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Review Article

Obstructive Sleep Apnea in Down Syndrome Children

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Abstract

Down Syndrome (DS) is the genetic alteration in chromosome 21 which presents specific characteristics and individual conditions significantly interfering with their stomatognathic system. Craniofacial aspect assessments are of substantial importance to the complete diagnosis and the interdisciplinary treatment planning provided to this syndrome carriers. Changes such as midface underdevelopment, micrognathia, muscle hypotonia, mouth breathing and macroglossia, narrowing at the nasopharynx and adenoid and tonsil hypertrophy are frequent respiratory obstructions during the sleep. The Obstructive Sleep Apnea Syndrome (OSAS) strongly interferes with these children quality of life and an interdisciplinary procedure including preventive approach and early orthodontic intervention seems to be useless to minimize or eliminate such problems. The current article aim to the main SD features, their correlation with OSAS as well as the SD diagnostic approach, treatment planning and orthodontic interventions. Search and analysis were performed in scientific articles and bibliographic databases concerning the link among Down syndrome, Syndrome Sleep Apnea and orthodontics. The following inclusion criteria were adopted: articles written in Portuguese, Spanish and English; indexed in LILACS, SciELO and PubMed databases and published between 2005 and 2015 (totaling 29 references). It was concluded that Down Syndrome children who have OSAS should be better assisted by trained professionals according to a bio-psycho-social context, an interdisciplinary health care system and to the therapeutic procedures that can be early applied to these patients. It must be done in order to improve their quality of life and to minimize losses that may become an irreversible issue.

Keywords: *Down syndrome Child; Obstructive Sleep Apnea; Orthodontics; Oral health; Quality of life*

Introduction

Down Syndrome (DS) or Trisomy 21 as the most common chromosomal abnormality found in the human species with an incidence of one in six hundred to one thousand births presents mental and physical impacts of defects on children health and especially on the stomatognathic system [1,2].

Actually, changes such as muscle hypotonia, midface underdevelopment, high-arched and atresic palate, relative macroglossia, narrowing at the nasopharynx, and adenoid and tonsil hypertrophy can be detected in these patients and they lead to frequent respiratory obstructions during the sleep [2,3].

The Obstructive Sleep Apnea Syndrome (OSAS) in children is characterized by recurrent episodes of partial or complete upper airway obstruction during the sleep which affects air flow between the environment and the lungs conducting to lack of oxygen supply to the body and sleep fragmentation. Accordingly, these episodes lead to systemic implications [4,5].

Sleep disorders have great impact on the lives of children, especially on those with intellectual disabilities. Decreased cognitive function, attention deficit, poor sleep and daytime sleepiness can be then observed in these patients [6]. In such cases, an early diagnosis and collaboration between a professional team is essential.

The evaluation of children with Obstructive Sleep Apnea Syndrome suspicion in dentistry depends on medical history, anamnesis, checklists, clinical details and physical examination [4,7].

Teleradiographic images requested for diagnostic reasons seem to be insufficient to define OSAS severity. The polysomnography, which is performed in the sleep laboratory, is considered to be the key to sleeping disorder diagnoses. It also works as treatment control [6,8,9].

Once OSAS is confirmed, the therapeutic approach must follow the etiology of the obstruction and the intensity of the clinical symptoms. Adenoid and tonsil hypertrophy are the most obstructive factors in children; despite the other associated conditions found in Down syndrome children. The absence of a multidisciplinary and knowledgeable professional team can favor therapeutic flaws [7,10].

It is worth understanding the obstruction etiologies to achieve the goals of the suitable treatment. Many surgical techniques such as the adenotonsillectomy, the uvulopalatopharyngoplasty, the tracheostomy and the maxillomandibular surgery may be indicated as OSAS treatment [11].

Faced on obstructive Sleep Apnea Syndrome, the orthodontist has the opportunity to early intervention in patients using orthopedic devices such as jaw expanders. In fact, it increases nasal cavity and jaw size in order to favor airflow [9,12].

