic problem besides the malocclusion itself which require special attention. The systemic disease can influence the orthodontic treatment, but the orthodontic treatment can worsen the general condition. Another problem is the mental retard condition and behavior disorder that can disturb the communication between the patient and dentist. Motivation to treatment usually not come from the patient but from the parents, so support from the surrounding people should be always provided. Because of that, several considerations must be taken during the treatment so the goal can be achieved.

The purpose of this literature study was to describe the problems of orthodontic treatment for Down syndrome patients and several considerations that should be taken to overcome the problem, so treatment result can be obtained and the patient can get a better life.

Dental problem on Down syndrome patients

Generally, Down syndrome patients have several dental problems. It is important how they can get good oral hygiene. Study about the oral health condition of individuals with Down syndrome in Nigeria showed that the subjects had poorer oral hygiene, with no significant sex difference. This condition need frequent oral health assessment and routine treatment is expected to be done.12 Study of dental treatment needs of 300 children (aged 9–13 years) with disabilities in Melbourne found that 41 % of them required simple treatment; including caries treatment, periodontal therapy and oral health promotion.13

Dental anomaly is often found in Down syndrome patient. The incidence is about 73%. Congenital missing teeth is about 10x times more common in individuals with Down syndrome than in general population.14,15 with a higher frequency in males than in females. Agenesis occurred more frequently in mandible than in maxilla and most oftenly on the left side. The main components in the pattern of agenesis observed in Down syndrome are supposed to be related to the peripheral nervous system and abnormal cartilagenous tissue.15 Research through longitudinal panoramic radiograph in Canada showed that hypodontia was found in 92% of samples when third molars were considered and in 56% when third molars were not considered. The most frequently agenetic teeth were maxillary and mandibular third molars, maxillary lateral incisors, mandibular second premolars, mandibular incisors, maxillary second premolars, and maxillary second molars. Hypodontia was more prevalent and severe in females.3 Another study in Netherland found 59.6% of 114 Down syndrome patients had missing teeth. Absence of both mandibular central incisors was a high predictor for oligodontia. Congenital heart disease and hypothyroidism are parameters involved in tooth agenesis.16 The incidence of canine impaction is ten times higher than normal individual.17

Delayed eruption of teeth is often occurred, with an abnormal sequence. Primary teeth may not appear until the age of two, with complete dentition delayed until age 4 or 5. Some primary teeth are then retained in some children until they are 14 or 15.6

Abnormality in tooth formation such as microdontia (35-55%) is also seen in people with Down syndrome. Crowns tend to be smaller, and roots are often small and conical, which can lead to tooth loss from periodontal disease. Severe illness or prolonged fevers can lead to hypoplasia and hypocalcification.6,18 Taurodontism is also found in Down syndrome patients with the prevalence of 0.54–5.6%.19 It is characterized by a large pulp chamber and it is most commonly seen in molars.20

Malocclusion on Down syndrome patients

Compare with general population, malocclusion in Down syndrome patients is more frequent, more severe and more skeletally based which can adversely affect functions.21 Malocclusion is often found in Down syndrome patients particularly because of delayed eruption of permanent teeth and underdevelopment of maxillary arch, leading to poor positioning of teeth.6 Study on 136 Chilean children with Down syndrome, showed a higher frequency
of malalignment in both the deciduous and permanent dentitions. The frequency was higher in the permanent teeth than in deciduous dentition. Higher frequency of malalignment was found in the upper central incisor, lateral incisor, and canine regions.  

The maxilla of Down syndrome patients showed hypoplasia in the vertical plane and the sagittal plane. Sagittal maxillary growth is relatively constant from the age of 8 to 18 years, with an average increase of 0.12 mm/year measured at the level of point A. In the vertical plane it grows at an average rate of 0.62 mm/year and 0.70 mm/year, measured at the level of the anterior nasal spine and posterior nasal spine respectively.  

Down syndrome may affect the size and shape of the palate. Palatal length, width, and height were significantly influenced. The palate of Down syndrome patients may appear highly vaulted and narrow, due to the unusual thickness of the sides of the hard palate. This thickness restricts the amount of space the tongue can occupy in the mouth and affects the ability to speak and chew. This V shaped palate is caused by deficient development of the midface, and it affects the length, height, and depth of the palate, but rarely affects the width. Cephalometric measurement held in Canada showed the reduced dimension height of maxilla and mandible, short teeth, maxilla and mandible, forward rotation of maxillary and mandibular plane that lead to deep bite and mandibular prognatism.  

