



Gateway

News from Huntington's New South Wales

Volume 19 No 1

Autumn 2016



HDYO Land is an interactive, colourful and fun program aimed to help children learn about Huntington's disease (HD). In HDYO Land they will be able to visit and explore 5 different areas in which they will meet some wonderful characters who will take them on an adventure through many locations, educating them about HD along the way!

In each area of HDYO Land children will learn the basics about different subjects such as:

- What is HD?
- How do people get HD?
- How does HD affect people?
- Living in a family with HD
- History of HD

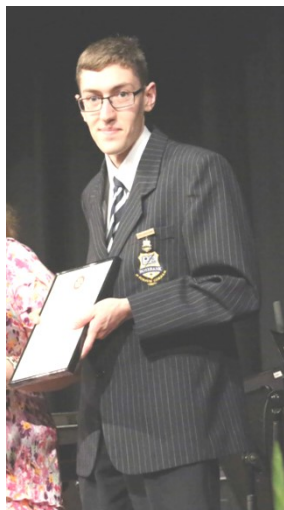
Each area is made up of fun characters, colourful illustrations, cool animations and voice actors bringing the characters to life! They can also find the hidden items that have been placed in each scene, the characters will be very thankful for their help!

HDYO Land is a great place for children to safely learn about HD. We do suggest that children explore HDYO Land with a parent or guardian who can help answer any questions they may have along the way. It's more fun to learn together. They may also find the colouring in pages from HDYO Land a fun and creative activity for all the family!

Visit HDYO Land at www.hdyo.org

Award for Volunteer

Eighteen year old Anthony Swales is an enthusiastic volunteer for Huntington's NSW. Anthony has raised funds and participated in both Walks 4 Hope at Parramatta. He has undertaken community service at the office, helping with a variety of tasks from cleaning out the store room to folding newsletters and stuffing envelopes. Anthony is currently in Year 12 at school and in recognition of his contribution to the community, especially HNSW, he received the 2015 Rotary Club of Five Dock Award for Community Service. Congratulations Anthony and thank you!



Fashion & Fizz Fundraiser in February

raises \$1,200 and over 30 ladies had a fun filled evening enjoying a fashion parade of beautiful clothes from blue illusion in Balmain, sipping on champagne and a little shopping!!



Brigitte Salden from SBS, Robyn Kapp, Pauline Keyvar, Penny Bachell and Anna Falkinder

'Super Sunday'

Sunday 6th March was 'Super Sunday' for Huntington's disease in NSW.

- Over 50 people enjoyed a fun afternoon of 'interesting and informative' Trivia, delicious finger food and a selection of drinks including boutique beers at the Petersham Inn. We raised \$3,500 which will go towards our goal to employ a Youth Worker. Special thanks go to Penny Mitchell and Pauline Keyvar for their organisation and we are so grateful to Anthony, the Manager of the Petersham Inn, for putting on the event.



*Winners of the Trivia—
Therese Alting from Westmead Hospital and friends*

- Members of the Faulkner Family from Newcastle and the Central Coast held a reunion and raised funds for HNSW. Over 150 people, representing four generations, enjoyed a BBQ, bare-foot bowls and catching up with family members. Close to \$5,000 was raised on the day and special thanks go to Alison Daskalovski who organised the reunion.
- NBN television attended the Faulkner Family Reunion and interviewed Holly Faulkner and her cousin Brittani Faulkner. The interview was aired on the evening news. Thanks Holly & Brittani for your willingness to share your stories.



Brittani and Holly

- Lisa Genova, whose latest book "Inside the O'Briens" tells the story of a family who discover the husband/father has HD, was interviewed on the ABC TV program One Plus One.



What could be more fun than getting some friends together and having a party?

So why not make it a fundraiser for Huntington's NSW?

Hosting a High Tea 4 HD during the month of May (or any month) is an entertaining and yummy way for you to show your support for Huntington's NSW.

Included with this newsletter is registration form and helpful tips for a successful High Tea 4 HD. When you register your High Tea you will receive a fundraising kit that will help you to make your High Tea 4 HD a winner!