Therefore, the current article addresses the main features and conditions of Down syndrome children as well as their correlation with the Obstrutive Sleep Apnea diagnosis. The medical management and the orthodontic interventions applied to Obstructive Sleep Apnea Syndrome are discussed through literature review as a way to find the best treatments to reduce the damages suffered by these children.

Methods

A search on published studies about the Down Syndrome genetic condition, the Obstructive Sleep Apnea Syndrome (SAOS) and about dental orthodontics expertise was conducted. The literature search strategy was based on the following meshes: Down syndrome, Obstructive Sleep Apnea Syndrome in children, and Obstructive Sleep Apnea Syndrome in Down syndrome patients and Orthodontics in Down syndrome children. The abstracts of the selected articles were analyzed in order to check if the articles would meet the inclusion criteria.

Inclusion criteria were: articles written in Portuguese, Spanish and English indexed in LILACS, SciELO and PubMed databases and published between 2005 and 2015. Exclusion criteria were: articles, theses and dissertations with no information about the herein analyzed theme. The search in the databases provided 29 references that have met the established inclusion criteria.

Literature review

Down Syndrome

Patients with special needs are those who have to live with diseases and/or health conditions that require special assistance due to the mental, physical, organic, social and/or behavioral changes caused by them [13].

Down syndrome (DS) or Trisomy 21 is the most common chromosomal abnormality found in humans. It shows incidence of one case for six hundred to one thousand births regardless of race and economic group. Great progress in the physical and mental treatments provided to children with this syndrome has been seen in the recent years and it resulted in significant increase in these patients survival and social integration [2,14].

These children face mental and physical retardation as well as physical changes due to their extra genetic material. These medical conditions can differ from child to child [1,13].

The dental-maxillofacial features found in Down syndrome children alter their entire stomatognathic system. Such changes have implications in different aspects such as speech, eating, posture, ventilation, aesthetics and social integration. Adenoids and tonsils hypertrophy, middle face floor underdevelopment (including the maxilla and the jaw) and craniofacial dysplasia (which gets worse with age) are found in these patients. They lead to decreased tonus in the anterior open bite, high-arched palate, small mouth and macroglossia (Figures 1, 2) [2,14].

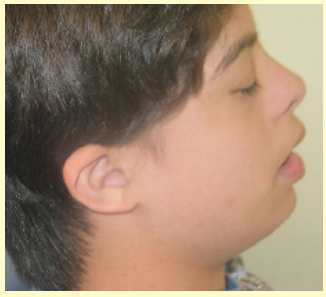

Figure 1: *Craniofacial features of a Down Syndrome child.*

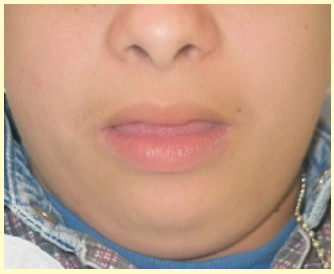

Figure 2: *Middle-third face features and tongue position in a Down Syndrome child.*

General Considerations about the Obstructive Sleep Apnea Syndrome (OSAS)

Sleep is a natural and recurrent detachment and unresponsiveness state from the organism towards the environment. Many neurobiological processes take place during the sleep to keep the physical integrity of the body [7,15].

Sleep disorders can affect any individuals in any age group, including children. They cause great impact, especially on intellectually disable children. These disorders include decreased cognitive function, attention and memory deficits, sleep deprivation, daytime sleepiness, bruxism, night drives, snoring and interrupted breathing [6,16,17].

The Obstructive Sleep Apnea Syndrome (OSAS) in children is characterized by recurrent partial or complete upper airway obstruction episodes during the sleep. Such episodes involve the functioning of 30 muscles. Sleep apnea is a combination of structural and neuromuscular factors that result in the relaxation and narrowing of the airways as well as in the reduction of the airflow (Figure 3) [4,17,18].

