HUNTINGTON'S DISEASE YOUTH ORGANIZATION

Being at Risk

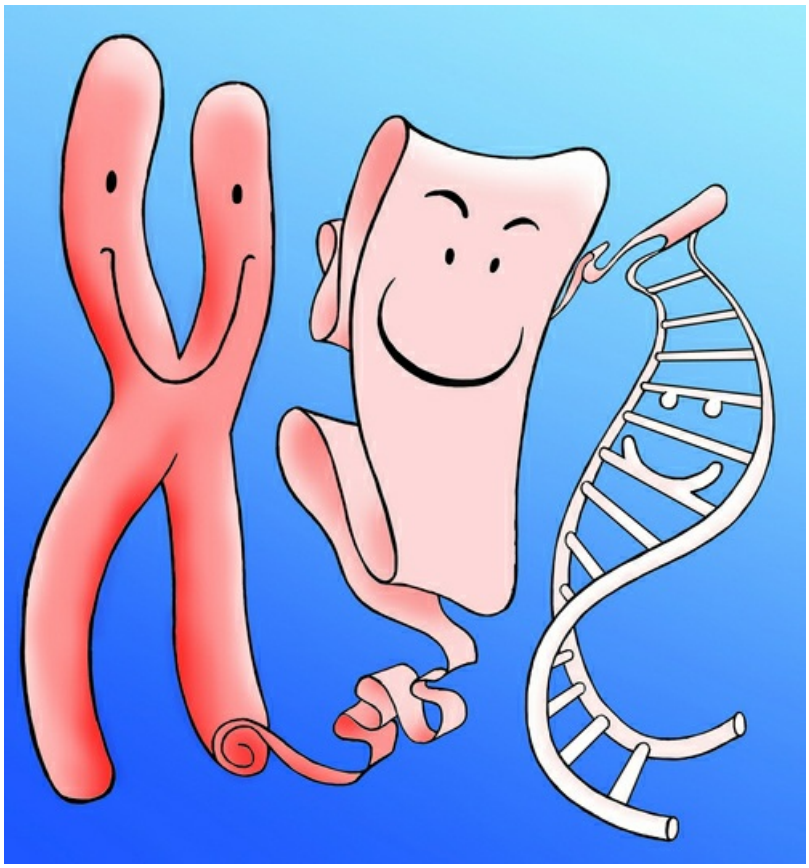
February 18, 2022

HDYO has more information about HD available for young people, parents and professionals on our site:

www.hdyo.org

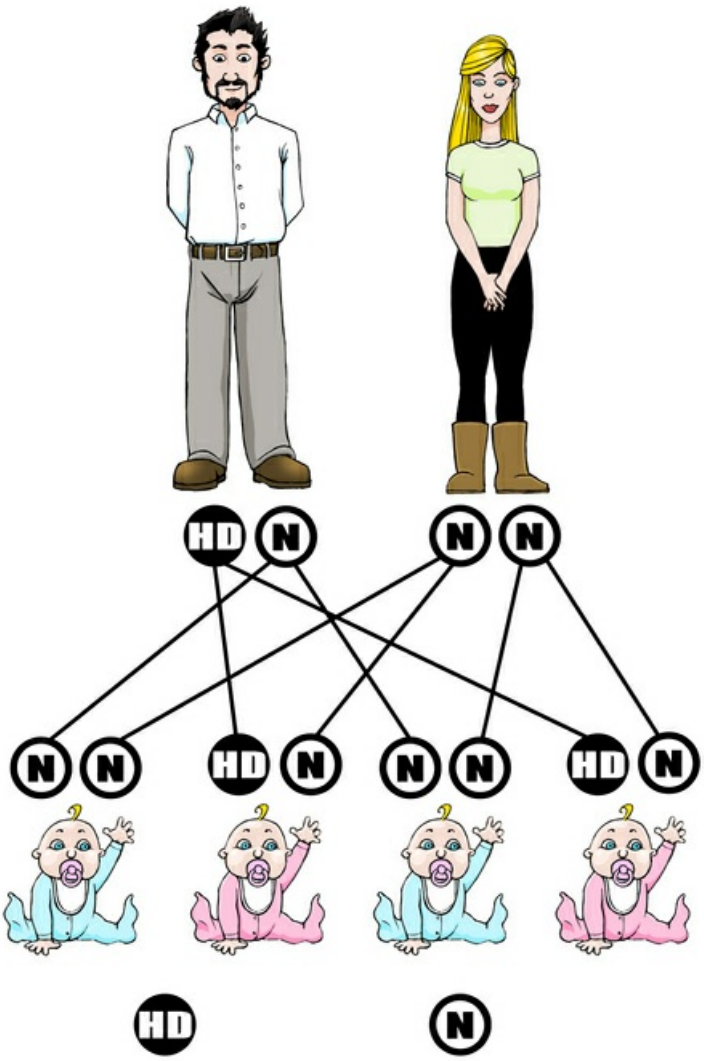
Being at risk of Huntington's disease can have a big impact on a young person's life. This section will aim to cover the worries experienced by young people with regards to being at risk, whilst providing advice and support on how to overcome these worries. But first, let's look into the science of being at risk.