If you have any questions or need any assistance, please don't hesitate to ring Pauline on 9874 9777 or email her at pauline@huntingtonsnsw.org.au

Save the Date

Be Seen Wear Pink & Green

Put the date in your diary and watch out on Facebook and the Huntington's NSW website to register.

Join us on the Walk 4 Hope.

We promise to have a few surprises in store for you this year.

10th September Speers Point

11th September Canberra

18th September Parramatta Park

**WALK4
HOPE**



Rural Family Support Worker—Update

Unfortunately we have not been able to fill the Rural Family Support Worker position due to circumstances beyond our control.

We are extremely disappointed that it has taken so long, however, in the meantime, if you need to talk to someone please contact Robyn on 9874 9777 or 1800 244 735 (Country NSW only). She is usually in the office Monday to Wednesday and Friday. Or you can email her at robyn.kapp@huntingtonsnsw.org.au. Thank you for your patience

Carers' Support Group

Huntington's New South Wales and
Huntington Disease Service – Westmead
Hospital

*invite you to
come along to our*

CARERS' SUPPORT GROUP

*for a get together with other carers who, like
yourself, are caring for a partner, a family
member or a friend with HD.*

*Come along and join us as we share our chatter,
laughter, tears and experiences.*

Sessions will be held at
Huntington's NSW Offices
21 Chatham Road, West Ryde NSW 2114

2016 Sessions

23 March, Wednesday, 10.30am
20 April, Wednesday, 10.30am*
18 May, Wednesday, 10.30am
15 June, Wednesday, 10.30am
13 July, Wednesday, 10.30am*
17 August, Wednesday, 10.30am
5 October, Wednesday, 10.30am*
9 Dec, Fri, Year-end Get-Together (TBC)

** Denotes School Holidays*

*To RSVP and for further information, please
contact either: Jet Aserios or Cecelia Lincoln, SW
Department, Westmead Hospital on 9845 6699 or
Robyn Kapp, Huntington's NSW on 9874 9777*

It's time for Carers to speak out

Carers NSW is calling on anyone who cares for a family member or friend to speak out and help advocate for unpaid carers by completing the Carers NSW 2016 Carer Survey. Every response will contribute directly to the support of carers and Carers NSW advocacy work.

There are over 850,000 unpaid family and friend carers living in NSW alone and findings from the Carers NSW 2014 Carer Survey indicate that one-in-four carers provide in excess of 70 hours of care per week. The 2014 Carer Survey also highlights the significant personal consequences of providing so many hours of care, including poorer wellbeing, increased need for support, and difficulty in accessing services.

The results from the 2014 Carer Survey did show several positive trends, for example, the number of carers who stated that they started caring because they wanted to was more than double those who felt they had no choice and the positive value of caring for a loved one was consistently reported to be greater than the negative impacts of caring.

The Carer Survey is being conducted by Carers NSW in collaboration with researchers from the University of Wollongong, UNSW Australia, Macquarie University, University of Sydney, Australian Catholic University, and University of Melbourne.

The survey is completely anonymous and is open to any unpaid family or friend carer over the age of 16, living in NSW. Results from the survey will be published in a publicly available Carers NSW report, a number of peer reviewed journals and will be used to influence advocacy work.

The Carers NSW 2016 Carer Survey is available online at <https://www.surveymonkey.com/r/2016CarerSurvey> until Saturday 30 April 2016, hard copies of the survey can be ordered by contacting research@carersnsw.org.au.



“My voice”

An art fellowship for people with HD

Through art people with HD can be given a new voice.
Art offers new ways to express ones thoughts and feelings.

* Stephen Democoure, a highly experienced art facilitator, guides participants with their creative communication through the use of visual and poetic art forms.

Participants enjoy the calmness of creativity, the pleasure of friendship and a renewed sense of well-being.

Interactive gatherings are held fortnightly on Mondays;

21st March — 26th April, 2016,

between 10 am & 12.30 pm

**Elsie Court Cottage
21 Chatham Rd, West Ryde**

A light snack is provided afterward.

More program detail:
<http://www.huntingtonsnsw.org.au/activities/art-program>

To join, please contact Amanda Dickey on
9874 9777 or amanda@huntingtonsnsw.org.au.

Participants require independence with daily living activities.

