



# Oz Buzz Video: Day 1

Day 1 of Oz Buzz: the nightly news, interviews and features presentation from the HD World Congress 2011



By Professor Ed Wild

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Edited by Dr Jeff Carroll

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**C**harles Sabine, Jeff Carroll and Ed Wild present Oz Buzz 1: a summary of the daily news headlines, in-depth interviews with top HD researchers, and entertaining features from the Huntington's disease World Congress 2011 in Melbourne. This is a draft copy that will be updated with better quality video soon.

**CHARLES:** G'Day! And for those, like me, visiting this island for the first time, welcome to the lucky country. And a very warm welcome also to the first, the inaugural, the debut Oz Buzz. Make no bones about it, tonight you witness history. Oz Buzz is born. Now there are two basic truths that we English, Poms, know about Australia. One is that we invent sports and they then regularly beat us at them. And two is that, despite the pain inflicted therein, we know that this is the most hospitable country in the world so thank you very much for having us here. Now those of you who were in Vancouver two years ago may recognise the people you'll see tonight responsible for the evening news there are indeed presenting tonight. But this is going to be very different, we do not stand still no flies on us. What we will do tonight is to reflect the new chapter we've reached in the evolution of the HD community. Now, unquestionably, a global community. Most of you will be familiar with the phenomenon that is HDBuzz and if you're not you certainly should be. It has revolutionized patient access to scientific news that is unprecedented in any field of medicine and I am proud to say that I hold the title of the consulting publisher of HDBuzz. Do you know how much work I put into that publication? Zip, Nada, Sweet Fanny Adams. If you wrote down all the hours that I put into my role as consulting publisher of HDBuzz it would be shorter than Muammar Gaddafi's social diary for the next month but, rest assured, when HDBuzz is rightly acclaimed with some sort of award for the new boundaries that it has pushed for public access to science I

will be right up on the stage pushing the others aside to hold the award No possums, I jest  
The astonishing work that is HDBuzz is all down to my two co-presenters tonight, who I  
would like to introduce to you now. The hosts with the most, the toast of researchers  
everywhere, who can boast that they are the best at telling it how it is First, from Boston,  
USA, but moving again, he's always moving, they call him the buzz because he's stinging  
like a bee: Dr Jeff Carroll And second, from London, England an intellectual beast, wild at  
heart and never afraid to unsheathe his sword Dr Ed Wild Now gentlemen, I am on  
tenterhooks, wound up like a kangaroo as hot as a dingo on heat tell me, tell me, tell me just  
how is Oz Buzz going to work?

**ED:** Well Charles I'd like to announce that like Batman and Transformers combined we've  
rebooted our franchise We're sticking with our formula of bringing the hottest science to the  
whole audience here in language that hopefully everyone can understand but this time  
around rather than try to digest every word that's been said all day we're starting with our  
news headlines feature stories that we think are the most exciting and relevant to the non-  
scientists in the audience

**CHARLES:** And Jeff I understand that we're going to see some researchers in a different  
light to usual?

**JEFF:** Well you might have noticed from the distinctive furniture over here that we've set up  
a comfortable space for scientists to let their hair down and talk science. We're calling it  
Chat-landia after rejecting Chat-ghanistan and Chat-mandu Now each night Ed and I will be  
interviewing three scientists hoping to probe behind their work, learn a little bit more about  
their work So this is for the audience in the room but we're uploading it so all of the world  
will be watching. The HD community have been alerted about this and can find out about  
what's happening right away, so we can reach out to the community in real time We've also  
requested and received questions from the HD community for these scientists

**CHARLES:** And so let's get straight on with the news headlines And they start with you, Jeff,  
where were you today?

**JEFF:** We started with the plenary session and heard from Peter Harper I'm going to try this  
clicker - nothing could possibly go wrong [laughter]