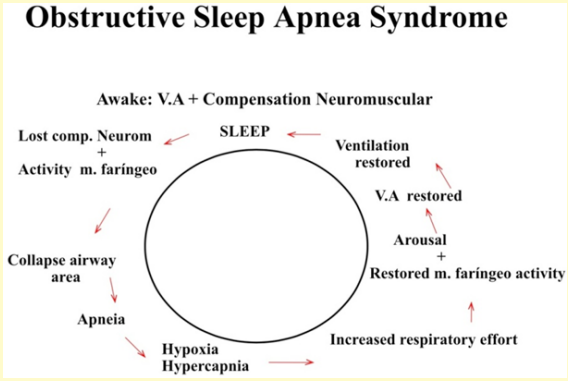

Figure 3: *The pathological sleep cycle in the Obstructive Sleep Apnea Syndrome.*

Children with neuromuscular and neurocognitive abnormalities present higher respiratory obstruction incidence due to in coordination, weakness or spasticity in their pharyngeal muscles, especially in the oropharynx. Other structural factors such as retrognathia and micrognathia, mandibular hypoplasia, atresia or high-arched palate, whenever found in Down Syndrome children increase their chance to suffer airway collapses during the sleep [3,5].

Sleep disorder diagnosis

The Obstructive Sleep Apnea Syndrome is considered as a disorder capable of fragmenting the sleep architecture, leading to functional, neurological and psychosocial consequences. A multidisciplinary professional team is extremely important to provide better and more complete assistance to patients affected by this syndrome [7,15].

To investigate sleep disorders, the diagnostic methods include specific questionnaires (Epworth), anthropometric and physical examinations, radiographic and CT scans, actigraphy, day or night polysomnography and oximetry. Actigraphy and oximetry are techniques applied through a device placed on the fist of the patient. These examinations assess the sleep-awake cycle, the total sleep time, the number of awakenings and the sleep latency [19-21].

None of these methods replace the polysomnography, which is a non-invasive procedure. In fact, it is the key to a successful OSAS diagnosis. The polysomnography is used to control parameters that must be measured during nightly spontaneous sleep [22].

However, taking Down syndrome or insomniac children to sleep in the laboratory may be a tricky strategy. Therefore, simpler methods are required to diagnose Obstructive Sleep Apnea Syndrome [9,10].

The diagnostic is confirmed when the apnea-hypopnea index is greater than one event per hour and the oxyhemoglobin desaturation is lower than 92%. It is extremely important to set the obstruction area, since such obstruction reduces the velopharyngeal space (soft-palate pharynx) and tends to occur in approximately 86% of the cases [8,23].

Treatment approach

The treatment given to sleep disorder children requires a multidisciplinary approach involving doctors, dentists and other health professionals [17]. Although there is no ultimate therapy against OSAS, the treatment must attain basic aims such as symptom relieve, morbidity reduction and improved quality of life [7,24].

When it comes to the therapeutic aspects, we should take care of obstruction etiology and the intensity of the clinical symptoms. These procedures include not just the surgical interventions (to rule out obstruction causes) but also the sleep hygiene advises (weight loss; no heavy food intake before going to sleep) [10,25].

Adenotonsillar hypertrophy is the most common OSAS cause in children [17]. But Down syndrome children present features which are associated to sleep disorder; therefore, a complete and comprehensive therapy is required. The association of surgical techniques such as tonsillectomy, uvulopalatopharyngoplasty, mandibular advancement and tongue reduction may improve these patients quality of life [12,26].

The adoption of less invasive measures such as the use of orthodontic appliances becomes a treatment option in case of early diagnosis. Intra-oral orthopedic devices such as jaw expanders and mandibular distractors enlarge the oral cavity boundaries (best tongue accommodation) and improve the airflow in the nasal cavity [27,28].

CPAP (a small compressor with naso-facial mask using positive pressure) is indicated to cases in which there is non-cooperation by patients or failure in other therapeutic procedures [11,29].

Results and Discussion

Sleep is a natural and necessary state for human beings who is highly valuable for health and where neurobiological processes take place keeping physical and cognitive aspects of the organism. As a consequence, it is very important to study its aspects and factors that can affect or interrupt this period of state such as OSAS [4,15].