If transport to and from the cottage is of concern, please advise Amanda so options can be explored.



Huntington's
New South Wales

Holiday Camp



Our annual camp on the Central Coast is fast approaching and we are inviting those with HD who might be interested in joining us for five days at Camp Breakaway, San Remo on the Central Coast, to contact us. The camp will be held from **Monday 2nd to Friday 6th May** inclusive.

The Camp provides an excellent opportunity for people with HD to connect with people who are in a similar situation to themselves, to talk, share experiences, participate in organised activities, indulge in great food, be with friends and have a great time.

Participants need to be able to walk, sit, stand, dress, undress, shower, eat, drink and use the toilet without requiring assistance.

For those who may need some assistance, a carer is very welcome to accompany them. Unfortunately we are unable to provide assistance with personal activities of daily living.

For all enquiries about the camp, to discuss eligibility and to obtain an application form please call us on 9874 9777 or email Amanda at amanda@huntingtonsnsw.org.au

Do you have a story to share?

If you have a story that you would like to share in "Gateway" please contact Robyn at robyn.kapp@huntingtonsnsw.org.au



As reported in our last edition of Gateway, the Enroll-HD study is now up and running at Westmead Hospital. This international study presents an opportunity for all family members affected by Huntington's disease to be involved in research. The study is an international collaboration, with study sites in the USA, Europe and South America. As of October 2015, close to 8,000 participants had been recruited world-wide.

The study is 'observational'. It is not a drug trial, but a way of gathering a large database of information to prepare for the next phase of research. Study visits are similar to usual clinic visits, comprising questionnaires, a physical examination, memory testing and blood collection. The main focus is on gene carriers, either affected or not, but the study is also open to the whole family.

The study involves a baseline visit followed by one visit annually. The first visit is the longest, and takes about two and a half hours. Studies after this should be around an hour. The Enroll-HD website is a great source of information about the study. You can find more information at the website, www.enroll-hd.org

The study organisers hope to recruit 20,000 people at risk or with HD worldwide. We are told that the study is designed with privacy concerns being paramount, and all information collected is de-identified. It is also possible to withdraw at any time.

If you are interested in participating in this exciting new study which represents the next phase of research into HD, you can contact the study coordinator, Therese Alting, at the Westmead HD Service. Telephone 9845 6310 or 0438 604 719 or email her at therese.aling@health.nsw.gov.au

Amanda's Story

My name is Amanda, I'm 28 years old and this is my story.

Our family didn't become aware of Huntington's disease until I was 14 years old when we found out dad had HD. Previous to that we thought dad was having a nervous breakdown as he started to behave a little different. Dad was a very lean, fit, handsome man who loved his daughters. He was the kind of man that would do anything for anyone and always lend a helping hand. He loved to run every day and participate in marathons and drink a lot of Coca Cola, they were his favourite things beside us girls, "his babies". We all had nick names. Shauna was Possum, Donna was Wombat, Kim was Bill and I was Weasel.

Our Mum and Dad separated when I was 4 years old. Dad would come and pick us up of a weekend and spend time with us then one day we thought he was arriving to pick us up only for him to tell us he cannot see us anymore and left. I was about 8 at this time and didn't understand and became very upset. Dad refused to see us again until he was admitted to hospital with a broken ankle he had been running on for months. I was 15 at this time. When we arrived at the hospital I didn't really know what to expect or if he would recognise any of his daughters but as soon as we walked in that room he yelled out "My Babies"! He was so happy to see us.

Some of Dads earlier symptoms included confusion, short temper, aggression, strict and odd routines and memory loss.

Dad moved around a few nursing homes which was quite unsettling but then made his home at Lady of Consolation in Sydney's west where he spent his last moments.

We then had our sister Donna tested for HD at the age of 23 because she also had unusual behaviour leading back to when she was a teenager and was previously diagnosed with schizophrenia. Some of Donna's earlier symptoms included seizures, learning difficulties, depression, hearing voices, paranoia. She was also quite the rebel but underneath all that there was still a gently spoken, giving, caring and loving beautiful young girl who loved to shop and loved to admire herself... A real girly girl.