**ED:** I think you'll have to imagine Peter Harper until we get the powerpoint up

**JEFF:** Peter did a fantastic job of highlighting some of the features of this community, you  
guys. He reminded us that in many ways HD has been, and continues to be, a pioneering  
effort for everyone suffering from other diseases, the things we learn and do are helping  
other people and I think we hope we can be pioneers in developing treatment, as well as  
coming together as a community so Peter's historical overview very nicely set the stage  
for... Professor Sarah Tabrizi, she highlighted the large number of potential therapies and  
really drove home the fact that in the next two years we really should hope to see some  
novel treatments with really exciting drugs, so they're exciting times - or as Sarah says - HD

- yes we can Elizabeth Aylward, from the University of Washington, did a really nice job describing the imaging results from the brain study in PREDICT and I think this did a remarkable job in telling HD patients that what they're doing in turning up for observational trials is really starting to help because she was able to show that the end-points that have been derived from this analysis, looking at HD patients, is providing things that we might be able to use to successfully run drug trials The afternoon session talked about animal models We're all pretty used to hearing about mouse models but what does that actually mean? That means scientists have modified the DNA of a mouse to have a mutant copy of the huntingtin gene somewhat like an HD patient Xiao-Jiang Li from Emory University brought up the fact that there are some interesting problems with modeling HD in mice There are things that scientists who work on mice, who shall remain nameless and are not on this stage, don't like to talk about like the fact that the mice don't lose a lot of neurons, the critical brain cells that die in Huntington's which causes symptoms, in the brain of a late stage HD patient there's millions of neurons that have died and gone away and that doesn't seem to happen in mice, and we don't know why, the mice get sick and have symptoms but they're not losing neurons and that's kind of curious Also human HD patients have hyperkinetic movement disorders, chorea we call it, mice don't get that and we don't know why These guys at Emory University are developing other models in other organisms like pigs and monkeys this is an artist's rendition, not the actual animal These models, the monkeys and the pigs, actually lose neurons like human HD patients but have other features that don't look like HD patients so the takeaway message was we need to use all these models and take away data from all of them

**CHARLES:** Thank you, and Ed where were you today?

**ED:** Well I hung around in the shared sessions this morning and then I was in the young people's sessions this afternoon The thing that really struck me this morning was Jim Gusella of Harvard Medical School who was talking about his work on genetic modifiers in Huntington's disease We know that HD is caused by a single mutation, in a single gene and we've known that since 1993 but we also know that there are big differences between individual patients that we don't understand and can't be explained by the mutation we know about things like what age someone is when the symptoms begin and some of those are certainly due to other genetic differences that we haven't found yet and that's what we call a genetic modifier these are important because any genetic modifier could become a really good drug target over the years several genetic modifiers have been reported in the literature but what Jim has done is quite remarkable he's re-analysed all of the old data, including his own data, and carefully looked at the statistics behind it amazingly he found that several of the genes we thought contributed to Huntington's disease and altered how the disease behaves turn out not to be exerting the effects we thought for example previously we thought that the smaller of a person's two CAG repeat counts affected, in a small way, the course of their HD but re-analysing the data in a systematic way found no evidence to support that now that might sound like a set back because we seem to have lost genetic modifiers but actually major efforts are under-way around the world to look at

