



Gateway

News from Huntington's New South Wales
Volume 14 No 1

Summer 2011

From the Executive Officer

Welcome to the first Gateway for 2011! At the start of any New Year it is a good idea to reflect on the events of the end of the previous year and look forward to upcoming events. With this in mind you will see this issue of Gateway is crammed with reports on the highlights of recent social and family events. As you can see, our work continues and the success of our events, programs and the development of our relationships and networks continue to go from strength to strength.

With respect to upcoming events, we have lots of exciting activities planned for the day respite program (social/lunch club) this year, the dates of which can be found in this Issue. The Camp is also coming up in March and preparations are well under way. Don't forget to register your interest early as there is already high demand for places this year.

Our Family Support team are planning trips to the various parts of NSW and ACT this year so that they can meet and talk with members of the HD community in those areas and find out how we can better meet your needs and provide you with support in the future. You can read about where we are planning to visit in this Issue. Please contact the members of the team and take the opportunity to meet them and talk to them if you can. I sincerely hope you and your family will be able to be part of some of the upcoming events. Of course, regardless of whether or not you can attend any of the events, we are always pleased to hear from you.

In this Issue we are also fortunate to be able to publish a contribution from Janet who is one of our members. I found her letter very hopeful and am sure that it will provide inspiration to others as it did to me. I want to thank Janet for taking the time to share her story with the HD community and would encourage others to send us contributions for future issues of this newsletter.

We want to remind you all that later in the year the World Congress on Huntington's Disease is being held here in Australia at the Melbourne Convention and Exhibition Centre from 11 – 14 September. This is a great opportunity for the members of the Australian HD community to be part of this bi-annual international event. To keep informed and up-dated on all developments as the Congress gets closer you can access the web site www.worldcongress-hd2011.org

In closing, we wish everyone who has been affected by the recent spate of natural disasters a speedy recovery back to 'normal' as soon as possible. As always, if you have any ideas for anything else that you might be interested in taking part in through the Association or you have ideas about how we could provide you with better services, as always please feel free to contact me either by phone at the Association or by email to ingrid@ahdansw.asn.au

Ingrid van Tongeren
Executive Officer

Huntington Happenings



HDBuzz is Up and Running

HDBuzz is a new internet portal that brings the latest news about Huntington's disease research to the global HD community. It is written in plain language, by HD scientists and provides easy-to-understand digests of scientific papers and conference reports. You can find this exciting new website at <http://hdbuzz.net/>

Brain Donation for Neuro-scientific Research

The Sydney Brain Bank, based at Neuroscience Research Australia, provides a research resource facility for the collection, characterisation, storage and distribution of human brain tissue for research purposes. Their primary focus is on various neurodegenerative conditions like Huntington's Disease, Alzheimer's Disease, Pick's Disease, Parkinson's Disease and other similar conditions.

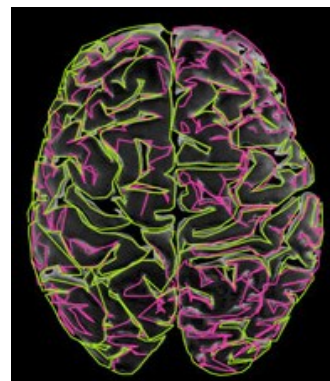
Researchers in Australia and overseas interested in investigating areas such as basic disease mechanisms, earlier and more accurate diagnosis, genetic contributions to diseases, better treatments, and ultimately, cure and prevention, can request this tissue for their research studies. To be successful in their goals, they rely on the generosity and altruism of people who donate their brains for research and also the scientists who request the tissue seeking to alleviate the suffering that neurodegenerative diseases cause.

Whilst they are interested in hearing from people with HD who want to participate, they are also interested in hearing from family members without HD who might want to participate as part of a comparison group for donors without neurodegenerative disease.

If you would like more information on this program, please call the Brain Donor Liaison Officer, Catherine O'Connor on **02) 9399 1107** and request an information package.



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Research Australia**
Discover. Conquer. Cure.



CAMP BREAKAWAY!

Our annual camp on the Central Coast is fast approaching. Interest is high! But we are still inviting members with HD who might be interested in joining us for 5 days at Camp Breakaway, San Remo on the Central Coast, Monday 14 March to Friday 18 March inclusive to apply to come to Camp.