By the age of 28 we were no longer able to care for Donna and mum had to make the heartbreaking decision to find her a home. We were lucky to find a fairly good nursing home for her even though it devastated us to see her to be surrounded by only elderly people and her decline.

In 2010 we had just lost our mum and 5 short months later in Feb 2011, Donna passed away too. She was 31.

Our eldest sister, Shauna decided to be tested after we found out dad had this debilitating disease. Shauna tested positive for the HD gene.

That didn't stop her from living life though. Shauna was a go getter, An outgoing, social person with a love for an adrenalin rush. Shauna has two beautiful daughters, Jasmine- 18 and Lakyn 2.

In October 2013 I moved down to Tasmania where Shauna and Lakyn were living with my sister Kim and her family. I made the decision to move my, what was free life in Sydney to come care for my sister and her baby, Lakyn because I couldn't have them split up and Shauna's symptoms were progressing and couldn't be left at home alone with Lakyn. Some of Shauna's symptoms included memory loss, confusion, irritability, depression, anxiety, swallowing difficulties, indecisiveness and lived on a specific routine. Some would say to look at Shauna, she looked normal but that is because they couldn't see her cognitive and behavioural symptoms of HD. Shauna passed away in May 2015

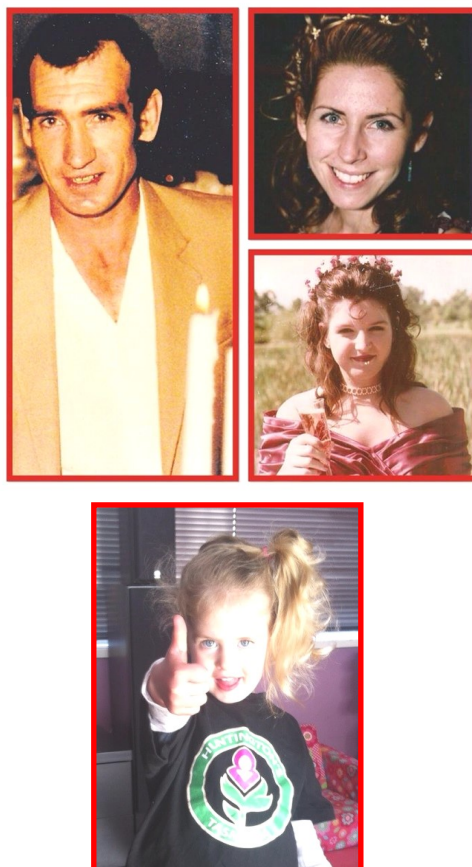
leaving behind two beautiful daughters. I am now Lakyn's legal guardian and will never let her forget what kind of woman, what kind of mum Shauna was.

Making the decision to be tested was really no choice for me. I needed to know and as scary as the whole process was right up to those few seconds before revealing the results I knew I needed to know for my future and I couldn't live in the unknown. I was fortunate enough to be gene negative, but after receiving that result I felt a lot of guilt for Shauna and Donna who both had the HD gene. At the end of the day we cannot change who we are but not let it stop us from living our life in the present because you just don't know what's around the corner!

For me, I am not going to lie, being a carer and being a sister was not an easy role. Especially with Shauna. I was old enough to see and remember her before the symptoms started. My big sister, someone I looked up to and confided in was going to be the one that was dependant on me. The last 2 years of being her carer has been such a roller coaster of emotions. I also had my own set of symptoms as someone being impacted by HD. I was depressed, isolated, started becoming an introvert, feelings of guilt, worry, anxiety, de-motivated just to name a few. Not to mention the stress it has on your relationships with others. I do believe it takes a special person to do it. Being a carer changes your being and you need to be willing to accept that.

I am sharing this with you all because I'm not ashamed of Huntington's disease or my dad for passing the gene on to my sisters and why? Because it's no one's FAULT!
If we don't speak up for them who will? This disease needs to get noticed and supported!