What does being 'at risk' mean?



Being 'at risk' basically means that a person has a chance of inheriting the condition from their affected parent. This is because Huntington's disease is a genetic condition, so anyone who has a parent with Huntington's disease is at risk of inheriting the condition themselves. The only exceptions are those that were born through procedures like PGD and other processes explained in the having children section.

How are people at risk?



Huntington's disease is hereditary - that means that it can be passed down, from parent to child, through genes in our DNA. Genes are passed to you from your parents - that's why you might have blonde hair like your dad, or brown eyes like your mum. We have thousands of genes, and they are all sectioned into different segments in our DNA. These segments are known as chromosomes and in total we have 23 pairs of chromosomes. The reason we have 'pairs' of chromosomes is because we inherit one from each parent. For your information, the gene that causes Huntington's disease was found on chromosome number 4, by scientists from around the world in 1993.

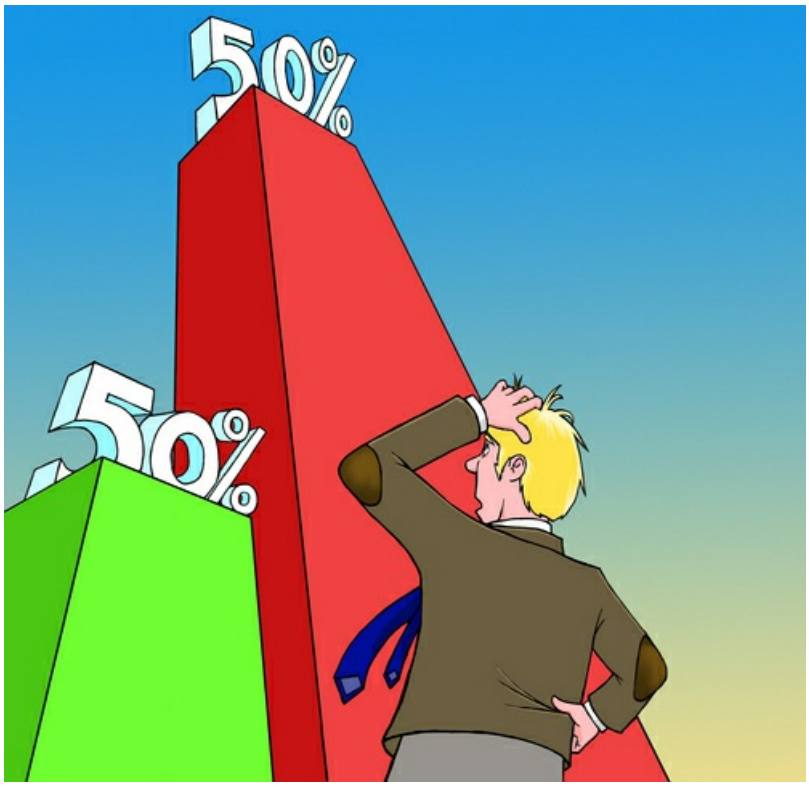
Because we inherit chromosomes from each parent, we end up with two copies of the each gene in our DNA - one from mum and the other from dad. Researchers discovered that the reason people develop Huntington's disease is because of one gene, in chromosome 4, that is longer than it should be. The fact this gene is longer means that it eventually causes symptoms of Huntington's disease. So, the gene can be described as 'expanded'. Usually a person with Huntington's disease has one expanded gene and one normal gene. A child of someone with Huntington's disease will either inherit the expanded or the normal gene from that parent - and a normal gene from the other parent. That's why there is a 50/50 chance (50% risk) of inheriting Huntington's disease. Whether a person inherits the expanded or normal gene is purely down to chance, and

that's why you may hear people compare the risk for Huntington's disease to the flip of a coin.

Passing the gene on

Unfortunately if someone does inherit the gene that causes Huntington's disease, then there is a risk of the gene being passed on to children they may have. This is a major concern for many at-risk, for quite obvious reasons. For a more detailed explanation of how people are at risk, I recommend a visit to our Huntington's disease gene section; you may also find the Having children section useful.