The characteristic of muscle hypotonia is found on perioral muscles, lip and chewing muscles and a protruding tounge, followed by active tounge protrusion or tounge thrush. A small oral cavity with a relatively large and fissured tongue leads to mouth breathing, which is a common cause of chronic periodontitis, xerostomia, drolling, angular cheilitis and halitosis.  

The prevalence of malocclusion on Down syndrome patients is quite high. Meštrović et al. found 92% children with Down syndrome had malocclusion, whereas study on 112 Down syndrome patients in Rio de Janeiro, Brazil showed a prevalence of malocclusion was 74%; cause by vertical and transversal occlusal alteration. Age, nail or finger biting habit, mouth posture, and cold or sore throat were the variables associated with the peavalence of malocclusion in these subjects. Maxillary anteroposterior hypoplasia makes 54% of Down syndrome patients have Angle class III tendencies. Craniofacial dysplasia that has already occurred at birth, become more severe with increasing age. An anterior open bite and class III malocclusion may be due to proclination of the incisor, under-development of the maxilla and a more anterior position of the hypoplastic mandible. Posterior crossbite occurred in 65% of patients due to maxillary transverse hypoplasia. A smaller maxilla also contributes to an open bite, poor positioning of the teeth and increases periodontal disease and dental caries. This finding is accordingly with study of Meštrović et al., who found class III malocclusion, crowding and anterior or posterior cross bite.

Systemic and mental problems in Down syndrome patients

About 40-50 % of the children with Down syndrome are born with a congenital heart disease, but most of them had received surgical correction within the first few years of life. The decrease in number of T cells can affects the immune system and contributes to a higher rate of infections and makes greater incidence of periodontal diseases, stomatitis, oral candida infection and gingivitis.  

History of the patients should be examined thoroughly. Patients can be asked to fill the questionnaire about their general health, because Down syndrome is often related to other health problems such as epilepsy, diabetes, leukemia, hypotiroidism, and also seven times higher as hepatitis virus carrier.  

Down syndrome patients have variation in intelligence condition. There are some patients with an IQ above 69, but the most typical is moderate or severe retardation (IQ 20-50). In early infancy they are in the range of low typical development, IQ decreases in the first decade of life and in adolescent years cognitive function reaches a plateau that continues into adulthood. Children with Down syndrome have more behavioral and psychiatric problems than other children. There are 17.6% of individuals with Down syndrome aged less than 20 years have a psychiatric disorder, most frequently a disruptive behavior such as hyperactivity disorder, oppositional disorder or aggressive behavior. Firstly, the parents are disappointed when they found their newborn is diagnosed with Down syndrome. However, within a few months, they become attached, because child with Down syndrome is happier and more loveable.  

Other problems are speech and language difficulties that affect communication, although research showed that there in no association existed between speech disorders and anterior open bite in Down syndrome Mexican children. There is individual variation in the delay of language acquisition in children with Down syndrome. Patients have great difficulty in communicating with people who do not know them well and their ability to develop the relationship with society is limited. Parents, patient’s family or caregiver will be able to help reaching better communication with the patients. It is important that the dentist communicate directly with the patient in order to build a level of trust to improve their confidence. Maybe it will need more time for explaining the orthodontic procedure, but if Down syndrome patients has already willing to be treated and trust the operator, they can be cooperative.  

DISCUSSION

A child with Down syndrome is a handicapped child with disabilities and special needs. Actually there should be a high orthodontic treatment need for Down syndrome patients because of an increased prevalence and severity of malocclusion. Nevertheless, orthodontic services for
Orthodontic treatment in Down syndrome patients is a specialized care that requires consideration of the unique needs and characteristics of the patients. The handicapped children, including those with Down syndrome, often require orthodontic treatment to address their dental and facial anomalies. This treatment involves a multidisciplinary approach, considering the overall health and social context of the patient.

Orthodontic treatment is benefiicial, but its effectiveness varies among patients. It is possible to achieve a significant orofacial function increase after treatment. However, the success rate of orthodontic treatment in Down syndrome patients is lower than in normal children, suggesting the need for a longer treatment time and more frequent visits. This may also affect the motivation of the patients themselves and their caregivers.

