

HD Awareness Day **Friday 30th March 2007**

Meeting the Care Needs of Younger People Seminar held 16th March 2007

The seminar for family members and health professionals held on Friday 16th March addressing the topic of 'Meeting the Care Needs of Younger People' was attended by approximately 40 people.

David Conry, Manager of Youngcare spoke about the obvious gap in service provision which has led to a conservative figure of approximately 6,000 younger people with high care needs in aged care nursing homes across Australia. Youngcare are advocating for younger people, promoting the need for a choice in living environments that can allow for an age appropriate lifestyle with dignity and opportunities to remain engaged in the broader community. To this end, Youngcare are very solution focused and are in the process of building a high care facility for younger people in Jindalee. This facility will have apartment style accommodation for 18 younger people and the hope is the develop a model of service that can be replicated by other government and non-government bodies. Youngcare also hope to develop an 'in-home' care service and a purpose built holiday accommodation facility.

David strongly advocates the need for partnerships with other organizations to achieve better outcomes for younger people. To realize the Jindalee facility, Youngcare have partnered with COAG (Council of Australian Governments), Wesley Mission and Disability Services Queensland.

Our second speakers were Elly Mander (Service Co-ordinator) and Vivienne Davis (Recreation and Volunteer Co-ordinator) from Arthur Preston Residential Services in Melbourne. Elly and Viv spoke about their Person Centered Approach to caring for people with Huntington's Disease in their Huntington's specific residential facility. The Arthur Preston

team not only identify what is important 'for' an individual, but also what is important 'to' the individual. In this way they can assist their residents to work towards a preferred future and make choices along the way.

During their speech, we viewed residents of the Arthur Preston centre doing some amazing activities like ballooning and parachuting. But, this framework is just as important for the day to day stuff. For example, it might be important 'for' Bill (fictitious name) a resident of the centre to have his food pureed. However, it is important 'to' Bill that he can have a glass of sherry after dinner.

It is attention to the lifestyle factors that can improve the quality of care for younger people with high care needs.

Posters

We would like to send a big thank-you to Robert, who was our 'model' for our Awareness Day Posters. Robert's willingness to participate and openness about his life has touched many people. The posters send a powerful message highlighting the lack of options in Queensland for younger people with high care needs. We would also like to thank his family for supporting his involvement.

Thank you also to Ted Hagemeyer of Sublime Photography (in Graceville), who donated his time to assist us with this project. Ted provided us with the wonderful images of Robert – and captured expressions that speak a thousand words.

The posters were mailed out to all Residential Facilities caring for HD clients in Queensland, Genetics Clinics, and Community Care Groups in the south east corner.

Banner

Our Awareness Day banner will be displayed outside our office from the 22nd March to the 30th March. Being situated on the very busy Ipswich Road at Annerley will provide us with good exposure.

Stickers

Our car stickers are now available. If you are keen to display a sticker, please call Barbara on 3391.8833. A sample of the sticker is on the back page of the newsletter. The sticker is in the Association's colours of green and red on a white background. Over 500 stickers have already been distributed.

Media Coverage

Gwen has been involved in 4 regional radio interviews as well as promoting the plight of the Vathke family in the Gold Coast Bulletin for which we have received a lot of phone calls from people in the area who have offered information and support. (Newspaper article attached)

We are very grateful to Judith and her granddaughters for opening their hearts and lives and assisting us in our Huntington's Disease awareness campaign.

Morning Teas

Three morning teas have been organized to help us to promote Awareness about Huntington's. They are being held in Caboolture, North Coast Region and Townsville.

Movie Night

Unfortunately, we did not get enough numbers to go ahead with our movie night as planned, so we have cancelled this event.

Julie Morrow, Welfare Officer

Do you care for a family member or friend who has a disability or who is frail aged?

Do you need practical assistance or information or just someone to talk to?

Is there a young carer in your family needing support?

Ring Carers Queensland on
1800 242 636

HD Lighthouse Contributing Editor's Comment – Marsha L. Miller, Ph. D

Posted to the HDL: 19 Sep 2006

<http://www.hdlighthouse.org/diagnosis/cag/updates/1299CAG.php>

It has been awhile since a review of CAG counts in Huntington's disease was published, so the new article (below) by Dr. Michael Hayden and colleagues is very helpful. The normal, intermediate, and reduced penetrance ranges have changed over time and there have also been errors in publications. The authors present the most current information in the table below.

