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POINT OF VIEW

## The importance of palliative care for children with terminal disease and the need to consider the role of sleep disturbances within this contexto

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Dear Editor,

It is estimated that 56 million children under 5 years of age will die between 2018 and 2030<sup>1</sup>. Although great progress has been made in controlling mortality since 1990, further improvement warranted to be made, particularly in Sub-Saharan Africa and south Asia<sup>2</sup>. Mortality statistics showed that of the 6.3 million children who died before the age of 5 years in 2013, 51.8% died of infectious diseases and 44% died in the neonatal period<sup>3</sup>. Palliative care is usually defined as an interdisciplinary approach provided by a team that includes not only doctors, but other health professionals.<sup>4</sup> The situation experienced by terminal patients and their families may have physical, emotional, spiritual, social and psychological effects<sup>4,5</sup>, and good palliative care can improve the quality of life of both patients and their relatives, helping them to face the problems frequently associated with life-limiting illnesses. Early identification and evaluation of the condition, followed by effective pain treatment, are the strategies required to relieve suffering. Palliative care and curative treatments that extend life may occur at the same time.

Pain management is crucial for children with terminal diseases. It is important to recognize that pain is a subjective perception to each individual and that even if the patient is unable to communicate, we cannot assume that they are not feeling pain<sup>4</sup>. It is essential that a multidisciplinary team that understands the clinical course of the disease and the most effective ways to control pain of these individuals in order to provide more comfortable end of life.

According to a systematic review, 4-40% of children and adolescents have chronic musculoskeletal pain<sup>6</sup>, which may deeply affect their sleep. Sleep quality is a major concern for terminally ill children and adolescents or their caretakers, since poor sleep and pain have a bidirectional relation and are believed to form a vicious cycle. However, these conditions are not well understood in children. Pediatricians must be aware and alert to the sleep quality of children or adolescents suffering from diseases, such as cancer, as the treatment of sleep problems can improve disease management. It is possible to understand the relationship between sleep and pain in pediatric populations within a biopsychosocial context as this interaction may be influenced by race, ethnicity, culture as well as quality of life, mood and physiology/biology<sup>7</sup>.

There is a hypothesis that sleep deprivation in children and adolescents can be the result of a modification to the hypothalamic-pituitary-adrenal axis (HPA) due to the brain's response to stress. One possible explanation could be that there is negative feedback inhibition controlled by the HPA axis. Thus, when glucocorticoid levels rise above a certain point, they inhibit the additional release of adrenocorticotrophic hormone and corticotropin-releasing factor, disrupting the response. The response of the organism when having sleep disorder and chronic pain may affect HPA axis, given that the HPA axis undergoes significant maturation during adolescence<sup>8</sup>.

Sleep problems are a common and serious issue in children with life-limiting conditions and severe psychomotor impairment, with a prevalence of 60-80%. Children with

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severe chronic illnesses can present sleep disturbances, such as difficulties in initiating and maintaining sleep, sleep-associated respiratory problems, daytime sleepiness, parasomnia, and irregular sleep-wake rhythm<sup>9</sup>. To evaluate the sleep quality of children with neurological disease the “Sleep Questionnaire for Children with Severe Psychomotor Impairment” (Schlaffragebogen für Kinder mit Neurologischen und Anderen Komplexen Erkrankungen-SNAKE) was developed. The SNAKE is multidimensional questionnaire that assesses five different dimensions of sleep associated problems (problems going to sleep, problems remaining asleep, arousal and breathing disorders, daytime sleepiness, and daytime behavior disorders), and collects important sleep information in respect of this vulnerable population<sup>10</sup>.

. Another important point to consider is the sleep quality of their mothers or caregivers. Children with severe neurological disease have profound sleep disturbances and the more severe the child’s impairment, the more nighttime care is needed, and that care is primarily delivered by the parents<sup>11</sup>. Over 60% of mothers caring for children suffering from Duchenne Muscular Dystrophy were reported to have poor sleep quality<sup>12</sup>. Studying parents of 214 children, adolescents, and young adults with severe psychomotor impairment, severe impairment of parental health status and quality of life were found<sup>11</sup>. More than 50% of the parents suffered from a sleep disorder (e.g. prolonged sleep latency, shortened sleep duration). Sleep disturbances in children, adolescents, and young adults correlated strongly with parental sleep disturbances, parental impairment of physical and mental functioning, parental social functioning, and parental working ability. Parents are severely affected in various aspects of daily living<sup>11</sup>. Therefore, health professionals need to include sleep assessment in their routine, considering that a good night’s sleep provides adequate function of the immune system and improved quality of life.

Pediatricians commonly need to provide information to the patient’s family about a range of topics, from details of palliative care to the death of a loved one. Understanding the course of the disease is very important for the patient, their family, and the caretakers. Ideally, direct language should be used when explaining the death process, with the use of words like “death” and “dying” being recommended. It is a difficult task, since this communication must occur at a very sensitive time<sup>13</sup>.

It is important to emphasize that the health team does not always anticipate the children’s death, missing the opportunity to discuss the matter in advance with the children’s family and help them to prepare emotionally. When the health team provides accurate information about the trajectory of the disease, caregivers are able to better face the problems encountered during the course of the palliative care, including children’s and family members emotional and spiritual support.

We would like to highlight the importance of developing an integrated relationship between the children, the health team, caretakers, and family members, to ensure a more dignified end of life for patients suffering with a terminal disease. Health professionals should be aware that direct or indirect sleep problems can influence the treatment. Caretakers and children should consider sleep hygiene strategies and interventions that may improve sleep and thus assist in the clinical improvement of the children with terminal disease. Moreover, future longitudinal studies should further investigate the role of sleep quality throughout the trajectory of a disease. This will provide more information on treatment, improving the quality of life of these patients.

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## CONFLICTS OF INTEREST

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The authors report no conflicts of interest.

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