Huntington's
The naked truth

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HUNTINGTON'S
THE NAKED TRUTH

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The information presented here is for educational purposes only. It is not to be taken as medical advice. The opinions and beliefs expressed are those of the author. People with medical problems or questions should consult a health professional.

Special dedication

To my wonderful late wife, Jan, for her courageous attitude in the face of many adversities. This courage was a demonstration of the kind of inner strength that very few of us possess. Jan's easy-going nature and acceptance of a life that would have driven most of us to total exasperation, was amazing. She has given all those who knew her the inspiration to face whatever may occur in their own lives, without complaint.

Sadly, Jan passed away on 3 May 1997. She remained defiantly independent to the end, and died without suffering. God bless you, Jan.

Acknowledgments

Thank you to all members of the Gene Family support group. This book could never have come about without the support of its wonderful members. If it hadn't been for their desire and real need for such a book, I would never have been able to achieve this major goal.

Special thanks go to the affected families who shared their lives with our readers.

Thanks also to the Genetic Centre at West Newcastle Hospital, for advice on the clinical information contained in this book.

For further information on Huntington's Disease, please contact the Huntington's Disease Association, or the Carers Association nearest to you. Offices are located in all capital cities. Check your local telephone book for location details.

Also check your telephone book to find out about any additional support groups that may already be in operation in your local area. We urge you to seek out your nearest support group for people with disabilities, and their carers, to gain the benefits of fellowship with members of these groups.

Foreword

Lack of information

Despite major breakthroughs in researching the defective Huntington's gene, there is still a culture of silence and ignorance from those who should be able to provide information about this frightening and cruel genetic disease. Many health professionals—doctors, nurses, social workers, even large support groups—do not provide enough written material and knowledge, on just what lies ahead for a family affected by Huntington's Disease. This is the first intention of this book: to help fill the gap in material on the disease for the benefit of sufferers and their families, carers, and other associated health professionals. The overall aim of this book is to provide personal accounts, as well as information, on the genetic basis of the disease, the expected course it can take, and the kinds of resources that are available to help people through each stage.

The most critical time in the lives of any Huntington's affected family is when the disease is originally diagnosed. So, it is imperative that up-to-date and comprehensive information is available to support each new diagnosis. Huntington's families need to know the prognosis for any symptoms and challenges, as well as the kind of support that can be

expected over the course of the disease. They also need to be reassured that there is usually a fairly lengthy period of time between diagnosis and extensive debilitation.

Being diagnosed with a common cold is simple in that some form of relief can immediately be prescribed, but with an illness like Huntington's, doctors and specialists seem ill-equipped to provide even the most basic accurate information. For someone newly diagnosed with the disease, knowledgeable reassurance is required immediately.

I have been told so many incorrect things about Huntington's that I have made a list of the more common misconceptions. Here are three of the most blatantly incorrect pieces of information that I have been served by health professionals:

~ only males get it;

~ it only appears in every third generation;

~ age of onset is *always* in the late 50's.

Surely it is the responsibility of health professionals to read widely, and remain informed about new developments in as many illnesses as possible, most especially those that are life-threatening.

This ignorance on the part of the medical profession may in part be due to a culture of superficiality, where general nurses and practitioners are content to be only generally versed in the vast number of illnesses—and even unversed in the less common ones. If the patient needs to know about anything that is 'different' to the usual kinds of illnesses that

are seen by the family doctor, then often the only choice is to consult an expensive specialist. However, this is not so easy with Huntington's—how many of these specialists exist? Even social workers are given only an overview of a wide number of different diseases, and the onus is on them if they want to know more.

Huntington's education should begin at university for prospective doctors and nurses. If they are given the latest, information, instead of over-simplified variations on George Huntington's nineteenth century interpretation, then those who are 'at risk' of being diagnosed with Huntington's, and those who have been diagnosed, can be given a better understanding of the likely course of the disease.

For sufferers and their carers

The idyllic lifestyle of a happy family at home, participating in each family member's normal growth and development, is lost to those families that are affected by Huntington's Disease. However, having Huntington's does not mean that your life ceases from that moment on. Quite the contrary; your life begins anew!

The first part of acceptance is the hardest. That is, to face the fact that the long-time dream of most couples—to graciously grow old together and be there to watch the grandchildren grow up—will not become a reality. Life has taken a major turn, and

your own mortality has never been more evident. In fact, someone has told you that there *will* be an end to your life.

