# Growing up with a Parent with Huntington's Disease:

# A Qualitative Study of Adversity, Conversations and Social Support

Submitted to the Department of Psychology

Faculty of Social Sciences

University of Oslo

for the degree of Doctor of Philosophy

by

Siri Hagen Kjølaas

Centre for Rare Disorders

Oslo University Hospital HF

October 2022

**Acknowledgements**

First, I would like to thank all the participants who so kindly shared their stories as part of this research. There would have been no study without you. The expertise you have in your own lives taught me so many important lessons about human strength, hope and optimism. I will take these lessons with me throughout my own life.

I also want to express my sincere gratitude to my supervisors. To my main supervisor, co-author and project manager, Kristin B. Feragen – I am forever grateful for your trust in me with handling this knowledge and for always being willing to share your invaluable expertise at every step of the way. You have been patient, wise and critical, in all the right ways to help me through. I have learned so much from you through this experience, for which I am very thankful.

To my co-supervisor and co-author Tine K. Jensen – thank you for giving me the right amount of patience, space and guidance to develop and learn at my own pace. You have gone out of your way to be of assistance when I would frequently be stuck. Most importantly, thank you for the invaluable expertise, insight and understanding you brought into this project. It has made all the difference to me, and to this research.

I also want to collectively thank my colleagues at the Centre for Rare Disorders, and our leader, Heidi Glosli. This research began with the desire to help young people in families with Huntington's Disease and the dedication of those who work with these families at the Centre for Rare Disorders. In particular, Gunvor A. Ruud – thank you for your patient guidance, for sharing your invaluable knowledge so willingly, and especially for being a good friend and mentor throughout my journey. Special thanks also needs to go to my PhD colleagues, Line Mediå, Anita Myhre Ramborg and Deniz Zelihic, for their friendship, discussions and support.

To the Norwegian Association for Huntington's Disease – thank you for the guidance and encouragement through this process; your efforts are always kind, inclusive and wise for those who need help and support from someone who understands. Any young person or family affected by HD is fortunate to have you as support in their lives. A special thanks must also go to the project manager, Ane Mygland. I am truly grateful for the inclusion, guidance and understanding you have shared with me through this process. May your story of strength, availability, persistence and hope be modelled by other families with HD in the future.

Several institutions must also be thanked – to Stiftelsen Dam, for granting me the financial support to do this research, to the Department of Psychology at the University of Oslo for accepting my enrolment with this project, and to Scribendi for academic proofreading assistance along the way.

I am fortunate to have had the love and support of my family throughout my life in general, and it has been invaluable through this process. I especially want to thank my parents for the unwavering support they always provide. You have always work to accommodate the unconventional ideas I have about where I want to go and what I want to accomplish. I could not have asked for better parents and supporters in life. From the bottom of my heart, thank you.

To my Australian family – this journey started with you and would not have taken place had I not been so fortunate to have you in my life. Although our paths no longer cross, our friendship has continued to be a guidance through every step of the way.

Last, but not least, to Finn, thank you for your love and patience during the entire processes, for the phone calls when I needed support and for letting me have my space to finish this thesis.

Siri Hagen Kjølaas

Oslo, October 2022

**Summary**

The aim of this thesis was to investigate the current and past experiences of young people who grew up with a parent with the progressive, inheritable, neurodegenerative condition Huntington's Disease (HD). Past research has identified a range of factors that can create challenges in these young people's lives, but they had not provided a sufficient understanding of the potentially dynamic nature these experiences.

The methodological approach used in this research is qualitative and includes an interpretative phenomenological framework and reflexive thematic analysis of data. A total of 36 participants with current or past experiences of growing up with a parent with HD, and 14 caregivers with experiences with having children with a partner with HD, participated in semi-structured interviews.

Paper I investigated adverse childhood experiences and the meanings made from these experiences in light of support, or lack of support, from caregivers without HD. The findings show that our participants experienced a range of adversity and distress in childhood, which led to ongoing feelings of unpredictability, loss, fear and a lack of love and care. The findings also show that the meanings young people made of these experiences were seemingly different when seen in light of the support, or lack of support, they felt they had from their caregivers without HD. In other words, participants who lacked support from caregivers were often overwhelmed by feelings of stress, whereas those with support described having coped and adjusted better in these experiences. These findings show the importance of strengthening resources and availability in caregivers without HD to promote coping and adjustment for this group.

Paper II investigated both offspring and caregivers' perspectives on having conversations about HD throughout childhood and revealed the many and often-difficult dilemmas both parts can encounter when talking about the disease. These dilemmas included when to tell, what to say, how often HD should be talked about on a day-to-day basis and whether or not to share disease-related information with people outside the family. Together, the paper highlights these young people's need to understand their parent's disease and their own life situation, and the need to build skills and strength in both the parent and the child, so that these conversations can be a tool of support according to each child's individual needs.

Paper III investigated young people's experiences with social support in relationships outside the parent–child context. The findings show the many and important ways relationships can be a resource in these young people's lives and included the provision of

love, care and a sense of belonging; facilitating coping skills; and helping to reduce or alleviate stressors at home. The findings also show the many ways support was hindered for these young people. Barriers to social support included the 'invisible' nature of symptoms of HD, the lack of understanding and acknowledgement from their parent with the disease and others, and the young people's own need to protect themselves or their family.

This research comes together to understand better how children experience living with a parent with HD and to make it understood as a risk for a range of complex adverse experiences that can be understood and addressed through having sufficient resources in social support. In the thesis, co-occurring risks that were found within several contexts or relationships at the same time are addressed, as were the potential for protection in adversity by relationships with others and themselves. The thesis also addresses the participants' experiences with risk and protection within broader systems of potential support, including potentially inherent dilemmas in the provision of support for this group found within the formal, ethical and legal contexts they grow up. The thesis identifies a potential to understand, conceptualise, and help these young people in similar manners and within similar frameworks as we would with other children who face risk and adversity in their developing lives.

## List of Papers

**Paper I**

Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2022). 'I knew it wasn't normal, I just didn't know what to do about it': adversity and caregiver support when growing up in a family with Huntington's disease. *Psychology & Health, 37*(2), 211-229. https://doi.org/0000-0003-3119-703X

**Paper II**

Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2022). Dilemmas when talking about Huntington's disease: A qualitative study of offspring and caregiver experiences in Norway. *Journal of Genetic Counseling.* 1-14. https://doi.org/10.1002/jgc4.1610

**Paper III**

Kjoelaas, S., Feragen, K. B., & Jensen, T. K. (2022). Social support experiences when growing up with a parent with Huntington's disease. *Health Psychology and Behavioral Medicine, 10*(1), 655-675. https://doi.org/10.1080/21642850.2022.2104286

## Contents

# 1. General Introduction: Setting the Stage

## 1.1. Any Child's Needs and Rights

> *Every childhood offers a critical window of opportunity to shape the trajectory of a child's holistic development and build a foundation for their future. For children to achieve their full potential, as is their human right, they need health care and nutrition, protection from harm and a sense of security, opportunities for early learning, and responsive caregiving – like talking, singing and playing – with parents and caregivers who love them. All of this is needed to nourish developing brains and fuel growing bodies.*
>
> (The United Nations Children's Fund [UNICEF], 2022, para. 3)

Any child has the right to have their basic needs met and to have opportunities to develop into healthy adults (United Nations Convention on the Rights of the Child, 1989). Growing up in contexts that cause severe distress, such as dysfunctional households or with abuse and neglect, violates such rights through the risk these experiences place on the possibilities that young people have to develop normally and live happy and healthy lives (Hughes et al., 2017). Caring for children and protecting them from risk and harm has become among the most important moral, economical and legal tasks of any society (Hughes et al., 2021; Shern et al., 2016). Even though most children in Norway grow up in safe and secure environments, many are still exposed to significant adversity throughout or during parts of their upbringing (National Institute of Public Health, 2022). Adversity can be brought into the lives of children in many ways, including having a caregiver who suffers from a severe disease. This doctoral thesis explores the experiences of being a young person growing up with a parent with the severe inheritable neurodegenerative condition of Huntington's disease (HD). This thesis and accompanying research will refer to this group in several ways, including *children, offspring* and *young people,* depending on the context. These terms all refer to the same group – that is, experiences of growing up in a family in which a parent has or had HD as children (0–18 years).

## 1.2. Who Cares? Young Carers and Children as Next of Kin

Two complementary terms are used to describe young people who have a parent with a severe disability or disease. In international research, they are typically referred to as *young carers*, a term that reflects the various forms of responsibilities that might be placed on young people by looking after a family member with a disease or disability (Pakenham et al., 2015).

Under Norwegian law, the group is referred to as *children as next of kin* (Health Personnel Act, 1999), a term that also encompasses the emotional and psychological impacts of having a parent with a severe disease.

Between two and eight percent of children and young adults have been estimated to provide informal care to a family member who is mentally or physically ill (Leu et al., 2019). In effect, this means that millions of young people worldwide have their childhoods affected by growing up with parents who suffer from severe diseases. There is a substantial amount of research on how parental disability and disease affect young people, particularly regarding links to adverse outcomes. For example, one systematic review of young carers of parents with chronic illness in general found that disability or disease in a parent can lead to role reversals and changes in family dynamics, psychological and physical health problems and limited social and educational opportunities for young carers (Chikhradze et al., 2017). However, being a young carer does not only produce negative outcomes. Several studies suggest that many young carers find ways to manage and maintain normalcy and experience positive effects of their young carer status (Chikhradze et al., 2017).

There are approximately 1.2 million children in Norway (National Institute of Public Health, 2022). Finding precise or current numbers of how many of these have a parent with a severe illness or disease is difficult. However, a national study that included 534 families affected by a variety of disabilities and diseases dating back to 2015 estimated that during the course of a year, 350 000 children will have at least one parent who is a patient in the public health sector because of mental illness, substance abuse or severe somatic disease (Ruud, 2015). This study showed mixed findings about the impact that being a parent with a severe disease can have on children who are next of kin. For example, children as next of kin generally scored their own mental health as the same as that of children without a parent with a severe disease (Ruud, 2015). However, the study also found a range of challenging aspects to having a parent with severe disease. For example, the children took on more carer tasks and household responsibilities, 18% had experienced trauma, children reported an unmet need to talk about their situation, and many had not been provided with the information needed to understand their parent's disease and its consequences. Moreover, although families were found to have an extensive need for practical help, they rarely experienced support from public services (Ruud, 2015). A range of dynamics influence how young people with a parent with a severe disease experience their upbringing. For instance, their experiences depend on the duration, development and symptoms of each illness or disease, if they are at genetic risk of inheriting the disease themselves, the broader contexts

in which they have these experiences and, importantly, the support they feel they have to help them cope with and adjust to potential challenges.

HD is one of the many severe conditions that can lead to a range of complex and enduring challenges for the affected person's children. The current research originated from a need to gain more knowledge about how young people in Norway experience growing up with a parent with this disease specifically.

## 2. Background

### 2.1. A Brief History of Huntington's Disease

HD was first recognised through the clinical descriptions of a Norwegian doctor, Johan Christian Lund, when he made his yearly medical reports and recorded symptoms of decline in motor functions and dementia that he observed accumulating within families in his area (Bhattacharyya, 2016). At the time, he referred to the disease as *Setesdalsrykkja*, a Norwegian term that describes uncontrolled jerk-like or dance-like movements. Not long after, the physician George Huntington published his observations of the same disease that would end up being named after him (Huntington, 1872). Sadly, what followed these initial descriptions was a history of maltreatment, stigmatisation and isolation of those affected by HD and their families by clinicians and the public (Wexler, 2010). For example, in 1934, a neurologist noted how all members in families affected by HD were 'liable to bear the marks of a grossly psychopathic taint, and the story of feeblemindedness, insanity, suicide, criminality, alcoholism and drug addiction becomes unfolded over and over again' (Wexler, 2010, p. 18). While much has evolved since then, both in terms of public perception and the way clinicians understand and help families affected by HD, research suggests that for many people at risk, issues such as perceived stigma and isolation still prevail to some extent today (e.g. van der Zwaan et al., 2022). A major breakthrough in how HD was understood occurred in 1993, when the expanded gene that cause the development of the disease was 'discovered', making it possible to test for the genetic error and to both diagnose and predict HD (MacDonald et al., 1993). Today, reproductive choices are also available to avoid transmission to future generations. While these genetic advances have provided a major breakthrough for families affected by HD, as well as scientists in search of a cure, for those at risk, the reward is purely knowledge as there is not yet any way to treat the disease (Ellison, 2017).

## 2.2. What is Huntington's Disease?

HD is an autosomal dominant neurodegenerative disease, which means that each biological child of someone with the condition has a 50% chance of inheriting the genetic mutation that will lead to the development of HD. HD results from an overproduction of a protein that we all produce and need, called Huntingtin (*Htt)*, which accumulates over time, resulting in toxic effects on the brain (MacDonald et al., 1993). While the repetition someone has of this genetic code determines if someone will develop HD or not, the actual development of the disease seems to happen in a complex interplay with other genetic and environmental factors that are not yet fully understood. When and how HD develops for each affected person is, therefore, highly individual.

In general, the disease progresses through different stages that, to different extents, disrupt all major functions, including motor, cognitive, psychiatric and behavioural disturbance (McColgan & Tabrizi, 2018). With time, the degeneration therefore severely changes how someone controls, understands and responds to themselves and those around them. The degeneration of motor functions is characterised by symptoms such as involuntary movement and trouble maintaining balance. The deterioration in cognitive functions are characterised by impairments in thinking, such as the ability to recognise others' emotions, and executive functions, such as flexible thinking, self-control and self-monitoring. The changes caused by the disease on neuropsychiatric functioning can come with a wide range of behavioural and psychological symptoms, including anxiety, irritability, aggression, apathy, depression, obsessive-compulsive behaviour and psychosis. Importantly, recent research shows that there is often a long phase of pre-symptomatic suffering, where both cognitive and neuropsychiatric functioning slowly and subtly changes up to 10–15 years prior to a diagnosis (McColgan & Tabrizi, 2018). However, as there are large individual variabilities in symptom development and duration, it is difficult, if not impossible, to ascertain a person's particular trajectory with the disease. For example, while some might primarily present with motor symptoms at earlier stages of the disease and little or no cognitive or psychiatric disturbance, others might experience mainly psychiatric or cognitive disturbance, with little or no visible motor symptoms.

HD is a rare disease, but will affect relatively many people. In Western populations, approximately 10.6–13.7 individuals per 100 000 are affected (McColgan & Tabrizi, 2018). In Norway, an estimated 300 people have HD and about 700 people are in the pre-symptomatic phases of the disease (Centre for Rare Disorders, 2019). It is also estimated that

over 1000 people are at risk of having inherited the gene that will lead them to develop HD, many of whom are young people with a parent who is affected. Although there are large variations in the timing of disease onset (between two and 85 years of age), symptoms of the deterioration caused by HD usually emerge between 30 and 50 years of age (Roos, 2010). Because of this typically late onset and devastating symptomatology, the disease has been called a 'genetic time bomb' (Huntington Society of Canada, 1996). Individuals are typically diagnosed when they have a confirmed family history of HD and present with obvious motor symptoms (McColgan & Tabrizi, 2018).

Although promising advances have been made towards a cure for HD, there is currently no treatment and few available options for symptom relief. The disease projection is often long, and on average, someone will live 17–20 years from the time they are diagnosed (Roos, 2010). By estimations made from Norway between 1986 and 2015, the average age of death for individuals with HD is 63.9 years, compared to a mean of 76.9 years in the general population (Solberg et al., 2018). The underlying cause of death in 73.5% of the cases was HD, with the most common immediate cause of death being respiratory diseases (44.2%). Suicide was also found to be a more common cause of death in the population with HD (2.3%) compared to the general population (1.3%; Solberg et al., 2018).

## 3. Research on Having a Parent with Huntington's Disease

### 3.1. How Does Huntington's Disease Affect Families?

Developing HD can be detrimental to those affected. They can feel a tremendous loss of self, autonomy and quality of life (Mahmood et al., 2022). They may not recognise themselves, the way they feel and the way they react, and they can find it difficult to be understood by others (Hartelius et al., 2010). They may experience fear of being a burden to others and feel guilty towards their children who might have their futures robbed by the disease too (Mahmood et al., 2022). Similar reactions might be found in the affected person's family members'. In the most obvious sense, this shared reaction refers to the fact that HD is a genetic disease. It may affect several members of a family and most certainly members of the extended family. However, HD affects families in many more ways than just the genetic sense. A family is an intricate system, and *family functioning* (the ability of a family to interact, overcome challenges and attend to the needs of its members) will change when one

of its members has a severe disease (Staccini et al., 2015). When HD is that disease, research indicates that family functioning can be severely challenged (e.g. Sparbel et al., 2008).

A main effect of the symptoms that follow HD is often dramatically changed interactions with others (Hartelius et al., 2010). For example, high levels of conflict frequently occur as symptoms such as aggression, inflexibility, irritability and obsessive-compulsiveness progress (Sparbel et al., 2008). In one of the first studies to report on the impact of HD on family functioning, families were described as living under widespread anxiety and suffering (Barette & Marsden, 1979). Another early Welsh study, titled 'Breakdown and stress in Huntington's chorea', described how both patients with HD and their families experienced poor quality of life, especially in their relationships (Tyler et al., 1983). Today, research from several countries reflects similar conflictual, stressful and complex family environments. In a study on the experiences of the psychosocial context of young people living in families affected by HD in Australia, most participants described a pattern of dysfunction, disruption and conflict (Mand et al., 2015). However, studies have also produced mixed findings, suggesting that having HD in the family is not just a story with breakdown as the only outcome. For example, one study found that although important family functions, such as interest, involvement and effective problem solving, were frequently disrupted by HD, most families actually reported being generally content with their functioning (Jona et al., 2017).

Although research on the experiences of family members of those affected by HD is generally lacking in Norway, a few studies contribute to valuable findings about individuals with HD and their families in the social and cultural context in which the research of this thesis took place. Generally speaking, in Norwegian families affected by HD, conflicts that may disrupt family systems seem to also inevitably arise (Røthing et al., 2014). Families here often struggle with balancing the needs of the family and the needs of the member affected by HD, often at the costs of meeting the needs of the family (Røthing et al., 2015). Moreover, neither the person with HD nor the family may be receiving the help and support they need (Røthing et al., 2015; van Walsem et al., 2015).

### 3.2. How Does Huntington's Disease Affect Young People?

As described in the previous section, HD can severely change and challenge the lives of those affected, as well as how their family environments function. These changes may be particularly difficult for children, who will undergo much of their childhood development within the earlier phases of the disease. Like many others, including children who grow up

with a parent with cancer, they will experience the burden of having a parent with a severe disease that leads to their premature death. However, given the inheritable and slowly progressive complex combination of disease symptoms, these young people may experience additional burdens. Over the past decades, a relatively small but significant body of research from several countries has shed light on what these burdens can be. In the following section, I will use examples from this body of research and highlight some main topics that have been found to be presented in these young people's lives.

Several studies suggest that this group can have overwhelming responsibilities at home – tasks that normally would and should be conducted by adults (Forrest Keenan et al., 2007; Kavanaugh, 2014; Lewit-Mendes et al., 2018; Mand et al., 2015; Williams et al., 2009). In one study conducted with participants across the United States, over half of the children and adolescents with a parent with HD reported providing personal care to their parent, such as visits to the toilet or helping with washing or bathing (Kavanaugh, 2014). The level of responsibilities that these young people assume may have the potential to damage the health and well-being of the young people (Forrest Keenan et al., 2007; Williams et al., 2009) and to correlate with conflicts with parents and poor psychological well-being (Kavanaugh, 2014).

Studies also suggest that young people's social and educational opportunities can be hindered by having a parent with HD. This is in part related to the level of responsibility that they may bear at home, which can limit the time and concentration young people have available for school and social activities (Forrest Keenan et al., 2007; Mand et al., 2015). Having a parent with HD can also create difficulties for these young people in connecting with their peers. For example, they may feel different from their peers, unlucky and isolated, and normal adolescent issues, such as the formation of peer and romantic relationships, can be overshadowed by the parent's disease (Mand et al., 2015; Røthing et al., 2014; Sparbel et al., 2008).

A few studies suggest that young people can be exposed to childhood adversity. In a Dutch quantitative study, the adverse childhood experiences (ACEs; measured as negative life events) of young people with a parent with HD were compared to those of a group of young people with a parent with another heritable condition. This study found that more than 50% of those with a parent with HD reported ACEs, including household dysfunction, such as the psychiatric problems of the parent (van der Meer et al., 2012). Other studies have also reported a range of distressing experiences, such as erratic behaviour from their parent with HD, including irritability and mood swings, worry about their own safety and experiences of

violence, aggression and even sexual abuse at home (Lewit-Mendes et al., 2018; Mand et al., 2015; Sparbel et al., 2008; van der Meer et al., 2012).

