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## Correspondence:

Peter Blair  
 FSID Research Unit, Level D  
 Southwell St., St. Michaels Hospital  
 BS2 8EG – Bristol – UK  
 Tel.: +44 (117) 928.5145  
 Fax: +44 (117) 928.5154  
 E-mail: p.s.blair@bris.ac.uk

## Sleep-disordered breathing in children: time to wake up!

Oliviero Bruni\*

The scientific literature in the last few decades stressed the importance of identifying and treating sleep-disordered breathing (SDB) in children, since it might determine serious neurobehavioral, cardiovascular, endocrine, and metabolic consequences.

Accurate identification of the prevalence of primary snoring and obstructive sleep apnea (OSA) in the pediatric population is critical for different reasons: a) estimate the magnitude of the problem; b) inference on the relationships with other emergent childhood health problems, such as obesity; c) possibility of preventing long-term consequences; d) identification of population subgroups that could be at risk for developing SDB; and e) guide for future investigations.

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One of the first worries of medical research when approaching a health problem is to clearly identify its magnitude, to evaluate how many subjects could be affected and how health related diseases could be prevented. This is increasingly more important if we care about children and if we are aware that we could avoid several long-standing consequences with early treatment of sleep breathing problems.

In this perspective, the very well conducted study of Petry et al.<sup>1</sup> in this issue of *Jornal de Pediatria* is of great importance because it gives for the first time a clear picture of SDB problems in children in a Brazilian region. This cross-sectional study was carried out in the city of Uruguaiiana, RS, in a large sample (about

\* Pediatric Sleep Center, Sapienza University, Rome, Italy.

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1,000) of schoolchildren aged 9 to 14 years, as part of a larger epidemiological study on asthma and allergies. The parents of 27.6% of the children reported habitual snoring (HS), while 0.8% reported apnea, 15.5% described daytime mouth breathing (MB) and 7.8% complained of excessive daytime sleepiness (EDS).

These results are in part similar and in part divergent from what is reported in the literature: HS ranges from 1.5 to 6%, apneic events during sleep from 0.2 to 4% and "generic" SDB from 4 to 11% of children. A recent meta-analysis study using generalized estimating equations to adjust for overdispersion resulting from heterogeneity of study results found an SDB prevalence of 7.45% (95%CI 5.75-9.61).<sup>2</sup>

In order to explain these discrepancies, we should take into account that there is a great variability of prevalence in different studies that could be attributed to several factors: different questions on the same argument (snoring, loud snoring, snorting, etc.), different time window (days or months); dichotomous questions or Likert scale; different meanings of the terms used, such as "often," "frequently," "sometimes," "seldom," and "occasionally." In addition, we should consider that race and ethnicity greatly influence prevalence: African-American children have a higher risk of developing SDB, or Hispanic children seem to be more affected by snoring or apnea than white children.<sup>3</sup>

Even considering all these potential confounding factors, one could be surprised by the elevated prevalence of HS (27.6%) in Uruguiana, which is significantly higher than rates that have been observed among similar-aged children in other countries. On the contrary, prevalence of apnea (0.8%) was lower than in other studies. Furthermore, 15.5% described daytime MB and 7.8% complained of EDS.

The point of clinical relevance is how many of these snoring children will have serious health consequences? The exact polysomnographic criteria that differentiate between innocent snoring (i.e., HS that does not lead to gas exchange abnormalities, sleep disruption, and/or to any morbid consequences), and snoring that is associated with adverse consequences have yet to be defined. Nevertheless, on the basis of the current estimate, of the many children with HS, approximately 2-3% will have clinically relevant disease.<sup>4,5</sup>