<i>Unaffected:</i>	
❖ <i>Normal</i>	26 and below
❖ <i>Intermediate</i>	27 - 35
<i>Affected:</i>	
❖ <i>Reduced Penetrance</i>	36 - 39
❖ <i>Full Penetrance</i>	40 and above

Individuals receive two copies of the huntingtin gene, one from each parent. A copy of the gene is called an allele. Since Huntington's disease is a dominant disorder, only one HD allele is necessary for the individual to develop the disease.

- ❖ If an individual has alleles in which the CAG repeats are 26 or below, then they will not develop Huntington's disease nor will any of their children.
- ❖ If an individual has an allele with 27 to 35 repeats, it falls within the intermediate range. The gene is normal and the individual will not develop Huntington's disease. *There have been no reported cases of HD in individuals with 35 CAG repeats or less.*

It is estimated that between 1 and 3.9 percent of the general population has a huntingtin gene with CAG repeats that fall into the intermediate range. However, the CAG repeats on an intermediate allele could increase if transmitted to one's offspring, possibly leading to Huntington's disease in the next or subsequent generations. Alleles with CAG repeats in the intermediate range are sometimes referred to as *normal mutable alleles*.

- ❖ Individuals with an allele with 36-39 repeats fall into the reduced penetrance range. Some people in this range will develop the disease and some will not.
- ❖ An allele with 40 or more CAG repeats is a full penetrance Huntington's disease gene. If the person lives long enough and doesn't die of another cause, for example, a traffic accident, he will develop Huntington's disease.
- ❖ A child of a parent with one HD allele and one normal allele has a 50 per cent chance of receiving the HD gene.

People who undergo gene testing and discover that they have an allele which falls into the intermediate category are presented with some concerns. On the one hand, they have been reassured about their own status, but they continue to have uncertainty about children they may have or are planning.

A major question is what is the likelihood of the CAG repeats expanding and causing HD in a child?

More research needs to be done to answer this question but several factors which affect likelihood are known at this time. One factor is the CAG count itself. The likelihood of expansion is greater with higher CAG counts. There has only been one known case of expansion from 27, the lower end of the range. A father with this count transmitted an allele with 38 CAG repeats to his child.

A second factor is gender. Expansions of CAG counts are more common when the parent is

male, regardless of whether he is in the intermediate, reduced penetrance, or full penetrance range. There has not yet been a case reported where this has happened from a mother and the likelihood of this happening is considered to be much less. There have been cases of maternal expansion from the reduced penetrance range, however.

A third factor is age. In the cases where an intermediate allele was transmitted into the affected range, the father was older with an average age of 36.7.

A fourth factor is family history. The risk of transmitting an allele in the HD range is higher if the person is from a family with new mutations rather than someone who has an intermediate allele from the 'general population'.

This requires some explanation.

There is no routine screening for the HD gene and people who are not from HD families do not request the test unless they are showing Huntington's disease-like symptoms, so the presence of intermediate alleles usually comes to light when someone who is at risk of testing. They may discover that they did not inherit their affected parent's HD gene but inherited an intermediate allele from their other parent who is from a family where HD has not occurred. For the purpose of discussion, the authors call them GP:IA (for general population intermediate alleles).

When someone without a family history is diagnosed with Huntington's Disease and the HD gene is confirmed through testing, siblings and parents may also seek testing to discover their own status. The transmitting parent with the intermediate allele or siblings who have received an intermediate allele would be classified as part of a family with new mutations – NM:IA.

Why would there be a difference?

The answer probably lies in haplotypes, or the actual coding of the whole gene. There are

other variations in the coding of the huntingtin gene that affect the likelihood of expansion. For example, the CAG repeats are usually followed by a CAA codon.

But in two families identified so far, there's a point mutation and the CAA has been replaced by another CAG repeat. This makes the CAG repeats more likely to expand.

There are other haplotypes which make expansion more likely. If there has already been one expansion in a family from an intermediate allele, the chances are greater that one of these haplotypes is found in the family.

So what are the person with an intermediate's actual chances of passing on a gene in the HD range to a child?

The best estimate at this time comes from analysis of sperm. The likelihood of an intermediate CAG count expanding to the HD range (36 and above) in a child is estimated at 6-10 percent if the parent is male with a CAG count of 35, 6 percent if it's a GP:IA and ten if it's a NM:IA (Chong et al., 1997). The odds for women aren't known but are thought to be much less.