the whole genome in a really systematic way for these genetic modifiers and Jim's robust analysis techniques will put us on the right footing to make sure we get from those studies, solid, important and true genetic modifiers and then just before lunch we heard from Colin Masters who's a local boy, made good and an Alzheimer's disease guy and he's trying to figure out what lessons that have been learnt in the huge field of Alzheimer's research might help us in the HD field and Prana Biotech, a local company, has been developing drugs that can alter how the proteins in the brain interact with metals in the brain, like copper, so atoms of copper can stick to some of the proteins and that can have an effect on how the proteins behave and Prana Biotech has a drug called PBT2 which affects the way the copper and the proteins interact which has been tested in Alzheimer's and is now lined up for a new trial in HD beginning as soon as the end of this year, here in Australia and the USA And then after lunch in the youth and young people's session where I felt very much at home Mike Orth from Ulm presented some fascinating results from a Euro-HD network study of young people's experiences in HD and the major message that I took from that is that young people are desperate to find out more and get more support from professionals and also from each other especially online instead of trooping into hospitals to be seen in HD clinics when they may feel fine And then as if by magic Matt Ellison appeared to announce the forthcoming launch of the HD youth organisation, HDYO So this is a site where kids and teenagers, young adults and parents and guardians will all be able to use the site to get reliable information, for young people, by young people, backed up by a huge international panel of experts it'll launch in January 2012 and it'll be translated into several languages and you can visit the site now at HDYO.org and sign up for email notification when it launches and speaking as a young person, which you can tell I am by my manner of dress and speech and hairline I am pleased to give HDYO my thumbs up

**CHARLES:** As the doctors make their way over to the very sexy chat-landia I will explain how our guests and I can win prizes Ed and Jeff have taken over with them to chat-landia the bell, which has travelled all around the world to join us here veterans of our past presentations will know that if anything is said that is too technical for non-researchers to understand they will hear this sound [bell] but if they get through their interview without a bell then they are awarded the no-bell prize and a rare almost priceless HDBuzz post-it note so now to introduce our first guest: Frank Bennett is the Senior Vice President of Research at Isis Pharmaceuticals, California He works with several academic collaborators on something called gene silencing They are therapies that many people talk of as one of the great hopes for us HD families Frank is related to two US presidents on his mother's side But more interestingly, by far, on his father's side he's related to a famous outlaw gang called the Dalton brothers who specialized in bank and train robberies he says you can reach you own conclusion on what happened when those gene pools mixed ladies and gentlemen: Frank Bennett

**JEFF:** Hi Frank, you look good on the couch Today you and Don Cleveland both talked about gene silencing I don't think anyone needs to be convinced that getting rid of the protein that causes HD would be a good therapeutic strategy There's some confusion, even in the

scientists, and among HD family members who've heard about different approaches to this therapeutic idea RNA interference, or RNAi, is something people hear about and also anti-sense or anti-sense oligonucleotides could you just briefly explain what the difference are in those approaches?

**FRANK:** First of all! They're all variations on a theme, I like to think of them as a genus or species, if you think about animal phylum and how you organize animals is that anti-sense technology is where you're designing drugs to bind to RNA and when they bind to RNA they modulate the function and what dictates the difference between the different anti-sense mechanisms you mentioned is what happens after the drug binds to the RNA and they recruit different enzymes that are involved in the degradation of the RNA

**JEFF:** So they're two different pathways to achieve the same effect?

**FRANK:** That's correct

**JEFF:** And your company, Isis Pharmaceuticals, you work with which of those?

**FRANK:** We actually work with both but the one that's been most advanced is the anti-sense approach where we're targeting degradation through an enzyme that's present in cells called RNase

**H JEFF:** So in addition to different chemistries there're also different targets so we've heard about so-called specific and non-specific approaches some people say allele specific allele non-specific could you briefly say what the difference is between those two target strategies?

**FRANK:** Sure, I think the audience is well aware that all humans, or all species, have two copies of a gene and in Huntington's disease one of the copies of the Huntington gene has a change in the base composition, an expansion of the CAG repeat whereas other copies, a normal allele, present in the most...

**JEFF:** An allele just means one copy of a gene

**FRANK:** A gene, that's correct

**JEFF:** That's ok, we'll let him off

**FRANK:** So there are two approaches we could take, one is to inhibit both copies of the gene so that we target a non-allele specific condition of huntingtin that means both the normal and the CAG expanded copy are affected or there's another approach we could take that's a little bit more complicated that would allow us to only inhibit the copy of the gene that has the CAG expansion

**JEFF:** And so at Isis you guys are working on both of these approaches in parallel?

**FRANK:** That's correct, and that one's a bit further ahead than the non-allele specific approach as far as the development of that approach, it's a little bit more advanced

**JEFF:** I know that nobody wants to get ahead of themselves in timelines and this is a complicated business and it's difficult to put numbers on something that's not engineering, it's science but do you have a qualitative sense for family members, is it twenty years away or happening next week?

