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"FIRES: When the brain of a healthy child suddenly "catches fire". An extremely rare syndrome that we are not able to stop, even today. A syndrome by definition is not just one disease but probably several diseases involving metabolic, immunological and neurophysiological disturbances which vary from child to child. In order to prevent these devastating conditions, we need to understand them first.

What is happening? What are the cells and antibodies involved in this self-sustaining spiral of cerebral hyperexcitability? To develop this understanding, fundamental research must be carried out on animal samples as well as the organisation of an international register of affected children. Such a register would allow researchers to carry out multicentre trials of potential new therapeutic agents for this syndrome. Current studies are looking into the role of innovative anti-epileptic medications i.e. those medications which prevent and control seizures, as well as immuno-modulating therapies, which focus on preventing the immune systems attack on the self. Not only will research assist in the development of new treatments, it also serves to map the course of this devastating syndrome. FIRES causes an intense epileptic state within the brain, the activity of which is comparable to an active volcano. The scars of constant seizures leave their mark on the child's brain resulting in a somewhat permanent intellectual and physical disability which is the most distressing aspect of this illness.

To better assist parents in coping with the effects of FIRES, research aims to map out its evolution thus providing information on what to expect in this stressful time. There is a long way to go, **but thanks to the association Paratonnerre, families, doctors, scientists, are united and FIRES is not a "rare" disease any longer.**

Participate

Parents of children affected by DESC or FIRES, we can share our experiences of the disease in complete confidentiality (its onset, how we live with it), exchange views on the background of our children before the disease (health, socialization, schooling) and exchange comments and hypotheses...

Healthcare workers, do you know of any children affected by DESC or FIRES? If so, enable these families to find help and be a part of the Paratonnerre association network. Thanks to your experience, you can contribute to the medical research and identification of the correct treatments.

Join up

If you wish to support the actions of the association and strengthen the support network which is built up around children struck by this disease, join the association! Contact us by email or by post and go to www.associationparatonnerre.org Thank you!

Paratonnerre

An association combatting FIRES syndrome



*Cope better
with a bolt
from the blue*



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Paratonnerre
Fight against FIRES syndrome

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The association

A bolt from the blue

On 10th July 2011, Anouchka, our five year old daughter who was in full health, suffered from an attack of epileptic seizures that were impossible to stop. After numerous tests, a diagnosis was made: FIRES syndrome, a rare disease (or rather a group of symptoms) of which little is known. "A bolt from the blue" the doctors told us. That's precisely how we felt. The bolt struck Anouchka, us and the closest to us. Our daughter has now since ceased to continuously convulse. As of today, she is handicapped and remains severely epileptic.

Questions, motivations, ambitions

Thanks to Anouchka and the support network which has surrounded her, the feelings of powerlessness and uncertainty that have accompanied us since 10th July have given way to a desire and hope to make things happen.

On 25th June 2012 we created the association Paratonnerre in order to organize our actions and obtain for ourselves and for the victims of this disease, the answers to persistent questions: What happened? What are the causes of the disease? How can we prevent it, treat it? How can we encourage the well-being and rehabilitation of these children? There are many questions and ambitious objectives. To achieve these objectives, we need you, your skills and your vigilance on this subject.

The objectives

- **Prevent** the onset of FIRES
- **Improve the lives of the children** and their families affected by the disease
- **Encourage research and good practices** in therapy in the medical and para-medical profession, during the acute phase and in the long term

FIRES SYNDROME

Fever Induced Refractory Epileptic encephalopathy in School aged children

"Between 4 and 10 years, a few days after a nonspecific fever illness, epileptic seizures appear at a child previously perfectly normal, these seizures increase in frequency and in duration up to a severe refractory epilepticus status. This acute phase is followed by a chronic phase with pharmacoresistant epilepsy and cognitive impairments."*

Until 2009, the FIRES was named DESC (Devastating Epileptic Encephalopathy in School-aged Children). Other naming had previously highlighted the imperceptible condition of the disease.

*Rima Nabhout^{1,2}, Olivier Dulac^{1,2}, Catherine Chiron^{1,2,3}
Extract of *Encéphalopathies épileptiques aiguës fébriles chez l'enfant*. Epilepsies. Vol 22, N°4, 294-9, Déc 2010, Épilepsie et inflammation

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Actions

The creation of a unique FIRES Resource Centre: www.associationparatonnerre.org

- **Information made clear and accessible** for families and their closest friends
- **The centralization of all the publications** concerning the syndrome: medical journals, press articles...
- **Family accounts of events, plus the accounts of doctors** (Intensive Care Unit doctors, paediatric neurologists, rehabilitation therapists, paediatricians...), therapists (speech therapists, physiotherapists, occupational therapists...)

Any raw or popularized medical information that is published online conforms to the charter of the association (see opposite).

Stimulate and support the medical research into FIRES

- **Create an impetus around the disease** by going to meet medical and para-medical professionals and care workers confronted or likely to be confronted with the disease
- **Pass on information about FIRES**, allowing parents and doctors to identify it as quickly as possible, treat it as best they can and provide the best well-being possible for the children affected
- **Maintain permanent contact** with specialists and institutions specialized in epilepsy, rare epilepsy, rare diseases and research
- **Look for funding** to galvanize the medical research

A family network

- **Share experiences** to improve the well-being of children and their families: rehabilitation, lifestyle, spare time, medicine, diet
- **Moral support** between families to help them cope better with the disease
- **Collect new information** for medical research

Disclaimer: children affected by FIRES suffer from a variety of after-effects and the objective of the association is in no way to replace the associations dealing with disability in general, nor the doctors, therapists, rehabilitation therapists...

CHARTER OF THE ASSOCIATION

- The association acts in connection with and in autonomy of the medical profession, with the aim of furthering medical knowledge about FIRES.
- Doctors approve the popularization of the medical texts.
- A peer review committee validates the information that is made available.
- In complete confidentiality, special importance is attached to the background of the children before the onset of FIRES.