A few studies indicate that children in families affected by HD can have compromised caregiving relationships in general. Working from the premise that early caregiving relationships shape social interactions through life, one study looked at attachment in adults at risk of HD and found that growing up with a parent with HD can negatively affect attachment representations (van der Meer et al., 2006). Interestingly, one quantitative study on the adverse parenting and quality of family functioning of a group of adults who grew up in families with HD found that adverse parenting practices, such as over-control and abuse, were reported from parents with and without HD (Vamos et al., 2007). Another study, which examined overall experiences across the United States and Canada, highlighted that young people felt the parent without HD was absent or unavailable, leaving them alone with caregiver tasks and financial responsibilities for their families (Sparbel et al., 2008). This study concluded that these young people may worry about not only the parent with HD but also their parent caregiver without HD and may struggle to find ways to meet the needs of both parents (Sparbel et al., 2008).

In addition to all these challenges, many young people are at risk of inheriting HD themselves. Two of the most common concerns for adolescents in families with HD are being at risk and deciding if they want to test to predict if they have inherited the genetic mutation that eventually will lead to them developing HD (Duncan et al., 2007). Research suggests that this worry can detrimentally affect these young people's daily lives (Forrest Keenan et al., 2007; Mand et al., 2015; Rolland & Williams, 2005). For example, one study, which investigated the lived experiences of young people at risk of HD internationally, found that as many as 80% experienced anxiety related to their own risk (Lewit-Mendes et al., 2018). Most research on being a young person in a family with HD has focused on the topic of pre-symptomatic genetic testing. This research highlights the many and often difficult questions that come with deciding whether or not to test for the disease (e.g. Forrest Keenan et al., 2015). Using the same data as employed in this thesis, another study, titled 'Psychological reactions to predictive genetic testing for Huntington's disease', investigated the participants' experiences of genetic risk and pre-symptomatic genetic testing and largely confirmed that this fear is present in the lives of many of the participants included in the present thesis too (Tillerås et al., 2020). The findings indicate that deciding to test for the disease before eventual symptoms occur is often a lifelong and difficult decision-making process and that

many young people grow up anticipating that they will develop the disease themselves and consequently limit their ideas about, and planning of, a normal future (Tillerås et al., 2020).

Many families conceal the existence of HD and their efforts to handle the stress that comes with it (Driessnack et al., 2012). Therefore, another major issue for these young people is that sufficient information is not shared with them about the disease as they grow up. For example, in one study, the parent's progressive losses compromised meaningful personal communication and family interaction – a loss that was experienced as a major source of sadness for the young people (Sparbel et al., 2008). Interestingly, one study suggests that knowing about HD from an early age helps participants cope and that those who worry about their own risk to the extent that it has significant negative impacts on their mental health had also found out that were they are at risk for HD later (Forrest Keenan et al., 2007).

Considering the variety of challenges that these young people may face, it is perhaps not surprising that several studies have highlighted the many and often profound emotional effects these experiences can have on them. For example, one study on experiences of being at risk of HD around the world found that those at risk frequently deal with difficult emotions, such as feeling frustrated, helpless and sad, and that they show high levels of anxiety and depression (Lewit-Mendes et al., 2018). Another study found that 48% of their sample had psychiatric disturbances of some kind, with affective, personality and behavioural changes being the most common and with many participants reporting more than one of these mental health issues (Vamos et al., 2007). These disturbances could, at least in part, be how young people in families with HD cope or are helped to cope with the challenges they face. For example, several studies suggest that these young people frequently attempt to cope through a range of less adaptive strategies, including emotional avoidance or risk-taking behaviours (Duncan et al., 2007; Forrest Keenan et al., 2015; Williams et al., 2013).

However, research also suggests that how these young people adjust to challenges varies. For example, one study found that many teens use well-known adaptive strategies to manage the issues they face, such as talking with others in similar situations and gathering information about their concerns (Williams et al., 2013). Another study indicated that when young people use strategies such as distancing, having a positive attitude, self-efficacy and flexibility, they cope better with the challenges brought into their lives by HD (Forrest Keenan et al., 2007). A general factor in coping successfully seems to be having protective factors, such as good systems of social support and strong attachments within relationships (Forrest Keenan et al., 2007).

Although research on support for this group is limited, it does suggest that offspring in families affected by HD can have a range of unmet needs. For example, one study examined young people in families with HD in relation to their caregiving tasks and identified several unmet needs, including information about the disease, support from friends and support groups (Kavanaugh et al., 2015). Another study suggests that young people generally struggle to find or access appropriate support (Lewit-Mendes et al., 2018). Due to their complex needs and often compromised caregiving relationships, they desire different types of help from others, indicating that to meet the needs of this potentially vulnerable group, support should be tailored from a range of resources (Forrest Keenan et al., 2007; Lewit-Mendes et al., 2018; Mand et al., 2015).

While the majority of available research on young people with a parent with HD is qualitative, most of these studies have reported on the experiences of these young people on a general level. This means that studies usually include many topics to form an overview of children's experiences, and few have looked at specific experiences in-depth. The in-depth research on specific topics, such as ACEs (van der Meer et al., 2012), attachment difficulties (van der Meer et al., 2006) and support (Lewit-Mendes et al., 2018), is quantitative and/or mainly focuses on the psychological understanding of experiences related to genetic risk and genetic testing. In this sense, the research provides valuable knowledge about *what* factors may present challenges in these young people's lives but does not necessarily provide an understanding of *how* these experiences come to be or how they are experienced. If we shift our understanding of these young people from asking what they experience to also asking how they understand these experiences, a good place to start formulating such questions is within frameworks of knowledge of how any child develops in contexts of adversity and distress.

The following section presents an overview of developmental psychological perspectives and concepts relevant to understanding why the research in this thesis was conducted and the framework in which this investigation can be understood. As each paper in this thesis provides information about relevant research within that specific frame of reference, this section mainly focuses on providing a broader description of the framework that has formed my understanding of how young people develop within a given context.

## 4. Theoretical Perspectives and Important Concepts

*Life is a story for which the beginning sets the tone.* (UNICEF, 2022, para.1)

### 4.1. Introducing a Transactional Understanding of Development

Scientists have been interested in how and why humans grow and develop as they do for well over a century. The first published paper in developmental psychology is accredited to Charles Darwin, who, in 1877, reported on the scientific observations of his own son from birth (Darwin, 1877). Since then, scientific interest in children's development has continued to grow. Today, developmental psychology represents the scientific study of how and why humans develop, change, cope and adapt across environments and transitions in life. Developmental psychology is interested in social, emotional, cognitive and physical development and examines the influences that nature and nurture have on the process of being and becoming a person. The ultimate goal of this field of study is perhaps to find ways to foster healthy lives and prevent factors that can cause harm to one's psychological or physical development.

Development rarely occurs in only the child or the context but in the relationship between the two. When we try to understand how young people develop in a given context, such as the context of having a parent with HD, we need to take into account both the individual and contextual influences that mould them (Fanti, 2011). Seeing development as transactional reflects how any child develops as a product of the continuous interactions the child has with others in the many contexts in which they live (Fanti, 2011; Sameroff, 2009). As the word 'transactional' reflects some form of exchange, a core idea is that people develop as active participants in their own lives; they are not only influenced by the environments in which they grow but also influence their environments (Fanti, 2011). A key way in which we actively participate in our own development is how we interpret and perceive events that form our experiences. This construction involves processes of giving meaning to our experiences, our relationships and ourselves, called *meaning-making* development (Kegan, 1980).

### 4.2. Adversity and Distress in Developmental Contexts

While childhood presents great possibilities for growth and flourishing, it can also come with huge risks. Throughout life, we are all faced with a range of demands and experiences that we need to learn to adjust to and overcome. Even from a young age, we experience challenges within the context in which we grow up. We may have difficulties in

peer relationships, at kindergarten or at school, and we may experience conflict with family members. To a certain extent, these are all normal, even healthy, parts of our childhoods and represent developmental demands that contribute to how we learn to regulate and adjust through experience. However, challenges can become overwhelming and interfere with normal development. These demands, often referred to as ACEs are presented by growing up in a setting with challenges that goes beyond what is considered normal, which risk overwhelming a child's resources and may consequently cause maladjustment in development.

ACEs refer to 'childhood events, varying in severity and often chronic, occurring within a child's family or social environment that cause harm or distress, thereby disrupting the child's physical or psychological health or development' (Kalmakis & Chandler, 2014, p. 1490). ACEs can include various experiences that fit this definition but traditionally fall under three main categories: *abuse, household challenges* and *neglect* (Boullier & Blair, 2018). The first category relates to experiences of emotional abuse, such as insults, being put down and fear of being physically hurt, physical abuse, such as being grabbed, slapped and having things thrown at one, and sexual abuse. The second category relates to experiences with household challenges, such as having family members with substance abuse problems, mental illness and parents' separation or divorce. The last category relates to experiences with emotional neglect, such as a lack of love, support and closeness in relationships, and physical neglect, such as not having someone to care for, protect, prepare food or provide clean clothes.

The first study to identify a strong link between the number of ACEs and the likelihood of negative health and behavioural outcomes later in life was conducted in the United States, in which more than 17 000 adults were asked about their childhood experiences (Felitti et al., 1998). Today, these results have been reproduced many times across the world (Hughes et al., 2017), including in Norway (Tomasdottir et al., 2015). Among the many unfavourable outcomes of ACEs are drug use, diabetes, heart disease, depression, autoimmune diseases and even early death (Hughes et al., 2017). This effect is graded, meaning that the more ACEs someone experiences, the greater the risk of negative health outcomes (Hughes et al., 2017). This relates not only to the individual but also to societies and future generations. For example, in Denmark, it has been estimated that 3–4% of yearly national costs can be traced back to ACEs (Hughes et al., 2021). Consequently, identifying groups of children who are at risk for ACEs, such as those who grow up with a parent with

HD, to minimise the risk to them, their futures, future generations and society in general should be of significant interest to any society.

To formulate an understanding of how such risk can transpire in children's lives, we also need to understand the process by which ACEs can transfer into poor mental and physical health. Here, stress seems to be a key element – or, to be more specific, toxic stress (Shonkoff et al., 2012). Stress can be defined as a physiological response that comes with arousal and negative affect (Folkman, 2020). It is not inherently harmful, and learning how to cope with stress is an essential part of any child's development. Our bodies are programmed to respond in certain ways to situations that we interpret as challenging or threatening. If there is a stressor in the environment, the body's stress response, called the 'fight or flight response', is triggered (Danese et al., 2012). We automatically shift our energy resources towards fighting off a threat or fleeing from an enemy. The response is physical, psychological and, importantly, adaptive, particularly in the short term and exists to keep us safe in emergencies (Danese et al., 2012). However, for children who experience ongoing and severe stress, called distress, the body's automatic response to stress might hurt more than it helps. That is because the activation that comes with stress is continuous, and the response to stress is maintained over time (Shonkoff et al., 2012). This type of prolonged stress activation, particularly in children who are vulnerable as their bodies and brains are still developing, may disrupt healthy and adaptive responses to stress and become toxic to the developing brain (Shonkoff et al., 2012). Toxic stress refers to excessive stress that is not regulated or removed and therefore accumulates over time. The term 'toxic' describes how accumulated stress in childhood can interfere with the development of healthy neural, immune and hormonal systems and even alter the expression of our DNA, in turn creating pathways that lead to poor health (Danese et al., 2012; Shonkoff et al., 2012).

This understanding offers a framework for investigating *how* risk threatens the health of any child, including those who grow up with a parent with HD. However, we know little about the nature of this group's ACEs. There could be several factors that differentiate this group's experiences from those captured by standard and quantitative measurements of ACEs, or by the experiences of young people with parents who suffer from other types of illness and disease. To this end, Paper I in this thesis aimed to qualitatively explore this group's experiences of adversity and distress throughout childhood. However, this investigation does not necessarily inform us about how these young people actually experience coping and adjustment. In fact, many children who grow up in adverse conditions end up as happy, healthy adults. Therefore, the outcome of adversity and toxic stress is not merely a primal

physiological reaction that remains uninfluenced by our surroundings. Understanding the meaning that young people who grow up with a parent with HD make of distressing experiences and situations, and how this can transfer into coping, may help explain why this is so.

### 4.3. Coping and Adapting through Stress

Coping refers to the efforts we make to manage stress (Lazarus, 1993). It is helpful to note that the term 'coping' is used regardless of whether the process is considered adaptive or maladaptive, successful or unsuccessful, and can be any attempt to manage our emotions, the problem itself and the situation by maintaining positive well-being (Folkman, 2020). Adaptive coping with stress reflects effectively regulating stress that, generally speaking, will lead to healthy development, such as mental and physical well-being or social functioning (Lazarus, 1993). In contrast, maladaptive coping reflects ineffective processes for managing stress. Maladaptive responses that do not efficiently regulate stress can lead to maladjustment, which refers to a negative developmental outcome, such as impaired functioning, distress and/ or poor physical or mental health (Gellman, 2020). A few studies about young people with a parent with HD highlight that this group can struggle with a range of manifestations of maladjustment, such as anxiety, depression and substance use (e.g. Lewit-Mendes et al., 2018; Vamos et al., 2007). In this sense, we know about factors that present risk and the possibilities of detrimental outcomes for the young people whose experiences we are investigating but little about the actual experiences that underlies such risk. The meaning young people with a parent with HD make of the distress they experience can be key to this understanding.

Making meaning is an ongoing evaluation of how our experiences compare to our goals, values and beliefs (Folkman, 2020). We make meaning through all situations whether they are normal everyday events, major life events or continuous stress (Folkman, 2020). This process serves as a core feature in how any child grows and evolves. Children constantly make meaning within their environments and change that meaning quickly as their experiences continue to grow (Ignelzi, 2000). They attach meaning to the words that they hear and the signs that they see. They make logic out of their social surroundings and the culture they experience. Last but not least, they use their experiences to make meaning of their lives, such as forming ideas about one's future and oneself (Ignelzi, 2000). A central concept in understanding how any stressful event or situation becomes overwhelming or manageable throughout development, including for young people who grow up with the stress of having a parent with a severe and life-threatening disease, also relates to the meaning they give to the

stressor, referred to as an appraisal in stress and coping theory (Lazarus & Folkman, 1984). When experiencing distress, the meaning we make of the situation determines the emotion that follows. With meaning comes the power to either regulate or increase the intensity of the emotional reaction we have to that stressor (Folkman, 2020). For example, in situations that we interpret as threatening, challenging or potentially harmful, we choose coping strategies that we believe will either help us regulate and reduce unpleasant emotions or manage the stressor itself. This framework opens up questions about young people who grow up with a parent with HD. How did the meaning they made from ACEs play a role in how they coped with and adjusted through such experiences?

In investigating how someone's experiences with meaning transfer to coping efforts that, in turn, can become developmental maladjustment or adjustment is an endeavour in which multiple influences need to be taken into account. As with development, the meaning we make and how we cope in contexts in which we experience adversity and distress are the result of an interplay between our characteristics, relationships, environments and experiences (Folkman, 2020). Together, they co-create how we manage and regulate stress. When these resources help mitigate or regulate responses to severe distress, they are referred to as promote factors, protective factors or buffers (e.g. Fergus & Zimmerman, 2005; Fraser et al., 1999). Throughout development, one of the most important resources promoting healthy development in light of risk is the resources in social support that children feel they have (Fergus & Zimmerman, 2005; Hughes et al., 2017; Shonkoff et al., 2012).

### 4.4. Social Support Offers Social Protection

Social support can be defined in many ways, but it generally includes having relationships that offer some form of emotional support (e.g. provision of values, love and sympathy), informational support (e.g. facts or advice) or instrumental support (e.g. behavioural and material assistance; Thoits, 2011). While the word 'regulation' in coping might give the idea that coping is something that happens within the individual, regulation can only take place if there is a social context involved in this process (Sameroff, 2009). This is true for humans as well as other social mammals, who all share the same distinct characteristic – being connected with others helps us recover from adverse experiences (Kikusui et al., 2006). Why is that? There are many reasons, ranging from biological to psychological explanations. On the one hand, neurobiological research suggests that social contact positively influences our reaction to severe distress by automatically lowering psychological and physiological activation (Kikusui et al., 2006). On the other hand,

compared to other social mammals, the long process we undertake to fully develop is not possible without the psychological aspect of protection, love and support from close adult relationships (Gopnik et al., 2017). Consequently, children are supported through stressful experiences when they have social and emotional bonds that buffer the effects of stress. The closer the relationship, the stronger this buffer or protection works (Fergus & Zimmerman, 2005; Thoits, 2011).

#### 4.4.1. Parent–child relationship as a pillar in development and a protector in stress

Across our lives, one of the most important arenas for social support is the relationships we have with our caregivers (Horstman et al., 2016). Naturally, therefore, when children experience adversity for any reason, the contexts provided by their caregivers are a particularly important resource in how they cope and adapt. On the one hand, having supportive relationships with close adults may protect children from the damaging effects of stress and adversity on their development in the short and long term (Shonkoff et al., 2012). On the other hand, not having sufficient support from caregivers can be devastating to children. Consequently, when children experience ongoing distress at home and do not have caregivers who they feel are available to support them, the child is at greater risk of developing psychopathology as well as other health issues (Thompson et al., 2015). Answers to how caregivers provide this type of support for young people in families with HD might be found by examining how they experience social support in these relationships. For any child, their interactions with caregivers create bonds that form the foundation for the majority of the functions needed for children to have healthy developmental trajectories (Horstman et al., 2016). These interactions facilitate feeling safe, loved, cared for and a sense of belonging, teach children how to manage conflicts and show how personal information is handled (Horstman et al., 2016). People, both young and old, are always actively constructing and re-constructing how they see the world based on the experiences they have (Sameroff, 2009). However, we know little about how these processes influence the ways young people experience the adversity that can result from growing up with a parent with HD. To this end, Paper I in this thesis explores how young people understand experiences of adversity and distress in light of the support they felt they had, or did not have, from their caregiver without HD.

The role of social support also recalls the importance of the meaning we make of stressful situations. Conversations with caregivers have a strong influence on guiding and

helping children create meaning from their life experiences and provide relief (Horstman et al., 2016). These supportive interactions can be particularly important for children who experience adversity, such as growing up in a family in which a parent is affected by a severe disease, as conversations with caregivers guide how children understand the changes and challenges happening to their close loved one and themselves (Dalton et al., 2019). However, understanding how young people with a parent with HD have made meaning of their experiences with the disease through interactions with their caregivers has not yet been explored. To this end, Paper II in this thesis explores the experiences of both caregivers without HD and offspring of having conversations about the many facets of growing up in a family with a parent with HD.

### 4.4.2. Social support outside the parent–child relationship

Because caregiving relationships exert such a powerful influence on childhood development, how social support can act as a buffer to stress and facilitate or impede development is perhaps most easily understood within this relationship. However, adaptive coping that leads to healthy development does not lie in the parent–child relationship alone (Sameroff, 2009). While at the beginning of life, our families influence our development the most, over time, other arenas grow in importance, including extended families, friendships and school. Any child is involved in a range of contexts and relationships that have a significant influence on them and that they, in turn, influence (Sameroff, 2009). All the contexts in which someone grows up are central to how they adapt to the challenges they encounter in life (Thompson et al., 2015).

Theoretically, social support can come from any part of a child's network (Taylor, 2011). Key to this idea could be the mechanisms by which social support works to help someone in adversity and distress. Social support theory suggests that relationships may work to buffer the effects of stress through two broad mechanisms; emotional sustenance and active coping assistance (Thoits, 2011). Relationships that provide emotional sustenance help children cope by indirectly reducing or "buffering" the physical and emotional arousal related to stressors, helping sustain the child's positive and adaptive emotions in stress (Thoits, 2011). Relationships that provide active coping assistance buffer or protect in stress by reducing the child's psychological or physiological stress-related arousal. In theory, various close relationships, such as extended family or friends, can therefore be supports and stress protectors through emotional sustenance, such as love, belonging or sympathy (Thoits, 2011). While past research highlights the importance of broad networks of social support for young

people with a parent with HD (e.g. Kavanaugh et al., 2015; Keenan et al., 2009; Lewit-Mendes et al., 2018), we know little about how these young people experience the support they find in relationships outside the parent–child context. For instance, theory suggests that relationships that do not necessarily involve close social contacts, such as with others with similar experiences, may still provide invaluable support by offering recognition and a sense of belonging (Thoits, 2011). Similarly, both close relationships and more peripheral parts of one's social network, such as health-care personnel, neighbours or teachers, can be a support and a protector in distress if they help the child reduce stress through active coping assistance, such as coping encouragement, information or advice (Thoits, 2011). To this end, Paper III in this thesis explores experiences with social support outside the parent–child context.