References:

Chong, S.S., Almqvist, E., Telenius, H., LaTray, L., Nichol, K., Bourdelat-Parks, B., Goldberg, Y.P., Haddad, B.R., Richards, F., Sillence, D., Greenberg, C.R., Ives, E., Van den Engh, G., Hughes, M.R., and Hayden, M.R. (1997). Contribution of DNA sequence and CAG size to mutation frequencies of intermediate alleles for Huntington Disease: Evidence from single sperm analyses. Human Molecular Genetics 6:302-309.

Collaboration and The Cure

*By Katie Reid, Communications Coordinator
Huntington Society of Canada*

Funding and seeding research is important in the search for a cure, but also integral to the search is the need for collaboration not only between HD families and researchers for

clinical trials, but between HD researchers and agencies around the world.

One of the most ardent proponents of collaboration within the HD community is the High Q Foundation and CHDI Inc. CHDI, Inc. and the High Q Foundation, Inc. (High Q) are non-profit organizations that share the mission of bringing together academia, industry, governmental agencies, and other funding organizations in the search for Huntington disease (HD) treatments.

CHDI, Inc. is pursuing a biotech approach to rapidly discover and develop drugs that prevent or slow HD. Through collaborations with industrial and academic partners, CHDI, Inc., participates in all aspects of drug discovery and development from high throughput screening to pre-clinical development. High Q supports HD research aimed at target identification and validation, the development and use of animal models, drug delivery, and the search for markers of disease progression.

In accordance with their mandate CHDI recently signed contracts with two different firms to help further HD research.

On August 1st CHDI Inc. and Edison Pharmaceuticals Inc. formed a partnership to develop analogs (a chemical compound that is structurally similar to another but differs slightly in composition) of Coenzyme Q10 selectively targeted to reach the brain and address the mitochondrial component of HD.

Edison Pharmaceuticals Inc. is focused on the development of drugs to treat inherited respirator chain diseases of the mitochondria - also referred to as energy impairment diseases. As recent research has suggested that alterations in energy metabolism may contribute to HD this is a strategic partnership.

CHDI has also selected to work with Evotec AG as a strategic partner. Since March 2006 four agreements have been signed between the two companies covering medicinal chemistry, assay development and medium-throughput screening (MTS), ultra-high-throughput

screening (uHTS) and library synthesis and management services. These contracts cover most of Evotec's integrated discovery offering. With access to these resources, CHDI has all the tools in place to help rapidly discover novel drugs against HD targets and further optimise them to the point of clinical development.

These partnerships combine the strength of each company improving the search for a treatment and cure.

Acknowledgement: "Horizon" Huntington Society of Canada Newsletter, No. 120, Fall 2006

Fourteen Changes That Could Signal Concern in HD

Phil Hardt wrote this article some years ago to inform and teach about some of the "softer" symptoms of HD so they could easily be understood, allowing everyone to recognize their subtleties early on so they can cope positively with them, instead of letting them ruin their lives. In addition, he said he got tired of everyone saying to him (when I would try to describe what was happening to me) – "I always forget where I leave my keys", or "Everyone does that!" He wanted to show the real HD-affected problems are much deeper than most realize, when compared with normal forgetfulness, emotions and behaviour. The idea came from an Alzheimer's handout he read, changed to fit those early cognitive, emotional and behavioural symptoms of HD. His goal was not to create mass hysteria, but to help eliminate so much of the distress and concern. The 14 Changes are:

1. Memory Loss that affects job or personal skills. It is normal to forget an assignment, deadline or a colleague's name, especially when under stress. However, frequent forgetfulness or confusion at home or in the workplace over an extended period is not typical of the person with HD and may signal concern.

2. Difficulty performing familiar tasks. Busy people pause occasionally to

think about what they are doing, or how to finish a project. However, concern is raised if tasks which used to be completed in two hours start taking all day, or if getting ready to go somewhere takes an hour instead of ten minutes.

3. Problems with speech and language. Everyone has trouble finding the right word sometimes, but a person with HD may forget simple words or substitute inappropriate words, such as, "place the pie in the toilet", instead of saying "in the oven". Speaking may be slow, with pauses between words and responses.

4. Disorientation to time and place. Anyone may momentarily forget the day of the week or what is needed from the store. But persons with HD can easily become distracted and completely lose track of time and tasks. They may remain disoriented until it is brought to their attention. They may get lost driving home, to work, or even a familiar store.