50/50 Odds



There are many different types of worries that young people have about the 50/50 risk associated with Huntington's disease. One common feeling amongst those at-risk is that, although they know scientifically the risk is 50%, it simply does not feel that way to them. Many people feel like the odds are stacked in favour of inheriting the expanded gene that causes Huntington's disease, rather than the normal gene that does not.

> **'Fifty per cent, no matter how often I get told "is a perfectly evenly balanced probability", seems a lot larger than it should be! I mean the 50% chance of having the (expanded) gene outweighs the 50% chance of not having it - it looms larger in my head at least. My maths teacher would kill me for saying that as it makes no sense logically.'** Luke

It is very common and understandable to feel this way. The worry can tend to feel larger than a 50/50 risk should, and the mind has a way of focusing on the worrying aspect of that 50/50 risk, rather than the more positive outlook that there is also a 50% chance of

not having Huntington's disease.

Feeling that the risk is more than 50% is not always a bad thing either, some people at-risk actually prefer to think and live as if they are going to get Huntington's disease at some point in their lives. People see it as a good way to cope - expecting the worst and hoping for the best - and also it can be seen as a good approach to living life because it can sometimes encourage people to do more with their time.

> **'Even though I am at-risk, I have always lived as if I'm gene positive and had already planned everything in my life to be done before I was 40, because I assumed I was going to get sick after that.'** Paula (who eventually tested negative)

50/50: Siblings - "One of us must have it"

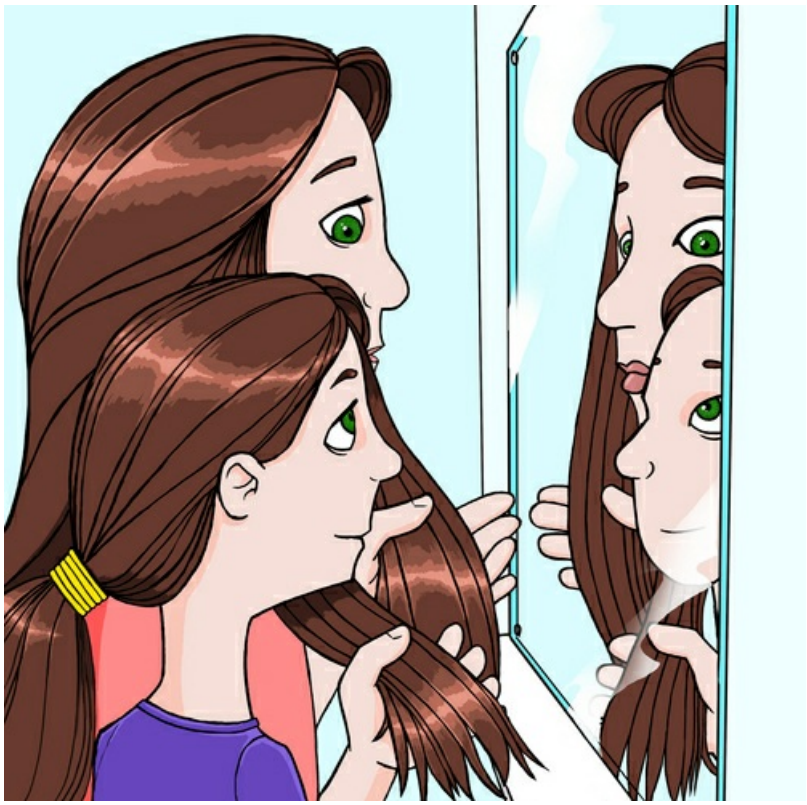
An extremely common feeling or misunderstanding among brothers and sisters at risk for Huntington's disease is that one of them **must** have it - as if it is a game of Russian roulette. But that's absolutely not the case: each individual has a separate 50/50 chance of inheriting Huntington's disease. For example, if there are 10 siblings in a family who all have a 50% risk, it does not mean that 5 of them must have the expanded Huntington's disease gene - the science does not work like that. Because each individual has their own 50/50 risk, there is absolutely no predicting how many out of those 10 siblings have or haven't inherit an expanded gene.

For some, this issue is a misunderstanding of the science for being at-risk, but for others it is more about the mind playing tricks again. Many young people who know and understand that each individual has their own 50/50 risk, find it doesn't stop them from thinking that siblings' risks must be linked.