## 5. Aims of the Research

The overall aim of this thesis was to explore the current and past experiences of being a young person with a parent with HD. The presented context, research and theoretical concepts guide the three main aims for the research in this thesis.

1. Paper I explores current and past ACEs of individuals with a parent with HD and how perceptions of caregiver support influenced these experiences.
2. Paper II explores the perspectives of both offspring and caregivers on the topic of talking about the disease throughout childhood.
3. Paper III investigates the current and past experiences of young people who grew up with a parent with HD with social support outside the parent–child context.

# 6. Methodology

## 6.1. A Qualitative Enquiry: Looking into the Subjective Experience of a Phenomenon

A qualitative methodology was chosen for this research and relied on data from transcribed interviews in the form of texts. This mode of collecting and understanding data belongs to the tradition of *phenomenology*, which refers to 'that which is seen or appears'. Accordingly, phenomenological research seeks to investigate and describe a phenomenon as it appears to those who have experienced it (Teherani et al., 2015). In this research specifically, I have attempted to convey our participants' experiences with the phenomenon of being a young person with a parent with HD.

## 6.2. The Research Team

The research was planned with the aim of reflecting topics of importance to our participant group. To meet this aim, several people with different formal or informal expertise were included as invaluable parts of one of the three groups that I broadly refer to as the 'research team'.

*The researchers* consist of the three authors of the published papers included in this thesis. To different extents, the researchers were responsible for the scientific process of planning, conducting, analysing and disseminating the research.

*Clinical experience experts* consist of counsellors at the Centre for Rare Disorders at Oslo University Hospital, who provided their perspectives from working with families with HD within the clinical setting. This group contributed by being discussants on broad research topics, contributing to the development of the interview guides, recruitment, and helping disseminate the results of the three papers.

*The member experience experts*, or reference group members, consisted of three individuals from the Norwegian Association for Huntington's Disease. This group provided perspectives from their personal experiences of growing up with a parent with HD or of having children with a partner with HD. They contributed at different steps of the research process, including providing feedback during the research planning when choosing which topics would be relevant to investigate, developing the interview guide and serving as discussants through the remaining research process.

## 6.3. Recruitment

To approach the research topics, we purposefully recruited two groups of participants. The first and main participant perspective we examined was that of a group of participants with current or past experiences of growing up with a parent with HD (included in the analyses of Papers I, II and III). The second group of participants included individuals with current or past experiences as caregivers who raised children with a parent with HD (included in the analysis of Paper II). To reach as many potential participants as possible, we distributed information about the study verbally and through separate information sheets in several settings where individuals in families affected by HD could be reached. These locations included educational courses held for families affected by HD during the recruitment phase, the Centre for Rare Disorders at Oslo University Hospital and genetic counselling services at Oslo and Haukeland University Hospitals and at St. Olav's Hospital. Information about the study was also distributed in print and online, primarily through the Norwegian Association for Huntington's Disease's homepage and Facebook page and bi-annual membership news letter. An information sheet about the study and a consent form were sent by post to those who contacted us expressing interest in participating or delivered in person when individuals approached one of the researchers directly. The information sheets outlined the study's purpose and method of study, provided information about the interview topics and included the main researcher's name and contact information. They also contained key ethical information, such as confidentiality and the participants' right to withdraw from the study at any time.

## 6.4. The Participants

### 6.4.1. Offspring

Anyone in Norway over the age of 12 years old who had current or previous experience of growing up with a parent with HD was invited to participate in this study. A total of 42 individuals contacted us wanting to participate. However, despite several attempts to set up interviews, six of these individuals could not be reached after this initial contact. A total of 36 participants, comprising 26 females and 10 males, were consequently interviewed and are included in the data analysis in Papers I, II and III. Participants ranged in age from 13 to 65 years ($M_{age}$ = 36.6 years). Seven participants were teenagers at the time of the interview (13–18 years), 10 were young adults (19–35 years) and 19 were adults (36–65 years). Nineteen participants grew up with a mother with HD, and 17 with a father with HD. Topics

that had the potential to come across as insensitive or invasive were discussed with our member and clinical experts when planning the research. Based on their recommendation, we explicitly did not enquire specifically about the genetic status of our participants (if they had tested for HD or what the a potential test result was). However, by their own accounts, three participants shared that they had been tested prenatally and were not at risk of having inherited HD from birth, and 19 participants shared that they had undergone pre-symptomatic testing, of which seven participants stated that they had or would develop HD sometime in the future and 12 stated that they would not develop HD themselves. The 36 interviews with participants with current or past experiences of growing up with a parent with HD took place between April 2018 and September 2018.

### 6.4.2. Caregivers

Fifteen participants who had current or past experiences as caregivers for children with a parent with HD contacted us, wanting to participate. One participant was not included in this study due to not providing written consent. Of the 14 caregivers interviewed, 11 were female and three male. They ranged from 42 to 69 years of age ($M_{age}$ = 54.9 years). At the time of the interview, three participants had children who were in late childhood (7–12 years), three had children who were teenagers (13–18 years), six had children who were young adults (19–35 years) and two had children who were adults (36–47 years). Nine participants reported currently being the main caregivers of their partner with HD. Five of these participants were no longer, or had not been, the affected partner's main caregiver due to their partner's death, admission to caretaking facilities or divorce/separation. While we explicitly did not enquire about the genetic status of the participants' children, three participants reported that their children were not at risk of having inherited HD from birth. The 14 interviews with caregivers took place between July 2019 and August 2020 and focused on the caregivers' experiences with their children's situation as they were growing up.

## 6.5. Qualitative Semi-Structured Interviews

A qualitative interview looks for details of someone's experiences on a specific topic and is a form of dialogue between the person being interviewed and the person interviewing (Brinkmann & Kvale, 2005; Moen & Middelthon, 2015). This research used semi-structured interviews to gather data on how young people experienced their upbringing with a parent with HD (included in the analyses of Papers I, II and II) and how caregivers with children with a parent with HD experience their children's situation (included in the analysis of Paper

II). The ‘semi’ part of the semi-structured interviews allowed for an open exploration of the participant’s experiences, while the ‘structure’ part helps the interviewer focus on specific topics. On average, the interviews lasted 60 minutes (range: 27–90 minutes). Face-to-face interviews were generally preferred ($n$ = 46); however, a few participants preferred telephone interviews ($n$ = 4). The interviews were conducted at the Centre for Rare Disorders at Oslo University Hospital, in the homes of a few participants or at other locations outside the home, including hotel rooms. The location where the interviews took place was based on the participant’s preference, and for one caregiver interview, a phone interview was necessary as a result of restrictions related to the Covid-19 pandemic. Most participants preferred to be interviewed outside their home setting as they were facing challenging home situations with partners, parents or other family members with HD and felt it was easier to talk in the privacy of our outpatient hospital department. All interviews were recorded and transcribed verbatim after completion.

Separate interview guides were developed for the two participant groups (offspring and caregivers). The interview topics were similar but took different approaches to the way questions were asked. We chose broad topics of questioning through a collaborative process between the researchers, clinical experience experts and member experience experts. The project manager and I used our theoretical backgrounds in constructing the interview guides. These ideas were subsequently discussed with both clinical experience experts and member experience experts for their relevance, interest and sensitivity to the research population. Obtaining this invaluable feedback on topics of questioning was important for several reasons. We wanted the research to represent the needs of the groups we were investigating and to have future implications in their lives. We also wanted to ensure that the questioning would be relevant to the actual needs and experiences of these groups and, equally importantly, be sensitive in not enquiring into topics that were not relevant or appropriate or would possibly break the trust or alliance between the interviewer and interviewee. As mentioned above, the genetic status of our participants was one topic that we were advised not to ask about unless the participants themselves brought it forth. Demographic information about offspring participants was gathered verbally at the beginning of the interviews. Demographic information about caregiver participants was gathered prior to the interviews using a questionnaire. The broad topics in the semi-structured interview guides included questions related to the participant’s childhood experiences and family situation during upbringing, young people’s relationships to their parents and others, communication and openness about the disease and experiences with support. It was important that the

participants were given the opportunity to provide nuanced narratives prompted by the questions we asked. The questions were therefore open-ended and focused on challenges as well as protective factors. The interviewers were encouraged to follow up on points of interest, thus encouraging rich and nuanced data material outside the pre-set questioning.

## 6.6. The Interview Setting

In qualitative research, an interview setting consists of a three-sided interaction in which the researcher, participant and research topic together produce the data (Moen & Middelthon, 2015). The data we gathered are therefore not only the product of our participant's accounts but also a co-creation resulting from several factors.

### 6.6.1. The interviewers

The interviews were conducted by seven different interviewers. Interviews of offspring were conducted by three postgraduate clinical psychology students from the University of Oslo and University of Bergen and by trained health professionals at Oslo University Hospital in Norway, including two clinical psychologists and a genetic counsellor. The postgraduate psychology students had no previous experience with HD, but the health professionals from the Centre for Rare Disorders had previous research experience with the disease. All interviewers undertook formal training in addressing and handling sensitive topics and had or received training in qualitative methods. The interviewers had no previous familiarity with the study participants. I conducted all 14 interviews with the caregivers. While the majority of these participants were unfamiliar to me, I had a past relationship with two of the participants through the Norwegian Association for Huntington's Disease.

### 6.6.2. Reflexivity and consistency during the interviews

The different types of characteristics any person brings to an interview setting, such as age, gender, title, education or style of communication, may influence the interview process in various ways. We took several steps to maximise the benefits of having multiple agents involved in the interview process while also attempting to minimise the possible negative impacts that this could have on data collection. To ensure fidelity and consistency across the interview practices, the project manager, an experienced and licenced clinical psychologist, contributed as a co-interviewer for the first interview in all the interviewers conducted. This allowed the project manager to provide supervision regarding interview techniques and clinical safeguards and offer the interviewers feedback and discussion about their interview

techniques. The project manager also supervised the need for a follow-up with participants after the interviews. Of our participants, five wished to receive a follow-up after the interviews and were referred to a clinical psychologist or counsellor working with families affected by HD, depending on their individual needs.

A key element in the production of qualitatively reliable and valid references is the researcher's ability to interact and learn with a degree of insight (Moen & Middelthon, 2015). As the main researcher responsible for analysing the data, I familiarised myself with the interviews I had not conducted personally by listening to the recordings and reading the interview transcripts. This way, I could reflect on the position of the researchers conducting the interviews and gain a better understanding of the subtleness of verbal tone and environment provided by both the interviewer and the interviewee, which is not easily captured by transcripts. Regarding the 14 interviews with caregivers that I conducted, I continuously tried to improve and reflect before, during and after the interviews. However, this was not necessarily a straightforward process. When conducting qualitative interviews, the interviewer needs to negotiate between the closeness needed for a beneficial relationship and keeping a productive distance (Kvale & Brinkmann, 2009). For me, in some of the interviews, this process felt quite easy. In other interviews, it required more work. For example, the first interview I conducted was in the home of one of the participants. The interview and conversation progressed easily; I was served tea and cake while the participant willingly talked about their experience, almost without any guidance from me. I remember feeling like I could not interrupt; after all, I was a guest in this person's house. Reflecting on this made me understand the significant influence that the context had on the alliance created during the interview. I was treated as a guest, and consequently, I acted as a guest. Knowingly, I took these experiences into the next interview that was conducted at home with participants, attempting to more actively adjust my role in this context. Was the next interview perfect? No. However, with this experience, as well as several others, I gradually improved my interviewing practices.

### 6.7. Data Analysis

Qualitative research requires a flexibility in methods of analysis that reflect the complexity of the phenomenon being studied (Tracy, 2010). To best address the different aims that I set out to explore, the research relied on two different analytical methods. In Paper I, we analysed the data using interpretative phenomenological analysis (IPA; Smith et al., 2009). In Papers II and III, we analysed the data using Braun and Clarke's method of

reflexive thematic analysis (RTA; Braun & Clarke, 2019, 2006). As the papers provide the specific processes of analysis, the following section elaborates on these processes by describing the general methodological approaches used in all three papers, and then the different methods of analysis and rationale for using these methods.

### 6.7.1. General analytical strategies

Although the three papers use different methodological approaches to analyse the data, several general analytical strategies are relatively consistent across them. I have worked from the premise that analysis is a process of making meaning of experiences and processes. In this process, I have shared the experiences of our participants through the lens of our interpretations and creations as researchers and not necessarily as a quest to find 'the truth' in our data (Braun & Clarke, 2019). The analytic processes were conducted inductively for all the papers, meaning that the analyses were guided by the topics and content of the participants' narratives as it was believed they experienced them (Braun & Clarke, 2019; Smith et al., 2009), or said in another way, were bottom-up as opposed to top-down.

The steps taken in the analysis of all three papers began with a similar approach. First, all three researchers familiarised themselves with the data by reading each transcribed interview and noting prevalent topics related to our participants' narratives. I then created initial codes and used these in discussions with the other researchers. In this process, we moved back and forth between the data, the codes I created and ideas of meaning, trying to find coherent and inclusive themes to encompass the content (Braun & Clarke, 2019). This process involved using different ideas of conceptualisations and theoretical assumptions latent in the mind, spending months in discussions, with agreements and disagreements, trial and error, and testing and re-testing ideas and assumptions against the data. Consequently, in all the papers, the development of themes reflects deep and prolonged immersion in the data related to each topic, thoughtfulness and reflection and a process that was active and generative. The final outcome of this process, presented as themes and sub-themes, is centred on a shared topic of the participants' experiences and our best attempt to capture the diversity of meaning related to that topic. The structure of the findings in all the papers reflects the process and outcome of the researchers as well as the data and is not something merely searched for and subsequently found. Instead, the findings were treated as a deliberate process that consistently involved decisions both individually and collectively. In all three papers, this extensive process is effectively summarised as *'themes were discussed until agreement was reached between all researchers'*. While this is true, the reality is also that the analyses reflect

an extinctive and time-consuming process that one rarely has the chance to elaborate on in published papers. In hindsight, I would perhaps describe the processes of analysis as similar to peeling off the layers an onion (the onion being the phenomenon I was studying) and carefully trying to understand, interpret and discuss each new layer that emerged. For each layer that was removed, I felt closer to 'the core' of the topics addressed by our participants, with 'core' product and description of themes reflecting the point at which all researchers were content with the depth and nuance of the meaning our findings provided.

### 6.7.2. Paper I: Using interpretative phenomenological analysis (IPA)

Analysis of the data in Paper I was guided by IPA (Smith et al., 2009). This paper aimed to explore ACEs related to growing up with a parent with HD and the meaning our participants had made of these experiences in light of the social support they felt they had from caregivers without HD. This analysis included all 36 interviews with young people's perspectives.

IPA was chosen to analyse the data in Paper I as I needed an analysis method that allowed for the description of the more categorical aspects of the participants' adverse experiences in childhood, while at the same time trying to make sense of and interpret the meaning of these claims and concerns from a psychological perspective. IPA is a particularly useful analytical method for examining topics that are complex, ambiguous and emotionally laden (Smith & Osborn, 2015), such as how ACEs may relate to experiences of social support. The method is designed to provide insight into how a person in a specific context describes a phenomenon, such as important life events or the development of relationships (Smith et al., 2009). Compared to other methodological approaches, including the thematic analysis used in Papers II and III (Braun & Clarke, 2006, 2019), the data in Paper I were dealt with in a more interpretive way and included reflection on what the participants experiences mean in this particular setting (Larkin et al., 2006).

IPA has a small core of defining concepts and has been developed as a flexible tool within different types of research (Larkin et al., 2006). First, it is phenomenological, meaning it explores peoples' experiences. Second, it is interpretative, meaning that the researcher seeks to understand how the participant makes sense of their life. Third, IPA is ideographic in that it analyses the case at hand and how the person makes sense of their situation. The data were approached with two focuses in mind (Larkin et al., 2006). First, I 'stayed close' to the data and the participants' accounts and then 'stepped back' to interpret these accounts. In the first focus, I highlighted and interpreted the participant's adverse experiences throughout

childhood and adolescence. In this sense, these experiences reflect a 'first-order analysis', while the interpretations about the role that the meaning of social support played in the participants' perceptions of adverse experiences reflect the analysis at a more interpretative level. I started the analysis by working thematically (as is common for several methods of qualitative research analysis), meaning that codes of data were used to search for common meanings, or themes, in the participants' experiences. The thematic starting point of the analysis may not always be part of the report in findings from qualitative research that use IPA, which often focuses on the interpretational outcomes of the analysis. However, when reporting on the findings in Paper I, I took the liberty of reporting both in the form of sequence as I believed this would provide readers with the most nuanced and accurate understanding of the participants' experiences. I then developed an interpretive position using the initial descriptions of adverse experiences to provide a form of commentary on how the participants had made sense of these experiences in light of the social support they felt they had, or did not have, in their caregiving relationships. At this interpretative level, the analysis in IPA involved a two-stage interpretation of the participants' accounts in which the process of analysis involved both the participants and me, the researcher, trying to make sense of their world, referred to as 'double hermeneutics' (Smith et al., 2009).

It is important to note that IPA was only used as the framework of analysis for Paper I and not as the underlying framework when planning the research or conducting the interviews. Therefore, our relatively flexible approach to this method diverts from perhaps more traditional uses. For example, to provide a more in-depth interpretation of data, it is common to include smaller sample sizes than found in the present research (Smith et al., 2009).

### 6.7.3. Papers II and III: Using (reflexive) thematic analysis

Data in both Papers II and III were analysed using Braun and Clarke's reflexive thematic analysis (RTA; Braun & Clarke, 2006, 2019). RTA is a robust and flexible qualitative method for identifying, analysing and reporting patterns or themes within the data (Braun & Clarke, 2006, 2019). In Paper II, all 36 interviews with offspring and all 14 interviews with caregivers were included in the analysis, with the aim of investigating experiences of talking about the disease throughout childhood. As part of the dynamics involving conversations with children, caregiver interviews were included in the analysis to complement and provide nuance to the experiences of young people in families with HD. In this analysis, RTA was considered robust enough to 'handle' the analysis of a relatively large

number of participant experiences, while being flexible in the sense that experiences from two perspectives were explored simultaneously. In Paper III, the 36 interviews with offspring were included to explore experiences related to social support and barriers to social support outside the parent–child relationship. Because both *how* people are helped by social support and what *hinders* support remain largely understudied phenomena, the flexibility and induction of RTA allowed patterns and themes among the patterns without underlying theoretical assumptions or pre-existing hypotheses to be found.

At its most basic application, thematic analysis offers a structural guidance of six steps to follow when analysing qualitative data (Braun & Clarke, 2006). However, the level of the patterns derived and interpretations made across those patterns rely on the aims of the research and the researcher (Braun & Clarke, 2019). The analyses in Papers II and III are therefore described by the later developments of thematic analysis. The reflexive approach in TA focuses on both reflexivity and a level of interpretation in the analysis (Braun & Clarke, 2019). Therefore, I did not follow the six-step process as outlined by thematic analysis in a linear or stringent manner, but instead used this outline as the foundation of a recursive process of analysis. In addition to providing a robust, systematic and flexible approach to data analysis, the RTA emphasises the active role of the researcher (Braun & Clarke, 2019). In the analyses in Papers II and III, this meant a continuous process of moving back and forth with the data, trying to be flexible and allowing the participants' accounts to be the main driving force of the outcome and focus of the interpretations.

### 6.8. Ethical Considerations

The research obtained ethical approval from the Regional Committee for Medical Research Ethics (Health region Southeast, Norway, reference number: 2017/1613). Participants were informed about the study and their right to withdraw at any time before giving written consent, and consent from caregivers was obtained for participants under the legal age for health consent (16 years in Norway).

Full confidentiality was ensured throughout the research process. The data were stored and handled in accordance with recommendations provided by the Data Protection Office (DPO) and in compliance with the applicable data protection rules. The main points of these recommendations and rules are that data need to be de-identified and stored so that they are inaccessible to unauthorised persons and only used in research as approved by the DPO and the Regional Committee for Medical Research Ethics. After being transcribed, all recordings were deleted. As the population under study is a relatively small group that could easily

identify one another, special attention was also given to safeguarding the identities of the participants when reporting the research. Moreover, in all presentations of the data, pseudonyms were used and any identifiable characteristics were altered.

