

HUNTINGTON'S DISEASE WHAT'S IT ALL ABOUT ?



A GUIDE FOR YOUNG PEOPLE

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HUNTINGTON'S DISEASE AND ME

You're probably reading this leaflet because someone in your family has Huntington's Disease (HD) - it might be your mum or dad, your granny or granddad, your aunt or uncle. You might have more than one person in your family who has HD.



WHAT IS HUNTINGTON'S DISEASE

Huntington's Disease (HD) is a hereditary illness that causes damage in a certain part of the brain. This stops the brain and body from working as well as they used to, and affects a person's movements, thoughts and behaviour. People generally start to develop symptoms of HD between the ages of 30 and 50. The part of the brain that is affected is called the basal ganglia. This is a very important part of the brain that co-ordinates a lot of the information that's whizzing around, and makes sure it gets to the right part of the brain. HD causes brain cells in the basal ganglia to deteriorate and die. This means that it can no longer co-ordinate information as efficiently, so the messages being passed through the brain don't always get to the right place, or get lost along the way the way.

Think of your mobile phone - sometimes you get a signal, no problem. But at other times, the signal gets cut off halfway through, just when you were getting to the good part of the gossip! At other times, you cannot get a signal at all.



Having HD is a bit like this, sometimes the messages get through and other times they only get a little of the way through. Other times they just don't get through at all. So some days are better than others. Some things affect the messages getting through - if the person is feeling stressed, this makes it harder for the messages to get through and things may become worse at times of stress. The brain cells in the basal ganglia deteriorate over a long period of time. This deterioration can take 15-20 years. During this time the affected person slowly loses different skills and abilities. This happens gradually and they can be reasonably well for a long time.

HOW DO YOU GET HD?

You might have more than one person in your family who has HD. This is because HD runs in families - it is hereditary. This means it can be passed from parents to their children. People might say that you look like other people in your family - this is because of your genes. Genes are passed to you from your parents - that's why you might have freckles like your dad or brown eyes like your mum. You have about 30,000 different genes in your body.

Genes are sections of DNA carried on your chromosomes. These genes are all different, and are programmed with different information. They decide things like the colour of your hair and your eyes, how tall you are, etc. They are like a recipe book - your genes are the ingredients that make you uniquely you. Everyone has a different recipe - that's why we're all different shapes and sizes. Most cells in your body have 23 pairs of chromosomes. Each parent gives you 1 chromosome in each pair.

Our genes are strung out along our chromosomes and there are 100s of genes in each chromosome. We have 2 copies of each gene as well - one from each parent. People can be identified because of the differences in their genes and the DNA that makes them. DNA can be extracted from one cell of our body, such as a single skin or blood cell, or even a hair root. That's why on your favourite crime dramas, they search the crime scene for DNA evidence. People can then be identified by their DNA.

Sometimes genes don't work properly. In people with HD, a faulty gene causes things to go wrong in the brain. If a parent has HD, they have 1 copy of the faulty gene and 1 good copy. They may pass either of these on to their children. Their children therefore have a 1 in 2, or 50%, chance of inheriting the faulty gene. You may hear people saying you are "at risk" - this just means you have the 50% chance of inheriting the faulty gene. If your grandparent has HD that means your parent is at 50% risk however if they do not have the faulty gene, then you are no longer at risk. The faulty gene does not "skip" from one generation to the next .

WHY DOES HD TAKE SO LONG TO DEVELOP?

No-one is 100% sure about what causes HD. We are fairly certain that the damage in the brain is caused by a build up of a protein (a type of body chemical) called *huntingtin*. The cells in a certain part of the brain slowly become sick due to a build up of this protein over a long period of time - it is when the build up of this protein becomes too much that the cells can no longer work as well as they used to and become sick and start to die. This damage to the brain cells is what causes all the changes in HD.

This is why most people only start becoming unwell when they're older - it takes that long for the protein to build up in the brain cells and make them sick. This means that people are well for a long time before any signs of the disease start to show. So they live normal lives like everyone else - they have careers, they get married, travel - do all the usual stuff basically!