5. Changes in mood or behaviour. Everyone experiences a broad range of emotions – it's part of being human. However, persons with HD may exhibit rapid mood swings for no apparent reason. These moods may be uncharacteristic changes from their usual temperament. They may show reduced or inappropriate emotional responses to any given situation.

6. Poor or decreased judgement. Everyone has gotten upset when they received a traffic ticket. However, a person with HD who is stopped and falsely arrested for being drunk may become extremely angry and insult or even hit the policeman because of poor judgement or lack of consequential thinking.

7. Problems with abstract thinking. Balancing a checkbook can be challenging for anyone, but for someone with HD, recognizing numbers or performing calculations may be extremely difficult and stressful. Diminishing concentration, focus and sound decision-making may signal problems if they continue for no apparent reasons.

8. Misplacing things. We all misplace a wallet or keys from time to time. However, a person with HD may put items in inappropriate places and not remember doing so, such as placing a carton of milk in the cupboard or a wristwatch in the sugar bowl.

9. Changes in personality. Personalities often change with age. A person with HD may experience uncharacteristic changes in their personality. For example, someone who has generally easygoing may become angry, paranoid or fearful and someone who was outgoing may become withdrawn from social interaction.

10. Loss of initiative. It's normal to tire of housework, business activities or social obligations. But for most people, this feeling is brief and enthusiasm and interest return. The person with HD may become apathetic and become indifferent towards activities which used to bring them satisfaction and happiness.

11. Depression. Tragedy saddens us all but the person with HD may not recover – they may show increased irritability or crying and may express feelings of hopelessness or guilt. They may lose interest in ordinary activities, such as sex, and may even experience disturbances in eating and/or sleeping patterns. Severely depressed individuals may even talk openly of suicide, saying things like, "I'm not needed anymore", or "Things would be better off without me".

12. Loss of social inhibitions. It is normal to change beliefs or values as you age. A person with HD may uncharacteristically start cussing, gambling, lying, cheating, stealing or being sexually inappropriate – things they would have never done before.

13. Loss of visual-spatial coordination. Anyone might misjudge a turn or hit a curb. However, a person with HD may lose the coordination or reflexes to avoid an accident, back out of a driveway, or shift the car. They may hit their elbows while walking through

doorways or bump into a wall while simply walking down a hallway.

14. Slowed comprehension. Everyone occasionally misses the punch line of a joke but a person with HD may have poor or slowed comprehension so they cannot grasp the meaning of a story or conversation. There may also be slowed interpretations or misinterpretations of facial expressions, such as approval or disgust, causing inappropriate responses and misunderstandings.

PLEASE NOTE: The changes noted above, subtle to severe, should be brought to your doctor's attention if they begin happening inexplicably or are not characteristic of your usual behaviour or normal abilities. Since some of these warning signs are so personal, only you or someone close to you, can help determine if they indicate areas for concern or are simply a result of stress or illness. What is typical for one person may not be considered "normal" for someone else.

Phil Hardt

*Link: <http://www/hdlighthouse.org/treatment-care/care/managinghd/updates/0002concern.shtml>
Reprinted: "Contact" AHDA (Vic) Inc. Number 32, August/September 2006*

Weighing up the Side Effects

All medicines potentially cause side effects, and side effects are one of the risks of taking medicines.

Deciding whether to take or continue taking a particular prescription medicine involves weighing up the anticipated benefits and potential side effects, in consultation with your doctor and pharmacist.

Your decision will be influenced by many factors, including your beliefs about health and medicines in general, the nature and severity of your condition, your feelings about the benefits or anticipated benefits of the medicine, the likelihood of experiencing side effects, and your tolerance of the side effects.

For example, someone taking a medicine for a life-limiting condition may be more likely to tolerate severe side effects than someone taking a medicine for a minor condition. Because of the seriousness of their condition, they are more likely to feel that the potential benefits of the medicine outweigh the impact of the side effects.

The more you know about the potential side effects, the more prepared you will be if you experience them.

Ask you doctor

Start the process of weighing up the benefits and side effects of a medicine when your doctor first suggests a new medicine.

No doctor can tell you with certainty what side effects you will or will not suffer, because everyone reacts differently to medicines. However, they can give you some guidance about your likelihood of having side effects based on their experience of treating other people with the medicine, and statistics about the medicine's side effects.

