

Perspective

Sleep EEG, Epilepsy and Polysomnogram in Autism and Autism Variants: Highlights of 2016 “Autism: Challenges and Solutions” International Conference in Moscow

Kenneth C. Sassower*

Department of Neurology, Massachusetts General Hospital, USA

An international conference on autism entitled “Autism: Challenges and Solutions,” took place from April 28 through April 30, 2016 in downtown Moscow, and was described by conference organizers as being one of the most well-attended international conferences on autism in all of Europe. The international conference was attended by both healthcare professionals and interested family members alike.

Throughout the three-day conference, co-sponsored by the Dr. Stephen Edelson’s Autism Research Institute (ARI) there were various clinical and research topics presented, including those on the subject of sleep EEG, epilepsy and the polysomnogram in the autism spectrum disorders and in certain autism variants.

Various presenters, including the undersigned, described the fact that autism itself is associated with increased risk of epilepsy. In addition, interictal epileptiform discharges (IEDs) are quite frequent (6–61%), and epilepsy is said to occur in approximately 5–30 percent of individuals on the spectrum. It is felt that the yield of epileptiform discharges increases during prolonged sleep. However, it is unclear as to whether or not these EEG discharges are in anyway causally related to the defects noted in the autism spectrum disorders.

In describing epilepsy, the sleep EEG and polysomnographic features of children with Rett Syndrome, it is felt that epilepsy can occur in up to 90% of affected individuals, and with seizures typically presenting between 3–5 years of age. Partial seizures are felt to be especially common. In addition, sleep has a particularly critical role, insofar as it has been said that upwards of 26% of affected individuals with Rett Syndrome who die do so in their sleep. While the exact causative etiology of sudden death is largely unknown, it has been postulated that hypoxia may lead to neurogenic hyperventilation as well as central apneas during the pre-sleep state. However, the exact role of these various

*Corresponding author

Kenneth C. Sassower, Department of Neurology, Massachusetts General Hospital, 55 Fruit St, Boston, MA 02114, United States, Tel: 1617-724-7426; Email: ksassower@mgh.harvard.edu

Submitted: 26 May 2016

Accepted: 01 June 2016

Published: 08 June 2016

Copyright

© 2016 Sassower

OPEN ACCESS

Keywords

- Epilepsy
- Autism
- Seizures
- Social Cognition
- Decision-making

processes in the select subset of children who die in their sleep remains largely unknown and speculative.

It has been noted that the EEG background of children with Rett Syndrome may be normal in affected children up to ages 2–3 years, but that centro-temporal spikes and sharp waves can occur in approximately 50% of children less than 3 years of age. In addition, focal seizures from the central and occipital regions have been described; as well as independent multifocal spikes; diffuse slow spike-wave discharges; rhythmic theta in the frontocentral regions; various periodic patterns; and, in rare cases, overt hypsarrhythmia.

In sleep, there is a lack of normal non-REM sleep features, as well as a derangement of daytime and nighttime sleep. EEG discharges are noted to increase during sustained sleep. However, distinct EEG stages are often lost over time, and sleep fragmentation can often occur, as well as a decreased percentage of stage REM sleep. In sharp contrast to the pre-sleep state, neurogenic hyperventilation is not evident during sustained sleep.

In addition, much of the time at the conference was spent describing characteristic sleep EEG and polysomnographic patterns in certain autism variants, including Angelman Syndrome, Landau-Kleffner syndrome, as well as Interstitial 15q11.2-q13 Reduplication syndrome [1].

With respect to Angel man syndrome, epilepsy and EEG abnormalities are said to occur in up to 80% of affected individuals. Seizures can present in 85% of patients within the first 3 years of life, and can include generalized Tonic-clonic convulsions, atypical absences, atonic seizures, as well as myoclonic seizures. In addition, sleep dysfunction may vary between 20–80% of affected individuals. Salient EEG findings may include so-called “notched delta” (bi-frontally evident);

diffuse theta (posterior localization); and posterior spikes and slow waves (visually reactive).

Landau-Kleffner syndrome is an acquired epileptic aphasia, which may eventually lead to autism. EEG abnormalities may include an unilateral slow-wave focus; bilateral independent spike-wave discharges; as well as so-called “continuous spike-wave discharges during slow sleep.”

A relatively new addition to the autism variants includes the so-called “Interstitial 15 q11.2–q13 Reduplication syndrome”. A recent publication of ours highlights the presence of autism and mild facial abnormalities, and the syndrome may account for 1-3% of all autism spectrum cases. With the assistance of comparative genomic hybridization, there may be an accelerated detection of submicroscopic micro-deletions and micro-duplications (copying number variants or CNVs), and it may be possible to identify genetic cause for autism in up to 20-25% of affected individuals. In addition, the EEG findings are especially striking, and include the presence of diffuse superimposed attenuated “fast” activity (beta) during both wakefulness and particularly during sustained sleep; i.e., resembling an “alpha-delta” sleep pattern. The excessive presence of diffuse superimposed attenuated beta activity can occur in affected individuals who are not on intercurrent psychotropic agents (such as the benzodiazepines or barbiturates), and can occur dramatically throughout the course

of wakefulness, but especially during sustained non-REM sleep.

CONCLUSION

In conclusion, this international conference on autism highlighted the fact that interictal Epileptiform discharges are quite common in children suffering from many of the autism variants. However, the exact relationship of these discharges to the presence of altered cognition and behavior is largely unknown. In addition, there are certain characteristic EEG patterns which may exist with respect to certain autism variants (such as Angelman’s syndrome; Landau- Kleffner Syndrome; Interstitial 15 q Reduplication Syndrome). Sudden death is a rare complication in the autism spectrum disorders overall, but may be relatively more common in the more severe subgroups (such as in Rett Syndrome), and with a conspicuously exacerbated presence during sleep. The conference also highlighted the need over time to better elucidate the role of these interictal Epileptiform discharges and seizures in the pathogenesis of the autism spectrum disorders and their clinical variants.

REFERENCES

1. Arkilo D, Devinsky O, Mudigoudar B, Boronat S, Jennesson M, Sassower K, et al. Electroencephalographic patterns during sleep in children with chromosome 15q11.2-13.1 duplications (Dup15q). *Epilepsy Behav.* 2016; 57: 133-136.

Cite this article

Sassower KC (2016) Sleep EEG, Epilepsy and Polysomnogram in Autism and Autism Variants: Highlights of 2016 "Autism: Challenges and Solutions" International Conference in Moscow. *J Autism Epilepsy* 1(1): 1002.