The Camp is an excellent opportunity for our members 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, and be with friends.

The camp will also provide much needed respite to carers (spouses, siblings, parents, friends) who care for people with HD and provide them the opportunity to relax, share ideas, problems and concerns.

Please call us at the Association on 9874 9777 or email Lily on lily@ahdansw.asn.au for a camp application form.



Camp Volunteers Needed

If you might be interested in coming with us to the beautiful Central Coast and helping us with the Camp we would love to hear from you. You can come for the whole 5 days or for just part of the week. If you want more information about volunteering at the camp, please call Ramona at the office on 9874 9777 or email her on ramona@ahdansw.asn.au

Family Support Program

Another busy year has flown by at the Huntington's Association. Social Club has thrived - we have been sad to see a few members leaving as their health deteriorated, but it has been lovely to welcome new members into our friendly group. Last year saw us bond through our lunch sessions, activities, trips to the Powerhouse Museum, Fish Market, Darling Harbour, Birkenhead Point, the movies, picnics and BBQ's. We look forward to many more fun and exciting sessions this year.

I had a lovely visit with the Illawarra Support Group late last year with Jet and Jeanette from the Huntington Disease Service Outreach Program and met some very courageous carers as well as people with HD. It was great to see familiar faces and to meet others I had spoken with but not met before. The group is going strong with some members trying to meet up on their own at times not scheduled by the outreach program or association. Please know that we will support you in any way that we can.

Ramona Watts



Do you live in Wagga Wagga, Albury or in or around the NSW South and Sapphire Coast Areas ?

The Association is planning to make a trip to the Wagga Wagga, Albury and Sapphire and South Coast areas in the first half of this year and would like to hear from members and service providers who feel they need support and education in the care of their relative, friend or client with Huntington's Disease. Please contact the Association on 9874 9777 or email me at ramona@ahdansw.asn.au to let us know what your needs are and we will endeavour to do our utmost in supporting you.

Sydney Family & Friends Support Group



The family & friend's support group's annual Christmas get together was a great success again this year. November 24th saw our group at the West Ryde Hotel eating, drinking and making merry leading up to the Christmas period.

It was a time where carers of relatives with HD were able to get away for a few hours in a relaxed and friendly atmosphere, indulge in a few drinks and engage in banter with other carers facing similar situations.

Jet and I thoroughly enjoyed the camaraderie and friendship we experienced within this non-structured environment and look forward to catching up with our special carers again this year.

Ramona Watts

Do you like this Newsletter?

Tell us what you do and don't like about this newsletter so we can keep on improving it.

You can call us at the Association offices or email Ingrid at ingrid@ahdansw.asn.au

Huntington's Disease Service
and
Huntington's New South Wales

*invite you to
come along to our*

FAMILY & FRIENDS 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.*

2011 Sessions

16 March, Wednesday, 10.30am
20 April, Wednesday, 10.30am
18 May, Wednesday, 10.30am
15 June, Wednesday, 10.30am
13 July, Wednesday, 10.30am
24 August, Wednesday, 10.30am
28 September, Wednesday, 10.30am
19 October, Wednesday, 10.30am
16 November, Wednesday, 10.30am
Dec - Christmas Get-Together - TBC

Sessions will be held
at

**Huntington's New South Wales
Elsie Court Cottage
21 Chatham Road
West Ryde NSW 2114**

For RSVP and further information,
please contact:

Jet Aserios: (02) 9845 7528
Social Work Department
Westmead Hospital

Family Support in the Mid North Coast

John Conaghan from Hunter Genetics and I have had significant contact with various families on the mid north coast, and we are planning a visit, most likely in May, to families and health professionals in the area.

In 2009 hampers for Huntington's families were funded by the Association through the mid north coast fund, and in 2010 we provided Shopping vouchers for affected families. This was much appreciated by the recipients, and I want to thank Naomi van Dijk for her work on this project.

On our last visit to Port Macquarie, we had a wonderful meeting with 2 family members who gave us their insight into needs of Huntington's families in regional and remote communities (some of which would be needs of people in city areas also).