There is hope, however, inasmuch as you can now plan for a new and more fruitful immediate future, because you have a sense of certainty about the path of your long-term future. Once diagnosed with Huntington's, you are adding the one thing to life that we usually only achieve with retirement, that is: *quality,* if not quantity, of life.

Huntington's, in most cases (unless it is juvenile Huntington's), progresses from onset to outcome over a period of between fifteen and twenty-five years. Why then, do so many people who have just been diagnosed, begin to wind down their lives and wait for the inevitable to happen? It always saddens me to hear of those affected who give up all hope of a normal existence to cocoon themselves within a walled environment of submission or denial. The reason for this seclusion is simple: sufferers are not told when diagnosed that they can lead a normal and productive life for many years, before Huntington's takes its toll.

Putting the quality back into life

No-one knows how long their life will be, or how it may end. Often as a result of this, we forget to include *quality* in our lives. This one element of life is frequently forgotten as we struggle each day to make

financial ends meet, and fulfil our commitments. Fortune-tellers make a lot of money from people who want to know what lies ahead; all sufferers affected by Huntington's know what lies ahead. Of course, the price they pay for this knowledge is the loss of years of life, as well as varying amounts of sickness for the time remaining. Yet, all knowledge provides a kind of freedom—in this case it is the freedom we can give ourselves to plan that new life we've always dreamed of, by maximising the time left to us.

Being diagnosed with Huntington's, or with any long-term illness, is a way of making us take stock of our present lives, and appreciate every moment from that point on. During the course of my late wife Jan's illness, we travelled through many parts of New South Wales, and even to the USA. Most of these trips would never have occurred were it not that she was ill—though her acceptance of the disease also helped us achieve many of our goals. This attitude was an inspiration for many people.

Not everyone can be so philosophical when it comes to facing their own mortality. This is despite the obvious fact that every person who is now alive is in fact slowly dying, anyway. However, this slow death occurs without us giving any real thought to 'meeting our Maker'. A wise doctor once told me: 'Life is a disease. It begins at birth and is only curable by death, so make every moment you have in-between count for something.'

Today is the most important day of our lives

because no-one knows for certain what the future holds, or even if we have a future. Life is not over because you have Huntington's, it is merely changing form. It is my aim to try and help you understand not only the clinical facts about Huntington's Disease, but to inspire you to see that each and every day can still be special, often in spite of, or even because of, Huntington's Disease. I will try to show you some ways to achieve peace within yourself, and find love in understanding the feelings of those around you. I will also relate some of the real life situations of sufferers and carers who are going through what you are probably already experiencing yourself. It can be uplifting and refreshing to know that you are not alone.

Facing a future without knowing how it is going to turn out is an accepted way of living our lives. Facing a future with the knowledge of what does lay ahead can instill a new, even positive, approach to life. I hope this book will not only give you a necessary understanding of Huntington's, but introduce you to a whole new way of living that will assist you in handling the complex, but wondrous, future that lays ahead.

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Jan and Peter: our story

Every member of a Huntington's diagnosed family is affected physically, mentally and emotionally. There is not a day that passes when they are able to escape from Huntington's. It lurks within every corner of the mind. It torments you by bringing out a kaleidoscope of cataclysmic emotions that other families may only ever experience once or twice in a lifetime. That is because these emotions are the kind that you usually only experience when tragedy strikes those you love.

Lifelong grieving is the sentence administered to anyone born into a Huntington's affected family. A similar sentence is passed on to anyone who falls in love with a Huntington's diagnosed person: they get to see someone die slowly, cruelly, without having any recourse but to stand by and watch. Even the friends of sufferers are admitted to this club of grieving that will continue throughout the life of the sufferer, and on through the lives of their children. It is too much for most to bear—obviously, not everyone is prepared to stand by and watch.

This is the cold, harsh reality of Huntington's Disease. Despite this, many families cover this gloom quite well, and remain fairly positive in the light of an ongoing and debilitating illness. However, the gloom is never really far from the surface. I know,

because I married a Huntington's sufferer, who had already been diagnosed. It is important that I tell this story, so you know that when I give advice I am doing so from experience, and out of love and compassion for other Huntington's sufferers.

