

# Newsletter

## to Health Professionals

Number 1

### Edito

## Recommendations from experts in the field of Ataxias and Cerebellar or Spinocerebellar Degeneration

(Medical council of the AFAF, CSC and ASL associations)

Created in January 2005, the medical and Health professionals council of the AFAF (Friedreich's ataxia), CSC (cerebellar degeneration) and ASL (spastic paraplegia) associations consists of neurologists, neurogeneticists, neuro paediatricians, rehabilitation physicians, neuro orthopaedic surgeons, psychologists, social workers, speech therapists, members of patients' associations and their families. It proposes answers to the most frequently asked questions in relation to ataxia and cerebellar or spinocerebellar degeneration. The primary aim of this newsletter is to facilitate the treatment of patients suffering from rare, uncommon, and therefore little known pathologies by informing health professionals about the issues involved. However, it must be emphasised that, in the absence of a confirmed diagnosis, extreme caution must be taken, particularly when prescribing certain medications.

## Frequently asked questions...



### 1 Flu vaccination in adults

There are no specific contraindications to vaccination against the influenza virus in the case of spastic paraparesis and cerebellar ataxia in adults. On the contrary, this vaccination is strongly recommended in ataxia patients who often have a reduced respiratory capacity due to scoliosis and swallowing disorders, with the risk of Mendelson's syndrome.

### 2 Treatment of anaemia and iron supplementation in Friedreich's ataxia

Documented iron-deficiency anaemia requires appropriate iron supplementation in addition to an etiological exploration of the deficiency. There is no contraindication to occasional treatment.

### 3 Vitamin E in ataxia

Some cerebellar ataxias are related to a genetic vitamin E deficiency (ataxia with vitamin E deficiency or AVED) which requires vitamin supplementation. In all other types of cerebellar ataxia, vitamin E has not proven its efficacy, despite its anti-oxidant properties.

### 4 Antibiotics

To our knowledge, there are no classes of antibiotics contraindicated in cerebellar ataxia and spastic paraparesis. However the following drugs must be used with caution in children: tetracyclines, including doxycycline (Vibramycine®, yellowing of teeth), and medication contraindicated in respiratory deficiencies, such as valproic acid (Depakine®).



## 5 General anaesthesia

To our knowledge, cerebellar ataxia and spastic paraparesis are not contraindications to general anaesthesia and no special precautions are required when using anaesthetic drugs, as opposed to other neuromuscular pathologies.

- a> It is recommended, however, to avoid the use of botulinum toxin injections and inform the anaesthetist of this type of treatment whenever applicable (limited use of curares).
- b> Confinement to bed and prolonged immobilisation prior to or during surgery can lead to aggravation of neurological difficulties which must not be unduly attributed to the anaesthesia.
- c> In the particular context of surgery, it is frequent that difficulties underestimated beforehand are brought to the fore and wrongly attributed to the actual anaesthesia.

## 6 Epidural anaesthesia

There are no known contraindications or cases of aggravation of the neurological disease in relation to epidural anaesthesia in patients with cerebellar ataxia or spastic paraparesia.

However, the possibility of technical difficulties has been indicated in patients with spasticity, particularly when spasms occur in flexion and/or in extension.

## 7 Bladder/sphincter problems

Any bladder/sphincter and/or sexual complaints which may develop during spastic paraparesia or in ataxia patients (in whom this aspect is sometimes little known) must be explored in a hospital service specialising in the treatment and assessment of bladder and sphincter disorders encountered in neurological diseases.

Depending on the town or region, these can be physical medicine and rehabilitation, urology, neurology or functional exploration services.

It should be remembered that once the assessment has been made, the symptoms can be effectively treated.

## 8 Local anaesthesia, dental anaesthesia

There are no known contraindications to this type of anaesthesia in patients with cerebellar ataxia, including Friedreich's ataxia, and spastic paraparesia.

## 9 Treatment of anxiety

- a> **In cerebellar ataxia**  
Benzodiazepines are not formally contraindicated, but it is important to remember that these treatments increase cerebellar symptoms. They should be avoided as first-line treatment, preferring long-term treatment with a serotonergic anti-depressant (several have marketing authorisation for anxiety) and to reserve them for complementary second-line treatment, prescribed with caution.
- b> **In spastic paraparesia**  
There is no greater limitation on the use of benzodiazepines in these pathologies than in the rest of the population. Long-term treatment with a serotonergic anxiolytic is also recommended as it is not addictive.

## 10 Anti-depressive treatments

- a> **In cerebellar ataxia**  
We do not know any contraindicated antidepressant drugs, but sedative treatments can be poorly tolerated (this is the case of tricyclic antidepressants, for example). We particularly recommend the use of serotonergic anti-depressants, with a minimal efficient dose (our clinical data shows that there is frequent hypersensitivity to this type of treatment which indicates that it should systematically begin with half-dose).
- b> **In spastic paraparesis**  
The same recommendations apply, but sedative treatments are often better tolerated than for ataxia and there is no hypersensitivity to serotonergic antidepressants. A gradual increase in dosage, however, results in better compliance in patients who are reticent about using this type of medication, as well as limiting the initial adverse effects.

" The next edition of experts' recommendations will deal with aftercare on the part of the medical and allied professions.

You can send your questions to [conseilmedicalataxie@yahoo.fr](mailto:conseilmedicalataxie@yahoo.fr), or 12 place Brisset, 02500 Hirson, France or to the associations' websites: [www.afaf.asso.fr](http://www.afaf.asso.fr) – [www.csc.asso.fr](http://www.csc.asso.fr) et [www.http://assoc.wanadoo.fr/asl.spastic](http://www.http://assoc.wanadoo.fr/asl.spastic). This document was written by the Medical and Health Professionals Council of AFAF, ASL and CSC. The council is chaired by A. Dürr and composed of M. L. Babonneau, P. Charles, F. Cottrel, P. Denormandie, M. Gargiulo, C. Goizet, T. Hergueta, P. de Lonlay, D. Mazevet, A. Munnich, MC. Nolen, M. Pelletier, C. Pinton, L. Vallée and K. Youssov. The file used to provide you with this document has been declared to the CNIL (French data protection watchdog). In accordance with the provisions of articles 39 et seq of the law of 6th January 1978 amended relating to computing, files and freedoms, you have the right to access and correct any data of which you are the data subject by applying to the presidents of the above associations. You may also, for legitimate reasons, oppose the processing of such data.