Some ideas discussed were: -

- ✦ Regular contact with Nursing Homes that have Huntington's residents, particularly with a view to helping families manage the situation when they have a person in care
- ✦ Fund raising possibilities to enable specific support to families
- ✦ Helping support people who feel isolated in Huntington's affected families
- ✦ Formation of 'loose self managing support groups' where there is need
- ✦ Support for young people (including young carers) in Huntington's families – linking them to other young people, and providing appropriate support
- ✦ Access to specialised health services (such as neurologists, psychiatrists, neurologists, nurses, youth workers) - such specialised services are often not readily available in regional, remote and rural areas
- ✦ Assist carers in relation to access to information
- ✦ Case management, and possible group support, for gene positive people – helping them to manage or cope with being gene positive
- ✦ Specific assistance to families managing the transition to nursing home care - choice, guilt, how carer life changes – helping the family through this whole process.

I am very keen to hear from families regarding their needs and ideas. So if you are on the mid north coast, I would love to hear from you. Email me - Mark Bevan at mark@ahdansw.asn.au or call me on 0410 629 850 – love to hear from you.

Mark Bevan

Developments on the North and Far North Coasts

I (Mark Bevan) am planning a trip to the far north coast of NSW in April this year. I will be linking up with the Queensland HD Association, who currently provide support to Huntington's families in the far north of NSW, and working with them on how the NSW Association can assist in providing support to Huntington's families in that area. On the way I will also be visiting other areas north of Kempsey and in and around Coffs Harbour.

The preliminary plan is to travel in the week beginning April 11, and visit as many people as possible, catch up with some Qld HD Association workers, meet with Huntington's families and related health professionals near the border, and those further south.

If you live in any of the areas I have mentioned, I would love to hear from you – you can email me at mark@ahdansw.asn.au or call me on 0410 629 850.

Central Coast Support Group

Central coast support group meetings have been going really well.

We have had a number of new people come along – good and bad side to that of course... great to make contact with new families and look at ways to support them, but that means that is another family affected by Huntington's.

There have been 2 great local fund raising efforts to provide funds to help support Huntington's families on the coast since the Mingara event in September. A Golf day at North Avoca raised \$1500 which was given as a gift to the Association for the central coast fund. A big thank you to Rick Parsons and friends for their generosity. Then the Central Coast Country Craft Fair on November 27 raised \$1950. Another big thank you, this time to Dorothy Phillips and her committee for selecting the Huntington's Association (Central Coast Fund) as their nominated charity for 2010.



The funds raised have led to the support group considering how best to utilise the funds to support families. It has been exciting to see the enthusiasm of the group as they consider what the priority areas of need for central coast families, and where we could potentially direct these funds. Watch this space for more news on this front.

On November 28, Letitia Quirk, Director of Care at Mathew John Aged Care Facility, very kindly opened her own home to the support group and families for an end of year gathering. This was very well attended, as people relaxed in the lovely grounds, enjoyed each others company and plenty of good food. Thanks again to Letitia for so actively supporting Huntington's families on the coast.

As we look toward further meetings this year – we had our first meeting back on January 17 – we are planning to have a range of visiting speakers, informative DVD's and great discussions over the year. Hopefully we will manage to arrange a number of social events as well..... we are a very sociable bunch.

Meetings are in 'The Education Room', Mathew John Aged Care Facility, 351 Terrigal Drive, Erina. The next meeting will be on **Monday March 21 at 7pm**. We have not yet locked in dates beyond that – but we will be meeting – so please contact me on 0410 629 850 if you need more information on meetings, dates or if you are on the coast and have other questions in relation to Huntington's. Remember – everyone is welcome at the meetings – love to see you there.

Mark Bevan



Day Respite Program (Social/Lunch Club)

We had a lovely and exciting Christmas celebration for our lunch club members on December 12th. Following years of tradition, Lily decorated the room with brightly coloured baubles, tinsel and beautifully wrapped presents under our tree – a welcome sight for our excited members. We had some morning tea and waited in anticipation for a group of wonderful children to come and sing some carols for us. Caleb, Abigail, Noah, Elisabeth, Isabelle and Keziah entertained our group with their cherubic renditions of traditional carols like "We Wish you a Merry Christmas" and "Silent Night" as well as funny versions of "Australian Bush Christmas" and "Jingle Bell Rock". They were met with loud clapping, laughter and many encores – thank you so much to the Bevan children for making our day so much fun. Lunch was a traditional fare of roast turkey, baked vegetables and a superb pavlova – there was enough food for seconds and thirds....We played some boisterous games, ate plenty of chocolate, and I think everyone went home tired, full and happy.