The treatment plan may need to be adapted to each individual's condition, considering the increased apprehension, short attention, and limited tolerance. Communication with the patients should be clear with the tell-show-do technique, so the children can understand what is happening. Patients and caregivers play an important role in keeping good oral health and hygiene well, and avoid hard and sticky child. Parent's agreement may be advantageous to the treatment.

Communication to the patients with Down syndrome patients is a challenge due to their increased apprehension, short attention, and limited tolerance. Motivation from the patients themselves is often not clear. It must be realized that there is still possibility of treatment failure due to patient's limitation. In this condition, maybe the objective must be changed from ideal condition to compromise, only to achieve esthetic improvement in order the child can be easily accepted in social community. Another treatment objective is to overcome the narrow palate which can influence the general health. It is reported that there was positive improvement on oral motoric and articulation function and also there was significant orofacial function increase after treatment with palatal plate.

To several persons, orthodontic treatment will be benefiicial, but to another persons may be not. Nevertheless, Down syndrome is not always a contra indication to orthodontic treatment. Orthodontic treatment still can be done in selected patients. If it has been decided to do orthodontic treatment, attention must not directed towards the malocclusion itself, but several considerations and good care should be taken particularly if associated with systemic, mental and behavior condition of the patients.

During orthodontic treatment there may be systemic condition that affects the oral environment. Child’s health history and clinical description should be well recorded and remembered to acknowledge the sensitivity of the patients. About half of the children with Down syndrome are born with a congenital cardiac anomaly. Orthodontist should be aware about the sign and symptom manifestation in oral cavity that probably associated with specific disease. Sometimes multidisciplinary approach is necessary to prevent bacterial endocarditis. There are always some possibilities of infections such as upper respiratory tract infections, hepatitis, epilepsy, leukemia, and other conditions, so medical consultation with the internist and pediatrician should be done. Premedication like antibiotic prophylaxis, vitamin and improved immune response are needed to be done.

Mental condition of the patients may be the most important factor that influences orthodontic treatment procedures. Parent's agreement may be advantageous to support their psychological condition. Parent’s ability to collaborate with the orthodontist is usually helpful for successful completion of treatment. It is important to always remember the wise advice that: “A person with a disability is still just a person who deserves to be treated with dignity and respect. We should never assume someone doesn’t understand what we are saying because they don’t speak as well as we do.” Based on that advice, better communication with the patients should be achieved. The hearing loss experienced by many of these individuals should be taken into consideration when communicating with the patient. Good doctor-patient relationship will increase the confidence level of the patients to overlook the difficult time during orthodontic treatment.

Behavior disorder is often a problematic in treating Down syndrome patients because of lack of understanding, increased apprehension, short attention, and limited tolerance. Communication to the patients with Down syndrome who usually has intelligence below average should be always developed, so good doctor-patient relation can be achieved. To overcome the communication problem, every phase should be done slowly, in order not to shock the patients. Every step that will be done should be explained clearly with tell-show-do technique, so the children brave enough to experience the treatment. In every step, operator should always think about the comfort of the patients. Maybe they require more chairside time and there will be an increased number of appointments.

Orthodontic treatment can be done if the patient is cooperative. This behavior is needed because orthodontic treatment is multivisit and need a long periods. The patients should keep their appointment to control, keep their oral hygiene well, and avoid hard and sticky child. Patient's ability to keep their oral hygiene is required to gain good treatment result. It is advised to take frequent drink or oral rinse to minimize the dry mouth sensation. Caries frequency in Down syndrome patients can be minimized with preventive measures such as fluoride topical application, fissure sealant, fluoride tooth paste suggestion and non cariogenic food and beverage consumption. Parents and caregivers play an important role in keeping good oral hygiene, maintaining the diet and consumption pattern, and going to the dental office to get routine treatment. Failure in keeping oral hygiene can increase the possibility of gingival hyperplasia, periodontal disease, and bacterial endocarditis risk.