With new medicines, the statistics are derived from clinical trials conducted on relatively small numbers of people, so usually only the more common side effects will have been seen. Rarer side effects that affect, say, less than 1 in 10,000 people may not have been seen.

Questions to Ask

Do ask your doctor and pharmacist any questions you have about the side effects of your new medicine. The more you know about the potential side effects, the more prepared you will be if you experience them.

The questions you might like to ask your doctor and the pharmacist include:

- ❖ What are the very common and common side effects?
- ❖ How often do the side effects affect people like me (ie. people of my age, state of health, etc).
- ❖ Are the side effects temporary or long lasting?
- ❖ Are there any serious side effects I should watch out for, and what should I do if I get them?

In contrast, more statistics will have been collected for medicines that have been used by thousands of people over many years. As a result, more will be known about the side effects, and the rarer side effects are likely to have been seen.

Similarly, no doctor can tell you about *all* the potential side effects of a medicine. However, they should tell you about the 'very common' and 'common' side effects (see box below) and about any side effects that may be particularly relevant to you.

If you are unsure whether to start a medicine, ask your doctor to go through the pros and cons of the medicine and any alternatives until you have the information you need to make a decision.

If you are considering stopping a medicine because of the side effects, tell your doctor. They may be able to prescribe another medicine instead, or adjust the dose of your existing medicine so the side effects disappear or become more tolerable.

What is the risk?

The terms used by doctors and pharmacists to describe the risk (likelihood) of particular side effects have quite specific meanings, and these meanings may be quite different to our personal interpretations.

The table below give the official meaning of each term used to describe the risk of getting a particular side effect. Thus, if your doctor says a particular side effect is rare, they mean that the side effect is likely to be experienced by 1-10 people in every 10,000 people or about 0.01%-0.1% of people.

Very common:

- ❖ More than 1 in 10 people
- ❖ More than 10% of people

Common:

- ❖ Between 1 and 10 people in every 100
- ❖ Between 1% and 10% of people

Uncommon:

- ❖ Between 1 and 10 people in every 1,000
- ❖ Between 0.1% and 1% of people

Rare:

- ❖ Between 1 and 10 people in every 10,000
- ❖ Between 0.01% of 0.1% of people

Very rare:

- ❖ Less than 1 in 10,000 people
- ❖ Less than 0.01% of people

Ask your pharmacist

Don't forget that pharmacists know a lot about medicines. They can usually answer any questions you may have, and give you further information about the medicine and its side effects.

If you are unsure whether a new symptom could be a side effect, consider talking to your pharmacist in the first instance. They will be able to tell you whether it could be a side effect and whether you should do something about it, such as going back to your doctor.

Read the CMI

Consumer Medicine Information (CMI) leaflets are specially prepared leaflets that give you comprehensive information about medicines: how they work, their side effects, how and

when to take them, and so on. A CMI leaflet has been written for every prescription medicine.

Doctors have CMI leaflets for most prescription medicines on their computers. When your doctor suggests a new medicine, ask them to print off the CMI leaflet. Both of you can then refer to it when discussing the potential benefits and side effects of the medicine. You can also highlight any important details on the leaflet, so you don't forget them when you get home.

Your pharmacist should give you a CMI leaflet when you have a prescription for a new medicine filled for the first time, and on request thereafter.

Many CMI leaflets are also available on the internet at www.nps.org.au (click on consumers).

At first glance, the list of possible side effects in a CMI leaflet may be quite daunting. However, if you understand what the terms 'very common', 'common', 'uncommon', 'rare' and 'very rare' mean (see box page 8), you can put the likelihood of side effects into perspective.

Ring Medicines Line

You can ring Medicines Line, Australia's national medicines information service for information and advice. The pharmacist on duty will answer your questions about medicines and their side effects. They can also post you the CMI leaflet for your medicine. Ring the service on 1300 888 763 Monday to Fridays 9am – 6pm EST.

How 'The System' Makes Medicines Available

Australia has a comprehensive system for regulating prescription medicines that is designed to give us access to safe, effective and affordable medicines when we need them.

Unfortunately, the complexity of the system means that few of us understand how it works. This article will endeavor to explain the processes involved in making new prescription medicines available to consumers. Please note that it does not explain the entire process.

Making new prescription medicines available to consumers involves two separate processes. One process decides which medicines can be sold in Australia. This other process decides which medicines will be listed on the Pharmaceutical Benefits Scheme.

