

# HJ News

AUTUMN  
2022

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Jaime Bodiam

## YOUNGER PEOPLE IN RESIDENTIAL AGED CARE (YPIRAC)

WORDS ABILITY FIRST AUSTRALIA

The Younger People in Residential Aged Care (YPIRAC) System Coordinator Program is a national initiative funded by the Commonwealth Government and operated by Ability First Australia.

It aims to give younger people (under 65 years) greater choice and control in understanding their options where they would like to live and what age-appropriate supports they need. With the consent of the younger person and/or their guardian, the YPIRAC team works collaboratively with government, local community and welfare organisations and service providers to help explore alternate home and support options.

**Who do they work with?** The program works with people under 65 living in residential aged care

as well as those in hospital or in the community who are at risk of entering permanent residential aged care.

**To make a referral for those at risk:** The My Aged Care Contact Centre is responsible for referring younger people at risk of entering residential aged care to the program if they are not an NDIS participant or they have not yet tested their eligibility.

With consent, YPIRAC will assist the person to test their NDIS eligibility, if appropriate, whilst they continue to explore age-appropriate home and support options to avoid entry to permanent residential aged care.

**For those under 65 years living in residential aged care:** the YPIRAC team will make direct contact with

residential aged care facilities who have been identified as having a younger person as a resident.

With consent, YPIRAC will assist the person to test their NDIS eligibility, if appropriate, and if the person would like to transition out of care, provide assistance to find alternative age-appropriate home and support options.

If the person is happy with their current situation and make the choice to stay, the YPIRAC team can then assist to explore additional supports, further enabling them to live their best life.

**ability first**  
AUSTRALIA

For more information about the program or to make a referral call Ability First Australia on 1800 771 663 (option 1).



## FROM THE DESK *Lewis Kaplan*

### LEWIS KAPLAN, CEO Huntington's NSW ACT

We are looking forward to some sort of return to normality over coming months with the expected slowing of the COVID-19 pandemic.

Plans for the year include the review and updating of our current strategic plan which was written in 2019 for 3 years.

We are aiming to start taking clients in our Illawarra HD Social Work project, supported by funding from the wonderful Co-Group. In an ideal world we would have an HD specialist social worker in each part of regional NSW and the ACT. But the cost of a full-time position would exceed \$150,000 per year and neither NSW Health nor local health services have shown any interest in finding this amount of money for an on-going regional service.

Our focus on supporting younger people with HD currently living in residential aged care is progressing on a number of fronts. We have established contact with Ability First (see page 1) whose role is to assist younger people in aged care. This organisation's first priority is to work with younger people in aged care who do not yet have an NDIS plan. Our research has shown that there are 44 people with HD under the age of 65 living in aged care without an NDIS plan, of whom 23 are in NSW and 1 is in the ACT. We don't know why such a high proportion are in NSW, but we will try to fix this urgently.

We have been lucky to win some funding from the NSW Government under the Social Sector Transformation Fund to upgrade our IT system. We will be working to integrate our client relations management system with our website, and we are looking at collaborating with other state HD associations to create efficiencies and streamline data management.

The next federal election will be called anytime (or may have already been called after we went to press). Two key issues for many people with HD are the quality and quantity of NDIS services they can access. We have been involved in an advocacy campaign called "Teamwork Works" led by National Disability Services. You can join by joining the campaign at <https://teamwork.org.au/> and/or sign an open letter [here](#).

We have in the last few months also met the Minister for the NDIS, Senator the Hon. Linda Reynolds and the Shadow Minister, the Hon Bill Shorten MP. We continue to make the key point that people with HD are different from the majority of NDIS participants as they have a degenerative and unpredictable condition. Most NDIS planners have no experience or understanding of HD and this reduces the likelihood of developing a quality NDIS plan.



### STOP PRESS

The Association's Board has recently approved the establishment of a new annual youth grants program with support from a generous benefactor. The grants will be made in the name of the benefactor's sister who died with HD: "the Ailsa Legacies", with four grants of up to \$2,000 plus some funding available in reserve for emergency support outside the annual grant cycle. More details will be available soon but first and foremost, many, many thanks to our anonymous benefactor for this generous gesture.