**FRANK:** It's years, but it's a couple of years, I think it's something that we're very close to having a compound that's ready to start clinical trials within a couple of years from now and the key test to get there is that we have to do a series of animal studies to evaluate the safety of the drug and that generally takes about nine months or so so we're hoping to start those animal studies next year and then a couple of years later start human trials

**JEFF:** What about that? Once you get to humans does Isis have experience with giving these types of drugs to people do you know how to deliver them? and how people will react when you expose them to them?

**FRANK:** Yes, so the focus of our company really is anti-sense technology, RNA targeting therapeutics but we're targeting a variety of different diseases we have two programs in neuroscience that are using a similar delivery method that we are using for the Huntington's drug and so we'll learn from those programs and it'll inform the Huntington's program the goal would be that it'll accelerate the human testing because we'll learn from our mistakes in the other program and so one is in clinical trials today: ALS, another neurological disease and one should start later this year: a childhood motor neuron disease called Spinal Muscular Atrophy

**CHARLES:** Thanks very much, so let's move on now, while Frank moves across the sofa to fit in a second guest Tony Hannan is the Associate Professor of the Florey Neuroscience Institutes the Melbourne brain center which means he is yet another Melbourne boy who seems to have done well for himself he works on something that I for one believe dispels the myth that there is nothing that gene-positive people can do about their fate: how environmental and lifestyle factors can influence Huntington's disease when he played rugby at college Tony says he was given the names kamikaze and the smiling assassin because of the nature of his tackle, sorry I mean tackling sorry typing problem there those world congress attendees who are not familiar with rugby by the way you can watch of course every day the current world cup being held in New Zealand Tony of course is supporting the second best team, Australia Tony Hannan

**ED:** Thank you very much for being a sport and hopping up on the gold sofa your talk was about environmental modifiers and if I'm not mistaken an environmental modifier is when an Australian cow farts and the methane gas gets into the atmosphere and the polar bears melt, am I right?

**TONY:** That's a kind of global version of an environmental modifier

**ED:** How would you define an environmental modifier?

**TONY:** So essentially all of us, include those of us from Huntington's families, are the product of nature and nurture a complex inter-relationship: genes and environment from conception onwards we're influenced in utero and then postnatally by more environmental factors an environmental modifier, for a disease, is anything that changes the disease relative to the gene effect so you've got genes and environment and any modifier is an environmental factors that can either delay onset or accelerate onset

**ED:** A lot of your work in this area has arisen from studies in mice dealing with something called environmental enrichment, what's meant by that?

**TONY:** So the concept of environmental enrichment is relative to standard conditions for mice standard conditions are groups of mice that have unlimited food and water, soft bedding, but it's generally a bit boring so environmental enrichment, relative to that - which varies between laboratories it's far more novelty, complexity, toys it's other objects like running wheels so it enhances sensory stimulation cognitive stimulation movements, and therefore enhances complex mental and physical activity

**ED:** So it's mice that are given a much more interesting environment?

**TONY:** Yes

**ED:** And what happens to HD mice when they're put in an environment like that?

**TONY:** What we found about a dozen years ago was that the ones that had environmental enrichment, these Huntington's mice that had a fragment of the human Huntington gene with the CAG repeat in their genome the environmental enrichment caused a delay in onset of the disease relative to those that had the standard, boring environment

**ED:** A pretty significant delay?

**TONY:** YES ED And since then you have been trying to figure out what it is that causes the mice to look better and suffer less?