Whilst we had a break over the holidays the program is now back up and running for 2011. As usual we are meeting every fortnight, alternating between lunch at Elsie Court Cottage and an outdoor excursion (picnic at a park, movies, mini golf etc).

If you have Huntington's Disease or know of anyone who might benefit from joining us over lunch, sharing of experiences and a few laughs, please contact us at the Association on 9874 9777 or email me at ramona@ahdansw.asn.au

Ramona Watts

2011 Day Respite (Social/Lunch Club) Program Dates

Month	Dates
March	1 14-18 Holiday Camp 29
April	12 School holidays
May	10 24
June	07 21
July	12 26
August	09 23
September	13 School holidays
October	11 25
November	15 29
December	13 Xmas Party

Do you live in or near Broken Hill? Armidale? Tamworth? Dubbo? Orange?

To say I am excited is an understatement! I am planning visits to Broken Hill, Tamworth, Armidale, Dubbo, and Orange in the first half of this year.

I will be contacting genetic counsellors, health centres, various medical professionals, and Association members in preparation for the trips, with a view to meeting on my trips as many Huntington's families and associated health professionals as possible, so that we can understand how to more effectively support them in the future.

I would love to hear from you if you live and/or work in these areas. Just email me at mark@ahdansw.asn.au or call me on 0410 629 850.

Mark Bevan

Taking Care of Yourself

Whether it's a resolution you set for January 1, or a goal you start mid-year to reach, you can make the next year of your life a healthier one. For many of us, the path to good health, both physical and mental, is not an easy one, with plenty of roadblocks along the way. Family obligations, caring and work demands, and lack of time and energy are only a few culprits that can stop the best of health resolutions in their tracks. Here we have some advice for a healthier mind and body...

Exercise

As if we needed another reason to get fit! Exercise isn't only for maintaining your weight and staying healthy. It's great for relieving the stresses of life. Plus, the effects of a good workout can last for several hours. Exercise also helps your mind by releasing those "feel good chemicals" that improve your mood.

Eat a Healthy Diet

What and when you eat has a great affect on your mood and energy. Avoid refined and processed foods (like white breads, rice, and sugar). Try to incorporate more complex carbohydrates (whole wheat breads, brown rice, veggies, fruit) and get your daily 8 cups of water. These healthy foods provide your body (and mind) with nutrients, and stabilize your blood sugar and your energy levels.

Get Some Sun

Most people know that sunlight provides us with Vitamin D. But did you know that it also improves your mood? Lack of sunlight can cause many people to become depressed—without knowing why! Similar to exercise, sunlight exposure releases neurotransmitters in the brain that affect mood. Try to spend a little more time outdoors. Keep your shades up during the day to let more light in. Try changing the light bulbs in your house to "full spectrum" bulbs. These mimic natural light and actually have the same affects on your mind as the real thing.

Act on your Resolutions

Studies have shown a strong link between healthy behaviours and depression. People who exhibited healthy behaviours (like exercising, not smoking, etc.) had less sad and depressed days than those whose behaviours were less than healthy.

Treat Yourself

Having something to look forward to can keep anyone motivated. But if you plan something exciting, your mood improves when you're anticipating it and when the event actually comes. Plan something that's exciting to you.

Relax!

You're busy! But even if you enjoy being busy, everyone needs some time off. Don't be afraid to say "No" to extra opportunities. Try to spend a few minutes each day doing nothing! Relaxation can alleviate stress and leave you with a calm energy.

Get Social Support

Don't underestimate the power of friends, family, mentors, co-workers, and neighbours. Who can you turn to when you're down and need a pick-me-up? Keep a mental list of these special people and don't be afraid to ask for help or encouragement when you need it.



Do you have a Story to Share?

If you have a contribution that you wish to make to the Newsletter please send it to us at the Association offices (see details on the back page) or by email to Ingrid at ingrid@ahdansw.asn.au The next Issue will be published in late May 2011. The deadline for articles is 4 May 2011.

Janet's Story

I found out I had Huntington's in late 2000 having known for many years that because my father had the disease I had a 50/50 chance of having it. The year 2000 also marked my 40th birthday and although we didn't know when my father's deterioration had began it seemed plausible his probably began in his 40s.



