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Management Protocol for Patients with Down syndrome associated with Alzheimer's disease

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Abstract

Down syndrome is one of the most common chromosomal alterations and due to the incremental in the oval life expectancy and its relationship with Alzheimer disease; the professionals have to be aware of the oral and behavioral characteristics to give patients an excellent treatment. The discovery of this recent relation between Down syndrome and Alzheimer disease, made us to evaluate 30 patients aged between 45 and 60 years old diagnosed with Down syndrome that were treated in a Dental Clinic for handicapped, in order to determine the changes associated with ageing in patients with this disease refereed by their relatives. 25% of the relatives observed behavioral and memory changes. With these patient's oral conditions and the health records it was designed a management protocol for give them and adequate treatment that increase their life quality.

Keywords: Down Syndrome; Alzheimer Disease; Dental Treatment

Introduction

Dr. John Langdon Down was the English physician who first described this syndrome in 1866. Nowadays, this is the most frequent chromosomal alteration; it occurs in one of 800 live births, in all races and economic groups. Down Syndrome (DS) involves an extra copy of chromosome 21 or part of it. Only a few decades ago, many young with DS were viewed as eternal children. Nevertheless, times have changed. Life expectancy has tremendously increased from 9 years in 1925 to over 40 years today. For that reason, the patients with DS have increased their needs and the professional must be prepared to face this fact [1-3].

Alzheimer's disease (AD) is a progressive neurodegenerative disorder characterized by synaptic dysfunction and accumulation of amyloid-beta (Aβ) peptide, which are responsible for the progressive loss of memory [4]. Historically, the discovery of the relationship between Alzheimer´s disease and Down syndrome evolved over 75 years, as results of various studies. The first scientific publication to call attention to the issue of premature aging and senility in individuals with Down syndrome by Fraiser and Michell who found that among a group of 62 adults with Down syndrome, a certain proportion died of precipitated senility. Jervis was the first to report a direct relationship between the neuropathological signs of Alzheimer's disease and dementia of Alzheimer´s type in adults with Down syndrome [5]. Nowadays, the researchers have shown that alterations in chromosome 21 are also seen in Alzheimer's disease (AD); in other words, this may be an early-onset which tremendously complicates the management of this type of patients. In DS, triplication of the Amyloid Precursor Protein (APP) gene results in increased brain levels of Aβ based upon evidence from human studies, mouse models, and induced human pluripotent stem cells [6,7].

Materials and Methods

An observational transversal study was made, in which 30 individuals with DS and their health records were evaluated to identify all the information of their oral health and their medical problems. This study was made at a Dentistry Center for Disabled People in Valencia, Spain. Also, an interview was made to their relatives for determinate the changes associated to AD. Every patient was older than 30 years old, so what we would be able to determine the relation between DS and the early developmental of AD. All patients who signed, or whose legal signed an informed consent accepting to be part of the study, were considered in the inclusion criteria. The ethical approval for the study was granted by the Bioethics Committee of the School of Dentistry of Valencia

University. After the evaluation of the patients and their medical records, it was made an extensive review of early papers related to the topic. Finally, with all the information obtained, it was designed a specific protocol for the management of this behavior and the treatment for pathologies associated.

Results

A variety of changes associated with ageing were identified in the stomatognathic system and the general heath of the patients studied. The relatives of the 25% of the patients described changes in the behavioral and the memories of the patients.

The Clinical Findings of the Oral Conditions Associated with DS and AD Were

- Area condition narrow palate
- Soft palate insufficiency oral opening angle of the mouth pulled down.
- Lower lip everted.
- Mouth breathing with drooling.
- Chapeled lower lip.
- Macroglossia, tongue scalloped and fissured.
- Protrusion and tongue thrusting.
- Desiccated tongue (result of mouth breathing)
- Number and size alterations
- Agenesia.
- Hypoplasia and hypocalcification.
- Delayed eruption.
- Periodontal Increased risk of periodontal disease.
- Frequent malocclusions.
- Frequent temporomandibular joint dysfunction.
- Bruxism.

