

# Newsletter

Issue 3



# ATAXIA

## Ireland

## Ireland will ratify UN Convention of rights for disabled by Christmas

The convention obliges states that are a party to it to “promote, protect and ensure the full and equal enjoyment of all human rights and fundamental freedoms by all persons with disabilities and to promote respect for their inherent dignity”

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### Housing

The IWA has produced an easy to follow guide on how to apply for social housing if you have a disability. It's called **Operation Sign-Up** and you can access more information here

<http://www.iwa.ie/house>

1. Contact local council for application form
2. Get supporting documentation (GP, OT, Tax office, Social Welfare and/or Employer's letter)
3. Return form and paperwork





**TEXT "ATAXIA" TO 50300 TO DONATE €4 NOW!**  
 People living with Ataxia benefit enormously from the many services provided by Ataxia Ireland.

*"Help us to continue helping them"* **PLEASE SEND A TEXT TO 50300**

Disclaimer - 100% of your donation goes to Ataxia Ireland across most network providers. Some providers apply VAT which means a minimum of €3.26 will go to Ataxia Ireland. Helpline: 01-4433890

## Two thirds of disability homes fail HIQA Inspection

The Health Information and Quality Authority (HIQA) has inspected 20 residential homes for people with disabilities and only 7 (35%) were deemed adequate for providing a safe and good quality of life.

Failures were highlighted in the area of staff training, resident privacy and safety. Three of the four HSE-run services inspected had shortcomings, also in the area of resident safety and medicines management.

Most centres emphasize the need for more funding to address these failings. Earlier this year a centre in Kildare was closed because of continued failure to address resident needs.

Unless improvements are made, more centres will be closed and forced to relocate residents.



Members who have appointments at the National Ataxia Clinic are encouraged to attend, if unable to do so, please notify the clinic so the appointment can be allocated to other patients

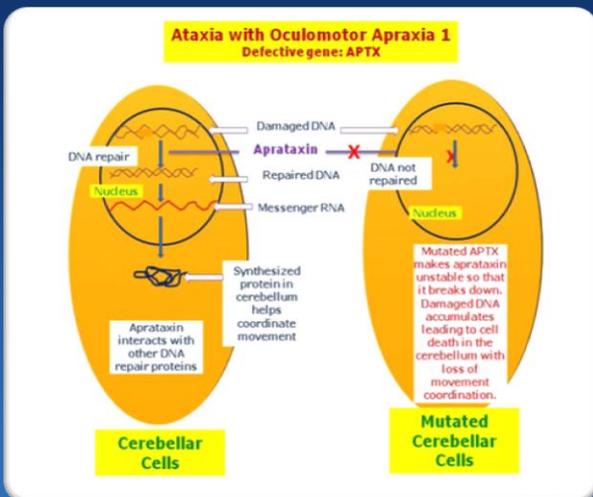
## In Focus: AOA1

Ataxia with oculomotor apraxia type 1 (AOA1) is an autosomal recessive cerebellar ataxia.

Age of onset is between 2-10 years with symptoms starting with gait imbalance, dysarthria (slurred speech) and limb dysmetria (lack of coordination). Oculomotor apraxia follows; this is when the head turns to targets before the eyes. Neuropathy dominates the clinical picture at a later stage and individuals have difficulty moving their extremities (fingers).

AOA1 is caused by mutations in the APTX gene. This codes aprataxin which plays a role in DNA-single-strand break repair (see picture).

Management of the condition involves physical therapy, speech therapy and a low-cholesterol diet is also recommended.



## RESEARCH

### [Metal Homeostasis improves motor performance in fly model of Friedreich's Ataxia](#)

Although frataxin expression is responsible for iron homeostasis, other metals such as copper and zinc also show altered levels in Friedreich's Ataxia patients which suggests that metal homeostasis may have a role in the disease.

Researchers were able to rescue the fly model by adding copper and zinc chelators which restored normal cell levels of these metals. This showed that the beneficial effect on motor performance was not dependent on iron levels as this metal was not altered.

[Soriano et al \(2016\) Plus One](#)



### [New kind of sunscreen which targets free iron in cells, could also treat Friedreich's Ataxia](#)

A new compound called the Mitoiron claw works by chelating or binding excess iron in the mitochondria. Researchers developed the compound as a sunscreen protector as UVA radiation reacts with this iron to produce toxic reactive oxygen species (ROS). The compound only targets mitochondrial iron so the cell itself does not become depleted. Although researchers in the UK were looking specifically at the UVA properties of the compound, they plan to look at applications of the compound in Ataxia treatment.

[Reelfs et al \(2016\) Journal of Investigative Dermatology](#)

### [New gene therapy given FDA Orphan Drug Designation for the treatment of FA](#)

Agilis Biotherapeutics research genetic diseases that affect the central nervous system. AGIL-FA is a gene therapy product that has just been granted approval for the potential treatment of FA. It is the first gene therapy to gain this status. AGIL-FA is a gene-therapy product consisting of a unique gene construct developed in partnership with Intrexon Corporation (NYSE: XON) delivered with adeno-associated virus technology. The virus acts as a vector for transporting the gene into the DNA.

[Access Press Release](#)



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The Disability Federation of Ireland DFI has made recommendations to the Department of Social Protection with regard to Budget 2017 so that it makes provisions for the disabled in line with this convention. The next budget will be the Government’s first opportunity to make social and economic reform for this cohort of people. Key issues include minimum income for people to address the added cost of living with a disability in terms of prescriptions, home adaptations etc.

The government also needs to equality proof payment policies during education & training so that disabled students can access Erasmus placements and work placements abroad on par with their peers.

They also need to introduce protocols for greater flexibility and timeliness for those who move in and out of employment due to their episodic conditions.

Another important issue is Eligibility for medical cards. This must be based on need and not income for those with long term conditions which are not going to go away.



**Coffee Morning**

**ATAXIA Ireland**

**September 25th**

**International Awareness Day**

Ataxia causes damage to the central nervous system. It affects an individual's ability to coordinate movements. It is often progressive and walking becomes so impaired, that a wheelchair is required. It can be acquired or inherited.

Join us and host a coffee morning to raise money and support our work.



Venue ----- Time -----

The National Support group for people affected by Ataxia. Founded in 1980. Registered Charity Number: 7076 CLG Number: 566322. 4 Leopardstown Business Centre, Ballyogan Avenue, Dublin 18. T: 01 299 9033 E: info@ataxia.ie www.ataxia.ie

This is a fundraising event but a limited number of tickets at concessionary rates for members will be available  
 Cost is €100 p.p, and €950 for a table of x10.