This disease has taken away half my family and now I'm fighting back!
Fighting for **HOPE**, Fighting for **DIGNITY**, Fighting for **AWARENESS** and Fighting for a **CURE!!**



Acknowledgement: Huntington's Tasmania Newsletter November 2015

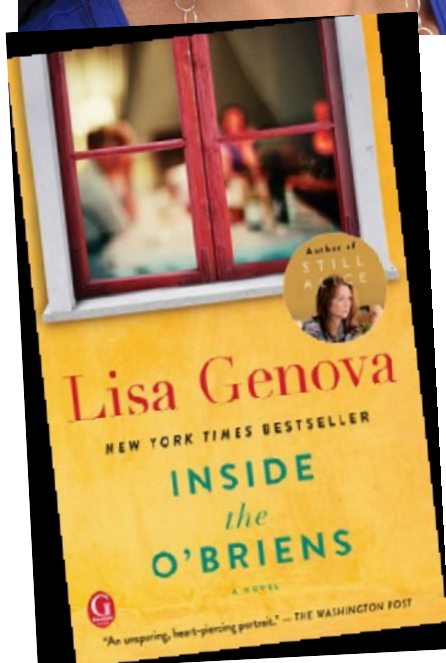
The Power of Fiction



In a piece written exclusively for EHDN News (European Huntington's Disease Network), March 2016, Issue 27, American neuroscientist and author Lisa Genova talks about why she uses fiction to explore the subject of neurological disease. Her first novel, the bestselling *Still Alice*, dealt with Alzheimer's disease and was made into an award-winning film starring Julianne Moore. Lisa's latest novel, another bestseller called *Inside the O'Briens* (Gallery Books, 2015), tells the story of a family, the O'Briens, affected by Huntington's disease.

My first year out of college, I worked as a lab technician in a neurobiology lab researching drug addiction. I was 22 years old in February 1993 when the scientists down the hall began celebrating. They had just isolated the genetic mutation that causes HD. I remember getting very still, the goose-bumps on my arms, knowing I was witnessing a historic moment in all of neuroscience. Only one thing causes HD, and these scientists had just discovered it. Surely, there would be a cure for HD. We are now 23 years later, and we still don't have a treatment or cure.

I've found that fiction is a powerful way in. Stories are accessible. Unless you're a geeky neuroscientist like me, you're probably not going to read the *Journal of Neuro-science* to learn about Huntington's disease. But you might read a novel called *Inside the O'Briens* (and I hope you do!). My role is to tell the truth under the imagined circumstances of my novels, to write informed fiction, to give the reader real medical information, but to package it in an accessible, human story that we can all relate to. I wrote *Inside the O'Briens* to hopefully create some much-needed awareness about a disease most people know little about.



And I've found that awareness and conversation are critically necessary steps in the march toward treatments and survivors. Historically, we've seen this with cancer and HIV. Awareness, open conversation, lifting the shame, secrecy, and stigma, acknowledging that the disease exists, are essential to developing treatments that lead to survivors. It's impossible to cure something that seemingly doesn't exist. A sense of urgency is needed. We're seeing this urgency happen with Alzheimer's, a disease we've all been terrified to openly talk about. *Still Alice* is playing a role in this, acting as a vehicle for conversation, for breaking down isolation and fear.

Inside the O'Briens is about Huntington's, but it's also about what's inside us and what gets passed down through the generations—not just our DNA, but also our faith, humour, resilience, love, gratitude. It's about how to find hope in a situation that appears hopeless. It's about finding courage when you're completely vulnerable. It's about learning to live in the moment. The future is a fantasy—only the present moment is real.

A review of *Inside the O'Briens*, can be found in Gateway, Vol 18, No 2, Winter 2015 or at <http://www.huntingtonsnsw.org.au/information/publications/gateway>

How NOT to write a news article about a clinical trial

A confusing story about a huntingtin lowering trial is published in the UK Telegraph, but cool new stuff is happening!

By Dr Jeff Carroll on March 11, 2016 Edited by Dr Tamara Maiuri

A recent article in the UK newspaper the Daily Telegraph has HD families very excited. The title, "First drug to reverse Huntington's disease begins human trials", certainly sounds exciting! But what's really going on? HDBuzz is here to help us untangle hope from hype in the huntingtin lowering world.

The typo heard round the world

The UK newspaper the Daily Telegraph recently posted an article titled: "First drug to reverse Huntingdon's disease begins human trials". Your first sign that an article about HD may not be well researched is the fact that they've misspelled *Huntington's disease* as *Huntingdon's disease*!