Marketing approval process

All prescription medicines must be evaluated for safety, effectiveness and manufacturing quality before the company can sell them in Australia.

Medicines must be evaluated for safety, effectiveness and quality before the company can sell them in Australia.

The body responsible for evaluating prescription medicines is the Therapeutic Goods Administration (TGA), which is part of the Department of Health and Ageing.

The TGA evaluates a medicine by weighing up the risks and benefits associated with its use. In doing so, it looks at the medicine's strength, side effects, toxicity and likelihood of harm with prolonged use, as well as the seriousness of the medical condition for which it will be used.

This approach ensures that safety is a prime concern when evaluating medicines used for less serious and long-term conditions. It also allows more toxic but potentially life-saving medicines to be approved for use in serious conditions, such as cancer, if the benefits outweigh the risks.

When evaluating a medicine, the TGA uses information derived from studies of the medicine's chemistry, studies of its effects on animals, and studies of its effects when used to treat people with the condition for which it will be used (clinical trials).

When the Therapeutic Goods Administration has completed its evaluation, it usually seeks the advice of an independent committee known as the Australian Drug Evaluation Committee (ADEC) before making a decision about its approval. Most of the committee's members are eminent medical doctors but a few are experts in other fields such as toxicology.

If approved, the medicine is listed on the Australian Register of Therapeutic Goods. It can then be sold by the manufacturer for use by Australian health consumers. However, the price charged will be determined by the manufacturer.

PBS listing process

After a medicine has been approved for sale in Australia, the manufacturer can apply to have the medicine listed on the Pharmaceutical Benefits Scheme (PBS). Listing on the PBS usually makes the medicine available to consumers at a cheaper price, because the Government subsidises the amount consumers pay.

Making a prescription medicine available on the PBS involves two stages. First, deciding if the medicine should be listed on the Scheme. Second, setting the price the Government pays the company for it.

The Pharmaceutical Benefits Advisory Committee (PBAC) is the body responsible for recommending whether a prescription medicine should be listed on the Pharmaceutical Benefits Scheme. When evaluating an application, it considers the medicine's cost, effectiveness and safety compared with the other treatments available for the condition for which the medicine is intended, including any non-drug treatments.

The Pharmaceutical Benefits Advisory Committee also recommends how much of the medicine should be available on a prescription and the number of repeat prescriptions allowable. It may also recommend that use of the medicine be restricted under the PBS if it feels such restrictions would enhance its safe and wise use, and ensure that it is used only for those people for whom its cost effectiveness (value for money) has been proven.

Evaluation for listing on PBS involves comparing the medicine's cost, effectiveness and safety with other treatments.

For example, the osteoporosis medicine Alendronate is available under the PBS only if the person has had an X-ray that shows they have had a fracture caused by osteoporosis. This is because it has been shown to have greater benefits for people who have already had a fracture compared with those who have not.

If the Pharmaceutical Benefits Advisory Committee recommends that a prescription medicine be listed on the PBS, it forwards its advice to the Pharmaceutical Benefits Pricing Authority (PBPA). The Authority then evaluates all the relevant information and recommends the price it thinks the Government should negotiate with the company for the medicine.

Both sets of recommendations then go to the Minister for Health for a final decision.

If listing is recommended and the Government can negotiate an agreed price with the company, the medicine is listed on the Pharmaceutical Benefits Scheme. It is then available to consumers for the price they normally pay for PBS prescriptions (see box page 11).

Pharmaceutical Benefits Scheme

The Pharmaceutical Benefits Scheme was set up in 1948 to ensure that the Australians have access to necessary prescription medicines at an affordable price. As of 1 December 2005, the scheme covered 804 medicines, which were available in 2,138 strengths and forms (tablets, capsules, injections, ointments, etc) and marketed as 3659 brand products.

Each time a medicine is dispensed, the Pharmaceutical Benefits Scheme pays the difference between the price paid by the consumer and price paid to the manufacturer, plus a fee to the pharmacist for dispensing the medicine.

A prescription of the medicine is then available to consumers for a maximum of \$4.80 for people on a concession card and a maximum of \$29.50 for people not on a concession card (2006 prices).

Implications

Australia's drug regulation system is recognised as being one of the best in the world. However, its comprehensiveness sometimes results in frustrations for consumers. In particular, the time needed to complete the evaluation processes can mean long waits for people wanting to use new medicines. In addition, not all medicines approved for sale end up being listed on the PBS, which makes them available only to consumers who can afford to pay the price charged by the company.