The study did not include children under the age of 12 years old. Children and young people have the right to express their views and be included in research (Hammersley, 2015). It was therefore important to include young people with current experiences of the phenomena under study. However, regarding the inclusion of younger children with a parent with HD, several ethical challenges needed to be considered. These issues were specifically related to the sensitivity of the topics addressed in the interviews and the distress that participating could create in multiple areas of life. Another major risk discussed when planning the study was the possibility that participation could create or loyalty conflicts within the interview setting or at home with parents with HD after the interview. It was determined that the possible risks of including younger children outweighed the possible benefits of their inclusion.

The research did not exclude participants with a pre-symptomatic or symptomatic diagnosis of HD. Given the genetic specificity of HD, some participants could have inherited the disease and might have symptoms of HD. However, in the many years over which HD develops HD, the disease does not primarily affect long-term memory, and affected individuals at earlier and mid-stages may recall their childhood without major interference from the disease. Participants with communication or cognitive functioning difficulties that might have affected their participation in the in-depth interviews would have been excluded from the data, but no participants were excluded on this basis.

## 7. Summary of the Papers

### 7.1. Paper I: 'I knew it wasn't normal; I just didn't know what to do about it': Adversity and caregiver support when growing up in a family with Huntington's disease

Paper I addresses questions about the subjective nature of the ACEs of young people who grew up with a parent with HD and how they made meaning of these experiences in light of the support they had found in their caregiver relationships. The ACEs of these young people may be both different and similar compared to other groups of children at risk and to standardised measures of these events. Qualitatively exploring these experiences can therefore provide a better understanding of the risk that this group of children specifically may face. To meet this aim, childhood experiences described in semi-structured interviews with 36 participants with current and past experiences as young people with a parent with HD were analysed using an interpretive phenomenological framework.

The findings move through two 'stages' of understanding about how adversity and distress were experienced within the parent–child context. The first overarching theme describes the nature of frequent and severely distressing adverse events growing up. These experiences lead to feelings of ongoing uncertainty or unpredictability at home; feelings of unresolved loss over slowly losing characteristics of their parent through the development of HD and the potential of losing one's own future by developing the same disease; fear from experiences with their caregiver with HD being aggressive, violent or belittling; and feelings of a lack of love and care. The second overarching theme included the meaning the participant's had made from these adverse and distressing experiences in light of the support they felt they had or did not have from their caregiver without HD. The findings show that participants without support were often overwhelmed by feelings of stress, whereas participants with support seemed to have coped with or better adjusted to adverse experiences.

The distress the young people experienced was discussed as fitting well within common categorisations of ACEs and also having features potentially specific to the disease. Importantly, the findings show that how young people experience adversity in childhood is influenced by the protection that they feel they can find from social support in their relationships with caregivers without HD. However, for many of our participants' caregivers, it seemed that the strain of being with a partner with HD, or having their own physical or mental challenges, could limit their availability to provide the support their children needed. In contrast, the findings also show the significant potential that caregivers without HD possess

to buffer or protect children by helping them create meaning from adverse experiences that aids them in coping and adjusting. Implications for practice from these findings included the need to recognise this as a vulnerable group of young people's with experiences that can be severely adverse and potentially traumatic. Moreover, to help with these experiences it is important to provide caregivers without HD with support throughout the progression of the disease. Health-care and support providers should be particularly aware of extra support needs when the caregiver without HD is also suffering from illness or experiences severe strain.

### 7.2. Paper II: Dilemmas when talking with children about HD: A qualitative study of offspring and caregiver experiences in Norway

Paper II addresses questions about how conversations within the parent–child context work to facilitate meaning from the many experiences that can come with having a parent with HD throughout childhood. While past research highlights the numerous benefits such conversations provide for children with a parent with a severe and life-threatening disease, family secrecy and insufficient information about genetic risk have been identified as experiences for this group of young people specifically. To meet these aims, the 14 semi-structured interviews with caregivers and 36 semi-structured interviews with offspring were analysed using RTA.

The findings in this paper are presented as dilemmas between the perspectives of offspring and caregivers. These dilemmas included questions about when to first talk about the disease, revealing discrepancies between the offspring's need to understand and the caregiver's wish to protect them. The dilemmas also included what to say about HD and how often the disease should be talked about on a day-to-day basis, underscoring the caregiver's insecurities regarding disease related topics and the negotiations between the offspring's and caregivers' individual needs to be available and approachable for these talks. The dilemmas also related to questions about how and if personal information should be shared outside the family, highlighting difficult choices between respecting the offspring's and other family members' autonomy to share their own information and the support that could be offered by talking with others about HD.

Together, the findings provide insight into the many difficult choices that both parents and their children may have to make as they try to create meaning of the many and long-lasting changes and challenges caused by HD. The findings offer a nuanced picture of the needs of young people with a parent with HD in relation to learning about the disease and receiving support from caregivers in these conversations. A difficult task for both offspring

and caregivers seemed to be balancing the many dilemmas that arise in conversations and how to use dialogue to best help offspring adapt to and cope with the many challenges that can come with HD. These findings related to each family's needs to find the right balance for each child between protection and preservation when they talk about HD. Implications for practice include preparing caregivers in families with HD by strengthen their ability to make conscious choices in current and future conversations with their offspring. Moreover, support providers should approach the topic of conversations about the disease with an understanding about the complex nature of these talks, and help to develop trust in their caregiver–child relationships so that they feel confident when exploring and addressing their child's individual and developmental needs for conversation about disease.

### 7.3. Paper III: Social support experiences when growing up with a parent with Huntington's disease

Paper III addresses questions about how young people with a parent with HD are helped by the support they find in relationships within the many contexts in which they inevitably grow up. The aim was to investigate the current and past experiences that young people who grow up with a parent with HD have with social support outside the parent–child context. To this end, the 36 semi-structured interviews with the participants who grew up with a parent with HD was analysed using RTA. To reflect on the actual processes involved in interactions in relationships that help or hinder young people from feeling supported, the paper investigated *how* social support outside the parent–child was experienced rather than *what* social support was provided.

The overarching finding of this paper was the many important resources that relationships outside the parent–child context can provide in helping these young people cope and adjust to childhoods with a parent with HD. Relationships were experienced as supportive when they provided a sense of love, care or belonging; when they provided coping skills; and when they reduced or alleviated stressors at home. In many ways, these roles seemed to compensate for young people's lack of support at home or with friends. However, the findings also show that experiences with how support was hindered was more common than experiences of how support was provided. Barriers to receiving and accepting support included their parents' and others' lack of acknowledgement and understanding of their situation and the young people's own need to protect themselves or their family from support that they were not sure would be of help.

These findings were discussed within theoretical understandings of the mechanisms by which social support work helps those who are experiencing distress. The practical implications of this research include encouraging support providers to help strengthen or build relationships that provide young people with emotional support, informational or instrumental support. To overcome the many barriers these young people may face, it is suggested that it is important to understand how HD affects young people and to provide support that aligns with the actual symptoms that come with the progressive nature of their parent's disease.

# 8. Discussion

This research project began with the overarching aim of investigating current and past experiences of growing up with a parent with HD. The findings of this research present a qualitative exploration of three broad topics. The first paper explored ACEs and the meaning these experiences had for young people when they were seen in the light of support, or a lack of support, from a caregiver(s) without HD. The second paper investigated both offspring and caregivers' perspectives on talking about the many experiences that could come with HD throughout childhood. The third paper investigated experiences with social support outside the parent-child context. The papers can be seen as an individual piece of a puzzle that comes together to provide knowledge that increases our understanding of how different relationships and contexts form the experiences of young people with a parent with HD. When we want to understand these experiences, empirical frameworks that inform about how both risk and protection influence the experiences of any child who faces adversity become particularly important (e.g., Fraser et al., 1999). Vulnerability or risk in this setting reflects the higher chances that someone will experience negative outcomes when exposed to severely adverse experiences (Hughes et al., 2017). Importantly, it reflects psychological risks and is not to be confused with the genetic risk of having inherited HD. Protection, or protective factors, reflects the resources someone has available to help them avoid potential issues that come with being vulnerable, such as social support (Fergus & Zimmerman, 2005; Fraser et al., 1999). How risk and protection co-create childhood experiences is generally seen as transactional, meaning that it involves a continuous process of exchange between risk factors or vulnerabilities and the resources a child finds in the closer and broader external relationships and contexts they interact with, as well as within themselves (e.g., Fanti, 2011; Fraser et al., 1999; Thompson et al., 2015). The following discussion focuses on the overall findings considering these broad concepts and frameworks of understanding.

## 8.1. Relationships in Adversity: Resource or Double Trouble

A natural place to start this discussion is to conceptualise the psychological risk or vulnerabilities in the participants' childhoods. Forming the background of this understanding is knowledge that children develop through continuous interactions with others in the different environments they grow up in (Fanti, 2011; Sameroff, 2009). When any of these relationships or contexts is the source of severely adverse or distressing childhood experiences, there is a significant risk that their current and future health and well-being will

be severely negatively affected (Hughes et al., 2017). As caregiving relationships provide the most important functions that innately and explicitly facilitate any child's development, such as love, care, belonging, and feelings of safety, children are also most vulnerable when adversity and distress take place within their caregiving contexts (e.g., Horstman et al., 2016; Masten, 2018; Thompson et al., 2015).

In possibly the most obvious sense, the findings can demonstrate aspects of the vulnerabilities or risks in caregiving relationships between a parent who develops HD and their children (Paper I). Here, the findings show how these caregiving relationships could be changed by the disease from being or having the potential to be a source of support to what may be a source of insecurity or fear. At the heart of these experiences were ongoing feelings of unpredictability, loss, fear, and a wish to have been loved and cared for in ways their parent with a progressing, severe disease had lost the ability to provide. However, within an understanding of these experiences as transactional, is also the concept that the outcome of such experiences is rarely only based on this risk alone. Instead, the resources a child can find in other relationships will influence the outcome of such risk in many ways. If a child does not have access to protective factors, such as social support, there is a higher chance of such risk leading to negative outcomes (Fergus & Zimmerman, 2005). Unfortunately, children who endure adversity within one relationship are at an increased risk of being exposed to adversity within other relationships in the same context or within other contexts (Finkelhor et al., 2007; Finkelhor et al., 2011). Finkelhor et al. (2011) suggest that this is because there are often common and similar underlying risk factors that accompany different types of childhood adversity – for example, the similarities inevitably found within a child's family environment. This understanding of co-occurring risk relates to the findings in this study because our participants often encountered the absence of resources in relationships with caregivers without HD that could potentially have helped them cope with and adjust to having a parent with severe disease (Papers I and II). At the heart of these experiences was the helplessness and hopelessness adverse experiences created when the young people generally felt they lacked love, safety, and protection from close adult relationships (Paper I). These feelings can be further understood as the confusion, self-blame, and disconnection that resulted from not having any caregiver who helped them understand and make meaning of the minor and major events and issues that would inevitably arise as their parent's disease progressed (Paper II). Moreover, in several instances, the absence of resources to cope did not necessarily stop with caregiving relationships. The feelings of helplessness and hopelessness had been further exacerbated by not finding needed support outside the parent-child context. For example, this

wider absence of support could result from how many members within their extended family had also been suffering from the disease, were preoccupied with caregiving tasks, were adhering to rules of family secrecy, or were disconnected from the participants' lives by ruptured relationships due to family conflict (Paper III).

Together, these findings can provide an important way to understand the nature of our participants' experiences within a framework of risk and protection. That is, risk for this group may not be only about the changes and challenges caused by their parent developing HD (although this presents a psychological risk in itself). Instead, these young people can also experience co-occurring or cumulative risk, which may unfold simultaneously within several relationships and contexts. This is important to recognise as a part of the complexity of psychological risk for this group because the outcome for children who experience severe adversity will generally be graded, meaning that the more types of adversity or the more contexts adversity occur at the same time, the higher the risk is for maladjustment in later life (Finkelhor et al., 2011; Hughes et al., 2017). Specifically, the more frequently any young person encounters severe distress, the higher the chances are that the stress becomes 'toxic' and impedes normal developmental processes that could or should have resulted in happy, healthy lives as children and as adults (Shonkoff et al., 2012). In light of the serious psychological suffering that some participants in this study had undergone, the need for future responses that minimise these co-occurring risks cannot be overstated. Such responses will require the attention of support systems that can promote safer overall developmental contexts for these young people. Work needs to be done, for example, to enhance the protection offered by caregivers together with extended family members so that they can be a resource to help their children cope with the distress associated with having a parent with this severe heritable disease. Importantly, the findings highlight the need for support systems to assess if these young people have relationships present in their lives that can promote coping and adaptation.

### 8.1.1. Every cloud has a silver lining

When pulling together the findings across this research, the role adversity played seemed to explain only one side of our participants' stories. Equally important when we try to understand the findings are the many and different ways these young people had found resources in their relationships with others that had helped them cope and adjust. In theory, every child who experiences adversity in one context or relationship has many opportunities for protection against the risks this pose on their lives through other relationships or contexts

(Fergus & Zimmerman, 2005; Thoits, 2011). This is because the resources found in meaningful relationships with others, such as social support and conversation, biologically and psychologically help regulate the child's responses to stress (Gopnik et al., 2017; Kikusui et al., 2006). A major part of this process is the meaning that is given to experiences of an adverse nature, which determines how someone copes with and adapts to challenges (Lazarus & Folkman, 1984). These meanings emerge from a complex interplay of various factors such as the child's characteristics, past experiences, and, particularly, the resources found in relationships (Folkman, 2020; Kegan, 1980). Of these factors, the resources found in caregiving relationships are the most consistently important for children who experience risk and include conversations and other types of social support (Fergus & Zimmerman, 2005).

Seen within this framework, the participants' descriptions also highlight how relationships can provide a resource when facing risk of adversity (Papers I, II, and III). At the heart of their experiences were how caregiving relationships that provided feelings of safety, love, and belonging could facilitate different meanings of ACEs, from hopeless and helpless without this support to manageable and controllable with this support (Paper I). Within contexts that were perceived as supportive, conversations with caregivers contributed to adaptive understandings that helped the participants cope with the disease and a sense of relief from stress (Paper II). In turn, this could facilitate how the young people understood the changes observed in their parent and their own risk of heritability from confusion and resignation to a sense of adaptation and a positive outlook on their futures. Similar supportive functions, or protection, could also be found when needed through relationships with persons outside the parent-child context. For example, love, care, and belonging when found in extended networks of social support seemingly compensated for not having found these protective factors in other contexts such as the participants' caregiving relationships or peer relationships (Paper III). Equally, others outside the parent-child context could form and reform understandings about the disease and instil hope regarding how these young people understood their lives and futures (Paper III).

Together, these findings provide another way to conceptualise the influences of risk and protection for these young people. That is, how the participants felt they coped with and adjusted to the adversity that came with having a parent with a severe disease seemed to be the product of whether they had relationships available to help them make meaning of and regulate responses to stress. I decided to call this part of the participants' stories 'silver lining', which represents a metaphor of optimism and emphasises the hopeful side of a situation that may seem only gloomy on the surface. This optimism calls for anyone who can

help strengthen or build social networks that can be a protection in these young people's lives. Building relationships that can be resourceful to these young people would require social support systems to act early, preventively, and continuously, as well as work to increase knowledge about the important role relationships have as potential protectors in risk of adversity related to having a parent with HD.

### 8.2. Inside Out: The Child as an Active Participant in Their Own Experiences

Risk and protection do not co-create childhood experiences as an either-or, but is instead a dynamic construct that will change with time and different experiences (Fergus & Zimmerman, 2005). In this mix, each child also brings their characteristics and behaviours into interactions with caregivers, friends, at school, with extended family members, or with public systems of support (e.g., Fanti, 2011; Fraser et al., 1999). Another important resource for any child who faces risk is therefore the protection they can find in themselves to cope with and adjust to challenges. For example, young people with high self-efficacy and social competency may show more positive outcomes regardless of risk (Fergus & Zimmerman, 2005). The research in this thesis also showed these strengths at the individual level. For example, this is demonstrated through the participants' reflections on their coping strategies for effectively navigating adverse experiences, including conflict, the optimistic cognitive representations they had formed of what the disease meant to them and their outlook on life, and the self-efficacy and independence they brought into their interactions with others (Papers I, II and III).

However, these individual assets are also influenced by the resources around the young people that facilitate this type of positive adjustment (Sameroff, 2009). This way, vulnerabilities on an individual level to cope with stress can be created when a child experiences adversity in one context, as this risks the development of characteristics such as lower self-esteem, learned helplessness, and dysfunctional thought patterns (Finkelhor et al., 2011). In this research, for example, having a parent with a severe and heritable disease, such as HD, can present a risk to a child's well-being that remain in their lives regardless of the strength and availability of the social networks that provide support (Paper II). Also, all three papers shed light on challenging experiences that may generate vulnerabilities for these young people, such as their accounts of perceived stigma, constant fear of having inherited the disease, or shame. These and similar challenges influenced how they perceived the potential of relationships with others as supportive and, in turn, could hinder them from being able to connect with friends or others for social support. For example, the fear of others searching for

symptoms if they found out that they could be at risk of inheriting the disease prevented them from reaching out to peers and others for support (Paper II). Additionally, uncertainty of potentially negative consequences for themselves and their families if they reached out for support also hindered them from sharing their home experiences (Paper III).

On top of these vulnerabilities, adversity will mean different things to different children regardless of whether someone is exposed to risk or not. For example, a child's emotional sensitivity and ability to approach emotionally latent issues and their stage of development all play a part in how a child approaches the social contexts in which they grow up (Maisel & Gable, 2009; Thoits, 2011). In this study, for example, some participants experienced their parents with HD moving away through feelings of abandonment and loss, while others experienced it as relief (Paper I and III). How children are a part of the dynamics of their relationships was especially evident when including the perspectives of caregivers on the topic of talking to children about HD (Paper II). For example, at the heart of some caregivers' experiences were feelings of rejection of their attempts to help, and insecurity about how to restore the dynamics in their relationships with their children to be of support. Moreover, some young people did not want to be supported through conversations for various reasons, including a desire to maintain normalcy (Paper III). Together, these and similar findings point to an important part in the mix of what can constitute risk and protection in these young people's lives. That is, the young people themselves needs to be considered when aiming to promote positive outcomes for this group. This requires the attention of available resources and support systems to be aware of the need to evaluate and enhance each child's individual strengths and coping mechanisms.

### 8.3. Risk and Potentials for Protection in Broader Social and Societal Contexts

The previous section focused on ways to conceptualise our participants' experiences with risk from adversity in light of the resources they find in relationships with others and themselves. In theory, while close relationships, such as with caregivers, peers, and extended family members, provide the most influential types of resources to promote healthy development and protect against risk, any child also develops within broader contexts of social and societal support (Bronfenbrenner & Morris, 2006; Sameroff, 2009). The resources a child or a family has available to cope and adjust when facing changes and adversity therefore also include the protection they find within the broader systems in which they inevitably interact (Masten, 2018). The following section discusses some of the vulnerabilities

or dilemmas that the findings suggest can come with being supported within these broader contexts for young people with a parent with HD.

### 8.3.1. The mask of sanity

One of the central themes in this research is the obvious discrepancies between how HD was experienced by children who lived with a parent with the disease on a day-to-day basis (Papers I and II) and how these experiences were understood and supported by others outside the home (Paper III). At the heart of these experiences were accounts of the emergence or presence of HD symptoms in their parent. For example, how unpredictability at home was connected to parental behaviours such as irritability, apathy, or rigidity, and how fear was connected to parental behaviours such as aggression and diminishing impulse control, lack of understanding, or missing emotional validation (Paper I). Additionally, parents were also described as unaware of the symptoms related to their disease and how it affected those around them and were, at the same time, often described as unwilling to receive help and support for themselves or their children (Paper III). For the sake of argument, I have called the risk this posed on the lives of these young people 'the mask of sanity'. While this is an expression originally used in the first clinical descriptions of a personality disorder (Cleckley, 1951), it can also illustrate the participants' experiences of how their parent's symptoms of a progressing severe disease remained masked or hidden, often for years, from others outside their family contexts.