The authors of the article also apparently have a time machine, because they reported that the research they were describing: "was presented at the American Academy of Neurology's 68th Annual Meeting in Vancouver, Canada, April 15 to 21, 2016".

Given that the article was posted online on February 26th, 2016, this is a pretty neat trick!

The logo for The Telegraph, featuring the word "The" in a smaller, elegant serif font above the word "Telegraph" in a larger, bold, blackletter-style serif font.

A garbled news story about HD in the UK Daily Telegraph might have distracted families from a very exciting achievement.

What's the article actually describe?

So what's the actual news behind this garbled news story? The HD drug trial being described is testing whether a *huntingtin lowering* approach using drugs called *antisense oligonucleotides* (or ASOs) is safe and effective for HD. If this sounds familiar, it's because you probably read about it here, on October 19th, 2015: <http://en.hdbuzz.net/204>.

Briefly, a company called Ionis, previously known as Isis, has developed drugs that reduce the levels of the harmful huntingtin protein in cells. In collaboration with the drug company Roche and a worldwide team of physicians, they're testing whether this drug is safe in early-stage HD patients. Of course, we hope that this drug will not only be safe but will also improve symptoms or even slow the progression of HD, but that answer will come from future trials.

"While the clinical trial described in the Telegraph's news story was already well known to HDBuzz readers, the field just had another major advance that you might not have heard about."

Ionis has previously shown that ASO drugs make HD mice look much healthier. This is very cool, but it's definitely not evidence that these drugs "reverse Huntington's disease", as stated in the article. Take it from someone who has studied HD mice for over 10 years - **only humans have HD!** The lab results with Ionis' drugs in HD mice are extremely exciting, but only trials in humans can tell us whether these drugs do anything for HD.

Why would a story like this get written at all? It's unfortunately become common for papers like the Daily Telegraph to rely on press releases from drug companies, universities and academic societies as source material for science news. A little digging in this case would have revealed that the trial supposedly being announced is already running, and that there really was no new story to get people excited about. Unfortunately, accurate science journalism doesn't seem to be a priority of many news agencies.

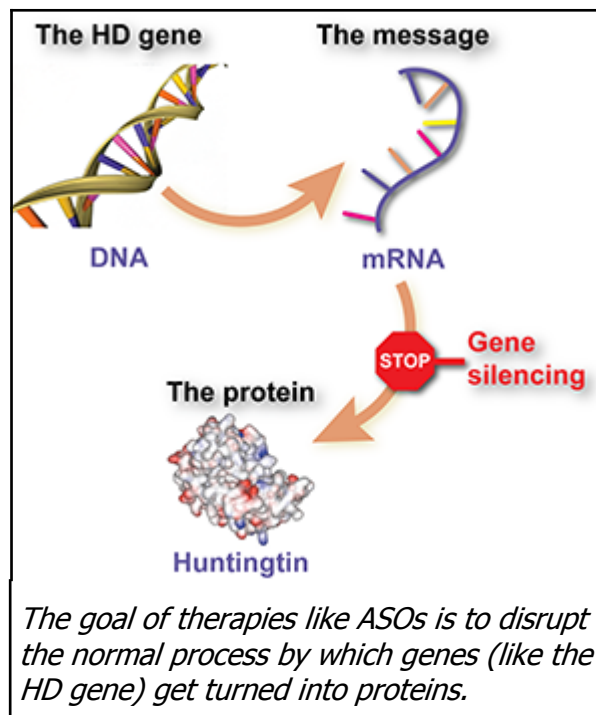
So, no news?

While the drug trial described in the Telegraph's news story was already well known to HDBuzz readers, the field just had another major advance that you might not have heard about.

Ionis is testing their ASO technology in other brain diseases, besides HD. One of these is a terrible disease of children called *spinal muscular atrophy*, which results in progressive loss of muscle function, commonly leading to permanent disability and early death.

One experimental treatment for spinal muscular atrophy is the delivery of ASO drugs to the fluid surrounding the brain and spinal cord. The ASOs being developed for spinal muscular atrophy have a different sequence than the ones Ionis is testing for HD, but are delivered in much the same way, and have a similar chemical structure.