For example, in a family with a brother and sister at-risk, if the brother decides to be tested and receives a negative result (meaning they won't get Huntington's disease) the sister may start to think that she must have inherited it instead.

As mentioned above, this is simply not the case so it's very important to remember that each person has their own separate 50/50 risk.

"You look just like your..."



Another common worry for young people at-risk is that they share the same characteristics as their family member (usually the parent or grandparent) who has Huntington's disease. This is down to the fact that the disease is genetic and people feel if, for example, they have the same hair, eye colour or features as their affected family member then they may have inherited the gene for Huntington's disease from them too.

> **'My mom has Huntington's disease and people used to say how much I looked like her and that I had my mom's hair. I used to smile but inside it would really worry me because, as much as I love her, I did not want to inherit everything from my mom!'** Tiffany

Fortunately, genetics does not work like this. Basically, each gene is passed down separately. Someone at risk could look, sound and behave exactly like their affected parent in every way - it doesn't matter: that person's chance of having inherited the gene that causes Huntington's disease is still 50/50.

The family 'script'

Some young people live in a family where there may appear to be a pattern to how the disease is inherited. For example, it may be the case that in the last few generations, in one particular family, they may have had only the women in the family inherit the disease, or the eldest siblings etc. This can lead people to believe that the pattern is certain to continue in exactly the same way for their generation of the family.

> **'I'm scared because it always seems to be the youngest sibling that gets Huntington's disease in my family. My mum was the youngest of her siblings, and so was my granddad. I'm scared because I'm the youngest sibling in my**

generation so I could get it too.' Leanne

This is an understandable conclusion and worry for a young person, but it's simply not true. No family has a 'pattern' or 'script' for how people inherit Huntington's disease. It is just coincidence that the family tree has worked out that way. Remember, each individual has a 50/50 risk - family patterns have no influence at all.

Predictive Testing

Another issue for those at risk is the decision to test or not. Since 1993, people at risk for Huntington's disease have been able to be tested to see whether they have the expanded gene that causes Huntington's disease.

The decision to test is down to the individual who is at-risk, and them alone. But making that decision either way is not often an easy one. Only about one in five people at risk decide to be tested. The majority either don't want to test, or remain undecided.

Spending time thinking about whether to test or not generates a lot of tension and worries for people at risk. Some worry about whether to test at all, or when the right time to test is; others about how they would react, how the family would take the news and whether they would be discriminated against if they decided to be tested.

'I've known I was at-risk for a few years now and I agonise over the decision of whether to test or not. It is such a difficult choice to make and there seems to be so much to consider when testing. It does cause quite a lot of worry on top of the worry I feel because of being at-risk.' Jacob

There are many issues associated with testing for those that are at-risk. If you would like to learn more, you'll find them in the Genetic Testing section.

Symptom Hunting

Perhaps the most common and biggest worry for those people at-risk comes in the form of 'symptom hunting'. Symptom hunting is when people at risk for Huntington's disease find they're searching for symptoms of the disease in themselves.

Usually this happens when someone trips or falls, is clumsy, or knocks something over. People immediately worry that this incident may be the involuntary movements associated with Huntington's disease. Many people will have seen their family member(s) with Huntington's disease drop things, break items or lose their balance. Because of this, when they themselves trip or break something, it is hard for them not to think and worry about the similarities between the two instances.

'Every time I dropped a cup, or my fingers moved on the steering wheel, or I tripped on a step I'd be thinking "oh my God, they are the early signs of Huntington's disease". I didn't know what it felt like to think, "I'm having a

bad day" or, "That step got in the way!", it was a classic case of symptom hunting and it was quite a regular worry for me.' Naomi

Almost everybody at risk symptom hunts - even some people who have had a negative genetic test, meaning they won't get Huntington's disease, still do it. Once again, it is very much an issue of the mind playing tricks and creating more worries than necessary.

'I used to symptom hunt all the time and I would end up getting really worried. But now I've almost grown tired of worrying all the time about something I have no control over. I figure that I cannot possibly tell if I have symptoms or not so I just think to myself "why bother to worry about it".' Jeff

It can be almost impossible to avoid the worry of symptom hunting, but attempting to take a similar approach to symptom hunting as Jeff does may help. Symptom hunting tends to come and go, and your mood can play an important role in how you cope. If you can find the ability to shrug or laugh off any trips, falls, spillages or breakages then you will spend far less time worrying about them in general.

Here's the thing: most people who are getting symptoms of Huntington's disease are actually not aware of changes in their movements or balance, because the disease causes loss of insight. So, if you're worrying about developing symptoms, it actually means it's very unlikely that you are.