When my late wife was diagnosed with Huntington's, her world turned upside down around her. Her marriage broke up. Jan's two teenage boys rebelled; only her daughter was too young to express how she felt. Jan took it all in her stride and was always there for her boys when they got into trouble, and cared wonderfully for her daughter. Then one day we met, and fell in love. Both of our lives changed direction instantly, and nothing was ever going to be the same again for either of us.

How we met

I was living in a mobile home park when my employer happened to mention something about another one of the residents, a Jeanette. Something about the fact that she was a single mother trying to look after a young daughter, whilst being ill, struck a chord with me. I kept asking questions about this lady, Jan, that my boss did not know the answers to.

On the Friday night of that week I had dinner with some friends. One of these friends mentioned that she had done a home competency assessment in the mobile home park where I lived—I immediately wondered if she was talking about Jan. My interest in Jan surprised my friends, but I did get some more

information about her. I discovered that her doctor was now trying to find a position for her in a nursing home—the assumption was that Jan could no longer look after herself, or her nine-year-old daughter. I decided that I wanted to meet this wonderful lady who was struggling with so many insurmountable problems.

The following day, Saturday, we finally met, rather romantically and by accident, in a phone booth. I went down to ring a friend, but there was a woman already using the phone, apparently with little success. Her car, which was parked nearby, contained both her young daughter and a white poodle, the latter of which was jumping around. I realised that this was the woman I had heard so much about. Giving up on her phone call, the woman left the booth, and telling me to go on ahead, said she would try again later. I replied, 'Thank you, Jeanette'. This surprised her, and she looked at me and asked whether we knew each other. Then everything I knew about her tumbled out, and from that moment on there were very few occasions that caused us to be apart. That very night we decided on marriage as the only reasonable option for our future. After finalising a divorce from her estranged husband, and the father of her three children, we tied the knot. This was nine months after we had met.

Her family knew her as Jeanette. Her friends called her Nettie, and to start a new life together she asked me to call her Jan. No matter what people called her,

they all loved her for her happy nature and ever-present smile. That smile could brighten up the darkest moment in anyone's life, and give joy to all that received it. Jan's genuine concern for others was always bigger than her own personal situation. Not once did she fall into self-pity: her way of dealing with Huntington's was to see it as a big 'bug', this way we all seemed to accept the 'bug' too, and manage to get on with life's smaller challenges.

Jan had endured an abusive marriage, and this had caused her to become alienated from her two boys, from the time they were in their teens. They no longer lived at home, and had constant brushes with the law as a result of their ongoing emotional turmoil. Rejected by their father, and blaming their mother for putting them at risk of Huntington's Disease, they naturally rebelled against family and society. Happily, they have now settled down to steady lifestyles.

In trying to understand exactly what I had accepted in marrying Jan, I began the long search for background information on the disease. This was in 1988, though it may as well have been the Dark Ages. No-one wanted to give specific technical information, or even provide descriptions for the likely course of Huntington's for the sufferer. The only relevant information available seemed to come from a book written in the sixties by a university lecturer, from a Huntington's affected family, which gave some insight into the various problems associated with the

disease. Sadly, Jan never actually fit into most of the examples presented in that book. It was not until a few years later that doctors acknowledged that the way in which Huntington's affects an individual is also individual.

When Jan and I met I was working irregular hours as a furniture removalist. This gave me some degree of freedom to spend more quality time with Jan and her daughter. Then I had a heart attack and had to quit the heavy lifting. Whilst recuperating, we all went for mid-week and weekend trips to show Jan the beautiful country towns of New South Wales. These first trips marked the start of a time-out pattern that was to continue for a number of years.

Also around this time, I started a small computer business—hoping for a new work avenue. Sadly, I chose the wrong partners, and the venture was doomed to fail. This brought me back to the core of my life again, my relationship with Jan. After much soul searching I found I had to come to terms with the issue of mortality. To help me do this I sought employment in the funeral industry; however, another heart attack shortened this job to only eight months. Fortunately this provided time enough to confront many of my fears, which were thus allayed.

In 1990 I went on a Carer's pension to take care of Jan full-time, as her physical condition was deteriorating rapidly. Continuing to put quality into our lives (in the way of trips) cost us both our life savings, but if I had to do it over again I would,

without a moment's hesitation. Each trip we took gave Jan a thrill, as she loved travelling, discovering new places, sharing new experiences; and watching her daughter grow into a very mature, independent young lady.

