Indian Journal of Pediatrics (October 2023) 90(10):955–956
https://doi.org/10.1007/s12098-023-04729-z

EDITORIAL COMMENTARY

What Should be the Next Choice After Failure of Hormonal and Vigabatrin Therapy in Infantile Epileptic Spasms Syndrome?

Parth Lal[1] · **Jitendra Kumar Sahu**[2]

Received: 3 June 2023 / Accepted: 7 June 2023 / Published online: 22 June 2023

The epidemiology of infantile epileptic spasms syndrome (IESS) in South Asia is peculiar because of a long treatment lag, preponderance of structural etiology, availability of synthetic adrenocorticotropic hormone therapy (ACTH), and limited availability of centers with expertise in ketogenic diet therapy and epilepsy surgery [1]. Many children with IESS fail to respond to hormonal therapy (oral steroids or intramuscular ACTH) and vigabatrin, whether tried sequentially or in combination. Then, the question arises of the suitable next choice and available options—another form of hormonal therapy, antiseizure medications (nitrazepam, topiramate, etc.), ketogenic diet, or epilepsy surgery. If oral steroids have been tried as hormonal therapy and failed, it may be worth giving a trial of ACTH—synthetic or natural, depending on availability and feasibility. However, if an option of ACTH therapy has been initially exhausted, there is no quality evidence on the effectiveness of subsequent trial of oral steroids therapy. However, it might be effective in some children with IESS based on the corresponding author's experience. The option of epilepsy surgery should be explored in children with IESS having lateralized pathologies. However, in children with non-lateralized pathologies where curative epilepsy surgery is not possible, there remain limited options — trial of the ketogenic diet or other antiseizure medications.

Different dietary therapies (ketogenic diet, modified Atkins diet, and low glycemic diet) have been studied in children with drug-resistant epilepsies and are considered established options now [2]. The modified Atkins diet possibly offers better tolerability than the classical ketogenic diet. In this context, the clinical trial published by Sharma and colleagues in the current issue of the Indian Journal of Pediatrics, is very pertinent [3]. They compared the modified Atkins diet vs. ketogenic diet in managing children with epileptic spasms refractory to first-line treatment. They found the short-term effectiveness of modified Atkins vs. ketogenic diet to achieve the cessation of epileptic spasms was 20% (4/20) and 15% (3/20), respectively. Inadequate power and lack of assessment of improvement in developmental quotient are the major limitations of the study. However, it provides valuable information and highlights the overall low efficacy of dietary therapies in their study cohort, which might be because of a long treatment lag and the late age of participants at enrollment. In observational studies from authors'center (PGIMER), the short-term effectiveness of nitrazepam and topiramate in achieving complete cessation of epileptic spasms was 51% and 23%, respectively [4, 5]. Although the indirect comparisons are not appropriate, these data might suggest nitrazepam or topiramate are also options, especially when the expertise of the ketogenic diet is not readily available.

Future studies with adequate sample size should provide a head-to-head comparison of diet vs. antiseizure medications among children with resistant IESS. A huge burden of children with IESS and the research potential of child neurologists in India, provide a unique opportunity to leverage cutting-edge collaborative research to drive meaningful improvements in clinical outcomes and advance the frontiers of IESS research. By fostering a collaborative research ecosystem, Indian child neurologists can lead the way to shape the future of IESS management.

✉ Jitendra Kumar Sahu
jsh2003@gmail.com

[1] Department of Pediatrics, Postgraduate Institute of Medical Education & Research, Chandigarh, India

[2] Pediatric Neurology Unit, Department of Pediatrics, Postgraduate Institute of Medical Education & Research, Chandigarh, India

Declarations

Conflict of Interest None.

References

1. Sahu JK, Madaan P, Prakash K. The landscape of infantile epileptic spasms syndrome in South Asia: Peculiarities, challenges, and way forward. Lancet Reg Health - Southeast Asia [Internet]. 2023. Available at: https://www.thelancet.com/journals/lansea/article/PIIS2772-3682(23)00030-6/fulltext. Accessed on 1 Jun 2023.
2. Devi N, Madaan P, Kandoth N, Bansal D, Sahu JK. Efficacy and safety of dietary therapies for childhood drug-resistant epilepsy: a systematic review and network meta-analysis. JAMA Pediatr. 2023;177:258–66.
3. Sharma S, Dabla S, Kaushik JS. Modified Atkins diet vs. ketogenic diet in the management of children with epileptic spasms refractory to first line treatment: an open labelled, randomized controlled trial. Indian J Pediatr. 2023. https://doi.org/10.1007/s12098-023-04527-7.
4. Zahan S, Sahu JK, Madaan P, et al. Effectiveness and safety of nitrazepam in children with resistant west syndrome. Indian J Pediatr. 2022;89:37–44.
5. Nadig PL, Sahu JK, Suthar R, Saini A, Sankhyan N. Topiramate as an adjunct in the management of west syndrome. Indian J Pediatr. 2020;87:6–11.