While our findings highlight how children experience the symptoms of HD, they do not necessarily explain why it appeared to have been difficult for others outside the family context to recognise these symptoms (Paper III). Bringing the findings back together with the understanding of how HD progressively affect the brain can offer an answer. Within this understanding, the subjective accounts of our participants align with neuroimaging research which show that structural changes are caused in the brain by HD from early in the disease trajectory (Epping et al., 2016; McColgan & Tabrizi, 2018). With descriptions of a sense of crisis in the family that could be present throughout childhood the findings also align with the length of time this deterioration will progress. While the most visible signs of HD are involuntary movements, the neuropsychiatric distruption that is an equal part of the symptomatology often starts many years before obvious motor symptoms occur (McColgan & Tabrizi, 2018). During this pre-symptomatic phase, a person with HD can still function independently in many areas of life. However, those who live with someone with HD on a day-to-day basis might have a different experience of the symptoms that are occurring. This

way, our findings might reflect a general issue regarding how HD can easily be misunderstood by general public health and support systems. For example, even though cognitive and psychiatric symptoms often present the largest burden on both the person with HD and their families, research and clinical diagnostics have mainly focused on the emergence of deterioration in motor function (Gibson et al., 2022; Papoutsi et al., 2014; Williams et al., 2007). However, when HD is understood and treated as a disease that primarily affects motor skills, without also taking the progressive psychiatric, behavioural and cognitive decline into account, the children who are next of kin may not receive the help and support they need. This may occur because support systems understand and help young carers based on the different needs they have if the parent suffers from mental versus physical illness. Commonly, the emotional effect of having a parent with mental disorder is considered larger. It includes experiencing more conflictual family relationships, having a lower quality of life, and experiencing more adverse effects in their daily lives compared to having a parent with a physical disability (Brockington et al., 2011; Van Loon et al., 2014). The findings of the research in this thesis suggests that although HD might present as a somatic disease on the surface, cognitive and neuropsychiatric symptoms are an integral part of the neurodegeneration related to the disease, particularly from the perspective of children.

The findings may also reflect issues related to how any disease with symptoms that progress unnoticeably is met by public health and support systems. Research suggests that the help and support patients and their families receive can be dependent on the visibility of their needs. For example, one study showed how patients received more help and support after a traumatic brain injury if their needs are particularly visible (Andelic et al., 2014). Also, another study show that young carers for parents with other neurodegenerative diseases also often feel invisible and unacknowledged (Bjorgvinsdottir & Halldorsdottir, 2014). In this context, it may be difficult for any child whose parent has a disease with an ‘invisible’ progression of symptoms to be understood and have their support needs to be met. However, the findings of this research show that having a parent with HD specifically can produce a range of factors that might complicate these underlying issues further. These factors include, for example, how HD can develop slowly and subtly with an ‘invisible’ neuropsychiatric and cognitive decline that may last for years before more evident symptoms present (Papers I, II and III). Also, how the parent may seem relatively unaffected by the disease in many settings of their lives, and how the parent may be unaware of their problems and the problems the disease creates for their children.

These findings highlight an important way to conceptualise risk and potential means of protection for this group of young people. It shows a potential vulnerability in broader contexts of resources when they are not equipped to understand or meet the needs of young people who have parents with complex progression, such as HD. Specifically, there is a risk that the adversity they experience may unfold while they remain 'invisible' to their surroundings, effectively hiding their support needs from resources that can be found in broader systems of support. In response to these findings, future support for this group should focus on strengthening resources that align with how the disease develop. This would require available support systems to identify and respond to potential needs for children related to all parts of the development of HD, including the potential for early emergence of neuropsychiatric and cognitive disruption. Importantly, available support should be adapted to the child's potentially changing needs in line with the progressive nature of their parent's disease.

### 8.3.2. The child's right to know and be safe versus the parent's right not to know or tell

In addition to resources that can be found in the public health-care system, other social or policy contexts also carry the responsibility of ensuring that children grow up within developmental settings in which they are safe from harm (United Nations Convention on the Rights of the Child, 1989). Within these broader contexts of resources, this research may first and foremost reveal something about dilemmas that can make it difficult for young people with a parent with HD to use the protection their legal rights should provide. For example, the young people's accounts showed how HD's slow and subtle symptoms seemingly crept in on their caregiving relationships, often over the years, without any viable explanation (Papers I and II). In terms of the caregivers, their accounts show how they had to keep the family secret about HD, as well as from their children because they were uncertain about whether the disease was present (Paper II). In some cases, the caregiver without HD knew about their partner's disease or risk but did not feel entitled to be the one to tell (Paper II). The accounts of both the offspring and caregivers also indicated how formal healthcare and support providers had generally not understood the issues they were facing or even been aware that the young people had a parent with a severe disease progressing behind the four walls of their homes (Papers II and III).

These accounts support one side of ethical and legal dilemmas – that is, the child's legal rights to grow up in circumstances in which they feel safe and are protected and to be

informed when their parent suffers from severe disease (e.g., Health Personnel Act, 1999 §10a; United Nations Convention on the Rights of the Child, 1989). However, our findings do not show or explain that running in parallel with the legal and ethical rights of the child are the rights of the parent with HD or who is at risk of having inherited HD. For their part, they are in their full ethical right not to know about their genetic information (Ost, 1984) and in their full legal right for health personnel not to share this information with others (e.g., Health Personnel Act, 1999). How genetic information is administered and how others' right to know, such as family members, should be considered has been a topic of debate and the focus of many legal battles (Lucassen & Clarke, 2022). The topic is fraught because, with the moral and legal rights of parents not knowing or sharing information about their potential to develop a severe genetic disease, the risk of serious harm to their children also lies as the child's ability to cope and adjust might depend on this information being shared to receive the support they need (Andorno, 2004). This may not be a dilemma for many children whose parents present with other types of disease, such as cancer or physical disabilities. However, diseases for which early changes in the brain present challenges with insight and perspective taking, such as neurodegenerative disease, traumatic brain injury, or psychosis-related disorders, inherent dilemmas between the rights of the child and the rights of the adult may emerge. By law, the child's rights outweigh any other concerns (The Human Rights Act, 1999). Nevertheless, this can present a practical dilemma for healthcare workers and those in other formal support channels. For example, public healthcare systems are obliged to protect the needs of children to be informed and safe but are, at the same time, also obliged to protect the rights of parents. Complicating the situation further is that what constitutes the actual development of HD is often blurred and at times not understood even by the parent who is developing the disease. Also, the disease seemed to frequently be a secret, both inside and outside the family, potentially making it difficult or impossible for formal support systems to be aware that these individuals' children might need help and support (Paper II and III).

These dilemmas raise questions and do not necessarily provide answers. However, based on the findings, it appears that when these dilemmas are not solved in favour of children, they can have severe negative consequences for that child (Papers I, II and III). The research in this thesis may therefore demonstrate some of the ways that the resources meant to protect this group of children can be compromised or hindered by the nature of the disease. In light of these dilemmas, future responses to promote protective factors for this group should include an increased understanding about how they can occur. Also, when possible, work

should be done preventively with parents that promote the importance of sharing information about disease in these broader contexts.

## 8.4. Implications

### 8.4.1. A call to minimise risk and maximise protection

The overarching message regarding the implications of this research is a call to minimise the chances of co-occurring risks in these young people's lives and maximise the resources and protection they can find in themselves and others to cope and adjust. In this sense, future interventions can be guided by risk and protective frameworks, and focus on minimising risk by strengthening individual and external factors that can work to protect these potentially vulnerable young people (Fraser et al., 1999). As with any child who grows up with a parent with severe disease, we cannot necessarily remove challenges related to these children's role as next of kin or the changes the disease will cause in their parents. However, much of the overwhelming feeling and maladjustment described by our participants arose from adverse experiences of not having support in relationships to cope and adjust. Such support environments have the potential to be purposely developed and transformed into strengthening contexts of social resources (Shern et al., 2016). The most urgent call for this type of intervention is to assess and build strength in their caregiving relationships. However, strengthening resources available at all levels, including formal systems of support, is needed, as is the enforcement of their legal and ethical rights.

### 8.4.2. Rare disease within understandable concepts

It is often difficult for any young person affected by a rare disease to obtain the help and support they need, as formal support systems tend to feel unequipped to approach conditions they know little about (von der Lippe et al., 2017). Therefore, a major contribution of this research is the potential benefits that can come with conceptualising the experiences of young people with a parent with HD as comparable to the experiences of any child facing adversity. Through this framework, these young people's experiences with both risk of adversity and potentials for protection can be understood by concepts that easily translate to support systems, such as child protective services, doctors, and psychologists. For example, terminology such as adversity, potentially traumatic experiences, co-occurring risk, overwhelming stress, and the need to strengthen social support resources can provide an already established understanding of the nature of what help and support these young people might need. These concepts also relate to empirically validated interventions to help children

with similar experiences cope and adjust. For example, as with other children at risk of adversity, interventions should strengthen resources within each child, each parent-child relationship, and each broader social context (Thompson et al., 2015). In this way, young people with a parent with HD may have much in common with other groups of children who grow up in developmental contexts that could lead to vulnerabilities. For other groups of vulnerable children, knowledge already exists to predict the outcomes if they do not have the resources available to cope and adjust, as well as the effectiveness of interventions that can be applied. Most importantly, the road to ensuring accurate and sufficient help and support for young people with a parent with HD may not be to come up with new interventions, but to connect these young people to resources found in systems of support that are already in place.

### 8.4.3. Future directions

The research in this thesis leaves several important questions that should be of interest to future studies. For example, more knowledge is needed to apply a framework of risk and protection to validate already existing interventions as potential means to support this group as well. These studies should also validate potential interventions for the whole family, particularly when it comes to measures that can build strength and skills in caregivers without HD. Based on the findings about the level of adversity many of these young people had experienced, it would also be interesting to explore how these experiences potentially impact their lives as adults. Because many of the young people who grow up with a parent with HD unfortunately are at risk of developing the disease themselves, research that asks questions about how overwhelming adversity and distress in childhood plays into the symptomatology and coping of adults who develops the disease could also be interesting. As a final point, Paper II shows how our understanding of a phenomena is changed and nuanced when it is investigated using different perspectives. Future research that continues to apply multiple perspectives is therefore needed to increase our understanding about the dynamic nature of how these young people grow up. For example, including the perspectives of parents with HD or at risk of developing HD could be of interest.

## 8.5. Methodological Issues

As with any research project, the research in this thesis has both strengths and challenges that should be addressed. The following section will focus on some of the main issues related to this research as a whole.

### 8.5.1. Ethical issues

Research needs to carefully consider how to avoid bringing extra burdens to the lives of vulnerable groups of people through their participation in the study. However, when research addresses vulnerable groups of participants, ethical dilemmas will inevitably be a part of the research process (Aldridge, 2014). The current research was conducted with potentially vulnerable young people and adults and involved talking about many aspects of their lives, including distressing experiences, thoughts and feelings. An ethical dilemma throughout the study has been the possibility of causing further negative psychological impact on participants. For example, interviewing people in times of crisis and stress and asking them to talk in detail about their experiences can be challenging for both the researcher and the participants. One of the ways this dilemma was navigated was conducting the interviews with flexibility and sensitivity, and by health personnel or students equipped to 'handle' the nature of the topics and emotional reactions that could follow. Also, relevant referrals or subsequent follow-ups were arranged if necessary.

The research was conducted to contribute to an understanding that would help children and families with HD and not to contribute to stigmatisation or further distress. All researchers have an ethical responsibility to avoid misunderstandings and enhance the possibilities of the findings being interpreted and used in valid ways (Kvale & Brinkmann, 2009). However, there is an inherent dilemma in our findings as this research reports on adverse experiences and risks for children related to their experiences with parents and families suffering from disease. In terms of this ethical dilemma, the intention of this research should be clarified. Giving a voice to children who are or have been next of kin to a parent with a severe disease is, in a sense, about empowering the expertise these children have developed from their experiences to allow others to benefit from this knowledge in the future (Aldridge, 2008). This research should therefore reflect the urgency for a person with HD, their families and their children to obtain the help and support they need, and not as creating stigma regarding the lives of parents who, by no fault of their own, suffer from a severe disease. Moreover, as this group has remained largely 'invisible' to health-care professionals and other systems of support, there is a need for scientific knowledge to promote the implementation of future policy that can help this vulnerable group. In light of the ethical dilemmas related to ensuring the dignity of parents and families with HD, I have tried to carefully consider how this research *reflects* on the participants and their families through data collection, analysis and the dissemination of findings. For example, it was important for me that the results would reflect nuances, and the interest has therefore been to describe the

meaning involved in the risk posed to these young people's lives as well as potential means to be protected from such risk through social support.

### 8.5.2. External validity: Transferability

External validity addresses the relevance that research can have to other settings and contexts (Malterud, 2001). This relates to the concept of transferability, which is perhaps most often known as *generalisability*, but is quite different in qualitative as opposed to quantitative research. In qualitative research, the derived knowledge can never effortlessly be generalised to explain the past or predict the future across all cases (Tracy & Hinrichs, 2017). Instead, qualitative research produces knowledge that will be found within people, contexts and cultures. However, this does not mean that the presented findings cannot be useful in other settings. In the implications, I stated how general conceptualisations of child development in contexts of adversity and distress could be used to inform our understanding of the developmental contexts of young people with a parent with HD. In this sense, the findings can also be used to generalise in the other direction. That is, by placing the results within well-known psychological frameworks of development, stress, risk, vulnerability and social support, the findings can also offer insights into other groups of children with similar experiences. For example, the findings from Paper I can be used to understand how any child who has a parent with any severe disease that affects their mental health and physical health can also experience adversity and a lack of support within other close relationships at the same time. Findings from Paper II can be used to unpick the complex dynamics involved in talking with children about difficult issues, such as disease, within any family. Similarly, findings from Paper III can be used to clarify the mechanisms by which any child who experiences adversity can be supported by relationships outside the caregiving context or be hindered from being supported in the ways that they need.

This research also increase the understanding about how symptoms of neurodegenerative disease is experienced by children with parents who are affected, and brings forth potential dilemmas that can come with meeting these young people's needs for support, both inside and outside the family context. These issues may be transferrable to better understand other children with other types of neurological disease. For example, about 10 000 children in Norway grow up with a parent with Multiple Sclerosis (MS) in Norway (Grytten et al., 2015). If we for example look at children impacted by having a parent with neurological disease in general, including MS, traumatic brain injury, and early onset dementia, the findings can have the potential to be used to better understand the experiences

and needs for strengthened systems of support for a relatively large and historically understudied group of vulnerable young people.

### 8.5.3. Internal validity: Trustworthiness

Internal validity addressed whether the methods used in a study appropriately investigated and reported on the research aims (Malterud, 2001). Said in another way, if the findings and the process to get to these findings are trustworthy.

#### *8.5.3.1.Credibility*

Credibility refers to whether results are plausible and whether the findings have been described in a way that makes them understandable to the reader (Tracy, 2010). Several steps were taken to ensure the credibility of this research. For example, to standardise, avoid vagueness and increase understanding and implementation of the analysis, data were approached in ways inspired by the guidelines of Consensual Qualitative Research (CQR), as suggested by Hill et al. (1997, 2005). Qualitative research is strengthened by having the process evaluated from different perspectives, which is referred to as triangulation (Patton, 1999). Two forms of triangulation were used to strengthen the validity of this research. First, different researchers served as discussants for the analyses to provide different perspectives on the data. Second, in Paper II, the topic of conversations about disease was explored from two different perspectives – that of the young person and of the caregiver.

Another central point in ensuring the credibility of the findings is that they correspond to how the group under study understands the same topics (Tracy, 2010). Therefore, feedback and collaboration with member experts and clinical experts have been invaluable to ensure credibility. This input provided an opportunity to test research findings but also for me to collaboratively develop my understanding of the phenomena I was studying (Tracy, 2010). Further, considering that the interviews were conducted in the participants' native language, Norwegian, the results had to be disseminated, and representative quotes had to be translated from the language they were originally presented in. To ensure credibility of this representation, it was important to make the least possible changes through this translation so that the nuances would not be lost and the individual's style was maintained. At the same time, because of the personal nature of the stories and how imbedded our understanding is in our culture and language, some local expressions were difficult to translate. To the best of my abilities, I have tried to present the meaning through translation to facilitate the credibility of the translated quotes.

#### *8.5.3.2. Informational power*

In qualitative research, as in any research, the research process requires planning and ascertaining how many participants should be included. This research included 50 participants (36 offspring and 14 caregivers), which can be considered as a relatively large sample size in qualitative research. The choices made regarding sample size included in all three papers to relate to what is called informational power. Informational power refers to the ability of the number of participants to inform us about the phenomena under investigation (Malterud et al., 2016). As in all studies, the number of people needed for inclusion in a study depends on the study's aims. When the aim and participants of a study are specific and strongly supported by established theory and the analysis wants to provide in-depth accounts of the experiences of single individuals, smaller sample sizes are needed (Malterud et al., 2016). In contrast, to provide informational power in studies which includes broad aims, with an innately more heterogeneous group and which looks to make inferences across participants, larger sample sizes are needed. For these reasons, I argue that a larger sample size is justified to provide the informational power needed in this study to ensure validity of the results.

#### *8.5.3.3. Can we trust memories?*

The presented findings rely on memories from both current and distant childhoods. While this broad selection of perspectives comes with various benefits, such as allowing inferences about the population under study that could be relevant across time, settings and place, it also leads to the question: Can these memories be trusted?

The validity of this type of retrospective data has been debated for several reasons. This is because the way anyone remembers events is as much a subjective construction as the account of any actual events happening. By nature, memories are filled with bias and will develop and change with time, just as we do (Schacter, 1999). Moreover, many factors influence how we remember experiences and events. For example, stress will affect almost all processes involves in creating memories, including how, and even if, something is remembered (Shields et al., 2017).

However, such scepticism to biases in using retrospective data should not necessarily apply, considering the methodological nature of this study. As a qualitative study, the aim was to explore the meaning of the participants' experiences in the ways that they recalled them and not necessarily to investigate the truthfulness or objective recall of the experienced events (Polkinghorne, 2005). A person will reflect differently on experiences when they are living through them compared to how they see these experiences with the passage of time. As this

research aimed to investigate a breadth of experiences of a phenomenon, I wanted to capture both reflections that changed with time and understanding and current childhood experiences. In this sense, our results could reflect a core of experiences that potentially are valid across time, place and context. Consequently, in this research all the participants' memories equally and validly reflect their true experiences and, importantly, the true meaning they subjectively made of these experiences.

### 8.5.4. Reflexivity

The most important tool within any field of research is arguably the researcher (Henwood et al., 1994; Malterud, 2001). Therefore, in qualitative research, the influence the researcher have on all steps of the research process should not be ignored. Rather, the researcher's possible impact on the study should be analysed and communicated so that subjectivity can be used as a resource rather than a threat. This process is referred to as *reflexivity* (Braun et al., 2019; Malterud, 2001). In the methodology chapter, I described some of the main ways reflexivity, fidelity and consistency were employed in the planning and gathering of interview data. However, reflexivity is a methodological tool that does not start or end with this process as the researcher will continue to influence the research process at each step.

Self-reflexivity encourages the researcher to be open about their own strengths and shortcomings and to address the characteristics and attitudes that they bring into the entire research process (Finlay & Gough, 2008; Tracy, 2010). In this sense, sincerity reflects research that is honest and transparent about the researcher's own biases and goals and how these could have played a role in the research process (Tracy, 2010). In this research, I have interpreted constructs and meanings from the participants' experiences. These interpretations relied upon abstractions and generalisations made with my ability to understand and reflect upon the topics I aimed to explore. For me as a researcher, the concept of reflexivity has been neither an easy nor a straightforward process. In hindsight, I can see how I was, in many ways, unprepared for the level of understanding and adaptability that working with people's life stories at the phenomenological level would demand of me as a researcher in producing this thesis. Having conducted mainly quantitative research in the past, as well as having worked within a hospital setting that deals with medical and genetic diagnoses, gaining a form of understanding of how the principles of reflexivity are applied in qualitative studies has taken both practice and time for me. What follows are some of the main issues that I have had to address regarding my role throughout this research process.

I, simply through my person, bring many characteristics into the research setting that have the possibility of swaying the way I interpret the data. I am a white female who grew up in a fairly isolated small town in Norway. I have not been exposed to disease, distress or despair in the way those whose narratives I interpret have. My exposure to any type of disease is largely from a clinical and medical perspective – that is, reading about disease and working with people affected by disease, not being one of those who live with it. I am, however, fortunate to be curious by nature and always open to new learning experiences, and I was fully invested in understanding this mystical research tool 'reflexivity', which seemed to come with no set recipe nor any step-by-step guidelines compared to the quantitative research tools I have used in the past. Thankfully, as a novice qualitative researcher, I was encouraged to always think, reflect, learn and evolve (Braun & Clarke, 2019). With openness of mind, and as I continued to gain knowledge about the phenomenon under study and the people who lived with it, I kept in mind that I needed to continue to ask questions about all that I learned and to remain reflexive. I asked questions about the foundation of the research, the research process, the analysis and myself. Examples of my quest to further my knowledge and insight into the topic under study included being an observer in different clinical settings that work with families with HD, such as genetic counselling sessions for individuals wanting to test for the genetic mutation and family advice sessions at Oslo University Hospital. I also had the pleasure of attending several meetings and social gatherings for members of the Norwegian Association of Huntington's Disease, becoming acquainted with people with HD, with the uncertainty of HD or with futures with HD and those that are affected as carers in different capacities outside an interview setting.

