Novel treatment approach to NORSE (New onset refractory status epilepticus) in children: Skipping pharmacological coma

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INTRODUCTION

NORSE(New onset refractory status epilepticus) is defined as a clinical condition, with new onset status epilepticus without any clearcut etiology in a patient without any pre-existing neurological disorder or epilepsy.

In majority of the cases of NORSE ;proven or possible either autoimmune or immune mediated etiopathogenesis is suspected.

Globally it has considerable implications in term of mortality and morbidity in survivors

OBJECTIVES

Our aim of the study is to discuss the implications of early and most importantly timely escalation of immunotherapy in children with NORSE.

MATERIALS AND METHODS

This study is retrospective observational study.

All the clinical data is accessed from medical records.

Study duration was from January 2020 to January 2022.

Total 13 children with NORSE were qualified and included in the study.

Their clinical, biochemical, radiological, electrophysiological and treatment details were recorded along with details of short term followup to 6 months were noted.

RESULTS

Out of total 116 children with status epilepticus 13 were presented with NORSE/FIRES. Mean age was 4.7 years.

History of preceding or current febrile illness was seen in 5 (38%) patients. All children had CSF study,EEG study and MRI study.

In CSF study pleocytosis was seen in 11(85%) children while rest biochemical parameters were normal in all samples. All CSF samples were negative for HSV PCR and CSF anti NMDA antibody.

MRI Brain study was done in 10(77%) children which was normal in all children.

All 13 children had abnormal EEG with 8(61%) of them had evidence of electrical status epilepticus. All the patients were given immunotherapy within 72 hours of presentation and it was timely/rapidly escalated as per combined clinical and EEG findings. ***The aim is to achieve clinical seizure freedom with normal physiological EEG background activity(unlike burst suppression with pharmacological coma) as per age***.

We used EEG background activity and spike burden as biomarker to assist the decision making of escalation of immunotherapy similarly as we use CRP in treatment of sepsis for antibiotics upgrade.

Seizure termination was achieved with median duration of 5 days. With this approach 11(85%) children didn't require ventilation and none was given pharmacological coma with zero mortality.

Immunotherapy was used initially combination of steroids and immunoglobulins followed by early use of rituximab (as early on day 5th) based on clinical and our EEG criteria.

CONCLUSONS

Early Immunotherapy has beneficial role in management of children with NORSE.

Our novel approach to treatment of NORSE/FIRES with early initiation of immunotherapy and rapidly escalation of it based on clinical and electrophysiological parameters in children seems very promising but multi-centre study is highly desirable for further validation.

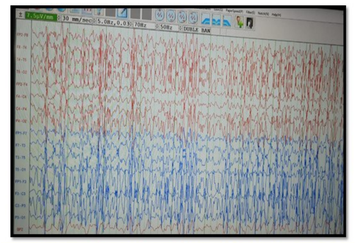

Fig 1: EEG is showing electrical status epilepticus without clinical events confirming NCSE(non convulsive status epilepticus),Day 1 OF IMMUNOTHERAPHY started (Inj Methylprednisolone and Inj Immuno globulin given),No pharmacological coma given

Fig 2: Same Patient Below shown EEG (done on Day 4 after onset of immunotherapy)shows generalised slowing of background activity with interspersed spike and wave activity, at this point immunotherapy escalated and Inj Rituximab given

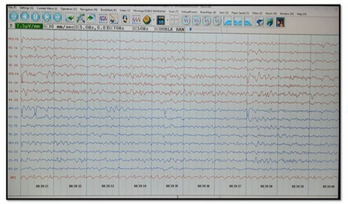

Fig 3(same patient): Day 8 after the onset of immunotherapy :-This EEG shows significant improvement in the background activity with only occasional sharp activity along with clinically substantial improvement in consciousness and cessation of clinical ictal activity.

Suggested reading

Wickstrom R, Taraschenko O, Dilena R, Payne ET, Specchio N, Nabbout R, Koh S, Gaspard N, Hirsch LJ; International NORSE Consensus Group. International consensus recommendations for management of New Onset Refractory Status Epilepticus (NORSE) including Febrile Infection-Related Epilepsy Syndrome (FIRES): Summary and Clinical Tools. Epilepsia. 2022 Aug 11. doi: 10.1111/epi.17391. Epub ahead of print. PMID: 35951466.

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