### Congratulations to the Scottish Huntington's Association!

A motion to expand specialist services for families in Scotland affected by HD received a historic level of cross-party support, backed by 98% of members of the Scottish Parliament (MSPs), the most supported in the country's parliamentary history.

The motion followed a recent study by Aberdeen University published in the Journal of Neurology that found HD cases in northern Scotland increased by nearly 50% over the last 30 years. Across the region, 14.6 per 100,000 individuals have been diagnosed with Huntington's. By contrast, 36 years ago that was 9.94 per 100,000.

# ORAL DRUG MAY CHANGE THE STORY FOR HUNTINGTIN LOWERING

**SOURCE** HD BUZZ

**WORDS** DR SARAH HERNANDEZ AND DR JEFF CARROLL



Huntingtin lowering has gained lots of attention in HD research, and for good reason. It was the first potential treatment designed to directly target the cause of HD – the huntingtin protein. But there are limitations to current huntingtin lowering approaches: they require delivery to the spinal fluid or brain surgery for delivery, can show limited distribution within the brain, and don't cross the blood-brain barrier (which is why they require a lumbar puncture or brain surgery). They also don't reduce huntingtin outside of the brain in "peripheral" tissue.

Scientists from PTC Therapeutics recently published their work in the prestigious journal *Nature Communications* describing a series of drug molecules that lower huntingtin which can be taken orally, and show distribution throughout the brain and body. These are results that would have sounded like science fiction even 5 years ago. But in a post-2020 world, wonders never cease! So let's dive into what their data show and what it means for huntingtin lowering.

## EFFECTS ARE ADJUSTABLE AND REVERSIBLE

All the previous experiments were done on cells in a dish. What happens when you give HTT-C2 to an entire organism? Does it have the same effect? Can it lower huntingtin in the brain? To answer those questions, PTC's researchers turned to mouse models of HD.

Mice were fed HTT-C2 every day. Yes, you read that correctly – the mice were given this drug orally. This is a huge difference from previous huntingtin lowering therapeutics! For those who have followed the huntingtin lowering

field, previous therapeutics have required either lumbar puncture or brain surgery, neither of which are ideal. This new type of approach, that accomplishes huntingtin lowering with a pill, could be a game changer for patients who have been hesitant about more invasive treatments.

Encouragingly, the more HTT-C2 the mice were given, the more huntingtin was lowered. This is great news because it suggests HTT-C2 dosage can be actively adjusted to change how much huntingtin is lowered. We don't yet know how much total huntingtin should be lowered in people to produce beneficial effects without being harmful, so this is a massive safety advantage – if huntingtin isn't lowered enough, more drug can be given, and if huntingtin is lowered too much, the dose can be reduced.

Another exciting finding is that the effects of HTT-C2 were quickly reversible. Just 10 days after treatment stopped, huntingtin levels returned to that which was observed prior to treatment. This is another major safety advantage – the "washout" of this drug is very fast, meaning the time it takes for the drug to leave a patient's system will be relatively quick. If a negative consequence is observed after HTT-C2 is given, the effects can be quickly reversed. However, 10 days is the washout timeline in mice and this will likely be different in people.

## REDUCES HUNTINGTIN IN THE BRAIN AND BODY

HTT-C2 treatment targets both the expanded and unexpanded copies of huntingtin, unlike current ASO-based approaches by Wave Life Sciences that will only target

the expanded copy. Because the unexpanded huntingtin copy is still needed to carry out its normal jobs inside cells, it's important to track how much of both copies are being lowered.

When researchers looked at the brains of HD mice dosed with HTT-C2, they saw about 50% reduction of both expanded and unexpanded huntingtin throughout the whole brain. This included in regions most sensitive to HD, which suggests HTT-C2 is having an effect in the areas where it's needed most.

Huntingtin is expressed in all cell types of the body, not just the brain. So while we know about the effects of HD in the brain because changes there control the most apparent parts of the disease like mood changes and chorea, there are also effects in other tissues like the heart and muscles. Because of this, it may also be beneficial to lower huntingtin in all tissues, not just the brain.