As the years passed away so too did our friends, or more to the point, Jan's friends. They put on their displays of affection and caring in the early stages, but then slowly the invitations became less frequent, until they completely stopped. Others were more blunt as they confessed they could not stand watching Jan decline. You know the saddest part of this was that they denied themselves the chance of seeing Jan's true strength of character. Only one lifetime friend and her family stuck with Jan throughout—many bumps in our lives were smoothed out by their love and support.

One of the special qualities that Jan possessed was her appreciation of any gift she received. Each and every card, piece of wrapping paper and gift was precious to Jan. If we knew a friend or relative was coming to visit, Jan made sure that whatever she had received from them was either on show, used in the course of the visit, or worn, as the case may be, to show them how much she thought of them. Once a neighbour gave her a bunch of flowers from her own garden, and Jan expressed her appreciation a thousand times over for the thoughtfulness and love shown her. I still have some of the old, empty boxes of chocolate given to her over the years.

'They say I have Huntington's. It worries them more than it does me.' This was Jan's favourite line, and it stunned many of the people who heard it. Jan always turned the conversation around to the other person when she perceived that they were uncomfortable in her presence. Their problems were always more important to Jan than her own: she always managed to encourage anyone with an uplifting smile that seemed so fixed, that I doubt even dynamite could have removed it.

Support

In the early 90's, we found out through a social worker that there was a Huntington's support group in Newcastle (about a hundred kilometres north of our home). From the very first time we attended one of their regular meetings we could feel the love and acceptance from the ten or so families in the room. It was almost a tangible kind of love, which immediately made us relax and enjoy the company of those present. The trepidation we had felt at the prospect of meeting people with more advanced Huntington's disappeared as we entered the meeting room.

Sharing our problems with others in the same boat provided us with new ways to deal with the day-to-day problems we faced. The little things we did to help us cope were shared with the group, as were the little things that others did to better cope. These little droplets of shared knowledge were revelatory for all members of the group. Anything that could help us

cope with the bad times, and enjoy more fully the good times, was to be welcomed. It wasn't long before I was busy producing a regular newsletter discussing Huntington's Disease and any related issues and problems—in fact anything that could affect us all. The Gene Family Huntington's Support Group was born.

Though we tried to live life to the full, and accept the presence of Jan's 'bug', her physical decline became even more restricting. Despite a decided lack in the particular symptom of chorea* of the arms and legs, Jan did experience increased rigidity in her limbs. This began to present problems in walking even short distances, and ended our much-loved short trips. The number of falls Jan suffered increased, and as a result, so too did her propensity for injury. Thank God this only resulted in stitches twice in ten years.

Jan's sleep pattern gradually changed over the first few years that we were together. By 1991 it was set in concrete. She would go to bed at two or three in the morning, and rise at nine. If we tried to get her to go to bed earlier then she would obstinately slow down any task she was engaged in, until the time she wanted to go to bed. Though I did not particularly like the hours she kept, it was impossible not to love and appreciate her stubborn streak.

One day we had been arguing for about four hours and our nerves were strung right out. Jan's daughter

* *Chorea* – uncontrollable, irregular and purposeless movements.

came home from school and innocently asked what we were arguing about. Then the foolishness of the situation became apparent to both of us, as we pointed to a biscuit crumb on the coffee table. From that day on I was very aware of the many varied and insignificant potential triggers for arguments. I made it a point to refer back to our 'biscuit crumb'. This saved us many unhappy and pointless hours of dispute.

Jan, bless her independent spirit, would insist on washing the dishes or cleaning up the kitchen whilst she thought she could manage. Unfortunately, it was not done to a satisfactory standard and irked my sensibilities. However, I learnt from the biscuit crumb and tackled things in a more peaceable way. After Jan had gone to bed at night I would wait for her to fall asleep, then go and redo the kitchen and wash the dishes again. We were both stubborn, but this way we could both be happy and in control.

Eventually, the lack of sleep wore me down. I had to seek help from service providers to look after Jan from 10 p.m. till bedtime, so I could catch up on some sleep. Jan, in her own inimitable way, loved the company and insisted I stay up to help entertain them. Only after they had left would she go to bed.