BEING AT RISK

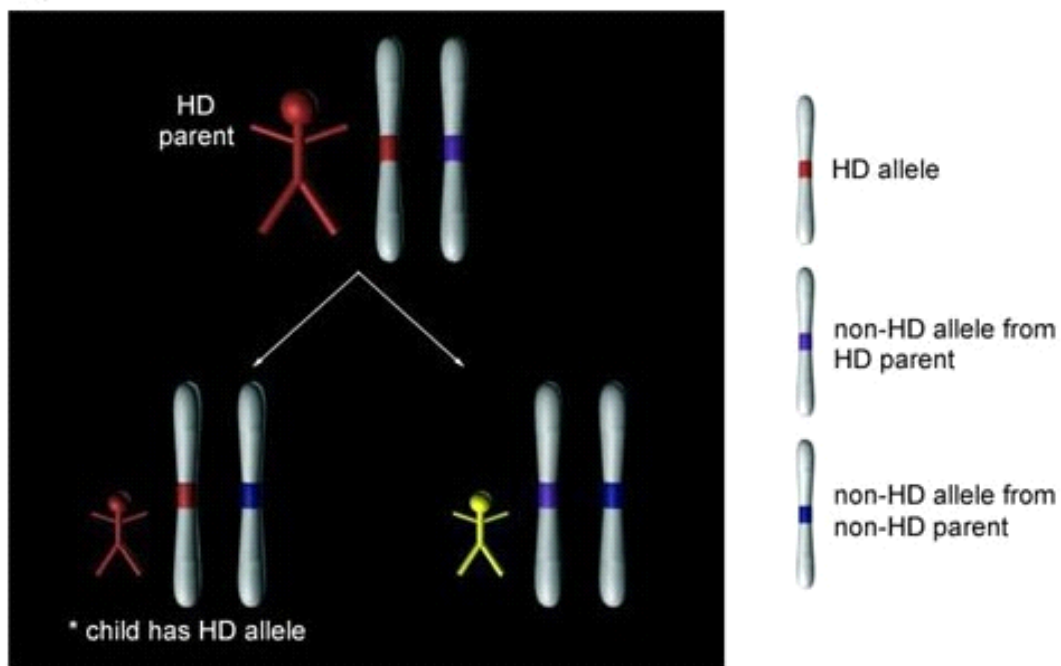
Having a parent or grandparent with HD makes life more complicated at times. You have to think about things most of your pals never have to. Being at risk doesn't mean that your life is over - it just means that you have another factor to think about when making important decisions about your life. You can still go to college, have a career, go traveling, get married - do whatever you want to do in fact!

It can be difficult to decide when to share this info with important people, and particularly when you are in a serious relationship. Maybe show them this leaflet so they can find out more about HD and what it's all about.



GENETIC TESTING

As you can see the person with HD has difficulty because now that scientists have discovered exactly what the fault is in the HD gene, it is possible to be tested to see if you have the faulty gene. **You have to be 18 to have the test** and it involves pre test counseling and giving a blood sample. Deciding to take the test is a very personal decision - some people want to know and others prefer to live with not knowing. It's up to each individual to think about what it means for them and how the knowledge of having or not having the faulty gene will affect their life. Talk through your thoughts with family and friends before making any decisions, and remember you can change your mind about it at any time, even after you've given your blood sample. Don't rush into anything until you've explored all the options and are ready to make an informed decision.



Each child has 1 in 2 chance of inheriting the non-HD allele. This is a 50% risk.

*In WA the **Predictive Testing Programme** is carried out by a group of health professionals and consists of Social workers, Geneticists, Neurologists, Psychiatrists, and Clinical Psychologists. For more information contact the **Neuroscience Unit (08) 9347 6464***

LIVING WITH HD

It can be hard when someone you love has HD. There can be lots of changes to cope with. Because of the damage to the brain, people with HD can change a lot as they become more unwell. Everyone in the family may have to do things they aren't used to doing. Most people have to give up their jobs at some stage when they become too unwell. This can have a big impact on everyone - there might not be as much money around and you may have to help out a bit more.

It is important for you to talk to people you trust about how you feel. People in your family, friends and teachers need to know what's going on so they can try and help you. The more people you have around you who understand what you're dealing with, the more help you can get. It's good to have someone to talk to!



The person with HD may live at home and need lots of looking after. You might help look after them - helping around the house, doing the cooking, looking after younger brothers and sisters, etc. This means you are a young carer - you help to care for someone in your family who is not well. It's good to help out at times, but make sure you still get time for yourself to do the things you enjoy. Even though you may worry about the person with HD when you're not there, you still need a break and some fun.



