

Through the unknown, find the new! -a retrospective study on Febrile Infection-Related Epilepsy Syndrome (FIRES)

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Objective: To summarize the clinical features and outcomes in children with FIRES and review on current evidence.

Methods: Retrospective cohort on children 1-16 years with a diagnosis of FIRES in a district hospital in south India over 1 year and literature review following consensus on FIRES in 2022.

What is already known?:

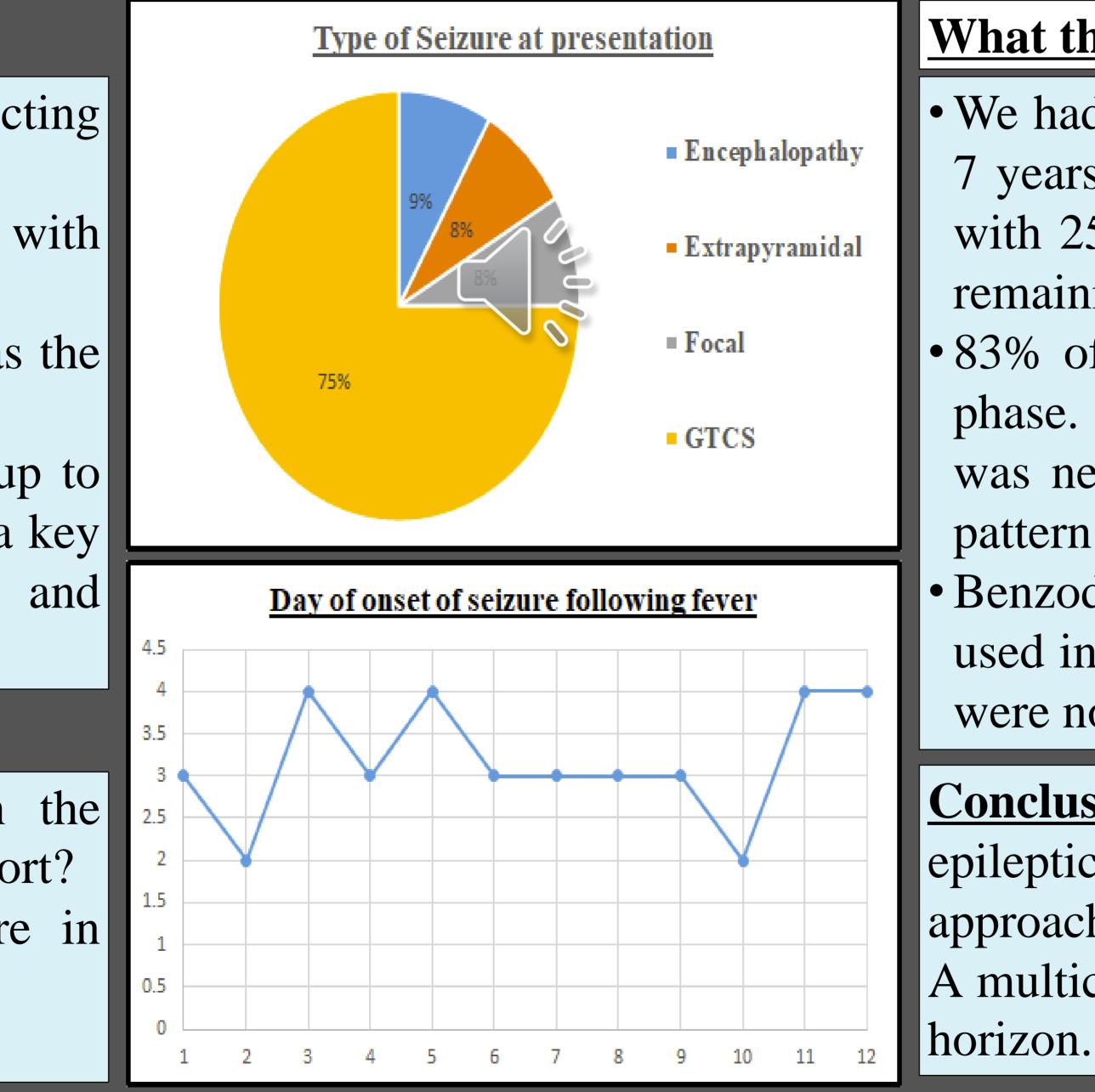
- FIRES is a catastrophic epileptic syndrome affecting previously healthy children of 3-15 years (1).
- The incidence is reported to be 1: 1,000,000 with male preponderance (2).
- Focal followed by GTCS has been mentioned as the common presentation by (3)
- The outcome varies but is usually poor, with up to 30% mortality with early management playing a key role. Ketogenic diet, Anakinra, Cannabidiol and Tocilizumab have been tried (1,3).

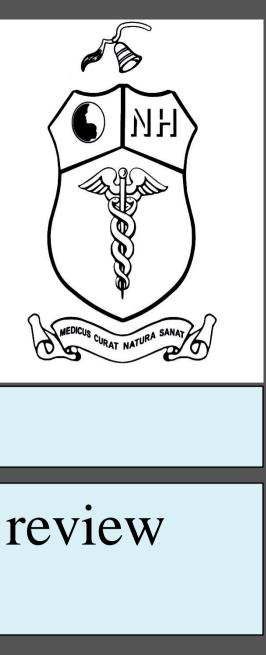
Further discussion:

Whether higher prevalence of infections in the 25 population related to higher incidence in the cohort? \triangleright Is there a structured investigation to explore in resource limited setting?

► Will genetic tests help in the future?

References: 1. Gaspard N, Hirsch LJ, Sculier C, Loddenkemper T, van Baalen A, Lancrenon J, Emmery M, Specchio N, Farias-Moeller R, Wong N, Nabbout R. New-onset refractory status epilepticus (NORSE) and febrile infection-related epilepsy syndrome (FIRES): State of the art and perspectives. Epilepsia. 2018 Apr;59(4):745-752. doi: 10.1111/epi.14022. Epub 2018 Feb 24. PMID: 29476535. 2. Serino D, Santarone ME, Caputo D, Fusco L. Febrile infection-related epilepsy syndrome (FIRES): prevalence, impact and management strategies. Neuropsychiatr Dis Treat. 2019 Jul 9;15:1897-1903. doi: 10.2147/NDT.S177803. PMID: 31371963; PMCID: PMC6635824. 3. Specchio N, Pietrafusa N. New-onset refractory status epilepticus and febrile infection-related epilepsy syndrome. Developmental Medicine & Child Neurology. 2020 Aug;62(8):897-905.





What this study adds:

• We had 12 children who were previously healthy from 3 to 7 years of age (70% male) in 1 year period in south India with 25% mortality in acute phase and severe disability in remaining children.

•83% of our cohort presented with GTCS during the acute phase. MRI was normal. CSF (including NMDA screen) was negative. EEG was abnormal in 91% but no definite pattern could be ascertained.

• Benzodiazepines and intravenous methyl prednisolone were used in all children. Ketogenic diet and immunomodulation were not tried due to limited resources.

Conclusion: FIRES is a diagnostic dilemma in spite of epileptic advances. Early clinical diagnosis with a structured approach to investigations might help in reducing mortality. A multicentric approach in the population might open a new