According to the literature, The Obstructive Sleep Apnea Syndrome is found in all age groups and presents different clinical aspects. There are many factors including: anatomical, functional, neural and genetic factors which interact in its occurrence [6,17,24]. According to Uema., *et al*. it reduces the cognitive function, thus leading to attention and memory deficit, sleep deprivation and daytime sleepiness and it may be associated with bruxism and night drives, as well. The mentioned syndrome is characterized by partial or complete obstruction in the upper airways during sleep and it leads to airflow reduction [4,16-18].

Matthew and Stevens, Kissman., *et al*. point out that syndromes, mainly the Down Syndrome (DS); as well as craniofacial and neuromuscular abnormalities predispose OSAS and respiratory infections and obstruction [3,5].

According to Rodrigues., *et al*. Chang and Chae this pathology is featured by apnea events, oxygen saturation fall, tachycardia bursts and hypertension. Thus, it is worth setting the obstruction point in the upper airways during the physical examination of apneic children [21,23].

Studies showed that the clear diagnosis of sleep disorders is based on gathering basic details and on the participation of several health professionals in the multidisciplinary treatment team [7,15].

Therefore, exams and tests are necessary to establish an accurate diagnosis and they include anamnesis, physical examination, the application of specific questionnaires (Epworth), radiographic images and sleep-wake records such as actigraphy, oximetry and polysomnography [10,20,25].

According to Asensi., *et al.* Ng., *et al*. Gondim., *et al*. and Kissmann., *et al.* , the polysomnography is considered to be the best method to assess sleep-disordered breathing and the elective method to evaluate these patients' quality of sleep. The thorough examination comprises ten monitoring steps: electroencephalogram, electromyogram, electro-oculogram, ECG, airflow, respiratory effort, snoring, patient and decubitus position, the movement of the lower limbs and oxygen saturation, video recording (if possible), esophageal manometry and capnography [5,9,11,22] .

Therefore, there are some alternative methods such as oximetry and pulse actigraphy which are measured in smaller and less complex displays and which do not affect the patients' sleep routine [20,21].

Asensi., et al. Souza and Cavalcanti point out the different factors associated to obstructive sleep disorders that have turned the OSAS treatment into a challenging task. Such task should take under consideration the anatomical and clinical changes, the individual cases as well as the severity and risk factors of the disease [10,24].

According to Levrini., *et al*. Asensi., *et al*. the treatment given to children with craniofacial abnormalities such as Down Syndrome, cerebral palsy or muscular hypotonia is more complex and involves surgery guidelines concerning the sleep hygiene [11,25].

According to Kissman., *et al*. the orthognathic surgery is presenting better results with success rate of approximately 92%. Patients need to be carefully evaluated for psychological aspects, mainly those presenting Down syndrome as well as family consent [5].

Less invasive options are the most used in early diagnosis. The positive pressure device (CPAP) is effective for patients who have not responded well to the surgical procedures or for more serious craniofacial anomaly cases, which are not recommended to surgical indications. Recent studies showed that the preliminary use of CPAPs is indicated for children who will be later subjected to any surgical procedure. The device is considered to be a continuous and undefined therapy [11,17,20,23,28].

According to Moreira., *et al*. Levrini., *et al*. Guimarães., *et al*. and Millán and Reyes, intra-oral appliances are excellent alternatives available for early and preventive treatments. The use of orthopedics as early as possible enlarges Down Syndrome patients' jaw bones, improves their nasal (airflow) and oral (tongue accommodation) cavities and helps the breathing process [12, 27-29].

From the analysis and interpretation of studies related to OSAS and Down syndrome, the best option and treatment indication is the realization of multidisciplinary planning with previous use of orthodontic appliances for these disabled children [2,7,10,11,14,17, 20,22,24,27,29].

Conclusion

Down syndrome in children resembles OSAS signs and symptoms; therefore, these children should be assisted by a multidisciplinary team which should base its actions on a bio-psycho-social context in order to improve these children quality of life.

Early diagnosis and treatment can reduce cardiovascular sequels, neurocognitive impairment and morbidity among these patients. Polysomnography is still considered to be the gold standard for OSAS diagnosis. Fixed orthodontic and orthopedic oral appliances have been successful in enlarging the oral cavity, in expanding the upper airways and in preventing future damages to these children.

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