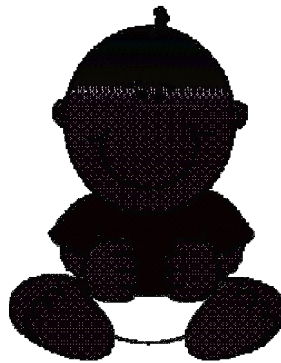
Sometimes they may go away to a special home to give you all a break and have some time-out. This is called "respite". It can be exhausting looking after someone all the time and everyone needs a break. After a while, they may move into the special home because they need looking after all the time and need specialist care. You can visit them in the home if you want to and it means they are being cared for all the time.

It's better to be open with the important people in your life as they can support you when things are rough. Keeping things secret is never easy and can make you unwell and stressed out. Even though it's difficult to talk about, you owe it to yourself to get the support you need and you might be surprised how understanding people can be! Make sure you choose a good time to bring it up - when you've got time to talk and are feeling calm. Remember though - it's up to you who you choose to tell and when. Trust your judgment - if it feels right for you, go for it!

HAVING A BABY

Later you may have to decide whether you want to have children. If you have the faulty HD gene, your children are at 50% risk of also having the gene and developing HD in the future.

Some people are prepared to take the risk in the hope that by the time their children become adults a cure will have been found or better medication will be available. Some people choose not to take that risk and decide not to have children.



There are options available these days to reduce the risks. Once a person is pregnant, the foetus can be tested to see if it has the faulty gene. This is called Prenatal Genetic Testing. There is also a procedure using In-Vitro Fertilisation (IVF) techniques that is available in some places. This is called Pre-implantation Genetic Diagnosis (PGD) and uses IVF techniques to fertilise the eggs in a laboratory and test them for the faulty gene.

Only embryos that are free of the faulty gene would then be implanted into the womb, thereby making sure the baby will be free of the faulty gene. Like all IVF procedures this can be expensive and can be a lengthy process. So there are options to consider when you're thinking of having children. Talk things through with your partner, family and friends, and speak to a member of the Predictive Testing Team to get more information so you can make the right decision for you.

HOW DOES HD AFFECT PEOPLE?

There are 3 different ways in which people are affected by HD—movements, thought and emotions and behaviour. Everyone is different and will be affected in different ways—some people have more problems with movement than others, some have more problems with behaviour.

MOVEMENT

People with HD often experience involuntary movements of their body, such as twitches and jerks. They also find it harder and harder to walk, and may fall over a lot. This is because moving our bodies requires a lot of co-ordination from the brain to move at the right speed, in the right direction and not bump into anything, and this generally goes on without us even noticing that our brain is working so hard.

Remember the basal ganglia? Well a lot of the messages whizzing about in our brain and passing through the basal ganglia involve moving our body. The basal ganglia makes sure the right message gets through to the right part of the brain to make the right part of our body move. So, when this no longer works as well and the messages are getting lost or not getting through properly, the person with HD cannot control their body as well any more.

Speech and swallowing also become difficult with HD. Again this is because these activities require a lot of co-ordination from the brain. There are people called Speech Pathologists who can help the person with HD to continue to communicate when speaking becomes too difficult. It can be hard to understand what the person is saying sometimes - be patient and persevere and don't pretend to understand if you don't. Ask them to spell the word or use different words to say what they mean.



Dieticians can provide advice and information on eating problems to make sure the person with HD is getting enough calories. It can be difficult for them to swallow, so often food needs to be liquidised. Also people with HD burn a lot of calories because of the movements they are constantly making, so they need a lot more calories to keep their weight up. Sometimes people can mistake some of these changes for other things—they might think the person with HD is drunk because of the way they walk or because their speech is slurred. Unfortunately people can jump to conclusions and you may have to put up with a lot of ignorance.

Lots of people have never heard of HD so make sure you give people the right information, show them this leaflet so they know what you're dealing with. This will help people understand and they'll be more able to give you the support you need.



THOUGHTS

The brain organises our thoughts and helps us get things done. When a person has HD, it can become harder to plan things and to remember things in the short-term. Again this goes back to the basal ganglia - it can no longer co-ordinate all the necessary information to do even simple tasks, so the affected person may find it hard to remember all the different steps of a task.