The on-going lack of sleep and heavy workload took its toll on me, and our doctor suggested regular respite care. This we did for several years, until 1996. One day, when Jan was in a nursing home, we were outside her room talking and joking as usual—then

everything went black. When I came around I had suffered a slight stroke, which affected my left side and deadened my left foot. Ironically, it was my stroke that caused Jan to be admitted to a nursing home full-time, and not her Huntington's Disease.

Two months later I suffered another slight stroke, and many of the things that I used to be able to do became impossible. The one thing I dearly wished to do was care competently for Jan, and this was denied me. I worked hard to overcome the problems associated with the strokes so that I would be able to take Jan back home to care for her. Sadly, before I could do this, Jan's beautiful, big heart gave out without warning, on 3 May 1997.

Two days prior to her passing I purchased a wheelchair for Jan, as her falls were increasing in number. She was so fiercely independent that she only sat in that chair once, the day before she died. I later realised that she only made this concession to please me, for the nurses told me that Jan got out of the chair immediately after I had gone, and refused to sit in it again. She could not accept all the things that the use of a wheelchair symbolised.

For Jan, the morning of her passing went as usual. She had breakfast, and then a casual shower with the assistance of a nurse. She was combing her hair when the nurse was called out for a moment. In the space of that moment, Jan collapsed and died. The ever-present smile remained on her face, a sign of her lovely, unconquerable nature.

When Jan was only nineteen, her mother passed away as a result of complications associated with Huntington's Disease. During our years together we would often drive to the Northern Suburbs Cemetery to visit her grave, and Jan would express a wish to be buried alongside her mother. This wish was granted, and now two wonderful people share in death the love they shared in life.

I consider myself very blessed to have been a part of Jan's life. I have been lucky to have shared the joys, hardships and heartbreaks with her. I have also had the privilege of watching and learning from her ability to cope with emotional, physical and environmental problems. Jan, with her wonderful hunger for life, taught me to cope with any problems in my own life, as well as with the thought of what may lay ahead.

It is a much sadder world without Jan, but no-one who knew her could ever wish her back to suffer the loss of her independence to Huntington's. She died before this could happen, and I will always be grateful for it. Jan may not be with us physically, but the memory of her smile still inspires all those who knew her. The Gene Family Huntington's Support Group members will always be encouraged by the memory of her independent attitude to life. Jan has inspired many Huntington's sufferers and their families to face their own futures with a smile and confident dignity. This is her legacy to others.

Living with Huntington's Disease—sufferers' stories

Sharing with other Huntington's families

Huntington's, as is the case with many long-term illnesses, has an unkind way of isolating families, friends and individuals. By sharing real experiences with you, the reader, I hope to show that isolation due to Huntington's can be overcome, as can most obstacles in life. Isolation can only exist when one is completely alone, but now that you have started to read this book, you can never be completely alone again. You have become part of the Huntington's group of families, and together we are one united family.

A commonly held belief is that Huntington's Disease is a rare condition. This is not true. Contact your local doctor, service provider or hospital and find out how many other families with Huntington's are in your area. Ask them to pass on your name and phone number or address to those families. Reach out and share with them and you shall find, as many of the sufferers in the stories have, the benefits of understanding and the love of fellowship.

The following case histories are related without reference to gender, to protect those persons who were good enough to share their personal experience. There is a mixture of hope and desperation in each of these cases, which exemplifies the myriad of emotions

that are typically experienced by a Huntington's sufferer. Although all cases have similarities, none are exactly alike. This is what makes it so hard for sufferers: they have to adapt to an individual type of illness. In many ways it would be a relief if Huntington's were predictable in its progression through the distinctive stages (explained later in the book). In reality, a sufferer will often show signs of progressing into the latter stages one day, then the next be back to the previous stage again.

Such fluctuations can be most upsetting for everybody concerned, and extremely hard to adapt to. It also makes it hard for a family to accurately explain a sufferer's condition, even to their own family doctor. Within families children will have different reactions to an affected parent's health, as they may imagine themselves looking at reflections of their own futures. Special counselling is essential for sufferers' children at such times. Sadly, Huntington's trained social workers are rarely available due to the wide dispersion of Huntington's families, and there are times when help is required, but not available. Families and professionals have to 'fly blind' at such times.

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