**TONY:** Yes

**ED:** So there's a question here from Victor Orozco in Colombia who's read your abstract and wants to know how is it that - given the facts we know about environmental enrichment what is the mechanism for those altering the progression of Huntington's disease?

**TONY:** So that's a big question and an excellent question there are two sides to this, and it's a whole area and this applies not just to Huntington's families but all of us so all of us will eventually, if we survive heart disease, diabetes and other diseases, if we live long enough pretty much everyone in this room will be exposed to a late life brain disease Alzheimer's or dementia, that kind of senile dementia so there's an element whereby this environmental enrichment changes a healthy mouse that doesn't have the Huntington's mutation and that's part of what we've been studying, and so have other groups around the world so if you take a healthy mouse and you expose it to this complex environment you can see the



birth of more neurons in the adult brain from adult stem cells in the brain you can see increased connections between different groups of brain cells and that's part of the effect, so you see all these in a healthy mouse there are other subsets of changes caused by the Huntington's disease mutation that are going wrong in Huntington's, a subset of those that seem to be reversed by this stimulation we're not sure how except it does get into the DNA and change this subset of genes that produce proteins but it may change some of the other processes that are going wrong in the cells in the brains of Huntington's mice and patients

**ED:** So there's some stuff about an enriched environment that's generally good for brains and some effects that seem to be specifically good for counteracting the effects of the Huntington's mutation?

**TONY:** Absolutely, and so this concept of brain reserve applies to Huntington's but also applies to the general population and applies to other diseases like Alzheimer's and Parkinson's

**JEFF:** So can I have a running wheel then?

**ED:** Back to the gym

**ED:** To put you on the spot slightly, a lot of people have been asking this online is there anything you can advise HD families, those who are at risk of the disease or have the mutation that comes from your scientific work in terms of things they can do in their everyday lives that might help to delay the onset?

**TONY:** A study that followed up the mouse study led by Nancy Wexler showed that in the Venezuelan population that environmental factors are there, we know they're there and a group led by Martin Delatycki in Melbourne also did a study that showed that being more active in patients, appears to delay onset but I can't say exactly what kind of activities so just like other diseases like Alzheimer's and there is some epidemiology, some clinical studies there it really comes down to those studies that say anything to do with the mental activity that is stimulating, that involves engaging, and this includes social interaction so that's the complex mental activity side and the other side, physical activity, it could be lots of things the clinical studies need to be done no one has the magic formula but the reality is, being more mentally and physically active, there's no downside if you're more physically active you reduce your risk of heart disease, certain cancers, diabetes if it's good for the body, it's good for the brain what it requires is people with the mental activity engaging in something for weeks and years, this is a chronic disease so it needs to be something people enjoy that they would do for years and decades it's very personal and until the clinical studies are done we won't know if one type of activity is better than another and like anything people need to be enjoying it or they won't keep doing it

**ED:** To answer Michaela in Stuttgart's question those studies, which you mentioned those are long studies comparing different types of people and how much exercise they do and so on those are being set up?

**TONY:** Yes

**ED:** We have to leave it there. Tony thank you very much.

**CHARLES:** Thank you Tony I see you are getting the hang of it with the sofa, good. Professor Leslie Thompson at the University of California Irvine leads a team of researchers looking at how cells modify their own DNA and the mutant huntingtin protein but there are other things less well known about the professor than that she wouldn't be here tonight if she hadn't lived in Mexico as a child and learned there Spanish which enabled her to join Nancy Wexler in Venezuela she was also a concert flautist and a triathlon competitor and one of my sources tells me to ask her how she got sperm samples in Venezuela and if she still has a subscription to Playboy Professor Leslie Thompson

**JEFF:** I'm glad that we could keep this so professional you guys look really cosy, the couch was smaller than we would like Leslie, you're interested in chemical changes to the Huntingtin protein and I think hopefully this audience, at least from today's talks that the Huntington gene makes the Huntingtin protein and it's actually the protein that's the problem but you've gone to the next step and are studying little chemical tags that are added to the protein and so just in general why would cells do that, why would they need to tag a protein they've already made?