When the authors assessed how much HTT-C2 lowered huntingtin in tissue outside of the brain, they found it was actually much higher than in the brain – it was lowered over 90%! While research suggests a 50% reduction would be tolerated, 90% is likely too much.

For safety reasons, the researchers at PTC further tweaked the drug by changing the chemical structure – resulting in another a drug they called HTT-D3. When given to mice, HTT-D3 showed huntingtin lowering in both the brain and body to equal levels of around 50%.

**To continue reading the story please visit the HD Buzz site <https://en.hdbuzz.net/317>.**

## WHY IS STORYTELLING SO IMPORTANT?

HD impacts every family differently and it's time that we tell our story. #LetsTalkAboutHD is a global effort to educate the world about the impact HD has on families. Huntington's NSW ACT is joining the campaign and would love you to join in too.

Our hope is to create a video montage, little snippets of families sharing their HD journey. If we can tell our stories, then we can get more help and support. Together we can work towards our long-

term objective of building regional hubs with social workers to support families.

We would love to have the videos ready to share in May when it is International Huntington's Disease Awareness Month. We are also hoping our community members can share stories and photos on social media using #LetsTalkAboutHD to educate people. If you would prefer you can send your stories to us, and we will share them on our social media.



## CHRISTMAS WRAPPED UP

Our Christmas appeal was sent out in December, and we want to say thank you to each person who donated. Your continued generosity allows us to support people and families affected by HD. Who would have thought that two years on we would be still facing this COVID-19 pandemic? **Even in this uncertain time our community managed to raise \$9,815.**

## HIGH TEA 4 HD

Having a High Tea 4 HD fundraiser is a great idea, particularly for people who enjoy an afternoon of yummy nibbles, warm drinks, and a chat.

Why not get your friends and family together for an afternoon of fine dining with delicious treats, a pot of tea and maybe a glass of bubbly or two? The next step is to find a venue. You could approach your local club, or function venue that is large enough and offers an atmosphere to complement the food and drink of the day.

Ideally you would charge people to attend, and you could also run additional fundraising activities during the afternoon.

**If you are keen to get a high tea underway please make contact with Alison by email [alison@hdnswact.org.au](mailto:alison@hdnswact.org.au) or call 0419 465 612.**



# HUNT 4 HUNTINGTON'S

An inspiring recap from Dan at F45 Singleton on their Hunt 4 Huntington's row held in January. This has been an incredible fundraiser! We would like to thank each and every person, business and community member who helped raise awareness and funds for Huntington's.

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F45 Singleton would like to thank Huntington's NSW ACT for allowing us to fundraise on their behalf and Bailey Property & Livestock who was our event major sponsor. Without either of these the event would not have been possible.

We couldn't be prouder of how the event unfolded and the support the community showed for Jaime's story and Huntington's Disease, with many families and friends joining in support for our rowers to cheer them through their 1hr time slot, which everyone completed. With a combined total rowed distance of 914,000m and a total amount raised of \$14,900 for Huntington's, it is fair to say this event was a success!

Now to extend a massive thankyou to all 26 local businesses who jumped in to sponsor a member of our community to row for 1 hour and the 48 members who donated and rowed to fill a total of 3 rowers every hour for 24 hours. Notable donations from 'Kind' supplying over 100 muesli bars and Nutrition Warehouse for goodie bags. You have all reminded us just how strong and giving our community is for raising awareness and funds for a great cause.

Jaime's courage and determination reminds us all that anything is possible when we put our mind to it. During the hard times, those around us can help carry us through. If you want to go fast, go alone. If you want to go far, go together. Together we did our part in supporting Huntington's NSW ACT with their incredible work.

## The F45 Singleton Team





# MEMBERSHIP SURVEY THE JOURNEY TOWARD EMPOWERMENT

**WORDS** AMANDA DICKEY

The next step in strengthening community participation saw us review our surveying approach. We discovered that the annual surveys lacked focus, and have tried to ask too much, of too broad an audience. This does not serve good decision making well.

A more purposeful method involved collaborating with our brains trust (the Community Advisory Committee) so we can ask better questions, in the right context, and to the right audience.

We will now conduct surveys as follows:

- an association membership survey - to capture member perceptions of our performance against our strategic goals and objectives.

