

INTRODUCTION

*Febrile infection-related epilepsy syndrome (FIRES) is a condition characterized by new onset super refractory status epilepticus triggered by a febrile infection in a previously healthy child.

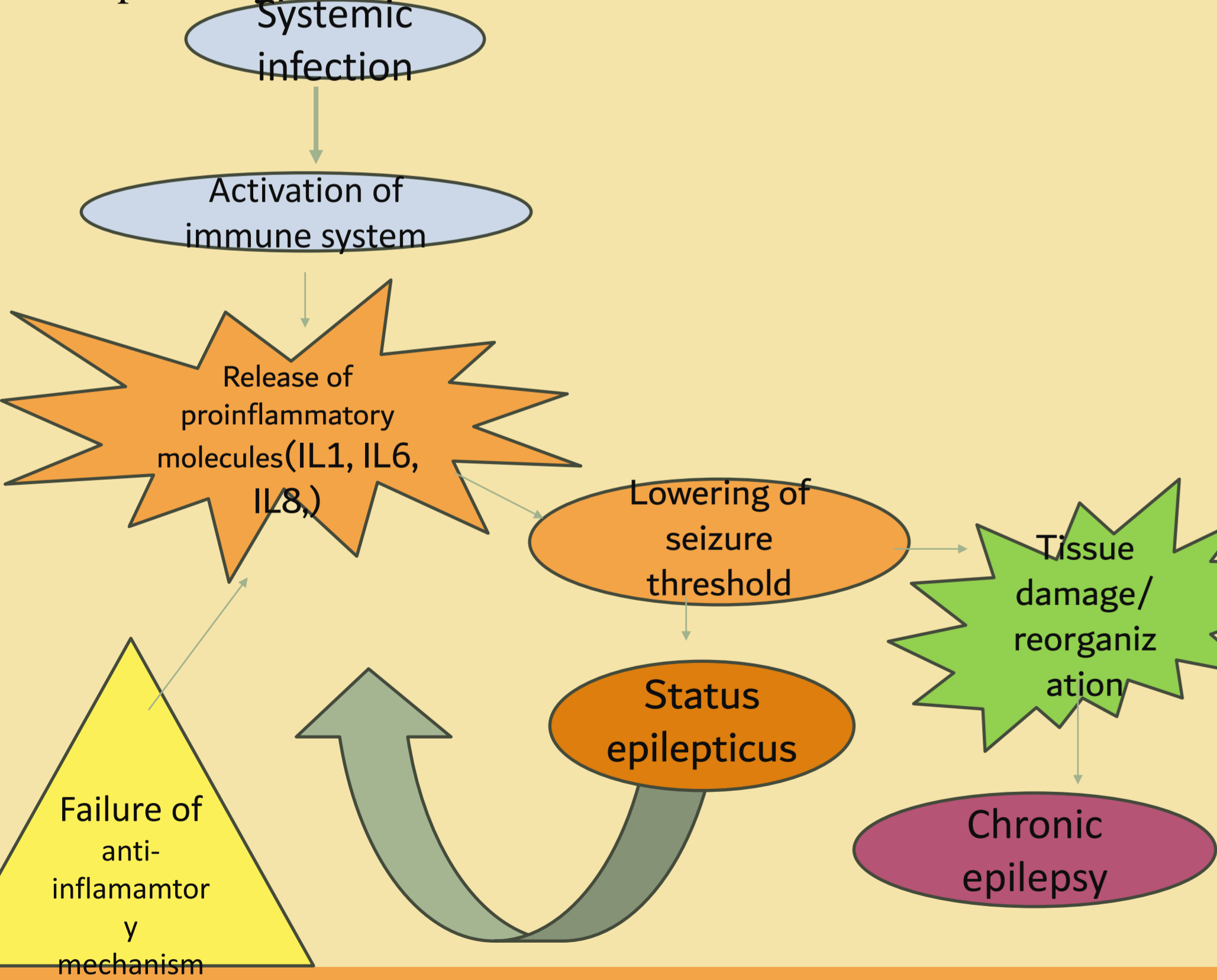
*Pathophysiology is largely unknown.

*Outcome is generally poor:

- mortality in 30%;
- refractory epilepsy with cognitive delay in 66–100% of the survivors.