**LESLIE:** First of all the proteins that are tagged have all sorts of different functions in the cell but they're all made the same way, they're encoded in the DNA, transcribed into RNA and made into proteins and then they have to do something, they have to go into mitochondria [bell] Oh!

**ED:** You can pull it back if you explain what a mitochondrion is

**LESLIE:** So the, energy making... heart of the cell

**JEFF:** The cell has made a protein it needs to get to the energy 'house'

**LESLIE:** Right so it will get a little tag on there, a little extra piece of protein on it and it will tell that protein to get into that part of the cell or it will cause that protein to interact with, to associate with, another protein in the cell

**ED:** To make friends with?

**LESLIE:** It will make friends with another protein in the cell! So these tags will tell the protein what to do, where to go what it's going to interact with etc.

**JEFF:** You guys have evidence that this process, not just making the protein but of tagging it and making it have different functions, that's messed up in Huntington's disease?

**LESLIE:** Yes, so in some ways its normal tags will be altered in other ways it will have new tags that will take on a new activity that it wouldn't normally have

**JEFF:** We got through that with only one bell, pretty impressive So not only are these tags on the protein but as you talked about today they're actually on the DNA the genetic information that encodes these proteins is tagged is it the same kind of tags? are they different tags?

**LESLIE:** They are similar kinds of tags: what a tag that would tell the Huntingtin protein that it needs to be cleared out of the cell, thrown in the trash will tell the DNA that it should make a particular protein So are cells all have the same DNA in them but because of a particular tag a skin cell will make certain proteins that make it a skin cell, a heart cell will make a heart cell because of these kinds of tags that say which genes will be turned off or turned on and will make specific proteins

**JEFF:** This is all very interesting but does it have a point from a therapeutic point of view? I don't mean does it have a point! But will it help HD patients?

**LESLIE:** Yes, it does. Because the enzymes, the proteins in the cell, that will put on or take off the tags those become therapeutic targets, so those have functions you can inhibit with a drug or activate with a drug we would like to, in some cases, enhance that ability or inhibit that ability

**JEFF:** So if you can fix this tagging and untagging...

**LESLIE:** Or at least restore it to the right balance, it's not necessarily an all or nothing thing you want to restore the balance

**JEFF:** There's drugs out there that do these kinds of things?

**LESLIE:** There are some that already exist in particular for cancer and other things that need to be evolved

**JEFF:** So we want to go to a video question from Ken Serbin who wants to talk to you about another aspect of your work we haven't a chance to talk about yet

**KEN:** Greetings from San Diego to all of the participants in the World Congress of Huntington's disease in Melbourne this is Gene Veritas the gene positive blogger on Huntington's disease aka Ken Serbin I would like to know how much more quickly your work in the field is moving along because of stem cell research and what are the most important discoveries you've come across with this stem cell research Thank you Dr Thompson, thank you to everyone for your efforts and a successful and wonderful congress to everybody

**ED:** You have ten seconds

**LESLIE:** Ok... so we're using stem cells as a large group, a collaborative effort to model disease, taking skin samples from patients with HD and using those in a dish, in a human cell, to model the disease you re-program them back to a very early state so they can become a heart cell, a brain cell, whatever the thing that I think is so exciting about this is

we can make them into the neurons that die in HD and study it in that way these same changes that we're talking about, gene death all these other things are showing up as different in this context

**JEFF:** Does this speak to what Xiao-Jiang Li was saying today about cells from a mouse are different from cells from humans? You could grow human neurons? Yes, you could grow human neurons in a dish and Virginia Mattis has a poster here that describes that work

**JEFF:** Fantastic, I think that was 12 seconds so we'd better turn back to Charles, thanks very much

**CHARLES:** Thank you to everyone over there in Chat-landia and thanks to our special guests Ed and Jeff we will see more of you tomorrow night

**CHARLES:** And now to our local reporter. And my is she local. She is more Melbourne than a tube of Foster's. She is so Melbourne that she's named after the town. Ladies and Gentlemen: Mel Brinsmead [applause and wobble board] So Mel, first of all what do we need to know about Melbourne.