The strength of Petry et al.'s study was not only the analysis of HS and OSA prevalence, but also of other clinical symptoms that are often not considered in different studies but that give a more complete picture of the child's health status, such as MB. Prevalence of daytime MB was 15.5%, although the age group considered is not at risk for adenotonsillar (AT) hypertrophy. Apart from the consideration that recurrent upper airway infections and exposition to passive smoking could be responsible for MB, we should consider that snoring and OSA in children are not only related to AT hypertrophy but also to structural craniofacial and orthodontic anomalies

that could persist after AT surgery and, obviously, require different treatments, such as orthodontic appliance or rapid maxillary expansion.<sup>6,7</sup>

Another relevant issue raised by the Petry's study is the evaluation of EDS. Prevalence of EDS in children with OSA is not clearly defined because children were not able to report sleepiness and also because sleepiness in children is often replaced by paradoxical behaviors.<sup>5</sup> Prevalence of symptoms compatible with EDS in OSA children is about 7%, but if we include other questions on behaviors associated with EDS (i.e. irritability, inattention and hyperactivity) the frequency increases up to 40-50%.<sup>8,9</sup> Data on sleepiness reported in the Brazilian population is similar to that in other countries and presence of snoring or apnea increased greatly the risk of having EDS.

Another interesting point is that Brazilian children with EDS secondary to SDB appear to have almost 10 times the risk of learning difficulties. This finding is in agreement with other reports and with a recent study that evaluated sleepiness in a sample of children with SDB using the pediatric daytime sleepiness scale (PDSS): frequent snorers had higher mean PDSS score and snoring or apneas and PDSS scores were significant predictors of academic failure after adjusting for age, sex, body mass index, specific school attended, and sleep habits.<sup>10</sup>

As the authors stated, "the relationship between SDB and poor performance at school detected in this study merits the attention of future studies, due to the impact that this finding could have on the community as a whole. The identification of these risk factors that can potentially be reversed by effective public health measures is the primary objective of studies such as this one."

Morbidity associated with SDB is only beginning to be understood. Further research into the epidemiology of childhood SDB and its consequences could play a key role in improving efforts to systematically diagnose and treat this condition.

We should now be aware that SDB in children may represent a major public health burden with many years of potential consequences for affected individuals, their families, and society; unfortunately, many cases remain undiagnosed, and/or untreated.<sup>11</sup>

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Correspondence:

Oliviero Bruni  
Pediatric Sleep Center  
Department of Developmental Neurology and Psychiatry  
Sapienza University  
Via dei Sabelli, 108  
00185 - Rome - Italy  
Tel.: +39 (064) 471.2257  
Fax: +39 (064) 95.7857  
E-mail: oliviero.bruni@uniroma1.it

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## Entering a new treatment age for mucopolysaccharidosis VI disease: a search for better markers of disease progression and response to treatment

Paul Harmatz\*

Mucopolysaccharidosis (MPS) VI is a rare autosomal recessive genetic disorder involving mutation and abnormal function of the lysosomal enzyme N-acetylgalactosamine (arylsulfatase B - ASB).<sup>1</sup> Decreased enzyme activity leads to incomplete degradation of the glycosaminoglycan (GAG) dermatan sulfate, and accumulation of breakdown products in cells and tissues. These breakdown products contribute to lysosome damage, cell death, and organ dysfunction. Although a wide spectrum of clinical severity occurs, the typical findings in a patient with significant disease include short stature, skeletal findings of dysostosis multiplex, joint dis-

ease, cardiac valve disease, obstructive and restrictive pulmonary disease, frequent respiratory infections and hearing loss, eye disease including corneal clouding and glaucoma, optic nerve disease, mild hepatosplenomegaly, and abdominal hernias. In contrast to other MPS diseases, primary central nervous system (CNS) disease and mental retardation are not a part of the MPS VI clinical spectrum. Until recently, treatment of patients with MPS VI has involved primarily supportive medical and surgical care.<sup>2</sup> Successful hematopoietic stem cell transplant (HSCT) has been described in a few case reports for MPS VI,<sup>3,4</sup> although the risk of transplant is signifi-

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\* MD. Pediatric Gastroenterology and Nutrition, Children's Hospital & Research Center Oakland, Oakland, CA, USA.

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