So there I was, a single mother with a young daughter knowing that at some point this awful disease was going to make my mind and body do things I had no control over. I am also a recovering Alcoholic and have been sober since 1992 so for a while I felt that I was only just beginning to live my life and now this was going to be my future (it had to be wrong). It took probably six months for the information to sink in and then began a long battle with depression. I still fight that battle today and I am told this is part of the disease.

I have spent the past 11 years going up and down with depression and wanting to end it all on many many occasions. The one and only thing that kept me here was my daughter. How could a put her through that, and if she discovered that she also had Huntington's she may take the same path. Because of that I have had to find ways to have some joy in my life (especially when the early symptom crept in). Sometimes when my emotions are all over the place and I just want it to stop I look to the sky and say I can't do this anymore. But I have to, I can't show my daughter that option. Then I'm angry at the sky because I can't do it.

So for me I chose to follow a spiritual path (not religious) to find some peace and happiness. Buying material things didn't work and holidays are lovely, but only if I have that peace inside to be able to enjoy my holiday. I keep my body as physically fit as possible by swimming. I enjoyed walking for many years but I have arthritis and my knees became a problem. So far the swimming is good but I must have discipline and go regularly. I have a great GP that I see regularly, and I have also spent the majority of those years with some form of counselling. I am on anti-depressants and my GP monitors that regularly.

When I found out I had the disease I gave up on love and I concentrated on being a mother. But as my daughter got older I got lonelier and I knew I would need more friends to do things with. I have some friends now and the past 3 1/2 years I have been in relationship with a man who shares similar outlooks and he got me started on meditation and we now regularly do that. He moved in with my daughter and I over a year ago, and it can be really hard but the we try our best to communicate and be as honest as possible. It is also one of the best experiences I have had in my life because I love him and he loves me. We even went on a 2 week driving holiday to South Australia last year and I had a great time stopping at different places along the way. This man knew about the Huntingtons from the beginning but has chosen to walk this path with me one day at time.



My beautiful daughter is almost 20, and at uni. We have 2 cats and a dog and I must say when I am feeling in the pits there is absolutely nothing better than pet therapy, you just got to smile at those big eyes looking up at you. There are very dark days but if anyone out there is struggling with their diagnosis I am here to assure you there can also be beauty and joy.

Janet

Speaking and Listening!!

Obviously, nothing is more important in your relationship with the person for whom you're caring than communicating with one another. This becomes more and more important as it becomes more and more difficult. The movement disorder affects speech in several ways. In the mid-stages of HD, people lose precision in making sounds, control of the volume of the sounds they make, and coordination of the speech and breathing mechanisms. This creates speech that is varied in volume, interrupted by grunting or breathing sounds, and hard to understand. In the most advanced stages, people express their range of needs and emotions with a few intelligible words or sounds.



Just as the movement disorder affects speech, the cognitive disorder affects the content of what is said. The ability to form ideas, organize thoughts, and present them in an orderly sequence is compromised in HD. Some people have difficulty starting a conversation, staying on the topic, or switching from one topic to another. Some get stuck on one topic and have difficulty getting off it.

As clear speech becomes more difficult, it takes great effort for people with HD to carry on a conversation. They will have a tendency to rely on a very small vocabulary of more easily understood words. This allows you to take on a more active role in a conversation, picking up on those key words, anticipating the idea, and expanding on it on their behalf. At the point where it is extremely difficult to be understood, some people simply stop talking. Your familiarity with a person's likes, dislikes, career, interests, hobbies, and relatives will keep the conversation going or allow you to become their "interpreter" with others.

It can be humiliating and frustrating for the person with HD, and embarrassing for you, when you have difficulty understanding their words. One way to show him respect is to put the burden of understanding firmly on you. Ask him for clarification. Ask their permission: "Do you mind if I repeat your words to you from time to time so you will know how I'm doing?"

Communication boards are commonly introduced to people who are having difficulty being understood. As well-intended as they may be, boards are not often adopted by people with HD as an alternative form of communication. Speaking, as impaired as it is, is easier than learning to use the unfamiliar board. As with other adaptive devices such as helmets and wheelchairs, introducing the communication board early, before it is actually needed, gives the user more time to learn how to use it, practice with it, grow fluent in its use, and possibly adopt it.