**MEL:** First of all Charles, everyone here needs to know, the Economist has recently rated Melbourne as the world's most livable city. And by the way Charles it's 'Mel-ben' I'm having clicker issues too

**CHARLES:** Melbern

**MEL:** Give it another go, Charles

**CHARLES:** Melben

**MEL:** Practically local

**CHARLES:** What has Melbourne given to the world?

**MEL:** Well, we invented, no not him, he is from Melbourne but we didn't invent him. There we go. The black box and the bionic ear, both from Melbourne. Geoffrey Rush, he wasn't born here but he lives here and he won an Oscar so we're going to claim him. Now we've already heard this young lady this evening, Ed's favorite, Kylie. Another bloke from Melbourne, we all love him, across the world, dear friend Rupert.

**CHARLES:** And on that note, with dear Mr Murdoch there, what do we we need to know about Australian culture?

**MEL:** Well despite how we're portrayed on telly we're not all like Mick Dundee or Steve Irwin We're pretty cas in Australia, pretty cas, none of this "pleased to meet you" business, it's "G'Day, how's it going?". This can be used at any time, when you first meet someone or when you're just passing by. Most importantly for everyone out there you need to know we drink pots, not pints or schooners. So when you go to the pub tonight order a pot of Foster's, it's the Aussie thing to do, when in Rome...

**CHARLES:** And now for our audience who might not be completely fluent in Australian could you give us a few key phrases that might be helpful?

**MEL:** I can: "Could all the blood nuts out there built like a brick shithouses, wearing thongs, please stand up now?" No one stood up, does that need some explaining. Well, our prime minister, perhaps the most famous blood nut. She's a ginger, a ranga, a red-head, get the picture? This is a brick shithouse, a prime specimen of a man, well toned, preferably someone you wouldn't want to meet in a dark alley?

**CHARLES:** Or you might, you never know [laughter]

**MEL:** And, yeah that's right, in Australia we wear our thongs on our feet

**CHARLES:** If someone wants to get out of their thongs, what can they do Mel?

**MEL:** Get yourself a fried dimmy with soy sauce, perhaps don't ask what's in it. Don't forget to try Vegemite. Spread it on your toast or perhaps pop it in bread with cheese, goes all right. Footie fever's also hit Melbourne, I perhaps won't say much about that, my team didn't make it. Now for those of you who like to explore the beach we've got St Kilda. We've got great coffee, and if it's a touch of Egypt head to our museum: Tutankhamun If art's more your thing head to the national gallery or the Ian Potter Centre for some real Aussie art. Or, if you look out the window there, take a stroll down the Yarra, there's a fire display on the hour, every hour.

**CHARLES:** I've seen on TV that it's always sunny in Australia, right?

**MEL:** It's Melbourne, be prepared for all seasons in one day. 17 today, 7 overnight, you're going to need a rug Charles, especially if you're going to the rooftop cinema tonight. Pop on your warm clothes, you're going to freeze.

**CHARLES:** I've already selected a particularly warm frock for the evening. Now I know the Economist said that Melbourne is now the finest place on earth to live but I understand Melbournians aren't..