With the patient guidance of supervisors, who helped by continuously challenging my processes of thought, I slowly but surely noticed changes in how I conceptualised the research – changes in my previous perceptions, the questions I had asked, the assumptions I had made and how I talked about and presented my research. I became more curious about all impressions and thoughts on the topics at hand. Sometimes I took two steps forward and one step back, but still, I moved in a direction I had not moved before. What I felt I had discovered was that my findings throughout the research process were not necessarily conclusions, but rather that everything that was discovered produced endless opportunities for more questions, further understanding and other points of view. I came to understand that there is no such thing as a perfect project plan, a perfect interview or conclusions made through analysis without exceptions. I realised that the way to go, instead, was to continue to revise all aspects of the research – and ourselves – as we discover new things and make new

meanings. In the wake of this project, I cannot say that the process could not have been carried out differently, nor can I say that I have provided every answer and every understanding to the phenomena I have explored. Luckily, I have come to learn that qualitative research is a continuous journey – one I hope to continue to learn from in the years to come.

### 8.5.5. Strengths and limitations

A major strength of the research is that it provides in-depth information on a generally understudied vulnerable group of children. The study included a broad range of participants who presented different perspectives (offspring and caregivers), reflecting on current and past experiences. However, this research also presents the experiences of participants who, in different ways, received an invitation to participate through channels by which families with HD receive formal or informal health-care services and/or support. The findings must therefore be considered within this context. For example, the participants may include a higher number of people with severe adverse experiences because without these issues, they may not have felt the need to be connected to formal or informal channels of support. Consequently, young people or caregivers in other families with HD may have different experiences from those conveyed in this research.

Perhaps the most significant strength of this thesis is the possibilities it offers to conceptualise the experiences of a rare, understudied group of potentially vulnerable children within a framework of risk and protection. However, as with the development of any child, these young people's experiences will always be much more complex than clear-cut frameworks of understanding. For example, the risk that comes with the potential to inherit HD will be a major source of distress in any young person's life, regardless of the systems of support available to help them cope with this.

I did not use participant or parent characteristics as an analytical tool to compare individual differences. Both the offspring and caregivers who participated in this study are, by nature, groups with common and diverse features. It is important to note that diversity in their experiences aligns with the nature of HD. For example, the disease affects people at a wide range of ages, and the symptoms will present differently for each person who develops the disease. However, variance that relates to the time in childhood when our participants' parents started to develop HD, strength and symptom manifestation and gender (of both the parent with HD and the child) have not been explored. Moreover, I included teenagers with current experiences and adults remembering experiences from the past. Although the experiences

recounted by older participants did not necessarily reflect on contemporary social, societal, legal or ethical frameworks of support, the participants consistently addressed similar topics, regardless of age. Nevertheless, these are elements that should be explored further by future research.

## 9. Concluding Remarks

This thesis aimed to investigate experiences of being a young person with a parent with HD. This aim was guided by knowledge about this group's unmet needs for support and an understanding of the many and often devastating issues that could be present in their lives by having a parent with this inheritable progressive life-threatening disease. Despite these contributions, there has not been sufficient research that qualitatively describes the potentially dynamic nature of these experiences. The research journey began by asking questions about this group based on theoretical and empirical frames of reference regarding any child's developmental processes when faced with adversity and distress. The research, which included three distinct, but connected topics, comes together to shed light on how children experience living with a parent with HD and to make it understood as a at risk for complex combinations of co-occurring adversity that can and should be addressed through existing support systems.

The nature of our participant's experiences related to adversity that could come with having a parent with HD, but also included the co-occurring risk of not having available support in other relationships and contexts. This adversity involved lack of support in relationships with caregivers without HD and extended family relationships, as well as the adverse experience of not being understood and not having their needs met within broader support contexts. However, the research also showed how resources when found within themselves and others could work as protective factors. Social support in caregiving relationships played a major role as a potentially protective factor in distress, although other relationships outside this context could also serve such a function. Together, the findings underscore important ways that psychological risk presented by having a parent with HD can mean different things to young people with a parent with the disease. That is, that this groups adverse experiences in childhood can be subject to the dynamics of interactions with others.

To advance much-needed support and help to this group, the findings highlight an urgent need for responses to minimise co-occurring risks in these young people's lives and to maximise the resources they can find in themselves and others. Suggestions on how to maximise this type of buffer or protection include strengthening caregiving relationships and adaptive coping mechanisms in children at an individual level. It includes providing support that aligns with how young people experience HD symptoms, which will involve addressing the neuropsychiatric and cognitive decline as a part of children's early and overall experiences with the disease. Suggestions also include acknowledging complex dilemmas related to the

availability of support for this group, in close as well as broad contexts of resources. Overall, the findings provide the potential to conceptualise these young people's experiences with a rare disease within frameworks of how we would understand any vulnerable child who grows up with risk of adversity. Thus, providing a potential to connect these young people and their families to systems of support and means of intervention that are already in place.

## References


Aldridge, J. (2008). All work and no play? Understanding the needs of children with caring responsibilities. *Children & Society*, *22*(4), 253–264. https://doi.org/10.1111/j.1099-0860.2007.00094.x

Aldridge, J. (2014). Working with vulnerable groups in social research: Dilemmas by default and design. *Qualitative Research*, *14*(1), 112–130. https://doi.org/10.1177/1468794112455041

Andelic, N., Soberg, H. L., Berntsen, S., Sigurdardottir, S., & Roe, C. (2014). Self-perceived health care needs and delivery of health care services 5 years after moderate-to-severe traumatic brain injury. *PM&R*, *6*(11), 1013–1021. https://doi.org/10.1016/j.pmrj.2014.05.005

Andorno, R. (2004). The right not to know: An autonomy based approach. *Journal of Medical Ethics*, *30*(5), 435–439. https://doi.org/10.1136/jme.2002.001578

Barette, J., & Marsden, C. D. (1979). Attitudes of families to some aspects of Huntington's chorea. *Psychological Medicine*, *9*(2), 327–336. https://doi.org/10.1017/S0033291700030841

Bhattacharyya, K. B. (2016). The story of George Huntington and his disease. *Annals of Indian Academy of Neurology*, *19*(1), 25-28. https://doi.org/10.4103/0972-2327.175425

Bjorgvinsdottir, K., & Halldorsdottir, S. (2014). Silent, invisible and unacknowledged: Experiences of young caregivers of single parents diagnosed with multiple sclerosis. *Scandinavian Journal of Caring Sciences*, *28*(1), 38–48. https://doi.org/10.1111/scs.12030

Boullier, M., & Blair, M. (2018). Adverse childhood experiences. *Paediatrics and Child Health*, *28*(3), 132–137. https://doi.org/10.1016/j.paed.2017.12.008

Braun, V., & Clarke, V. (2019). Reflecting on reflexive thematic analysis. *Qualitative Research in Sport, Exercise and Health*, *11*(4), 589–597. https://doi.org/10.1080/2159676X.2019.1628806

Braun, V., & Clarke, V. (2016). (Mis)conceptualising themes, thematic analysis, and other problems with Fugard and Potts' (2015) sample-size tool for thematic analysis. *International Journal of Social Research Methodology*, *19*(6), 739–743. https://doi.org/10.1080/13645579.2016.1195588

Braun, V., & Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative Research in Psychology*, *3*(2), 77–101. https://doi.org/10.1191/1478088706qp063oa

Brinkmann, S., & Kvale, S. (2005). Confronting the ethics of qualitative research. *Journal of Constructivist Psychology, 18*(2), 157–181. https://doi.org/10.1080/10720530590914789

Brockington, I., Chandra, P., Dubowitz, H., Jones, D., Moussa, S., Nakku, J., & Ferre, I. Q. (2011). WPA guidance on the protection and promotion of mental health in children of persons with severe mental disorders. *World Psychiatry, 10*(2), 93–102. https://doi.org/10.1002/j.2051-5545.2011.tb00023.x

Bronfenbrenner, U., & Morris, P. A. (2006). The bioecological model of human development. In Damon W, Lerner RM, editors. Handbook of Child Psychology. New Jersey (US): John Wiley & Sons; 2007. doi:10.1002/9780470147658.chpsy0114

Centre for Rare Disorders. (2019). *Huntingtons sykdom*. https://oslo-universitetssykehus.no/seksjon/Senter-for-sjeldne-diagnoser/Documents/Huntingtons%20sykdom/Huntington-veileder_2019.pdf

Chikhradze, N., Knecht, C., & Metzing, S. (2017). Young carers: Growing up with chronic illness in the family – A systematic review 2007–2017. *Journal of Compassionate Health Care, 4*(1), 1–16. https://doi.org/10.1186/s40639-017-0041-3

Cleckley, H. M. (1951). The mask of sanity. *Postgraduate Medicine*, *9*(3), 193–197. https://doi.org/10.1080/00325481.1951.11694097

Dalton, L., Rapa, E., Ziebland, S., Rochat, T., Kelly, B., Hanington, L., Bland, R., Yousafzai, A., Stein, A., Betancourt, T., Bluebond-Langner, M., D'Souza, C., Fazel, M., Fredman-Stein, K., Harrop, E., Hochhauser, D., Kolucki, B., Lowney, A. C., Netsi, E., & Richter, L. (2019). Communication with children and adolescents about the diagnosis of a life-threatening condition in their parent. *The Lancet, 393*(10176), 1164–1176. https://doi.org/10.1016/S0140-6736(18)33202-1

Danese, A., McEwen, B. S. (2012). Adverse childhood experiences, allostasis, allostatic load, and age-related disease. *Physiology & Behavior, 106*(1), 29–39. https://doi.org/10.1016/j.physbeh.2011.08.019

Darwin, C. (1877). A Biographical Sketch of an Infant. *Scientific American, 4*(86), 1372-1373. https://doi.org/10.1038/scientificamerican08251877-1372bsupp

Driessnack, M., Williams, J. K., Barnette, J. J., Sparbel, K. J., & Paulsen, J. S. (2012). Development of the HD-teen inventory. *Clinical Nursing Research, 21*(2), 213–223. https://doi.org/10.1177/1054773811409397

Duncan, R. E., Gillam, L., Savulescu, J., Williamson, R., Rogers, J. G., & Delatycki, M. B. (2007). "Holding your breath": Interviews with young people who have undergone

predictive genetic testing for Huntington disease. *American Journal of Medical Genetics, 143*(17), 1984–1989. https://doi.org/10.1002/ajmg.a.31720

Ellison, M. (2017). The impact of Huntington disease on young people. *Handbook of Clinical Neurology, 144*, 179 –182. https://doi.org/10.1016/B978-0-12-801893-4.00015-8

Epping, E. A., Kim, J.-I., Craufurd, D., Brashers-Krug, T. M., Anderson, K. E., McCusker, E., & PREDICT-HD Investigators and Coordinators of the Huntington Study Group. (2016). Longitudinal psychiatric symptoms in prodromal Huntington's disease: A decade of data. *American Journal of Psychiatry*, *173*(2), 184–192. https://doi.org/10.1176/appi.ajp.2015.14121551

Fanti, K. A. (2011). Transactional Models. In R. J. R. Levesque (Ed.), *Encyclopedia of adolescence* (pp. 3003–3013). Springer.

Felitti, V. J., Anda, R. F., Nordenberg, D., Williamson, D. F., Spitz, A. M., Edwards, V., & Marks, J. S. (1998). Relationship of childhood abuse and household dysfunction to many of the leading causes of death in adults: The adverse childhood experiences (ACE) study. *American Journal of Precentive Medicine, 14*(4), 245–258. https://doi.org/10.1016/S0749-3797(98)00017-8

Fergus, S., & Zimmerman, M. A. (2005). Adolescent resilience: A framework for understanding healthy development in face of risk. *Annual Review of Public Health, 26*(1), 399–419. https://doi.org/10.1146/annurev.publhealth.26.021304.144357

Finkelhor, D., Ormrod, R. K., & Turner, H. A. (2007). Poly-victimization: A neglected component in child victimization. *Child Abuse & Neglect*, *31*(1), 7–26. https://doi.org/

Finkelhor, D., Shattuck, A., Turner, H. A., Ormrod, R., & Hamby, S. L. (2011). Polyvictimization in developmental context. *Journal of Child & Adolescent Trauma, 4*(4), 291–300. https://doi.org/10.1016/j.chiabu.2006.06.008

Finlay, L., & Gough, B. (2008). *Reflexivity: A practical guide for researchers in health and social sciences*. John Wiley & Sons.

Folkman, S. (2020). Stress: Appraisal and coping. In *Encyclopedia of behavioral medicine* (pp. 2177–2179). Springer.

Forrest Keenan, K., McKee, L., & Miedzybrodzka, Z. (2015). Help or hindrance: young people's experiences of predictive testing for Huntington's disease. *Clinical Genetics, 87*(6), 563–569. https://doi.org/10.1111/cge.12439

Forrest Keenan, K., Miedzybrodzka, Z., Van Teijlingen, E., McKee, L., & Simpson, S. A. (2007). Young people's experiences of growing up in a family affected by

Huntington's disease. *Clinical Genetics, 71*(2), 120–129. https://doi.org/ 0.1111/j.1399-0004.2006.00702.x

Fraser, M. W., Galinsky, M. J., & Richman, J. M. (1999). Risk, protection, and resilience: Toward a conceptual framework for social work practice. *Social Work Research, 23*(3), 131–143. https://doi.org/ https://doi.org/10.1093/swr/23.3.131

Gellman, M. D. (2020). Behavioral Medicine. In *Encyclopedia of behavioral medicine* (pp-223-226). Springer International Publishing. https://doi.org/10.1007/978-3-030-39903-0_1660

Gibson, J. S., Rhoten, B. A., Ridner, S. H., & Claassen, D. O. (2022). Perceived effects of neuropsychiatric symptoms on functional status in early-stage Huntington disease. *Western Journal of Nursing Research, 44*(2), 141–150. https://doi.org/ https://doi.org/10.1177/0193945921992545

Gopnik, A., O'Grady, S., Lucas, C. G., Griffiths, T. L., Wente, A., Bridgers, S., Aboody, R., Fung, H., & Dahl, R. E. (2017). Changes in cognitive flexibility and hypothesis search across human life history from childhood to adolescence to adulthood. *Proceedings of the National Academy of Sciences, 114*(30), 7892–7899. https://doi.org/10.1073/pnas.1700811114

Grytten, N., Torkildsen, Ø., & Myhr, K. M. (2015). Time trends in the incidence and prevalence of multiple sclerosis in Norway during eight decades. *Acta Neurologica Scandinavica*, 132, 29-36. https://doi.org/10.1111/ane.12428

Hammersley, M. (2015). Research ethics and the concept of children′s rights. *Children & Society*, *29*(6), 569–582. https://doi.org/10.1111/chso.12077

Hartelius, L., Jonsson, M., Rickeberg, A., Laakso, K. (2010). Communication and Huntington′s disease: Qualitative interviews and focus groups with persons with Huntington′s disease, family members, and carers. *International Journal of Language & Communication Disorders, 45*(3), 381–393. https://doi.org/10.3109/13682820903105145

Health Personnel Act. (1999). https://lovdata.no/

Henwood, K. (1994). Beyond the qualitative paradigm: A framework for introducing diversity within qualitative psychology. *Journal of Community & Applied Social Psychology, 4*(4), 225–238. https://doi.org/ 1052-9284/94/040225-14

Horstman, H. K., Hays, A., & Maliski, R. (2016). Parent-child interaction. In J. F. Nussbaum (Ed.), *Oxford research encyclopedia of communication.* Retrieved from http://communication.oxfordre.com/view/10.1093/acrefore

Hughes, K., Bellis, M. A., Hardcastle, K. A., Sethi, D., Butchart, A., Mikton, C., Jones, L., & Dunne, M. P. (2017). The effect of multiple adverse childhood experiences on health: A systematic review and meta-analysis. *The Lancet Public Health, 2*(8), 356–366. https://doi.org/10.1016/S2468-2667(17)30118-4

Hughes, K., Ford, K., Bellis, M. A., Glendinning, F., Harrison, E., & Passmore, J. (2021). Health and financial costs of adverse childhood experiences in 28 European countries: a systematic review and meta-analysis. *The Lancet Public Health, 6*(11), 848–857. https://doi.org/10.1016/S2468-2667(21)00232-2

Huntington, G. (1872). *On chorea. The Medical and Surgical Reporter of Philadelphia.* Butler.

Huntington Society of Canada. (1996). *Understanding Huntington's disease: A resource for families.* Huntington Society of Canada.

Ignelzi, M. (2000). Meaning-making in the learning and teaching process. *New Directions for Teaching and Learning*, *2000*(82), 5–14. https://doi.org/ doi.org/10.1002/tl.8201

Jona, C. M., Labuschagne, I., Mercieca, E.-C., Fisher, F., Gluyas, C., Stout, J. C., & Andrews, S. C. (2017). Families affected by Huntington's disease report difficulties in communication, emotional Involvement, and problem solving. *Journal of Huntington's Disease*, *6*(3), 169–177. https://doi.org/10.3233/JHD-170250

Kalmakis, K. A., & Chandler, G. E. (2014). Adverse childhood experiences: Towards a clear conceptual meaning. *Journal of Advanced Nursing*, *70*(7), 1489–1501. https://doi.org/ 10.1111/jan.12329

Kavanaugh, M. S. (2014). Children and adolescents providing care to a parent with Huntington′s disease: Disease symptoms, caregiving tasks and young carer well-being. *Child & Youth Care Forum, 43*(6), 675–690. https://doi.org/ 10.1007/10566-014-9258-x

Kavanaugh, M. S., Noh, H., & Studer, L. (2015). "It'd be nice if someone asked me how I was doing. Like, 'cause I will have an answer": Exploring support needs of young carers of a parent with Huntington's disease. *Vulnerable Children and Youth Studies, 10*(1), 12–25. https://doi.org/10.1080/17450128.2014.980370

Keenan, K. F., van Teijlingen, E., McKee, L., Miedzybrodzka, Z., & Simpson, S. A. (2009). How young people find out about their family history of Huntington′s disease. *Social Science & Medicine, 68*(10), 1892–1900. https://doi.org/10.1016/j.socscimed.2009.02.049

Kegan, R. (1980). Making meaning: The constructive-developmental approach to persons and practice. *The Personnel and Guidance Journal, 58*(5), 373–380. https://doi.org/ 10.1002/j.2164-4918.1980.tb00416.x

Kikusui, T., Winslow, J. T., & Mori, Y. (2006). Social buffering: Relief from stress and anxiety. *Philosophical Transactions, 361*(1476), 2215–2228. https://doi.org/ 10.1098/rstb.2006.1941

Kvale, S., & Brinkmann, S. (2009). *Interviews: Learning the craft of qualitative research interviewing*. Sage.

Larkin, M., Watts, S., & Clifton, E. (2006). Giving voice and making sense in interpretative phenomenological analysis, *3*(2), 102–120. https://doi.org/10.1191/1478088706qp062oa

Lazarus, R. S. (1993). Coping theory and research: Past, present, and future. https://www.sunswlondon.nhs.uk/wp-content/uploads/2013/01/Coping-Process-Theory.pdf

Lazarus, R. S., & Folkman, S. (1984). *Stress, appraisal, and coping*. Springer.