Some families find it helpful to assemble a book or picture album full of photographs that represent their interests, hobbies, family, career, and preferences. Since non-family caregivers may first meet the person when they have difficulty expressing themselves or recalling events from the past, the album serves two purposes. First, the album is a communication aid which allows them or you to point to pictures when you don't understand each other. Second, it serves as a treasury of interests, children, grandchildren, relatives, hobbies, achievements, pets, home or apartment, and favourite sports teams so you can better know who this person is and can communicate with them for years to come.

Please remember that people with HD can comprehend our speech and understand all that's going on around them to a far greater degree than most people may at first suspect. How effectively they communicate through spoken words is not an accurate predictor of how well they understand what you say.

Family members and caregivers agree that people, even in the most advanced stages of HD, somehow manage to communicate with their caregivers very effectively through facial expressions, eye gazes, and other subtle movements that may only be understood by those closest to them. Look and listen carefully!

Caregiver Communication Tips

Have the person with HD: -

- Slow down, especially if their speech has a "racing" quality to it.
- Repeat/rephrase.
- Say the main word.
- Spell the word.
- Write the word, even if they can write only a few letters.
- Show you.

You, the caregiver should: -

- Try to rephrase the main idea.
- Use short sentences.
- Ask for feedback.
- Allow plenty of time.
- Wait... for up to a few minutes for a reply.
- Try not to repeat or rephrase a question while you're waiting for a response.
- Use touch to help keep them focused on the conversation.
- Ask for help from others when needed.
- Never pretend to understand!
- Consider using a simple communication board.

The family can: -

- Make a scrapbook or memory book.
- Tell you about facial expressions or phrases that they understand.
- Continue to call or write even if they cannot respond clearly.

From: *A Caregiver's Handbook for Advanced-Stage Huntington Disease*

Author and Editor: Jim Pollard, MA (pictured right) published by the Huntington Society of Canada, 1999. Reprinted, 2000.

This publication is available from the Association. If you are interested in a copy please contact us on 02) 9874 9777 or email Lily at lily@ahdansw.asn.au



HD Research Update

Novel Compounds protect Brain Cells from Neurodegeneration

by Marsha L. Miller, Ph.D., January 27, 2011

Researchers from Southern Methodist University and the University of Texas at Dallas have collaborated to develop new benzoxazine compounds to prevent neurodegeneration. Similar indolone compounds had been found to inhibit cell death but were toxic at higher concentrations. The benzoxazine compounds were synthesized by altering the ring core structure, preventing toxicity even at high doses. Benzoxazines are the reaction products of an amine, a phenol and formaldehyde.

The researchers tested one of these compounds, HSB-13, in a toxin model of HD where mice are administered 3-nitropropionic acid (3-NP), a neurotoxin which causes striatal cell death. 3-NP causes an acute injury rather than the slow degeneration seen in HD; this model was widely used prior to the development of genetic models. Mice pre-treated with HSB-13 had fewer striatal lesions and improved motor performance. In addition, mouse neuroblastoma cells were treated with homocysteic acid to induce oxidative stress-mediated apoptosis; the cells that were pre-treated with HSB-13 survived. In another experiment, HSB-13 increased survival time in an Alzheimer's fruitfly model.



Dr. Ed Biehl (centre) with Drs. Sukanta Kamila (left) & Haribabu Ankati

"Our compounds protect against neurodegeneration in mice," said synthetic organic chemist Edward R. Biehl, the Department of Chemistry professor who led development of the compounds at SMU. "Given successful development of the compounds into drug therapies, they would serve as an effective treatment for patients with degenerative brain diseases."

"Additional research needs to be done, but these compounds have the potential for stopping or slowing the relentless loss of brain cells in diseases such as Alzheimer's and Parkinson's," said collaborator Santosh D'Mello, professor of molecular and cell biology who led the team at UT Dallas. "The protective effect that they display in tissue culture and animal models of neurodegenerative disease provides strong evidence of their promise as drugs to treat neurodegenerative disorders."

Dallas-based startup EncephRx, Inc. was granted the worldwide license to the jointly owned compounds. A biotechnology and therapeutics company, EncephRx will develop drug therapies based on the new class of compounds as a pharmaceutical for preventing nerve-cell damage, delaying onset of degenerative nerve disease and improving symptoms.