Imagine that every time you have to do something your brain runs a 'program' of how to do that task. Every task is broken down into smaller steps -just like the steps in a computer program.

EXAMPLE

Correct

- Put water in kettle
- Switch kettle on
- Get cups
- Put milk in cups
- Put teabags in pot
- Wait until kettle boiled, pour into teapot
- Wait until tea brewed, pour into cups
- Put right amount of sugar in tea

Person with HD

- put water in kettle
- forget to switch kettle on
- get cups
- forget to put milk in cups
- put teabags in pot realises kettle not turned on, switch kettle on wait until boiled, pour into pot
- wait until brewed, pour into cups
- realises there's no milk in cups
- adds milk
- wait to cool, drink tea, realises there's no sugar
- adds sugar



As you can see the person with HD has difficulty because

- They may do things in the wrong order
- It might take more steps and a longer time to do a task
- The person may get frustrated while doing the task

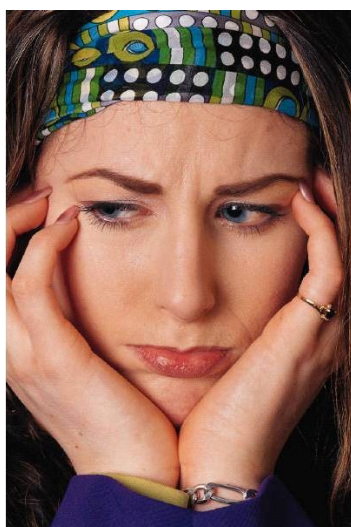
People with HD can also find it difficult to concentrate on things and find it hard to make decisions. They may become impatient and want things done immediately. It can also be difficult for them to deal with a change in plans.

Even though they may struggle to do some tasks, this doesn't mean they don't know what's going on around them. They still know who you are, and that they love you, and want to hear about how things are going with you - so keep talking!



EMOTIONS AND BEHAVIOUR

You might have noticed that the person with HD is more irritable, angry or depressed than before. They may behave differently to other people and not be as aware of social rules anymore. These changes may have been happening gradually over a long period of time. They may not be able to do things with you the way they used to. They may sometimes get angry with you and be rude to you. They may take different medicines to help control their mood and these can be very helpful



Some people with HD can get frustrated and aggressive - this may be because they cannot do things as easily as they used to. Some people find it hard to feel sympathy or understand someone else's point of view, and it can seem as if they're being selfish and self-centered.

Try to remember that it is the illness that causes these changes. IT IS NOT YOUR FAULT - or theirs. They still love you but cannot show it as well because of the changes in their brain. This can be the hardest part of HD to deal with.

Remember - HD progresses in a different way and at a different rate in each person. Everyone is unique - these are just some general ways in which HD affects people. The person in your family may only have some of these symptoms.

FEELINGS

Sometimes it can feel unfair that you have to deal with all this HD stuff when you just want to be like your friends. It can be hard to cope with all the changes HD can bring. You might feel:

ANGRY

It's okay to be angry - we all get angry sometimes. Just try to let your anger out in a safe way - punching your pillow, screaming at the top of your voice, throwing a ball against the wall as hard as you can, going for a run - whatever works for you, as long as you don't hurt yourself or any one else. It feels better when you let it out!



SAD

If you're feeling blue, let someone know - a big hug can make you feel better! Crying also helps to let go of some of those sad feelings. Having good people around when you need to talk also helps.



WORRIED

Let someone know what's on your mind. You may be worried about the person with HD, or feeling anxious about the future, or thinking you've got the gene because you were a bit clumsy today. Worries feel bigger when they just sit in your head - let them out with someone you can trust.



CONFUSED

All this information can be confusing. If you have any questions, just ask! Other people in your family might know the answer or can help you to find out. You can also contact Huntington's WA or the Neurosciences Unit to speak to someone in confidence.



EMBARRASSED

People with HD can behave differently. Other people might not understand why they do this. Try and explain to your friends what HD is - show them this leaflet. It'll help them to help you too if they understand more about it. If things are difficult at school, maybe you can show your teacher this leaflet and have a class discussion about HD, or check out <http://ourspace.org/>



GUILTY

None of this is your fault. If you need sometime out, don't feel bad about this - you need to look after yourself and have a break from HD. Explain that you need some time out for yourself - and do something that makes you feel good!