Orthodontic treatment is started with impression and radiographic taking. The first problem is how to get the study models. Down syndrome patients often have strong gag reflex due to tongue condition (macroGLOSSIA) and anxiety or phobia. This condition usually can be overcome with simple explanation and good communication. Impression must be taken as quickly as possible, with fast set type or low viscosity impression material. This kind of material needs skillful and fast working operator. Failure in the first impression may increase the difficulty to get the second impression and so on. If it is not possible, impression can be taken under conscious sedation, intravenous sedation or even with general anesthesia.
Panoramic radiograph is considered to be useful for orthodontic assessment. However, it requires patient’s cooperation, the patients stay still minimally during the rotation of the tube of the panoramic radiographic machine. Down syndrome patients often have difficulty to stay still because of uncontrolled head and limb movements. Problems will also arise if frightened child should be restricted in a cephalostat since it will increase their fear and even generate panic. Periapical photo maybe more difficult to obtain since the film must be held intra orally during the x-ray exposure and this procedure may be uncomfortable and increase the anxiety of the patients. In certain cases, computed tomography (CT) scan can be performed under sedation.21

Generally, orthodontic appliance is bulky, uncomfortable and painful. On the other hand, the appliance needs day-to-day maintenance.28 These conditions will make the patients reluctant wearing the appliance. The treatment should be planned in a simple way, not in a hurry to reach the goal, slowly but sure to get the improvement.

It was reported that orthodontic treatment problems with fixed appliances were more difficult than with removable appliances. Treatment period with fixed appliance was harder on 47% patients, compared with 11.8% removable appliance users.28 The problems of wearing removable orthodontic appliances are mastication and speech difficulties. It is beneficial that adjustment of removable orthodontic appliances can be done extra orally. Furthermore, oral health maintaining is better with removable orthodontic appliances. Successful orthodontic treatment with removable orthodontic appliances has already reported by Becker and Shapira.33 In severe malocclusion cases, combination treatment with fixed appliance may be needed. From the doctor’s point of view, orthodontic treatment with fixed appliance is hard to be done. The patients should sit in dental chair for a long enough periods of time and the teeth must be dry during bracket placement. Sedation or general anesthesia will make the procedures easier to be done, as started by Jackson in 1967.30 The problem is that the equipments for sedation are rarely provided in dental private practice. It should be done in a clinic or more sophisticated hospital.

It will be easier if bracket placement is done indirectly, since it can be accurate and the process will be less time consuming. Thus, the most time consuming step should be done out of the mouth including steps in dental laboratory. Light forces should be given to move the teeth. It’s not advisable to give heavier forces, since it will make the patient more uncomfortable. Straight wire technique can be chosen because it has minimal wire changes. Treatment with self ligating bracket is also beneficial since it can reduce the visiting time and no ligature wire or elastomeric modules make teeth brushing easier to be done.

Maintaining oral hygiene is more difficult with fixed than with removable orthodontic appliances. Because of that reason, it is recommended for Down syndrome patients to wear removable orthodontic appliance rather than fixed appliances. Fixed appliances should be worn only in limited period. Besides of that, bonding procedure must be done correctly so it is not easily debond. Replacement of bracket will difficult and need more time.21 Activation, wire changing, and other procedures can give unpleasant sensation because of many instruments used inside the mouth.

Fixed appliance is the only choice of treatment for non cooperative Down syndrome patients, since the patients are unable to remove the appliance by their own efforts. Nevertheless, the immature condition can make the patients do something that damage the appliance, so it can not work properly and even hurt the oral mucosa.

After active treatment has finished, retention period is needed in order to maintain the treatment result. In children with skeletal discrepancies, or with large tongue, non elimited bad habits, treatment result can not be maintained and tends to relapse. To achieve long period of stability, removable retainer can be used, but it depends on the willingness of the patient to wear it. Retainer such as Hawley retainer, wraparound, or clear retainer can be used.34,35 If the cooperation is doubtful, it is better to use permanent retainer such as bonded lingual retainer. In cases with hypodontia, if there are still residual spaces after the treatment is completed, prosthodontic approach is needed for planning the suitable denture. Successful orthodontic treatment is not only increase the esthetic factor but also improve the other function including swallowing, speech and mastication.

Failures can occur because of unpredicted condition such as: the patients become ill, uncontrolled behavior and inadequate oral hygiene.31 It is recommended to remove the appliance in these conditions, although the treatment result has not achieved yet in order to prevent further negative effects.

It is concluded that orthodontic treatment still can be done on selective Down syndrome patients, although some difficulties may occur. Medical, mental and behavior condition besides the malocclusion itself will affect the treatment result. Several considerations, skillful orthodontist and chairside, specific effort and facilities such as sedation equipment and well trained operator are needed to gain the treatment objectives.

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