Awareness of Sleep-Disordered Breathing in Children with Down Syndrome

Marco Zaffanello* and Giorgio Piacentini
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Pediatric Division, University of Verona, Verona, Italy

Keywords
Children; Down syndrome; In-laboratory overnight respiratory polygraph; Obstructive sleep apnea; Sleep Disordered Breathing

Abbreviations
AHI: Apnea-Hypopnea Index; DS: Down Syndrome; OSA: Obstructive Sleep Apnea; ODI: Oxygen Desaturation Index; OSAS: Obstructive Sleep Apnea Syndrome; SDB: Sleep Disordered Breathing

Editorial
Obstructive Sleep Apnea (OSA) is characterized by partial or complete cessation of airflow and oxygen desaturation during sleep owing to upper airway collapse [1,2]. The severity of sleep apnea depends on the abnormal size of the airway [3]. In children, the major contributor to upper airway narrowing is hyperplasia of pharyngeal tonsils and adenoids [4]. Moreover, pediatric craniofacial disharmony was strongly associated with sleep-disordered breathing (SDB) [5]. History of premature birth and a family history of OSAS as well as obesity and African-American ethnicity were associated with increased risk for SDB in childhood [6]. Polysomnography is indicated when the clinical assessment suggests the diagnosis of obstructive sleep apnea syndrome (OSAS) [7].

We report the case of an African, Down syndrome (DS), 7-year-old female child with obesity (BMI 27 kg/m²) and SDB. Adenotonsillectomy was performed at the age of 5 years. No congenital heart disease. On admission, it was referred by parents a difficulty breathing during sleep, habitual snoring, nocturnal awakenings, gets up from bed at night. On examination, she showed prognathism, macroglossia, scoliosis nasal septum and nasal turbinate hypertrophy. A pediatric sleep questionnaire was positive [8]. Laboratory exams demonstrated vitamin D deficiency (8 ng/ml). The in-laboratory overnight respiratory polygraph study (SOMNOscreenTM PSG, SOMNOmedics GmbH, Randersacker, Germany) was performed [2,9]. She showed (estimated Total Sleep Time - TST 10.1 h): Apnea-Hypopnea Index (AHI) of 40.5 events/h, Obstructive Apnea Index (OAI) of 29.2 events/h, Central Apnea Index (CAI) of 4.2 events/h, oxygen desaturation index (ODI; SpO2≤3%) of 14.4 events/h, mean SpO2 98%, Minimum SpO2 82%, mean Heart Rate (HR) 94 bpm (minimum HR 59 bpm), phase angle 37 degrees and snoring 14.9% of TST. Figure 1 shows a representative picture of relevant obstructions of the high airways in this child. For this reason, nasal C-PAP began. The study was performed in accordance with the Declaration of Helsinki and under the terms of all relevant local legislation.

SDB, specifically OSAS, is recognized as a consistent problem in children with DS. The prevalence of DS varies between 1/650 and 1/1000 worldwide. In children with DS, Churchill “et al.” [10] reported a prevalence rate of OSAS between 24% and 59%. Recently, the high prevalence of OSAS in DS population has been confirmed by new studies in the topics. In particular, Brockmann “et al.” [11], by unattended home polysomnography performed in 44 children (mean age 3.6, min 0.1– max 10 years), reported that OSAS was present in 61% of them. Maris “et al.” [12] diagnosed OSA in 57.1% of 54 children, aged 7.5 (5.4–11.6) years. Konstantinopoulou “et al.” [13] reported that 87% of 23 children, aged 8–19 years, had OSAS. Basil “et al.” [14] found that 74% of 177 children (among pooled 303 patients: age range of 2.1-19.1 years) had OSAS and the obese individuals were more likely to have the moderate or severe condition [15].

Patients with DS have many predisposing factors for developing OSAS, including obesity, midfacial hypoplasia, an abnormally small upper airway with superficially positioned tonsils and...
The picture shows a 5-minute polysomnographic recording of the case patient. She had frequent apnoas, oxygen desaturations, snoring, autonomic activation and arousals.

relative tonsillar and adenoidal encroachment [16]. Moreover, these children are also at increased risk for comorbidities, such as congenital heart disease, pulmonary hypertension, ear infections and scoliosis [17]. In particular, OSAS risk was elevated in obese DS children [14]. Trois "et al." showed a positive correlation between OSA severity and body mass index (BMI) [18]. Furthermore, OSAS has been associated with cardiovascular complication (pulmonary hypertension) [16]. Finally, Konstantinopoulou et al reported Left Ventricular diastolic dysfunction, with improvement with CPAP use [13]. The tonsill hypertrophy has a major role in addressing OSAS in DS children. Although tonsillectomy resolved 30 -50% of OSA [10], monotherapy was also reported insufficient [19]. Treatment of OSA involves the use of CPAP, upper airway surgery, and dental appliances, along with weight-reduction strategies, nasal steroids, and oral leukotriene modifiers as adjunctive treatments [20].

In children with DS, a polysomnographic screening is mandatory, considering the potential overall consequences of untreated OSAS [11]. A referral to a pediatric sleep laboratory for all children with Down syndrome by 4 years of age was recommended by the American Academy of Pediatrics [15]. In particular, these children are more susceptible to the additional negative impact of sleep respiratory disturbance since they have frequent pre-existing medical and neurocognitive disabilities. The early diagnosis and treatment are not only aimed to ameliorate the sleep quality of these children but also to obtain positive physical effects. Unfortunately, it is also recognized that the access to a sleep specialist may be limited in some geographic areas.

References