So it's big news that on March 8th, a group of researchers published results describing the successful completion of a safety study of an ASO drug in spinal muscular atrophy! That study, very similar to the Ionis/Roche one being done currently in HD, was designed to establish whether the treatment was safe and well-tolerated by kids with spinal muscular atrophy.



As in the HD trial, ASOs for spinal muscular atrophy have to be delivered directly into the spinal fluid. This is a relatively routine procedure for neurologists and is very similar to the epidural anesthesia that many women have during childbirth, but is obviously much more complicated than taking a pill. So it's huge news that kids with spinal muscular atrophy were able to have the drugs delivered in this way without any major complications.

The spinal muscular atrophy study also gives us very important new information about how long ASOs last in the fluid surrounding the brain. Researchers found that even 4-5 months after a single injection, half of the injected drug was still around. This means we might be able to dose people less frequently in future HD trials of ASOs, which is great news.

Another major finding of the spinal muscular atrophy study was that kids who received the highest dose of drug actually had improved symptoms, which is not the expected outcome for spinal muscular atrophy. The numbers are small (only 10 kids in the group that showed improvements), but this is an incredibly exciting result for families afflicted by spinal muscular atrophy, and it bolsters our hope for ASOs in HD.

Ignore the noise, keep your eyes on the prize!

This busy HD news week is a reminder of the ups and downs of trying to follow HD research. Sometimes what seems to be a big advance is really something that we already knew about, re-packaged by a news outlet looking for a story. But remember, really important advances are being made in the clinic and labs around the world that give us realistic hope for effective treatments against HD. Our advice - try to tune out the noise, when big stuff happens, you'll hear about it here!

Acknowledgement: <http://en.hdbuzz.net/215>





Huntington's New South Wales

PO Box 178, West Ryde, NSW 1685
21 Chatham Road, West Ryde, NSW 2114
Telephone: (02) 9874 9777 Facsimile: (02) 9874 9177
Free Call: 1800 244 735 (Country NSW only)
Email: info@huntingtonsnsw.org.au
Web Site: www.huntingtonsnsw.org.au

AHDA (NSW) Inc

The Australian Huntington's Disease Association (NSW) Inc is a not-for-profit organisation established in 1975.

Our Mission

The energies and resources of the Australian Huntington's Disease Association (NSW) Inc are directed towards satisfying the needs of people with or at risk for Huntington's Disease and their families in NSW and the ACT by providing and/or facilitating delivery of a range of quality services.

Our Philosophy

People with Huntington's Disease and their families are individuals with equal value to all other members of Australian society, with the right to treatment and care by knowledgeable professionals and care givers, the right to appropriate support services and the right to have the best quality of life possible.

Our Services

These include education and information; advocacy; counselling and referral; holiday programs; family support; rural outreach and client services.

Our Board

President: Brian Rumbold
Vice President: Deb Cockrell
Treasurer: Richard Bobbitt
Secretary: Therese Alting
Members: Felicity O'Neil & Katy Clymo

Association and Other Useful Contacts

Huntington's NSW

Robyn Kapp OAM
Executive Officer

Pauline Keyvar
Fundraising & Marketing

Stewart Swales
Administration Co-ordinator

Amanda Dickey
Programs Officer

Huntington Disease Service

Dr Clement Loy
Director
Westmead Hospital
(02) 9845 6793

Outreach Service
Colleen McKinnon
Westmead Hospital
(02) 9845 9960

Dr Sam Kim
Neurologist
Westmead Hospital
(02) 9845 6793

Huntington's Unit
St Joseph's Hospital
(02) 9749 0215

Research Queries
Dr Elizabeth McCusker
(02) 9845 6793

Predictive Testing

Fiona Richards
Social Worker
The Children's Hospital
Westmead
(02) 9845 3273

HD Clinic Appointments
Outpatients Department
Westmead Hospital
(02) 9845 6544

Jet Aserios
Social Worker
Westmead Hospital
(02) 9845 6699

Hunter HD Service

John Conaghan
Social Worker
John Hunter Hospital
(02) 4922 3076

Cecelia Lincoln
Social Worker
Westmead Hospital
(02) 9845 6699