- a voice of the community survey - to gather people's thoughts, opinions, and feelings aiming to understand our HD community's concerns and aspirations better.

To kick this all off we will invite association members to complete a short survey. Members will be emailed online access to the survey in March. To request a hard copy please call the office.

The information members provide will be used confidentially and will not be shared with anyone outside our organisation. Their views will be considered when planning our strategy and support services.

Want to be involved but not a member?

Sign up now on our website or call the office for assistance.

**More information about how we'll capture the voice of the wider community will come in a later edition.**

## A GIFT OF HOPE

**Have you made a Will?**

**Is your Will up to date?**

A gift in your Will is a gift of hope that allows you to make a difference in the fight against Huntington's disease beyond your lifetime. We need your help to ensure that we can keep on being the voice for current and future generations of families facing Huntington's disease.

Our goal is to continue to reach out and be there in those tough times when everything may seem so hopeless. Lobbying government, providing support and education, and raising awareness to remove the social isolation and fear for families who have been impacted by Huntington's disease.

Leaving something behind is a lovely legacy that can help others.

**For more information or to obtain a copy of our Gift of Hope flyer please email [info@hdnswact.org.au](mailto:info@hdnswact.org.au) or call the office on 9874 9777.**

## CARER GATEWAY An Australian Government initiative that is making some changes to better help Australian carers.

**WORDS** AMANDA DICKEY

In the tail end of 2021, we collaborated with the 5 Carer Gateway service providers for NSW and ACT. This was to cut through the confusion felt by our community and form a better understanding of the latest supports and services available to carers.

We then hosted two carer information sessions (with Q&A) for the purpose of introducing this national initiative to our carer community. Three more sessions are planned for later this year.

So, what is the Carer Gateway?

In a nutshell, the program:

- provides practical information and advice to carers
- helps carers get the services and support they need
- provides free counselling services
- provides free coaching to help with the caring role
- connects carers together through a community forum.

**Feel ready to register for Carer Gateway support? Call 1800 422 737.**

**Want to know more? Visit our website at: <https://huntingtonsnswact.org.au/service/carers-gateway/> and register for a future information session.**



# THE HD SPOTLIGHT ON

## Jaime Bodiam

Community member  
and Juvenile HD mum



### Q Tell us about your HD Journey?

**A** My daughter Lily was just 10 years old when diagnosed with Juvenile Huntington's disease (JHD). We had spent the previous 6 years investigating what was happening to her. Lily went through multiple diagnoses including speech delay, sensory processing disorder, Asperger's, autism, and mitochondrial disease. In doing so, Lily went through a multitude of unnecessary procedures including liver biopsies, muscle biopsies and multiple MRIs under general anaesthetic. A simple blood test was all it took in the end for an automatic diagnosis of HD for Lily and her Dad.

By this stage Lily was already in a wheelchair, had a feeding tube and a full-time carer at her school. Fortunately, I was able to care for Lily right until the very end at home with the help of her Palliative Care Team. This was important for us, seeing as Lily was only 16 and didn't want to be in hospital away from her siblings and her puppies.



During the last 12 months we received the devastating news that my two sons aged 15 & 18 also had the positive gene for HD. My 12-year-old daughter hasn't been tested.

### Q What unique barriers do you think JHD families face?

**A** JHD is extremely rare and it was not only my first experience with JHD but the first time almost everyone in our lives had even heard of HD. I felt very alone as I was navigating the changes and trying to work out what the future would be for Lily.

After Lily's diagnosis of JHD, I had to navigate my way through supporting Lily, facing the way the disease progressed, modifying our house to accommodate the required equipment, building and coordinating an amazing team around me to help to support Lily and basically learning how to become a nurse.

### Q If you had time with the Prime Minister what would you like him to change to improve the lives of HD families?

**A** There are a number of things the PM has the power to do in order to improve the lives of HD families:

- Ensuring NDIS funding is sufficient for the needs of those with HD and ensuring it is scalable as the disease progresses. For us, the NDIS was an absolute nightmare to begin with. It was time consuming with so many

barriers just to get the equipment that every person with HD should be provided.

