Where there is smoke……

General Medicine Team C
Lisa Barrow, Harishan Tharmarajah, Penelope Wittick
With thanks to Dr Mark Medway, Dr Kannan and the Neurology Team
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Fires (Fever Induced Refractory Epilepsy Syndrome)

• Rare childhood epileptic syndrome characterised by:
  • Refractory status epilepticus during or after fever
  • No evidence of central nervous system infection

• Underlying mechanisms proposed:
  • Genetic predisposition
  • Inflammation: role in precipitation and recurrence of seizures and neuronal damage
  • Brain maturation


Epidemiological features

• Age 4-9 years old
• More common in males
• At RCH 7 children admitted between 1991-2010
• In Europe
  • Prevalence: 1/100,000
  • Annual Incidence: 1/1,000,000


Clinical features

• Febrile illness, often simple URTI preceding
• Timing between fever and seizures
• Multiple seizure types
• Altered conscious state between seizures
• Seizures refractory to multiple anti epileptic medications

Important conditions to exclude

- Infection:
  - Viral
  - Bacterial
  - Other
  - Metabolic
  - Toxins
- Autoimmune/inflammatory
- ADEM
- NMDA receptor encephalopathy
- Limbic encephalitis

Investigations

- Bloods
- Lumbar puncture
- Infectious and metabolic work up
- Electroencephalogram
- MRI

Treatment options 1

- Intravenous immunoglobulin
- Ketogenic diet

Treatment options 2

- Barbiturate anaesthesia
  - Most studies this was the only anticonvulsant to temporarily stop seizure activity in acute phase. Howell, K. (2012). Long term follow up of febrile infection related epilepsy syndrome. Epilepsia, 101-110.
- Therapeutic hypothermia

Outcomes

- Mortality 10 - 30%
- Refractory epilepsy and intellectual impairment in 66-100% of survivors
Take home messages

- FIRES increasingly described in the literature
- Consider in previous well school aged child presenting with encephalopathy after a trivial febrile illness with refractory seizures
- No clear evidence of efficacious treatments → need for further studies, particularly with defined trial of treatment and responses documented

References