Leu, A., Frech, M., Wepf, H., Sempik, J., Joseph, S., Helbling, L., Moser, U., Becker, S., & Jung, C. (2019). Counting young carers in Switzerland – A study of prevalence. *Children & Society*, *33*(1), 53–67. https://doi.org/ 10.1111/chso.12296

Lewit-Mendes, M. F., Lowe, G. C., Lewis, S., Corben, L. A., & Delatycki, M. B. (2018a). Young people living at risk of Huntington's disease: The lived experience. *Journal of Huntington's Disease*, *7*(4), 391–402. https://doi.org/10.3233/JHD-180308

Lucassen, A., & Clarke, A. (2022). In the family: Access to, and communication of, familial information in clinical practice. *Human Genetics*, *141*(5), 1053–1058. https://doi.org/10.1007/s00439-021-02401-0

MacDonald, M. E. Ambrose, C. M., Duyao, M. P., Myers, R. H., Lin, C., Srinidhi, L., Barnes, G., Taylor, S., A., James, M., Groot, N., MacFarlane, H., Jenkins, B., Anderson, M. A., Wexler, N., Gusella, J. F., Bates, G. P., Baxendale, S., Hummerich, H., Kirby, S., … Harper, P. S. (1993). A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. *Cell, 72*(6), 971–983. https://doi.org/ https://doi.org/10.1016/0092-8674(93)90585-e

Mahmood, S., Law, S., & Bombard, Y. (2022). "I have to start learning how to live with becoming sick": A scoping review of the lived experiences of people with Huntington's disease. *Clinical Genetics, 101*(1), 3–19. https://doi.org/10.1111/cge.14024

Maisel, N. C., & Gable, S. L. (2009). The paradox of received social support: The importance of responsiveness. *Psychological Science, 20*(8), 928–932. https://doi.org/10.1111/j.1467-9280.2009.02388.x

Malterud, K. (2001). Qualitative research: Standards, challenges, and guidelines. *The Lancet, 358*(9280), 483–488. https://doi.org/10.1016/s0140-6736(01)05627-6

Malterud, K., Siersma, V. D., & Guassora, A. D. (2016). Sample size in qualitative interview studies: guided by information power. *Qualitative Health Research, 26*(13), 1753–1760. https://doi.org/10.1177/1049732315617444

Mand, C. M., Gillam, L., Duncan, R. E., & Delatycki, M. B. (2015a). "I'm scared of being like mum": The experience of adolescents living in families with Huntington disease. *Journal of Huntington's Disease, 4*(3), 209–217. https://doi.org/10.3233/jhd-150148

Masten, A. S. (2018). Resilience theory and research on children and families: Past, present, and promise. *Journal of Family Theory & Review, 10*(1), 12–31. https://doi.org/10.1111/jftr.12255

McColgan, P., & Tabrizi, S. J. (2018). Huntington's disease: A clinical review. *European Journal of Neurology, 25*(1), 24–34. https://doi.org/10.1111/ene.13413

Moen, K., & Middelthon, A. L. (2015). Qualitative research methods. In *Research in Medical and Biological Sciences*, 321–337. Academic Press.

National Institute of Public Health. (2022). *Public Health Report*. https://www.fhi.no/en/op/hin/

Ost, D. E. (1984). The 'right' not to know. *The Journal of Medicine and Philosophy, 9*(3), 301–312. https://doi.org/10.1093/jmp/9.3.301

Pakenham, K. I., & Cox, S. (2015). The effects of parental illness and other ill family members on youth caregiving experiences. *Psychology & Health, 30*(7), 857–878. https://doi.org/10.1080/08870446.2014.1001390

Papoutsi, M., Labuschagne, I., Tabrizi, S. J., & Stout, J. C. (2014). The cognitive burden in Huntington's disease: Pathology, phenotype, and mechanisms of compensation. *Movement Disorders, 29*(5), 673–683. https://doi.org/10.1002/mds.25864

Patton, M. Q. (1999). Enhancing the quality and credibility of qualitative analysis. *Health Services Research, 34*(5), 1189–1208. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1089059/pdf/hsresearch00022-0112.pdf

Pierce, M., Hope, H. F., Kolade, A., Gellatly, J., Osam, C. S., Perchard, R., Kosidou, K., Dalman, C., Morgan, V., Prinzio, P., & Abel, K. M. (2020). Effects of parental mental

illness on children's physical health: Systematic review and meta-analysis. *The British Journal of Psychiatry, 217*(1), 354–363. https://doi.org/0.1192/bjp.2019.216

Polkinghorne, D. E. (2005). Language and meaning: Data collection in qualitative research. *Journal of Counseling Psychology, 52*(2), 137. https://doi.org/ 10.1037/0022-0167.52.2.137

Rolland, J. S., & Williams, J. K. (2005). Toward a biopsychosocial model for 21st-century genetics. *Family Process, 44*(1), 3–24. https://doi.org/10.1111/j.1545-5300.2005.00039.x

Roos, R. A. (2010). Huntington's disease: A clinical review. *Orphanet Journal of Rare Diseases, 5*(1), 1–8. https://doi.org/10.1186/1750-1172-5-40

Ruud, T. (2015). *Barn som pårørende: resultater fra en multisenterstudie.* Akershus universitetssykehus HF.

Røthing, M., Malterud, K., & Frich, J. C. (2014). Caregiver roles in families affected by Huntington's disease: A qualitative interview study. *Scandinavian Journal of Caring Sciences, 28*(4), 700–705. https://doi.org/10.1111/scs.12098

Røthing, M., Malterud, K., & Frich, J. C. (2015a). Balancing needs as a family caregiver in Huntington's disease: A qualitative interview study. *Health & Social Care in the Community, 23*(5), 569–576. https://doi.org/10.1111/hsc.12174

Røthing, M., Malterud, K., & Frich, J. C. (2015b). Family caregivers' views on coordination of care in Huntington's disease: A qualitative study. *Scandinavian Journal of Caring Sciences, 29*(4), 803–809. https://doi.org/10.1111/scs.12212

Sameroff, A. (2009). *The transactional model*. American Psychological Association.

Schacter, D. L. (1999). The seven sins of memory: Insights from psychology and cognitive neuroscience. *American Psychologist, 54*(3), 182-203. https://doi.org/ 0003-066X799/S2.00

Shern, D. L., Blanch, A. K., & Steverman, S. M. (2016). Toxic stress, behavioral health, and the next major era in public health. *American Journal of Orthopsychiatry, 86*(2), 109-123. https://www.mhanational.org/sites/default/files/Toxic%20Stress%20Final_0.pdf

Shields, G. S., Sazma, M. A., McCullough, A. M., & Yonelinas, A. P. (2017). The effects of acute stress on episodic memory: A meta-analysis and integrative review. *Psychological Bulletin, 143*(6), 636-675. https://doi.org/10.1037/bul0000100

Shonkoff, J. P., Garner, A. S., Siegel, B. S., Dobbins, M. I., Earls, M. F., McGuinn, L., Pascoe, J. Wood, D. L. (2012). The lifelong effects of early childhood adversity and toxic stress. *Pediatrics, 129*(1), 232–246. https://doi.org/10.1542/peds.2011-2663

Smith, J. A., Flowers, P., & Larkin, M. (2009). *Interpretative phenomenological analysis: Theory, method and research.* Sage.

Smith, J. A., & Osborn, M. (2015). Interpretative phenomenological analysis as a useful methodology for research on the lived experience of pain. *British Journal of Pain, 9*(1), 41–42. https://doi.org/10.1177/2049463714541642

Solberg, O. K., Filkuková, P., Frich, J. C., & Feragen, K. J. B. (2018). Age at death and causes of death in patients with Huntington disease in Norway in 1986–2015. *Journal of Huntington's disease, 7*(1), 77–86. https://doi.org/10.3233/JHD-170270

Sparbel, K. J. H., Driessnack, M., Williams, J. K., Schutte, D. L., Tripp-Reimer, T., McGonigal-Kenney, M., Jarmon, L., & Paulsen, J. S. (2008). Experiences of teens living in the shadow of Huntington disease. *Journal of Genetic Counseling, 17*(4), 327–335. https://doi.org/10.1007/s10897-008-9151-6

Staccini, L., Tomba, E., Grandi, S., & Keitner, G. I. (2015). The evaluation of family functioning by the family assessment device: A systematic review of studies in adult clinical populations. *Family Process, 54*(1), 94–115. https://doi.org/10.1111/famp.12098

Teherani, A., Martimianakis, T., Stenfors-Hayes, T., Wadhwa, A., & Varpio, L. (2015). Choosing a qualitative research approach. *Journal of Graduate Medical Education, 7*(4), 669–670. https://doi.org/10.4300/JGME-D-15-00414.1

The Human Rights Act (1999). https://lovdata.no/

The United Nations Children's Fund [UNICEF]. (2022). *Early childhood development.* https://www.unicef.org/early-childhood-development

Thoits, P. A. (2011). Mechanisms linking social ties and support to physical and mental health. *Journal of Health and Social Behaviour, 52*(2), 145–161. https://doi.org/10.1177/0022146510395592

Thompson, R. A., Flood, M. F., & Goodvin, R. (2015). Social support and developmental psychopathology. In D. Cicchetti & D. J. Cohen (Eds.), *Developmental psychopathology, risk disorder and adaptation* (pp. 1–37). John Wiley & Sons, Inc.

Tillerås, K. H., Kjoelaas, S. H., Dramstad, E., Feragen, K. B., & von der Lippe, C. (2020). Psychological reactions to predictive genetic testing for Huntington's disease: A qualitative study. *Journal of Genetic Counseling, 29*(6), 1093–1105. https://doi.org/10.1002/jgc4.1245

Tomasdottir, M. O., Sigurdsson, J. A., Petursson, H., Kirkengen, A. L., Krokstad, S., McEwen, B., Hetlevik, I., & Getz, L. (2015). Self reported childhood difficulties, adult

multimorbidity and allostatic load. A cross-sectional analysis of the Norwegian HUNT study. *PloS One, 10*(6), 1–16. https://doi.org/10.1371/journal.pone.0130591

Tracy, S. J., & Hinrichs, M. M. (2017). Big tent criteria for qualitative quality. *The International Encyclopedia of Communication Research Methods*, 1–10. https://doi.org/10.1002/9781118901731.iecrm0016

Tracy, S. J. (2010). Qualitative quality: Eight "big-tent" criteria for excellent qualitative research. *Qualitative Inquiery, 16*(10), 837–851. https://doi.org/10.1177/1077800410383121

Tyler, A., Harper, P., Davies, K., & Newcome, R. G. (1983). Family break-down and stress in Huntington's chorea. *Journal of Biosocial Science, 15*(2), 127–138. https://doi.org/ 10.1017/S0021932000014413

United Nations Convention on the Rights of the Child, November 20, 1989, https://www.ohchr.org/en/instruments-mechanisms/instruments/convention-rights-child

Vamos, M., Hambridge, J., Edwards, M., & Conaghan, J. (2007). The impact of Huntington's disease on family life. *Psychosomatics*, *Psychosomatics*, *48*(5), 400–404. https://doi.org/10.1176/appi.psy.48.5.400

van der Meer, L., Timman, R., Trijsburg, W., Duisterhof, M., Erdman, R., Van Elderen, T., & Tibben, A. (2006). Attachment in families with Huntington's disease: A paradigm in clinical genetics. *Patient Education and Counseling*, *63*(1–2), 246–254. https://doi.org/10.1016/j.pec.2005.11.019

van der Meer, L., van Duijn, E., Wolterbeek, R., & Tibben, A. (2012). Adverse childhood experiences of persons at risk for Huntington's disease or BRCA1/2 hereditary breast/ovarian cancer. *Clinical Genetics*, *81*(1), 18–23. https://doi.org/doi:10.1111/j.1399-0004.2011.01778.x

van der Zwaan, K. F., Mentink, M. D., Jacobs, M., Roos, R. A., & Susanne, T. (2022). Huntington's disease influences employment before and during clinical manifestation: A systematic review. *Parkinsonism & Related Disorders*, *96*(2022), 100–108. https://doi.org/10.1016/j.parkreldis.2022.02.022

Van Loon, L., Van de Ven, M. O., Van Doesum, K., Witteman, C. L., & Hosman, C. M. (2014). The relation between parental mental illness and adolescent mental health: The role of family factors. *Journal of Child and Family Studies*, *23*(7), 1201–1214. https://doi.org/ 10.1007/s10826-013-9781-7

van Walsem, M. R., Howe, E. I., Iversen, K., Frich, J. C., & Andelic, N. (2015). Unmet needs for healthcare and social support services in patients with Huntington's disease: A cross-sectional population-based study. *Orphanet Journal of Rare Diseases, 10*(1), 124. https://doi.org/10.1186/s13023-015-0324-8

von der Lippe, C., Diesen, P. S., & Feragen, K. B. (2017). Living with a rare disorder: A systematic review of the qualitative literature. *Molecular Genetics & Genomic Medicine*, *5*(6), 758–773. https://doi.org/10.1002/mgg3.315

Wexler, A. (2010). Stigma, history, and Huntington′s disease. *The Lancet, 376*(9734), 18–19. https://doi.org/10.1016/S0140-6736(10)60957-9

Williams, J. K., Ayres, L., Specht, J., Sparbel, K., & Klimek, M. L. (2009). Caregiving by teens for family members with Huntington disease. *Journal of Family Nursing, 15*(3), 273–294. https://doi.org/ 10.1177/1074840709337126

Williams, J. K., Driessnack, M., Barnette, J. J., Sparbel, K. J., Leserman, A., Thompson, S., & Paulsen, J. S. (2013). Strategies used by teens growing up in families with Huntington disease. *Journal of Pediatric Nursing, 28*(5), 464–469.

Williams, J. K., Hamilton, R., Nehl, C., McGonigal-Kenney, M., Schutte, D. L., Sparbel, K., Birrer, E., Tripp-Reimer, T., Friedrich, R., Penziner, E., Jarmon, L., & Paulsen, J. (2007). "No one else sees the difference": Family members′ perceptions of changes in persons with preclinical Huntington disease. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, *144*(5), 636–641. https://doi.org/10.1002/ajmg.b.30479

# I

Psychology & Health

ISSN: (Print) (Online) Journal homepage: https://www.tandfonline.com/loi/gpsh20

# ‘I knew it wasn’t normal, I just didn’t know what to do about it’: adversity and caregiver support when growing up in a family with Huntington’s disease

Siri Kjoelaas, Tine K. Jensen & Kristin B. Feragen

**To cite this article:** Siri Kjoelaas, Tine K. Jensen & Kristin B. Feragen (2022) ‘I knew it wasn’t normal, I just didn’t know what to do about it’: adversity and caregiver support when growing up in a family with Huntington’s disease, Psychology & Health, 37:2, 211-229, DOI: 10.1080/08870446.2021.1907387

**To link to this article:** https://doi.org/10.1080/08870446.2021.1907387



Published online: 22 Apr 2021.

PSYCHOLOGY & HEALTH
2022, VOL. 37, NO. 2, 211–229
https://doi.org/10.1080/08870446.2021.1907387

Routledge
Taylor & Francis Group



# 'I knew it wasn't normal, I just didn't know what to do about it': adversity and caregiver support when growing up in a family with Huntington's disease

Siri Kjoelaas[a], Tine K. Jensen[b,c] and Kristin B. Feragen[a]

[a]Centre for Rare Disorders, Oslo University Hospital HF, Norway; [b]Department of Psychology, University of Oslo, Norway; [c]Norwegian Centre for Violence and Traumatic Stress Studies, Oslo, Norway

**ABSTRACT**

**Objective:** Excessive adversity in childhood can have long-term consequences on health and well-being. One group of children that may be at risk are those who grow up with a parent with Huntington's disease (HD). Despite this, there is little knowledge about how these children are impacted by adversity. We aimed to explore adverse childhood experiences (ACEs) of individuals who grew up with a parent with HD and their perceptions of caregiver support.

**Design:** Semi-structured qualitative interviews of 36 adults and adolescents were analysed using interpretative phenomenological analysis (IPA).

**Results:** First, the analysis revealed a range of frequent adverse events during participants' childhoods, leading to feelings of uncertainty, loss, fear, and a lack of care. Next, how the presence or absence of support had impacted participants' perceptions of these experiences was analysed. Results showed that participants without support were often overwhelmed by feelings of stress, whereas participants with support tolerated adversity better.

**Conclusion:** Findings suggest that children in families with HD can be exposed to a range of adverse experiences. Findings also suggest that sufficient support from a caregiver without HD can buffer or protect against negative consequences of these experiences.

**ARTICLE HISTORY**
Received 23 June 2020
Accepted 7 March 2021

**KEYWORDS**
Adverse childhood experiences; Huntington's disease; qualitative; support; coping; stress

## Introduction

Children who grow up with a parent with Huntington's disease (HD) can be exposed to a range of adversities. HD is a progressive neurodegenerative disease with a 50% risk of genetic transmission; it currently has no cure, and limited interventions for symptom relief are available (Roos, 2010). The typical onset of HD is between 30 and 50 years, a time when many of those affected will have caretaking responsibilities for children and families. During this time, the

**CONTACT** Siri Kjoelaas hagsir@ous-hf.no Centre for Rare Disorders, Oslo University Hospital HF, Rikshospitalet, P.O. Box 4950, 0424 Nydalen, Oslo, Norway

disease will progress through different stages, affecting physical, psychological, and cognitive functioning, and it leads to death after an average of 17–20 years (Roos, 2010).

HD causes several changes that can affect a person's capacity to provide appropriate care for children. For instance, neurobiological and behavioural markers of the disease are often characterised by changes in personality, apathy, aggression, and decreased empathetic understanding (Duff et al., 2007). In one study, parents with HD scored significantly higher on a range of indicators of dysfunctional parenting, including over-control, indifference, and abuse, compared to caregivers without any illness and caregivers suffering from depression (Vamos et al., 2007). Research has also suggested that children who grow up with a parent with HD often do not receive adequate support or help and are left with overwhelming responsibilities at home (e.g. Dondanville et al., 2019; Kjoelaas et al., 2020; Lewit-Mendes et al., 2018). To date, only one study has systematically explored the presence of adverse childhood experiences in relation to HD, indicating that this group of children experiences significantly more adversities, such as domestic violence and parental dysfunction, than children growing up with parents with another inheritable genetic disease (van der Meer et al., 2012).

Children and adolescents who have frequent adverse experiences appear to have significantly poorer mental and physical health outcomes than those who experience little or no adversity growing up (Hughes et al., 2017). Preventing children from having to experience frequent or co-occurring adversity is therefore an essential responsibility of any society. Adverse childhood experiences (ACEs) can be defined as 'childhood events, varying in severity and often chronic, occurring within a child's family or social environment that cause harm or distress, thereby disrupting the child's physical or psychological health or development' (Kalmakis & Chandler, 2014). Associations have been found between ACEs and a range of unfavourable outcomes, including changes in functions essential for decision-making and emotional control, drug use, diabetes, heart disease, and even early death (Felitti et al., 1998; Hughes et al., 2017). Studies have also indicated that the risk of poor mental and physical health outcomes increases with the number of ACEs experienced (Hughes et al., 2017). Stress is a key element of understanding how ACEs affect health and development (Shonkoff et al., 2012). Stress is not inherently harmful and learning how to cope with stress and adversity is an essential part of any child's development. However, exposure to severe and frequent adversity may disrupt healthy and adaptive responses to stress, so that stress becomes toxic to the developing brain. Toxic stress can disrupt healthy developmental trajectories by inhibiting the normal development of neurological, immune, and endocrine systems, which in turn can place children at risk of future health-related challenges and disability (Hughes et al., 2017; Shonkoff et al., 2012). However, how children adapt depends on several factors other than adverse experiences. One of the most important predictors of how adversity will impact a child is the nature of their caregiving relationships and the support they receive. Having close, supportive relationships with adults may protect children from the damaging effects of stress and adversity on their development (Shonkoff et al., 2012). In contrast, children who experience frequent and severe adversity and grow up without supportive adults will be at higher risk of damaging long-term effects.

Despite indications that HD may affect parenting skills (Vamos et al., 2007) and that children who grow up in families with HD can be exposed to a range of negative

life events (van der Meer et al., 2012), there is little knowledge about how these children are impacted by adversity. The current study therefore explored the impact of ACEs on individuals with previous and current experiences of growing up with a parent with HD as well as their experiences of caregiver support.

## Method

### Sample and setting

With the overall aim of describing the experiences of growing up in a family impacted by HD, we analysed interviews collected in 2018. Anyone in Norway over the age of 12 years who had current or previous experiences of growing up with a parent with HD was invited to participate in the study. Information was distributed verbally and through information sheets in several settings where individuals who had grown up in a family affected by HD could be reached. Locations included educational courses for families affected by HD, counselling services at Oslo and Haukeland University Hospitals and at St. Olavs Hospital, the Norwegian Association for Huntington's Disease, and Facebook. Information sheets outlined the study's purpose, provided information about the interview topics, and included the main researcher's name and contact information. In response to the formal and informal invitations, 42 people gave their initial written consent to participate. Of these, six people could not be reached when subsequently contacted for an interview. A total of 36 participants were therefore interviewed and included in the analysis. Participants ranged in age from 13 to 65 years ($M_{age}$ = 36.6 years). Table 1 summarises the participants demographic information.

### Data collection procedures

An interview guide was created based on the relevant literature and feedback from clinical experts and a group of user representatives. Individual semi-structured interviews were conducted, focusing on both challenges and protective factors. The semi-structured nature of the interviews permitted an open exploration of the interviewees' experiences while helping the interviewer focus on specific issues. Broad topics included the participants' childhood experiences, family relationships, and experiences of support.