LOOKING AFTER YOURSELF

☀ It's very important to take good care of yourself when difficult things are happening.

☀ Make time for things you enjoy - hanging out with your pals, a steaming bath with loads of bubbles, shopping, going for a run, playing your computer - remember to still have fun! !

☀ Talk to people about how you feel - good friends, family, people you can trust. Don't keep your worries to yourself.

☀ If you help out a lot at home, see if there's a Young Carer's Group in your area. You can meet regularly with other young people who help to look after a family member - they won't all have HD in their family but they'll still have some similar stuff to talk about. Plus you get some time out and the chance to have some fun!

☀ If you're thinking about taking the test for HD, get all the info you can and talk it through with the people who are important to you. Take your time in making a decision - there's no rush.



FREQUENTLY ASKED QUESTIONS

These are some of the most commonly asked questions from children and young people:

1. CAN I CATCH HD?

You can't catch HD but if your mum or dad has it there is a chance you may get it when you grow up. But there's the same chance that you won't get HD at all and you may live until old age without getting it.

2. IS IT IN YOUR BRAIN OR IN YOUR BLOOD?

Huntington's disease is an illness that affects the brain. Our brain works a bit like a computer, sending messages to other parts of the body that control how we think, eat, walk and talk. If it is not working properly the messages are not clear and our body cannot work properly.

3. HOW DO I GET HD?

You might have more than one person in your family who has HD. This is because HD runs in families – it is hereditary. This means it can be passed from parents to their children.

4. CAN MY BROTHER/SISTER GET HD?

If your mum or dad has it there is a chance that your brother or sister may get it when they grow up. But there is the same chance that they might live until old age without getting Huntington's disease.

5. WHAT HAPPENS IF I DON'T HAVE IT?

When you are older you may find out that you will not have HD. This means that you will not get the disease nor can it be passed on to any children you might have. You will still have all the choices to make about your life like you did before e.g. what job to do, where to live, whether to have children or get married. It just means you will be able to make these decisions without worrying about getting HD in the future.

6. WHY MY FAMILY? WHY NOT SOMEONE ELSE?

It is natural to wonder why this illness has happened to you and your family. It can feel unfair that you have to deal with all this stuff when other people don't. You may feel many different things about all the changes HD can bring – confused, worried, angry, sad, embarrassed, alone. But you and your family are not to blame for HD and **NONE OF THIS IS YOUR FAULT**. It is a disease we still have a lot to learn about and no one knows exactly where it came from or why.

7. HOW DID THE VERY FIRST PERSON GET HD?

No one knows just where Huntington's disease comes from. Usually it is passed down from parents to their children, and in very very rare cases it can just happen to someone out of the blue (this is called a spontaneous genetic mutation).

8. WHY IS IT CALLED HUNTINGTON'S DISEASE?

The earliest writing about HD dates back as far as the sixteenth century but in 1872 a doctor from New York wrote one of the best descriptions of HD. He was called George Huntington. It used to be called Huntington's chorea because of the movements most people with HD develop. Now people know that the disease causes other changes so the name was changed to Huntington's Disease. HD for short.

9. ARE DOCTORS ABLE TO HELP A PERSON WITH HD ANYWAY?

Although doctors can't make the illness completely better, they can give medicines to make it easier for people with Huntington's Disease to walk, talk and think.

10. HOW CAN I HELP A PERSON WITH HD?

There are different ways in which you can help the person with HD. Here are some:

HELP OUT AT HOME

The person with HD may live at home and need looking after. This may mean that everyone in the family has to do things they aren't used to doing, or didn't do as much before the person with HD became unwell. You might be helping to look after them too e.g. by helping around the house, looking after younger brothers and sisters etc. This means you are a Young Carer – you help to care for someone in your family who is not well. It is good to help but make sure you get time for yourself to do things you enjoy.

If you help out a lot at home, see if there is a Young Carer's Group in your area. If you join one of these groups you will meet other young people who help to look after a family member – they won't all have HD in their family but they'll still have some similar stuff to talk about. Plus you get some time out and the chance to have some fun!

Check out this site: <http://www.youngcarers.net.au/> for more info about young carers



LOOK AFTER YOURSELF

It's very important to look after yourself when difficult things are happening. Learning how to take care of yourself can be hard and you need to learn when to ask for help or when to take some time out. See some of the other sections in this booklet.