**MEL:** Melbonians

**CHARLES:** Melbonians, aren't entirely happy, is that correct?

**MEL:** That's right Charles, this week in a local Melbourne newspaper, children were asked the question: 'How would you make Melbourne a better place?' Now young Lily thought you'd do that by being a barbie or a princess and playing with lots of arts and crafts Abdujabar thought there should be more zoos with lions in them, and swimming with the sharks and dolphins would make Melbourne alright Young Magnus, he thinks more magnets would do it [laughter]

**CHARLES:** Thank you very much Mel, you can go on the promise that you'll be back tomorrow night

**MEL:** I will [applause]

**CHARLES:** And now something to remind you of why you are all here. The people who are watching this on the net around the world: some have Huntington's now, some will have it in the future and many more are the unsung heroes who care for those people. They are a breed apart, truly special people. Now a group of teenagers from Adelaide in South Australia who have to look after relatives who have HD got together in a workshop to co-write a song about their lives. Here, set to pictures of carers around the world who pray that you scientists here can help them, and the people they love, is the extraordinary song: Tell someone we care. This was actually put together by these children, I say children - teenagers, who put their experiences together, talked about it, wrote the song, the words to the song express their experience for caring for a loved one: Tell someone we care. [song] [applause] Isn't that extraordinary? Thank you to all of the families, especially here in Australia for providing those pictures. Now I have one small piece of housekeeping to do as we near the end of our presentation. Alice Wexler has asked the representative of the HD family organizations to come to a brief informal meeting after Oz Buzz here. The purpose of which will be to update on their activities, problems and needs. I want to just tell you an extraordinary anecdote about something that happened about two hours ago in this building which I think speaks volumes. First, about the reality of the prevalence of this disease which is much greater, we all know, than the statistics that have been given out in the past. And also about how much we have to do to draw people into organizations like this. Just two hours ago in this building Professor Alan Tobin was in the lift, elevator, of the Hilton hotel and he saw some people who he assumed were delegates at the conference. He said "How do you do? I'm Professor Tobin" and they realized that they were actually at a completely different conference, totally unrelated. Any they asked "what are you here for?", and he replied "an HD conference going on here." And they said well our family is absolutely riddled with Huntington's disease but we didn't know there was any way of getting support. Is there any way of getting advice? That actually happened two hours ago in this building, incredible. So this is Oz Buzz's premier, I hope that you will be able to join us tomorrow night. Thank you, the audience here, and at home. It's because of all of you that we get a chance of hope, that we can dream of a day soon when this disease does not scare us anymore. And on that note I will leave you with the words of an Aboriginal proverb: "Those who lose dreaming, are lost", goodnight

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*The authors have no conflicts of interest to declare. [For more information about our disclosure policy see our FAQ...](#)*

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## **GLOSSARY**

**motor neuron disease** A progressive neurological disease in which motor (movement) neurons die. Also known as ALS or Lou Gehrig's disease.

**huntingtin protein** The protein produced by the HD gene.

**RNA interference** A type of gene silencing treatment in which specially designed RNA molecules are used to switch off a gene

**gene silencing** An approach to treating HD that uses targeted molecules to tell cells not to produce the harmful huntingtin protein

**observational** A study in which measurements are made in human volunteers but no experimental drug or treatment is given

**therapeutics** treatments

**mitochondria** tiny machines inside our cells that process fuel into energy, enabling cells to function

**CAG repeat** The stretch of DNA at the beginning of the HD gene, which contains the sequence CAG repeated many times, and is abnormally long in people who will develop HD

**prevalence** A figure estimating how many people there are in a particular population who have a certain medical condition.

**stem cells** Cells that can divide into cells of different types

**anti-sense** the half of the DNA double-helix that is mostly used as a backup, but sometimes produces message molecules

**neuron** Brain cells that store and transmit information

**chorea** Involuntary, irregular 'fidgety' movements that are common in HD

**Allele** one of the two copies of a gene

**genome** the name given to all the genes that contain the complete instructions for making a person or other organism

**RNA** the chemical, similar to DNA, that makes up the 'message' molecules that cells use as working copies of genes, when manufacturing proteins.

**Cas** The bit of a gene-editing system that cuts the DNA. It's a type of protein called a nuclease. Cas is short for 'CRISPR-associated'.

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