- Increase education and awareness of HD. Because of the degeneration of the brain, many people with HD end up making bad decisions or becoming increasingly frustrated when attempting to communicate and end up in situations that can be avoided with better awareness of HD and its symptoms.
- Significant money needs to be invested into research and trials for HD so that treatments can be developed to arrest the progression of HD.

### Q Why do you think it is important for HD families to share their HD story and fundraise?

**A** Although we live in a beautiful caring and tight knit community, many of the people who took part in the fundraiser, many who have known my family almost our entire lives, just didn't know the full extent of what happened to Lily and the devastation the disease causes. Fundraising not only secures much needed funds for research and support but ensures a greater understanding of not only HD but other rare diseases and the people who have them.

### Q What is your ideal Sunday?

**A** A sleep in, a coffee then either a day at the beach with the kids or working in my garden.



# Huntington's NSW ACT

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## Our team

### LEWIS KAPLAN

Chief Executive Officer  
0407 108 667

### GISELLE BEAUMONT

NDIS Senior Support  
Coordinator  
0422 604 737

### MALISHA FERNANDO

NDIS Support Coordinator

### AMANDA DICKEY

Community Programs Officer  
0456 013 612

### VITA SOLANO

Illawarra HD Social Worker  
0413 195 398

### AMY HALE

Youth Social Worker  
0499 031 231

### STEWART SWALES

Administration Co-ordinator

### ALISON WEIR

Fundraising & Marketing  
0419 465 612

## HD SERVICE

### Westmead Hospital

**DR CLEMENT LOY  
(DIRECTOR), DR SAM KIM,  
DR FLORENCE CHANG**

Neurologists  
(02) 8890 6793

**CECELIA LINCOLN &  
FELICITY STEHOUSER**

Social Workers  
(02) 8890 6699

### PETER KWONG

Clinical Nurse Specialist  
(02) 8890 9960

HD Clinic Appointments  
Outpatients Department  
(02) 8890 6544

## HUNTER HD SERVICE

### John Hunter Hospital

### JOHN CONAGHAN

Social Worker  
(02) 4922 3076

## HUNTINGTON'S UNIT

### St Joseph's Hospital

(02) 9749 0215

## PREDICTIVE TESTING

Find your nearest Genetics  
Clinic at [www.genetics.edu.au/genetic-services](http://www.genetics.edu.au/genetic-services)

## EDUCATION RESOURCES

### Huntington Study Group

Global HD cooperative  
therapeutic research org.  
<https://huntingtonstudygroup.org>

### HDYO

Support and education to  
young people (aged up  
to 35) impacted by HD  
around the world.  
<https://en.hdyo.org/>

## RESEARCH QUERIES

### Dr Therese Alting

Enroll-HD Study  
(Westmead Hospital)  
(02) 8890 6310 or  
0438 604 719

### HDBuzz

Research news. In plain  
language. Written by  
scientists. For the global  
HD community.  
<https://en.hdbuzz.net/>

# WHAT'S HAPPENING?



Due to the COVID-19 pandemic and changing government restrictions, our support groups are meeting in a variety of ways. RSVPs are now essential for all meetings as there are COVID protocols that need to be followed to keep everyone safe. Take a look at how your region is choosing to connect.

## CARER SUPPORT

### CENTRAL COAST

Postponed. Seeking interest.

### COFFS HARBOUR

Group closed to new registrations due to latest COVID outbreak.

### NEWCASTLE

Jesmond, 10:00am - 12:00pm  
2nd Thursday monthly.

Note: Meeting room limited to 12 attendees

### SYDNEY

Westmead Outreach are not running a face-to-face group in 2022.

### Online Carer Support group

This group is open to both metro and regional carers.

## COMMUNITY SUPPORT

### ACT

Postponed due to latest COVID outbreak.

### CENTRAL COAST

Seeking interest.

### ORANGE & CENTRAL WEST

Orange, 10:30am - 12:30pm  
1st Saturday monthly.

### PORT MACQUARIE

Group closed to new registrations.

### WOLLONGONG

Online, 10:00am - 11:00am  
1st Monday monthly.

**Latest details and to register your interest can be found on our website at: <https://huntingtonsnswact.org.au/support-groups/> or contact us on 02 9874 9777.**