Being at risk: The impact on life

Being at risk, and the worry that comes with it, can also have an impact on people's personal lives, whether that be educationally, career-wise or relationships. If you watched the video clip above you would have heard Raphael talking about how he feels being at-risk 'blocks' him career-wise. Raphael doesn't say whether this is due to his own feelings and worries about being at-risk, or the views of others that he may work with, but this is not an unusual feeling for somebody at risk to have.

'I feel like I have no time to build a career, I feel like I must achieve everything straight away before I get old and perhaps become ill. But then I end up feeling that I can't achieve my goals in that timeframe and what's the point? ' Emma

Another point that Raphael makes is that he does not want to 'waste time' because he may get ill when he is older. This is another classic example of the impact being at-risk has on the way people think and plan their lives. That sense of 'wasting time' is something that people at-risk may be able to relate to with regards to their own lives. Some people may feel they are wasting time doing the job they currently do or the university course they are enrolled in. Anybody can feel this way, but being at-risk can really make it seem vital not to be wasting time and as a result people tend to second guess the decisions they make in their lives.

> **'I was working at a job which was completely boring, I hated it. I used to look at the clock in the office and think to myself "I am wasting my life away here, I may get ill and still be stuck in this place".'** George

The impact of being at-risk on people's lives can be very difficult to cope with. But the impact, although difficult, does not have to be a negative one. Rather than letting the fact you are at-risk hold you back or bring you down, it can be used to encourage yourself to achieve what you want to achieve in life, whatever that may be.

> **'I really did not like the job I was in; I told myself that I needed to change things. So I started doing a part-time degree in a course that actually interested me. Eventually I left my job, got my degree, went travelling around North America for a few months, came home and started a new job that I am far happier doing. I feel like I'm living the way I want to live now and I don't look at the clock in the office as much as I used to!'** George (again)

George used the fact he was at-risk to encourage himself to achieve the things he wanted to achieve. Sometimes, achieving what you want to achieve can mean making a lot of changes in life, or being brave and determined about what you want to do in life. The point is that being at-risk does not always have to be a 'dark cloud' hanging over people - it can be the encouragement to go on in life and achieve the things you want to achieve.

Support

Being at risk, as has been highlighted, causes a lot of worries and concerns for young people. So it is important that young people at risk are supported by those around them. As a family, the best way to cope with Huntington's disease is to discuss it freely and be open, so that young people feel comfortable and know that they can talk about any worries they have with the family.

If open dialogue about Huntington's disease is missing in the family, young people can end up feeling very isolated with their concerns. Often, just being able to talk about their worries is a great relief for them.

As a young person, your family should always be the first place to turn to when you want to discuss how you feel with regards to Huntington's disease. But if you don't feel that you can talk to your family about Huntington's disease or you don't want to talk to them, then you may find support through HDYO on our Facebook Group. There, you will find many young people going through similar concerns who will be able to share experiences. Talking to others in similar situations can be a great support.

You may also be able to find support through your national Huntington's disease organisation, which may run youth camps, support groups, or young adult conferences. Not all organisations will be able to provide support specifically for young people, but it is certainly worth asking them what they have to offer for you. However, if you do feel you are struggling with being at-risk (quite a normal feeling) and would like professional support, then contact HDYO and we will be able to direct you to those that can help.

Staying informed, getting involved

Learning about Huntington's disease is a great way to help come to terms with being at risk. By finding the HDYO website, you've already taken a big step, because knowledge is power. Another thing a lot of people find helpful is to remember that scientists around the world are working 24 hours a day to develop treatments that will prevent the disease or slow it down.

Some people devote time to fundraising for Huntington's disease research and care; some take part in scientific research projects that need at-risk volunteers; others find that reading about the latest research on sites like HDBuzz gives them enough information to carry on with their lives, knowing that progress is being made. Being involved in fundraising, research or both can be a powerful way to turn round, face the disease eye-to-eye and take a stand against it.

There's no right or wrong way to spend your life if you're at risk, and it's perfectly OK to not want to think about Huntington's disease at all. But getting involved has proven to be beneficial for many young people and the more people that do get involved in fundraising and research, the closer those treatments will become.

Accepting the risk

No-one wants to be at risk for a disease, but facing that risk and accepting it will help you move forwards in the long run. It is never easy accepting such a risk, and it will take some time, and effort to come to terms with it. But through educating yourself on the disease, finding a support network that is there for you, and understanding that this risk does not stop you from achieving the things you want to achieve in life, you can learn to

accept, and even embrace, the risk of Huntington's disease.