Acknowledgement:

Medicines Talk, Autumn 2006

This is Dorothy's Story

My name is Dorothy and I am the youngest member of a Huntington's disease family. As my father was 45 when I was born I never knew him before he developed the condition.

For most of my life I was ignorant of the fact that I was 50% at risk of developing HD.

As a child I considered life was pretty well 'normal' – but looking back now I can see it was far from that. We lived in fear of Dad and had to tread softly for fear of rousing his vile temper. Even though he never physically abused us the emotional and psychological abuse left me very much traumatised.

After leaving school I began my nursing training at Latrobe. It was at this time I became aware of the HD stigma as I was refused entry to the superannuation scheme as perhaps, someday in the future, I might develop HD. After 6 months my training was terminated and I was never informed as to why this happened. Even my mother was not told the reason did it have anything to do with my family history of HD???

Some people even treated me as though it was inevitable that I would develop HD. I discovered that my surname and HD were synonymous – that my connection to an HD family was automatic. People back then, and even many today, are not aware that HD only affects families who have inherited the gene.

Later on I undertook my Mothercraft training in Hobart and then went on to work in this field at several maternity hospitals and life was pretty good. Eventually I completed my general nursing training in Victoria and went on to obtain my midwifery certificate. I worked in maternity until I went to New Guinea for 2 years – no one knew my HD history there!

On returning to Tasmania I worked in midwifery again – then came the time when the ward where I was working was facing closure and began the worst period of my life and could only be described as 'hell on earth'.

I was forced into a 'retraining course' even though the course co-ordinator did not want me and only grudgingly accepted me into the course. I don't know how she connected me to an HD family but somehow she did, and

told other members of the course that I was showing signs of HD. I had just previously been through a presymptomatic test and knew that I was not displaying any such signs. Later I was informed that it had been reported that I had HD but was denying it. I expect this was the reason my retraining was terminated. This news left me feeling like I had been hit by a ten ton truck and splattered all over the universe.

I went back to the hospital to work and as the ward was closing, eventually took a forced redundancy. I discovered Hospice Care and decided that was something I could do – but it didn't work out either.

Then came the good part of my life! That same year I trained as a volunteer with a group that was organised by the local council and was lovingly accepted into the group. There followed almost 13 wonderful years of helping the frail elderly and disabled and loving it. As I said to one dear lady that while they get pleasure out of what we do for them, we also gain much pleasure in being able to help.

I took the predictive test in 1993 and received the best result I could have hoped for – I was not going to develop HD. Yet how could I tell my brother as he had a positive result and I was negative? He was delighted!

I have been a member of the Huntington's Disease Association for 20 years and have held the position of branch secretary for 10 years.

These have been good years as it is great to be free from the shackles of HD. I am healthy and have found my niche in life. I also enjoy helping out with the Grief Share support groups run twice a year at our local Church. Another ministry I have is writing poetry which I once kept to myself but I now share with others. I am no longer the timid little insecure person I used to be and hope to go on helping others, with or without HD, for as long as possible.

Acknowledgement: HD Newsletter, AHDA (Tas) Inc., August 2006

Fundraising

Upcoming events:

Movie Night – Friday 30th March: See attached flyer for details.

**Sausage Sizzle – Bunnings, 51 Kingston Street, Underwood – Saturday 3rd March, and
Bunnings at Maroochydore – Sunday 18th March.**

Please phone the HD Office if you are able to assist at either of these events. Each Sausage Sizzle usually raises between \$700 and \$1000 in funds, so it is important that we take up Bunnings' generous offer.

Community Assistance – We have received, and gratefully acknowledge major financial assistance from the following donors:

AI Interior Linings

G. Cheyne

K. Fox

R. Hirsimaki

J. & G. Philp

M. Sevil

Corks– Our cork supply seems to have dwindled off. Contact the HD Office for information on this fundraiser.

IGA Community Chest

By shopping in an IGA Store and buying IGA branded products or selected lines marked with the Community Chest logo, a guaranteed percentage from each of these products sold is set aside. At the end of each year, your local IGA store selects a local group or charity to donate all of the money that has been collected at the store.

Each outlet may differ in how the scheme operates, so if you shop at your local IGA store, why not approach management requesting details of their Community Chest with a view to nominating the HD Association as your charity.