11. WILL THE PERSON WITH HD DIE?

Once people develop HD they have it until they die. However, people do not die from HD but from other related causes, in particular many people die of pneumonia.

12. WHEN DO YOU THINK THE PERSON WITH HD WILL DIE?

Some people have lived for 30 years with the disease but most people live from 15 to 25 years after the disease begins to show.

13. WHY DOES THE PERSON WITH HD BEHAVE AND MOVE THE WAY THEY DO?

These are some of the questions children have asked:

Why do they say horrible hurtful things? Why do they move around all the time? Why do they get angry so quickly? Why do they do silly things like cross the road, when they shouldn't? Why do they sometimes ignore me when I talk to them? Why do they forget things in the past?

Because of the way HD affects the brain, people with HD can change a lot as they become more unwell. There can be lots of changes to cope with. They may not be able to do things with you the way they used to. They may sometimes get angry for what seems like no reason and be rude to you. Try to remember it is the illness that makes them do this.

HD can make people change in three main ways: physically, emotionally and mentally. But remember that everybody is different so the person you know with HD may not have changed in all of these ways.

14. WILL MY MUM/DAD BE THE SAME AS MY GRANDMA/GRANDPA OR NOT?

Huntington's disease does not affect everybody in the same way and no-one can predict exactly how it will affect somebody or when. However, there are typical ways in which HD affects people.

15. WILL THE PERSON WITH HD EVER FORGET ME?

It is unlikely that the person with HD will forget you, but when the illness gets worse it can become harder for them to communicate or remember things in the short term. And even though they may struggle to do some tasks this doesn't mean they do not know what's going on around them. They will still know who you are, and want to hear about how things are going with you!

16. I SOMETIMES THINK HORRIBLE THINGS ABOUT THE PERSON WITH HD? CAN YOU HELP ME STOP?

Even when you know that the person with HD is ill you can still feel hurt, angry, embarrassed and upset by them. It is natural to have these feelings, especially if the person with HD is very different from the way they were before. Try to remember that it is the illness that causes these changes. It is not your fault, or theirs. There are things which you can do to help you cope with your own feelings and take care of yourself.

17. SHOULD I TELL OTHER PEOPLE E.G. FRIENDS/TEACHERS ABOUT HD IN MY FAMILY?

It can be hard to know if you should tell other people about HD in the family. Most people have never heard of it. So some people will understand and some won't. It's probably better to be open with the important people in your life because they can help you when things are tough. Make sure you choose a time when you've got time to talk and are feeling calm. But remember it's up to you who you choose to tell and when. You might decide to tell a few now.

USEFUL WEBSITES

Huntington's WA
www.huntingtonswa.org.au

Huntington's Queensland
<http://www.qahda.com>

Australian Huntington's Disease Association (NSW)
<http://www.ahdansw.asn.au>

Huntington's Victoria
<http://www.huntingtonsvic.org.au>

Huntington's SA/NT
<http://www.huntingtonssa.org>

Australian Huntington's Disease Association (Tasmania)
<http://www.huntingtonsastralia.asn.au/tasmania.html>

Huntington Society of Canada
<http://www.huntingtonociety.ca/english/index.asp>

Huntington's Disease Society of America
<http://www.hdsa.org>

Huntington's Disease Association (UK)
<http://www.hda.org.uk>

Huntington's Disease Association Scotland
<http://www.hdscotland.org>

The Huntington's Disease Lighthouse
www.hdlighthouse.org

Euro HD
www.euro-hd.net

Hopes A lay persons Guide
<http://www.stanford.edu/group/hopes/>

NEED HELP?

OUR HD SPACE

Helping young people affected by HD

<http://ourhdspace.org/>

emailus@ourspace.org

or

If you want to talk to someone about living with HD in your family, or if you need more information about HD, get in touch with:

HUNTINGTON'S WA (INC)
THE NICHE, SUITE B
11 ABERDARE ROAD
NEDLANDS
WA 6009

08 9346 7599
admin@huntingtonswa.org.au
www.huntingtonswa.org.au

NEUROSCIENCES UNIT
CNR MOORO DRIVE &
JOHN XX111 AVENUE
MT CLAREMONT
WA 6010

08 93476464
neurosciences@health.wa.gov.au

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