RESPONSE TO TREATMENT WITH TOCILIZUMAB IN FIRES, EXPERIENCE OF TWO CASES

Alamo R., Angulo Manchola D., Mejía Zuñiga M. F., Julián E., Carullo M.P., Arancibia M., Muñoz J., Vidal C., Abregu M., Godnjavec J., Rivera M., Vázquez G., Bongiorni L., Schteinschnaider A.

Neuropediatric Departament, FLENI. Buenos Aires, Argentina. - Neuropediatric Departament, Güemes Sanatorium. Buenos Aires, Argentina.

439

18th ICNC 2024 Footprints to the future INTERNATIONAL CHILD NEUROLOGY CONGRESS Cape Town, South Africa I May 6-10, 2024

INTRODUCTION

Febrile Infection - Related Epilepsy Syndrome (FIRES) is a serious neurological condition that affects healthy children. It leads to a super-refractory status epilepticus, with a high mortality rate, neurocognitive sequelae and/or intractable epilepsy. The mechanisms that generate it are inflammatory and autoimmune, for this reason immunomodulatory therapy is indicated.

CASES REPORTS

Two previously healthy male patients, 6-y and 5-y old, who presented with febrile symptoms and subsequently Super-Refractory Status Epilepticus, received sedation and barbiturate coma, remaining in a Paroxysm-Suppression pattern for 70 and 65 days, respectively.

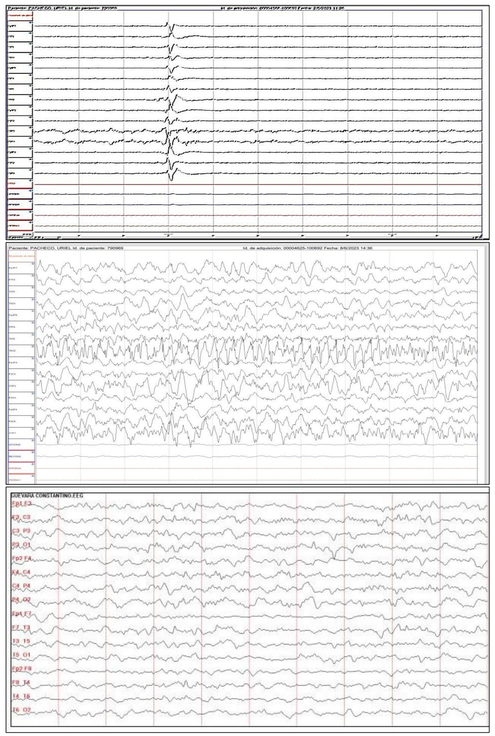
In studies carried out, infectious, metabolic and structural causes were ruled out. A search for antibodies for immune-mediated encephalitis was performed, with Anti-GAD being positive for the youngest of the patients.

They received multiple antiseizure drugs in addition to immunomodulatory treatment with corticosteroids, gammaglobulin and rituximab, the second also plasmapheresis, without a positive response. Ketogenic therapy was added, in the first case suspended due to adverse effects and in the second one, with partial response.

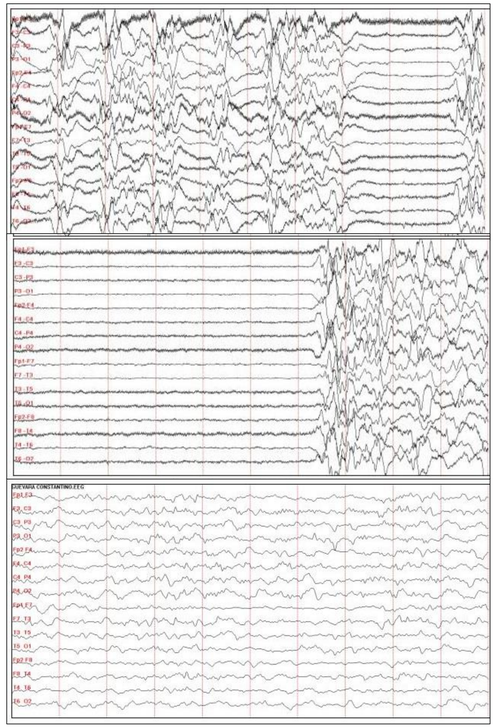
After treatment with Tocilizumab, they progressed with remarkable electrographic and clinical improvement and extubation at 78 and 56 days respectively.

Patient's age	6 years old	5 years old
Febrile infectious previous	YES	
Anti seizure drugs	BDZ, DFH, LEV, CLB, STM, LCS, CBD, PER, FBM. #1: + FBM - #2: + TPM	
Ketogenic Diet	YES #1: not tolerated	
Inmuno Therapy	Corticosteroids, IgGIV, RTX, Tocilizumab. #2: PLEX	

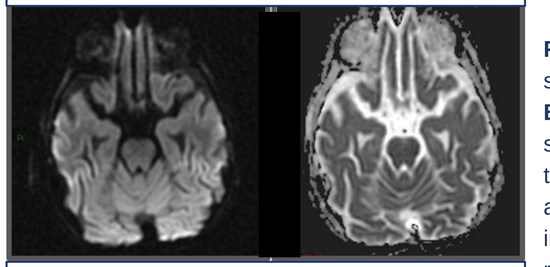
PATIENT 1: EEG evolution



PATIENT 2: EEG evolution



PATIENT 1: Brain MRI



PATIENT 2: Brain MRI

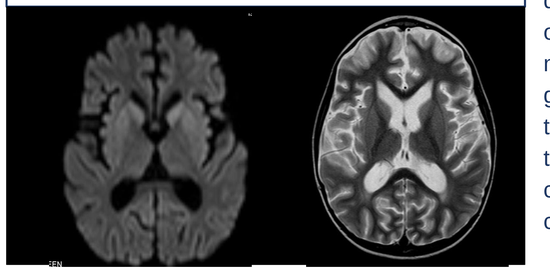


Figure A: Axial section T2. **Figure B**: T2 coronal section. In the left temporal lobe, an area of alteration in gray-white matter differentiation is observed in the middle temporal gyrus that extends to the superior temporal gyrus, compatible with cortical dysplasia.

CONCLUSION

The combination of clinical presentation features and therapeutic response to tocilizumab in our cases supports the key role of cytokine-mediated neuroinflammation in FIRES.

This strengthens the use of tocilizumab as a therapeutic modality in refractory ES.

More prospective controlled studies are needed to validate the efficacy and safety of tocilizumab in the treatment of FIRES.

Contact: angeles@fleni.org.ar