Aaron Heifetz, president and chief executive officer of EncephRx, Inc., said, "We believe this library of novel neuroprotective compounds will prove an important step in the effort to improve the health for patients with neurodegenerative diseases, such as Huntington's disease, Alzheimer's disease and Parkinson's disease." EncephRx initially will focus its development and testing efforts toward Huntington's disease and potentially will have medications ready for human trials in two years, said Heifetz. Chris Jeffers, managing partner of FirstStage Bioventures, the parent company of EncephRx, added, "FirstStage is very excited about this technology and looks forward to helping EncephRx quickly move these compounds into the clinic."

References:

- Southern Methodist University press release, December 7, 2010.
- Texas University at Dallas press release, December 10, 2010.
- Lulu Wang, Haribabu Ankati, Shashidhar Kumar Akubathini, Michael Balderamos, Chelsey A. Storey, Anish V. Patel, Valerie Price, Doris Kretschmar, Edward R. Biehl, Santosh R. D'Mello. "Identification of novel 1,4-benzoxazine compounds that are protective in tissue culture and in vivo models of neurodegeneration." *Journal of Neuroscience Research* Volume 88, Issue 9, pages 1970-1984, July 2010

Capase Inhibitors with Therapeutic Potential Identified

by Marsha L. Miller, Ph.D., January 21, 2011

The HD community has been hoping to hear news of a safe and effective caspase six inhibitor ever since Dr. Michael Hayden reported that caspase six resistant mice with the HD gene do not develop Huntington's Disease. A research team led by Buck Institute faculty member Lisa Ellerby, Ph.D. and Yale University faculty member Jonathan Ellman, Ph.D. has developed three novel pan-caspase inhibitors that block proteolysis (fragmentation) of the HD protein at caspase 3 and caspase 6 sites.

The word caspase comes from cysteine-aspartic-acid-proteases. Caspases are enzymes which are used in apoptosis, programmed cell death. There are various caspases that initiate the process, that cleave proteins and that actually 'execute' the cell. Apoptosis is a necessary process in development and also in destroying tumours. Unfortunately, apoptosis is also implicated in neurodegenerative disorders. Apoptosis is triggered by cellular stress, especially mitochondrial stress, and this is known to occur with Huntington's and the various other neurodegenerative disorders. Caspase six cleaves the HD protein into toxic fragments which enter the nucleus of neurons and interfere with gene transcription.

In the Hayden study, HD mice were also engineered to be resistant to caspase six cleavage and they did not experience neurodegeneration. This indirect strategy was used rather than testing them with a caspase six inhibitor since the existing ones were based on peptides and did not cross the blood brain barrier. The Ellerby-Ellman research team used a substrate based fragment approach called substrate activity screening (SAS) to identify weak-binding nonpeptide fragments that they then optimized to increase potency.

These molecules show particular promise. They cross the blood-brain barrier and act selectively to block the processes involved in HD.

Dr. Ellerby

Dr. Ellerby said that the inhibitors are based on properties of a drug which had entered Phase I clinical trials for the treatment of human liver preservation injury. "These molecules show particular promise. They cross the blood-brain barrier and act selectively to block the processes involved in HD." Dr. Ellerby said the caspase inhibitors both suppressed the proteolysis of Htt and rescued HD neurons that have begun to undergo cell death.

The study looked at striatal and cortical neurons from an HD rat model. Dr. Ellerby is already testing these compounds in animal models of Huntington's Disease. "We believe this is going to help us move the field forward because now we can test these compounds in live animals," said Dr. Ellerby. "Up until this point we have not identified a caspase inhibitor that has acted selectively against the toxic effects of the Htt mutation."

If Dr. Ellerby finds one of the inhibitors to be safe and effective in an HD mouse model, this would be proof of principle that the therapeutic strategy is worth pursuing for people with the HD gene.

References:

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Huntington's New South Wales

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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 10/11

President: Robyn Kapp OAM
Vice President: Anne Low
Treasurer: Richard Bobbitt
Secretary: Viki Moraitis
Don Ayres
Keith Dingeldei
Jim Finn
John Conaghan
Karen Bevan

Association and Other Useful Contacts

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Toni Ling Zhang
Administration Officer

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