For that reason, a specific protocol was designed to guide the dentist to an appropriated schedule of treatment. To comprehensively assess a patient with DS and AD, the stage of the AD must be evaluated and should be classified as early, moderate or advance as can described below:

Early Stage AD

- Demonstrate to patient and/or caregiver how to perform preventive dental procedures such as brushing, flossing and application of topical fluoride gels and chlorhexidine.
- Establish an aggressive recall program for the patient, including the use of topical fluoride
- Short-acting benzodiazepines may be needed to provide dental care.
- Treatment plan should anticipate a decline in the patient's oral health over time. Possible sources of acute or chronic pain should be treated.
- Most dental procedures can be performed; however, fixed prosthesis are generally preferred over removal ones to avoid injuries.
- Antibiotic cover for surgical procedures:
  Amoxicillin 500 mg one hour before the treatment (50mk/kg)
  Clindamicin 600 mg one hour before the treatment (30mk/kg)
- Positioning of the patient: Protection against concussions. Patients could become rigid, often requiring a nursing facility.
- Timing of appointments: short appointments are recommended.

Moderate Stage AD

- Patient may become more uncooperative due to progression of the disease.
- Sedation with a short-acting benzodiazepine often will be indicated.
- Short appointments are usually indicated.
- Treatment becomes more maintenance – oriented than completed rehabilitation.
- Provide instructions and prescriptions for the treatment of xerostomia, candidiasis, or acute infection f they occur.
- Avoid drug interactions with drugs the patients may be taking.

Advance Stage AD

- IV sedation may need to be used for the dental treatment.
- Complex or time-consuming dental treatment should be avoided.
- Diagnose and treat oral conditions associated with the patient's physician regarding their status and presence of other medical conditions.

After the development of this protocol, it was applied for a period of six months with all elderly patients with diagnosis of Down syndrome who attended the Center for Patients with disabilities. Each one of them was located according to their characteristics within the stage that corresponded to him and the steps indicated in the protocol for the development of the treatment were follow. This facilitates to the dentist, the management of the patient with this condition.

Discussion

Down syndrome is the most frequent chromosomal alteration in humans due to an extra copy of the chromosome 21. These patients suffer from many diseases, such as cardiovascular diseases, endocrine alterations, increased susceptibility to infections, immune defects and mental retardation [1-3]. Most recent researchers have discovered that the alterations of chromosome 21 are also seen in AD, together with several of the neuropathological features also present in DS such as reduction of nerve cells, changes in the phospholipid composition membranes, neuritic plaques, neurofibrillary tangles, degeneration of the basal forebrain cholinergic neurons and dementia [7-10]. In fact, People with DS develop Alzheimer's disease at higher rates and a younger age of onset compared to the general population [11]. Clinical features of dementia in people with DS are like dementia in general population: loss of memory, cognitive decline, changes in adaptive behavior, neurological changes and language difficulties [8,9]. The prevalence of dementia in people with DS is about 3.4% in their 30s, 10.3% in their 40s, and 40% in those over 50 [12,13]. The consensus from a number of studies is that 50-70% of DS individuals will develop dementia by ages 60-70 years [14]. DS and AD share some of the physiopathological characteristics of ageing and DS is considered a precocious and/or accelerated model of senescence. The dementia in patients with DS appears after the age of 35 and occurs only in 15 to 45% of people with DS with a mean age at onset for dementia of 51.7 years [15]. One of the limitations of the study was that all patients were taken from the same center of patients with disabilities, which limits generalizability. Therefore, it is recommended that the sample be extended in a next research. Oral characteristics in patients with DS are a narrow palate, mandibular protrusion that origins a lot of malocclusions; macroglossia, dental agenesis, number and size alterations, bruxism and an increased risk of periodontal diseases. These conditions are most difficult to treat when the DS and AD are shown together. The medical complications in ageing patients create the necessity of efficient treatment of the oral conditions because the life of the patient is in stake [16].

Conclusion

The World Health Organization estimates that there are approximate 170 million people with mental retardation worldwide. In other words, nearly 3% of the world's population has some form of mental retardation, being Down syndrome the most common genetic condition. It has been estimated that 2/3 of this population remains without an adequate dental treatment. Their life expectancy has increase tremendously over the recent years as a result the development of the typical neuropathologic hallmarks of Alzheimer's disease. Dentists should be prepared to treat AD in patients with DS contribute to increase dental incidence of caries and periodontal disease. To treat a patient with this condition and not having an accurate knowledge might put the life of the patient on risk. By following a specific protocol of treatment, the professional can realize success in the general treatment of these patients. The goals of the treatment planning for these patients are to maintain oral health, comfort and function, none the less, to prevent and control oral diseases.

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