• Early diagnosis crucial for optimal management.

• Proposed hypothesis

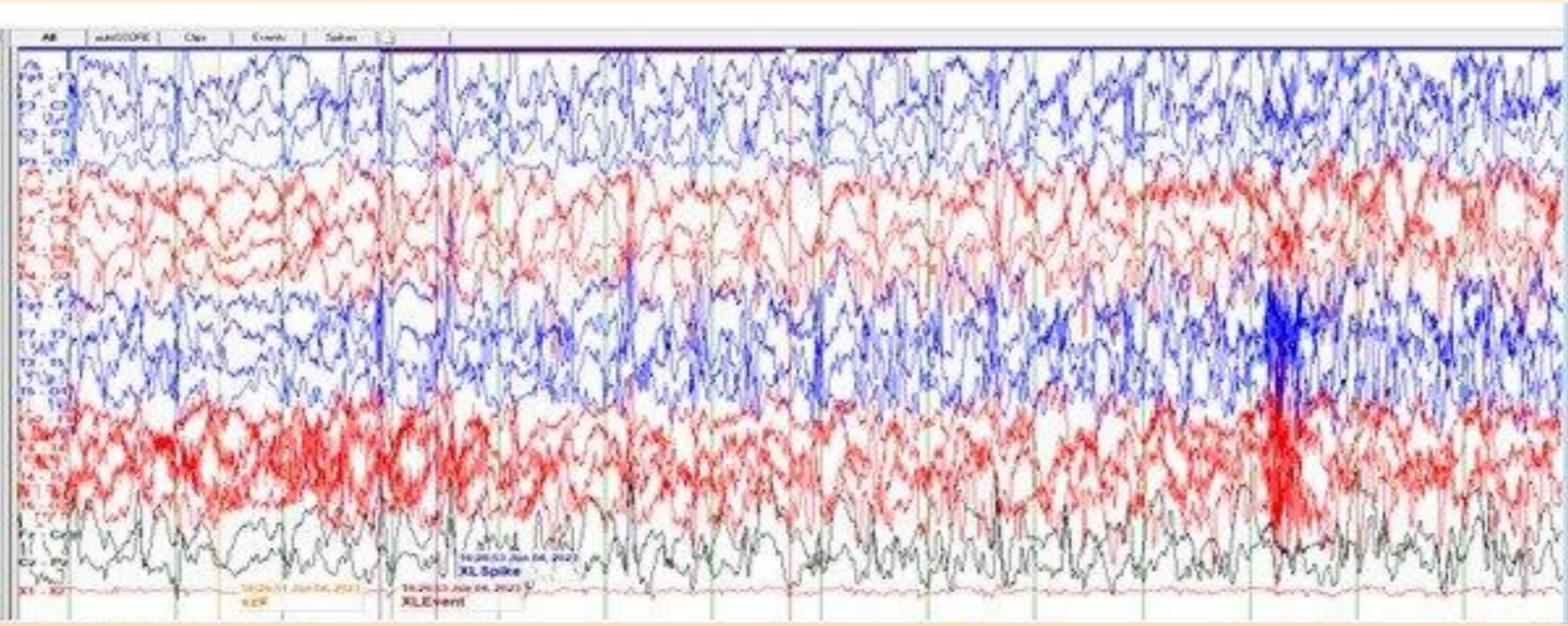


OBJECTIVE

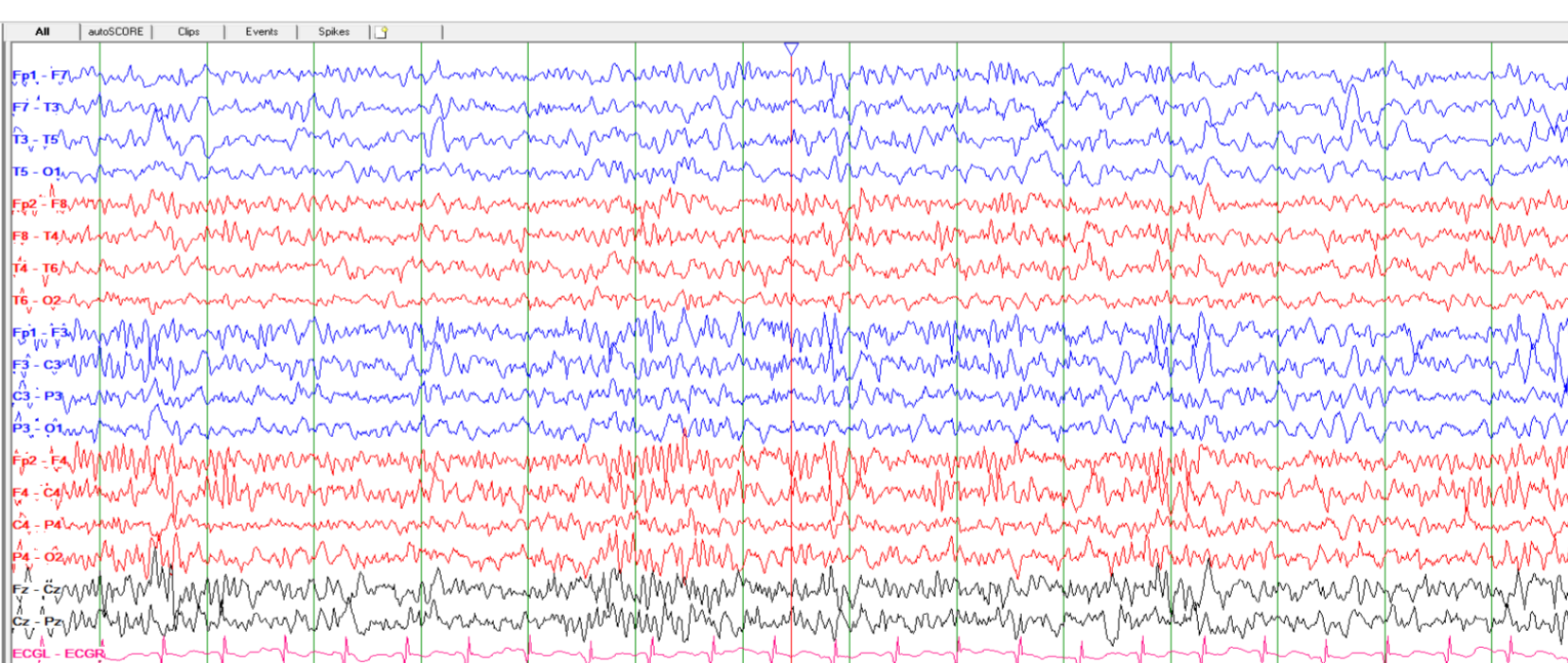
To report the case of FIRES in a previously healthy 8-year-old girl with a favourable outcome due to early institution of the interleukin-6 inhibitor Tocilizumab.

METHOD

An 8year old previously healthy girl child was admitted with acute onset focal seizures progressing to super refractory status epilepticus (SRSE) after a three day history of fever and sore throat. Her initial EEG showed frequent high amplitude bilateral epileptiform spike and wave discharges with left predominance, that rapidly culminated into non convulsive status epilepticus with coma within subsequent 24hrs. CSF analysis, autoimmune panel and infection work-up were negative. Despite escalating treatment with multiple anti-seizure medications(ASMs) along with high dose bolus phenobarbitone, midazolam and ketamine infusions alongside immunotherapy, seizures remained refractory. Interleukin(IL) levels were tested on D18 and her serum IL-6 levels were reported to be significantly elevated [IL-6: 120pg/ml (*Range <7pg/ml*)] while IL-1 was normal. On day 20 of hospitalization, Tocilizumab was started at a dose of 12mg/kg every 2weeks, with a total of three doses given over 6weeks.



EEG at admission



EEG at the time of discharge

RESULT

Tocilizumab led to a significant reduction in electroclinical seizure burden within one week of initiation, allowing the gradual discontinuation of anesthetic infusions. The patient was discharged home in good health after 3weeks. At 1year follow-up, she remains well and active on 3ASMs with good cognitive function and independently functioning status, online school attendance with no motor deficits and only rare focal unaware seizures at a frequency of 1 to 2 brief seizures per month.

CONCLUSION

Early use of Tocilizumab treatment in FIRES cases is associated with favorable patient outcome.

REFERENCES

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