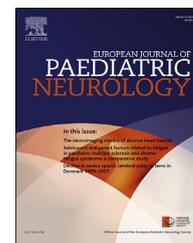




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Editorial

Editorial relating to paper by Schoonjans et al. EJPN 2019; A good night's sleep in Dravet syndrome – an unmet need

Dravet syndrome (DS), a developmental and epileptic encephalopathy, is considered a 'model' disease in genetic epilepsy resulting in difficult to control seizures, learning disability, behavioural comorbidities and motor disorder. It is caused by widespread SCN1A (Nav1.1) dysfunction throughout the brain and is now understood as a channelopathy where a genetic change will present according to its severity and the genetic background impacting on a variety of neuronal networks.¹

The focus of previous research has mainly been centred around seizure control; however, Dravet syndrome has significant impact on cognition and behaviour contributing to high levels of parental stress. Sleep problems are common in Dravet syndrome, affecting the vast majority of patients.² They may be related to nocturnal seizures, co-sleeping or fear of life-threatening seizures in their child's sleep or sudden unexplained death in epilepsy (SUDEP). Consequently, parents and carers lack restful sleep themselves with many co-sleeping with their child.

In this issue Schoonjans et al. focus on exploring sleep behaviour and prevalence of different types of sleep problems in DS. Using validated sleep questionnaires, the authors compared a group of 56 DS patients with 66 epilepsy controls.³ Although significantly more DS patients reported sleep difficulties in general, severe sleep problems were equally frequent in both groups, affecting one third of DS patients and epilepsy controls.

Increased daytime sleepiness and worse parental-reported quality of sleep were particularly affected in Dravet syndrome patients compared to epilepsy controls. Not surprisingly, side effects from medication such as stiripentol were thought to account for the increase in daytime sleepiness. The majority of DS patients had mild night waking difficulties, possibly related to nocturnal seizures resulting in overall lower quality of sleep. Parental perception of sleep problems appears to be significantly worse in DS parents compared to epilepsy controls. As a result, parents themselves complain of interrupted sleep, increased daytime sleepiness, poor concentration and worse quality of life. However, the authors found that most parents (61.5%) never received any advice or treatment for their child's sleep problems, despite there being a substantial

body of evidence for behavioural and medical treatment strategies addressing sleep problems in children with learning disability such as DS.

In clinical practice the findings of this study highlight that sleep difficulties are common in DS, particularly daytime sleepiness and poor-quality sleep. These problems can have a profound impact on quality of life for families. While it is important to firstly identify these difficulties, medication side effects have to be considered and lastly emphasis should be paid to offering professional sleep advice including behavioural and medical treatment strategies.

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