On average, the interviews lasted 60 minutes (range: 27 to 90 minutes). Face-to-face interviews were preferred ($n = 33$), but telephone interviews were an option and were chosen by some participants ($n = 3$). The interviews were conducted the Centre for

**Table 1.** Demographic characteristics of the participants (N = 36).

| Variable | | Label | N |
|---|---|---|---|
| Age | 13–18 years | Teenager | 7 |
| | 19–35 years | Young adult | 10 |
| | 36–65 years | Adult | 19 |
| Gender | Female | | 26 |
| | Male | | 10 |
| Parent with HD | Mother | | 19 |
| | Father | | 17 |

**Table 2.** Main topics and sample items applied in the semi-structured interview guide.

| Interview topic | Sample question |
|---|---|
| Background information | *What motivated you to participate in this study?* |
| | *Please describe the family you grew up in?* |
| Childhood experience | *Tell me about your childhood?* |
| | *How did your parents' disease affect your family?* |
| | *How has growing up with a parent with HD affected you?* |
| Disease- and self-disclosure | *What is your experience of disclosing information about HD?* |
| | *How do you think parents should inform their children about HD?* |
| Resources and support | *What sources of support did you have growing up?* |
| | *If you had the chance, what would you say to someone in a similar situation as you are/ were?* |

Rare Disorders at Oslo University Hospital or in the homes of a few participants. Counsellors from the Centre for Rare Disorders were involved in planning the study, but they were not involved in the data analyses due to their involvement with potential participants. Several researchers conducted the interviews, including postgraduate psychology students and trained health professionals at Oslo University Hospital in Norway. The students had no previous experience with HD, whereas the health professionals had previous research experience with the disease. All the interviewers had or received formal training in qualitative methods before conducting the interviews. The interviewers had no previous familiarity with the study participants. Supervision regarding interview techniques was provided if needed. The project manager, an experienced licenced clinical psychologist, participated in at least two interviews conducted by novice researchers to ensure the reliability and consistency of all interviewers' practices. Table 2 displays interview topics and sample questions.

## Analyses

The interviews were recorded and transcribed verbatim. The project manager performed sample tests on the correspondence between the recorded interviews and transcripts to ensure accuracy between transcripts produced by different members of the research team. Because we were interested in the participants' own perspectives on growing up with a parent with HD, interpretative phenomenological analysis (IPA; Smith et al., 2009) was used. IPA consists of three core features. First, it is phenomenological, meaning it explores peoples' experiences. Second, it is interpretative, which means the researcher seeks to understand how the participant makes sense of his or her life (double hermeneutics). Third, IPA is ideographic in that it analyses the case at hand and how the person is making sense of his/her situation. In the analysis, the researcher first 'stays close' to the data and the participants' accounts and then 'steps back' and interprets these accounts.

During the first analytic step, we read and re-read all the interviews independently to become familiar with the data, and we started the initial coding. The authors then discussed their preliminary thoughts. During this part of the analysis, we noted that the participants' exposure to a range of ACEs overshadowed many of the interviews. Therefore, in the next step of the analysis, ACEs was used as a sensitising concept to see how these experiences shaped the participants' upbringing and childhood. Within this framework, we worked inductively. Through a dynamic and interactive process,

**Table 3.** Superordinate and subordinate themes.

| Superordinate theme | Subordinate themes |
|---|---|
| Adverse childhood experiences | *I never knew what to expect* |
| | *You lose them over and over again* |
| | *I was scared all the time* |
| | *I had to take care of myself* |
| The role of support | *Nowhere to turn* |
| | *Having a lightning rod* |

the variety of adverse experiences were analysed case by case, then across cases, to bring forth events and meanings. Within the overarching theme related to ACEs, four subordinate themes were identified as follows: *I never knew what to expect; You lose them over and over again; I was scared all the time*; and *I had to take care of myself*. The themes are linked in that they are all related to events that caused significant distress but are separated in the meanings or feelings that the different experiences generated. During the next step of the analysis, we observed that participants consistently described adversity in the context of their caregiving relationships and home environments. Therefore, we extended the scope of the analysis to investigate participants' support from caregivers without HD in relation to their adverse experiences. Caregivers were defined as adults in a close relationship with the participants who were responsible for their care; they lived in the same home and included biological parents and/or stepparents. We revisited all the interviews, extracted text, and coded the meaning of caregiver support in the same dynamic and interactive manner as was used in the first step of the analysis – case by case and then across cases. For this second superordinate theme, we identified two subordinate themes describing how participants perceived support in their caregiving relationships. One in which participants felt they did not have close, supportive relationships, and one in which they felt they had close supportive relationships with a caregiver without HD, respectively labelled *nowhere to turn* and *having a lightning rod*. Table 3 summarises the superordinate themes and corresponding subordinate themes.

Reflexivity was emphasized throughout the analysis. Two of the authors (the first and the last) have experience with HD as researchers. Theoretical knowledge about HD was helpful during the analytic phase as it helped us understand and recognise elements in the participants experiences specific to HD. However, this also had the potential to create biased interpretations of the interviews. Inspired by the consensual qualitative research model (Hill et al., 1997), and to enhance validity and counteract group thinking and researcher bias, the first and last author formed the primary analytic team. The second author, who has extensive experience working with trauma in clinical and research settings but no previous experience or knowledge of HD, served as a discussant and analysed the interviews independently. In this process, we were able to both capitalise on the different positions we had in relation to the data and challenge the theoretical and professional assumptions in the interpretations made from the data. We encouraged reflections on our different positions and experiences, and the first author kept a journal of these reflections. Consensus was obtained after repeated rounds of independent reading, sharing of notes and open discussions, then re-reading and re-discussing the interviews. Some participants were young when their parents

developed symptoms of HD, whereas others reported memories from late adolescence. The interviewers asked participants to provide the approximate age at which they experienced these events. Given this study's focus, only adverse experiences that had occurred in childhood and adolescence were included in the analysis.

In the presentation of the findings, the frequency labels *general*, *typical*, and *variant*, as suggested by Hill et al. (2005), are used to indicate the degree of representativeness across individual cases. The topics or themes that were general in the sense that they applied to all but one participant are referred to in the text as *all participants*. Topics were considered typical if they applied to more than half of the cases and are referred to as *most participants*. Topics were variant if they were represented in less than half of the sample but in more than two cases; they are referred to in the text as *some participants.* Quotes that illustrate the subordinate themes were selected and translated from the original language into English. Participants were given pseudonyms and identifying information have been omitted.

### Ethical considerations

We obtained ethical approval for the study from the Regional Committee for Medical Research Ethics (Health region South-East, Norway, reference number: 2017/1613). Participants were informed about the study, gave written consent, and were informed of their right to withdraw at any time. The confidentiality of participant information was ensured throughout the research process. Parental consent was obtained for participants under the legal age for health consent (16 years in Norway). Due to the sensitivity of the topics discussed during the interviews, relevant referrals or subsequent follow-ups were arranged by a clinical psychologist and project manager, if necessary. All the participants received a follow-up call within two weeks after the interview to assess the need for referral to a clinical psychologist and to obtain their feedback about how they experienced being interviewed about the topics in question. Three participants wished to receive follow-up after the interviews and were referred to a clinical psychologist.

## Results

In this study, we focused on adverse events and distressing experiences described by the participants. The participants' parents had developed symptoms of HD at different times and in different ways, and the severity of the disease as well as its impact on their family and childhood varied. Some participants reported undergoing adverse experiences throughout their childhood. Others described having few or no ACEs in their early childhood years but increasing difficulties as they grew older.

### Adverse childhood experiences

Four themes that reflected participants' experiences of different types of childhood adversity were identified as follows: I never knew what to expect; You lose them over and over again; I was scared all the time; I had to take care of myself.

### *I never knew what to expect*

Most participants provided stories of how changes in their parent caused by HD affected their *sense of stability and security*. Even though there were individual differences in the severity of HD and its impact on their parent, participants gave similar descriptions of how HD had impacted and changed their parent's mood, way of thinking, and behaviour. That is, the affected parent was described as becoming increasingly irritable and angry, apathetic, and distant, being anxious or depressed, and often having problems with remembering things and responding appropriately. At home, the atmosphere was described as increasingly unpredictable, because their parent's mood and behaviours changed constantly, often within short periods of time. One participant reflected on how she had noticed strange fluctuations in her mother's mood from an early age:

> We would be sitting there, having a good time, and my mother would just suddenly start to cry, and I did not understand it at all. And then she would get herself together somehow, disguise it with laughter. It impacted me, like: 'What was that?' And what that did to me? It probably made me feel responsible for her well-being from an early age (Nancy; adult female, mother with HD).

In this context, the participant highlighted how strongly these experiences had affected her as a child. She described her mother's changes in mood as if the light had been turned off in the room, suddenly altering the atmosphere at home. This participant, along with several others, had not known how to interpret the changes they observed. In turn, unpredictable reactions from a parent left them feeling responsible for the changes that occurred, thinking they had done something wrong. They had to be sensitive to the needs of their parent, adjusting their behaviours. Conflicts at home were frequent and for some of the participants a daily experience. They never knew what to expect. There had been a constantly negative atmosphere at home and a general feeling that something was off; they could sense this mood as they walked into a room. In an attempt to create stability, some accommodated their behaviours to the needs of their parent with HD. One participant described: 'I became very insecure, I didn't know, always offering to help, unsure of what I had done wrong, not knowing when the next episode would happen' (Dorothy; adult female, mother with HD).

### *You lose them over and over again*

Changes in their parent's personality, mood, and behaviour not only created unpredictability but were also described as a *significant loss* for the participants. Most participants felt that HD took away the true nature of their caregiver, transforming them into a person they no longer recognised. The feelings of loss described by participants seemed to be different from the feelings of unpredictability described in the previous theme. That is, participants' descriptions of unpredictability appeared to be connected to anxiety, the feelings of loss seemed to be more connected to sadness. One participant described: 'The worst part about the disease is the personality changes. It's like you lose them over and over again' (Donna; adult female, mother with HD). Others also described the significant loss they felt as a result of the

progression of their parents' disease and the sadness that came with this loss. Even though they grieved, they did not have the opportunity for closure, because the feelings of loss were continuous and progressive. Some genuinely longed for an emotional bond with their parent, feeling that the connection they should have had, or used to have, was no longer there. Knowing their parent had an ultimately deadly disease left some participants fearful of losing them in the future, and the diagnosis led to the experience of a profound hopelessness. One participant talked about his recent experience of coming to terms with his mother's diagnosis. Within this context, he explained how time with his mother had been turned into a continuous wait for changes and the inevitable outcome of the disease. These thoughts preoccupied him and had left him struggling with depression, anger, and resentment:

> It's a hopeless situation. With a divorce, you can think that maybe they will get back together. Maybe, maybe, maybe. But there is no maybe here. It is black and white. She will become sick, and she will die. There is nothing you can do about it. A blessing from the king will not help. The world's best doctors will not help. It is not like with cancer, where you have a chance. You have no chance. You are 110% without any chance (Jordan; male teenager, mother with HD).

They experienced complex and persistent distress because they were unsure of how to interpret and understand the multiple feelings of loss in their caregiving relationship. Some expressed love for their parent but felt torn by the unpredictability they had to experience as a result of HD. The well-being of those who were unable to reconcile with the complicated feelings of loss was profoundly affected. A few described acting out, through aggression or substance misuse, to cope with what they felt was profoundly lacking and unfair in their life. Others avoided situations and interactions with their parent with HD. They used avoidance to protect themselves from sadness or disappointment and from the fear of losing their caregiver to the disease.

Some also described how their parent's partnership was impacted by HD, often resulting in separation or divorce. Others were separated from their parent due to their increased need for care, such as moving to a nursing home. As such, the disease led to physical separation from a caregiver, intensifying the loss they already felt. Some also experienced having new stepparents in their families because of divorce, which was a positive contribution for some but a new source of adversity and distress for others. Although they feared or even dreaded the presence of an unpredictable parent at home, their physical separation from the affected parent led to feelings of loss or abandonment. For a few participants, however, divorce or separation was a positive experience that reduced the level of stress and unpredictability they felt at home and improved their relationship to both caregivers.

### *I was scared all the time*

In addition to growing up with unpredictability and loss, most participants also experienced a variety of distressing adverse events. Several categories of events were created that both were and were not connected to HD. The most common experience was having *witnessed or been subjected to physical violence or threats of physical violence*. These events were often severe and ranged from verbal abuse and threats to physical assaults. Some participants endured physical assaults or threats of immediate harm;

however, most witnessed domestic abuse between caregivers. Those who experienced frequent and/or severe violence and threats lived under tremendous stress. They were constantly afraid of one or both caregivers and had not felt safe at home. One participant recalled how her father with HD had exposed her and her mother to physical and verbal abuse for as long as she could remember: 'I think I was scared all the time. I was always prepared for the worst. It could be something as small as a word that would trigger the anger' (Shirley; adult female, father with HD). Within this context, she explained how fear created by the abuse had affected her as a child:

> I became a quiet and invisible child. I tried to avoid any source of conflict. I sort of erased myself and surrendered to it. There were a few times when I was a teenager that I spoke up, but in general, I stayed quiet. I remember how learning to read made my life better. It was a way to escape (Shirley; adult female, father with HD).

The impact and meaning portrayed in her story were similar to the accounts of several other participants who also had experienced verbal and physical abuse. Fear of being hurt and fear of others in their family being hurt were present during the events themselves and a constant presence in the form of anticipation of future events. To cope with the presence of domestic violence, some either avoided their caregiver or attempted to avoid triggering violent events with their caregiver.

Being *embarrassed, insulted, or rejected* by their caregiver was also experienced by most of the participants. One participant recounted how her mother with HD would casually tell the children about her sex life with their father. Such embarrassing experiences were highly uncomfortable and had often eroded the trust they extended to their caregiver. Several participants also described nagging or belittling behaviours from a caregiver. One participant talked about the callous demeanour of his father, which he had experienced throughout his upbringing. In this context, he questioned whether these characteristics reflected his father's personality or reflected the development of HD:

> It was uncomfortable, and there was always nagging, I can't… I can hardly remember him saying one positive thing to me. You were not supposed to think you were capable of anything. He was good at putting us down and suspicious about everything that we did (Mark; adult male, father with HD).

As this passage demonstrates, rejection and embarrassment severely compromised some participants' self-esteem and sense of security. Out of fear of being embarrassed or humiliated, they withdrew emotionally from their caregiver and often from social and public settings where others might witness this behaviour. Trying to make sense of the rejection and insults, they felt guilty and ashamed, thinking there was something wrong with them. Some made extensive attempts to please their parent in order to win their trust, get their acknowledgement, and demonstrate that they were worthy of their love.

Some of the participants witnessed their parent *talking about or attempting to commit suicide*. One adult female participant described how her mother with HD attempted to take her own life several times and how she and her siblings had to watch her in shifts to make sure she would not be successful. Suicide attempts were

a highly disturbing childhood experience that severely impacted the participants' sense of security. They felt powerless over their parent's decision to end their life, often feared new suicide attempts, and endured complex emotions and reactions that were brought about by their parent wanting to die. One participant reflected on how his mother's suicide attempt had been the catalyst for a range of complex emotions, including reflections about his own possibility to inherit the disease:

> She attempted suicide. But she survived. (…) I think it [taking one's life] might be for the best, and I plan to do the same if I get the disease. It does not get better; this only goes in one direction. Some may say it is a coward solution, but I want to say it's the best solution for the family, so you don't have to be the burden she is. Not that it is her fault, and not that I want her to kill herself, but that's what I would do (Gabriel; male teenager, mother with HD).

Some participants also described having a caregiver who *abused substances*, mainly alcohol but in some instances prescription drugs as well. Substance abuse was described not necessarily as the leading cause of adverse events, but as a catalyst of other events. Having a parent who abused substances appeared to create more frequent and often more severe adverse experiences, such as physical assaults between parents or suicide attempts. One participant described: 'My dad was an alcoholic. The night we moved away from him, he was going to shoot my mom. I woke up and saw that' (Diane; adult female, mother with HD). For this participant, as for several others, substance abuse had dramatically increased the feelings of instability and unpredictability, as well as fear.

### *I had to take care of myself*

Some of the participants also felt they *lacked appropriate care* and had to be self-sufficient growing up. Experiences of neglect often accompanied their experiences of violence and abuse. A few were not provided with adequate food and had no one to prepare food for them as children. Others did not have caregivers who cleaned, did laundry, or provided them with clean clothes. A few reported that they endured the messiness and lack of care by providing their own food and clothes. Others had taken on an adult role, shouldering responsibility for the household as well as taking care of their parent with HD. They had to be creative, figuring out ways to manage the multitude of tasks at home. One participant described how she constantly had to put out burning cigarettes her mother dropped or failed to put out herself, living in constant fear that their house would burn down. Asking a caregiver to help with responsibilities such as household chores, cooking dinner, and caring for others in the family was also a potential source of conflict. Taking responsibility for their own care was, therefore, also a way to accommodate the needs of the parent(s) in order to avoid potential disputes. One participant, who grew up with a mother with HD and a father who suffered from alcohol abuse, described: 'We lived in a mess. It did not get cleaned. Mom didn't clean herself, so I had to make sure she would shower, and there was dirty laundry everywhere' (Evelyn; adult female, mother with HD). In this context, she explained how HD had affected her mother's ability to see the needs of others, including the needs of her daughter. She had planned to run away several times, but had stayed out of a sense of obligation to take care of her mother:

> I lost my youth this way. I wasn't given the opportunity to rebel. There was no room for that. I went from being a child to being an adult. Had to take responsibility, take care of myself. And of course, most people can do that, but you sacrifice a lot along the way (Evelyn; adult female, mother with HD).

Not receiving appropriate care also came at a high cost to other participants and severely limited their autonomy. This had felt normal at the time; but as they grew older they reflected upon the lack of normality and feelings of neglect. Seeing that other peers had caregivers who provided for them and protected them, some felt envious, at times leaving them resentful of their parents for failing to provide them with the care they needed.

## *The role of support*

Descriptions of adversity varied depending on whether participants felt they received support from their caregiver(s) without HD. The study's second aim, therefore, was to explore participants' perceptions of support from their caregiver(s) without HD and how the presence or absence of support influenced their experiences of adversity and their childhoods. Based on participants' descriptions of support, two themes were generated: *nowhere to turn* and *having a lightning rod*, describing the experiences of those who felt they did not have support from their caregiver without HD and those who felt they did.

### *Nowhere to turn*

Most of the participants grew up in households where, at some point in time during their childhood, both caregivers were a source of adversity. In addition to growing up with a parent with HD, some participants had a caregiver without HD whom they experienced as aggressive, malicious, unpredictable, and/or suffering from severe mental disease. These caregivers were described as triggering one another, often leading to severe physical or emotional abuse between the adults. One participant described turbulent adolescent years marked by repeated physical abuse between his father and stepmother. In this context, he highlighted the responsibility of both caregivers in the domestic violence he had witnessed:

> The relationship between my dad and stepmom was… They were fighting and crazy. I remember a few times she would stick a fork in his back and hit him with a carving board. And suddenly, she would have a black eye, and he would say she was hitting herself. But it was both of them (Charles; adult male, father with HD).

Others described how their caregiver without HD was under severe mental strain from the toll HD had taken on their family and, therefore, was not emotionally available to support their children. These caregivers were often described as profoundly unhappy, and exhausted, distant, or tired. One participant described how her father with HD had repeatedly abused her mother. In this context, she told about a mother who had done the best she could, but that had not been able to spare her children from the emotional consequences of the abuse:

> There are those specific episodes that stick. I would come into the kitchen, she would lay on the floor, exhausted, not crying normally, but sobbing in this strange way. And he would prance around, back and forth, over her, as if everything was normal, loading the dishwasher. I would bend down, trying to help her up, and she would pull away… And I remember that intense feeling of fear, and that I didn't know how to make it right again (Abigail; adult female, father with HD).

Experiencing multiple adults in charge of their care as highly unpredictable or unavailable had a profoundly negative impact on the participants. Those who should have been their source of comfort were instead a source of fear. Some felt utterly alone and overlooked by those in charge of their care. They were left on their own in highly distressing situations with a sense that they had no one to protect them. They felt anger or resentment towards their caregivers for not acting to prevent or alleviate their distress. They felt as though they did not have a close relationship with either of their caregivers, and they felt a lack of love, closeness, and support from any adult. There was an overall feeling that something fundamentally important was lacking in their childhood. Some reported that they had no one to help them understand or make sense of the adversities they had to endure. Many felt invisible, scared, and lonely. One participant grew up with a father who had exposed both her and her mother to physical and psychological abuse. She described how her father had failed to provide them with any support, even deliberately providing her with a false explanation for the changes she noticed when her mother developed HD. Telling this story, she cried and expressed anger over the consequences of being lied to by the person who was supposed to be her source of support when she was only a child:

> The only thing my stepfather told me was that I had made my mother sick and that it was my fault. (…) I felt scared, right? When you are not told what is going on, and you think, what is happening, did I do anything? That guilt (Diane; adult female, mother with HD).

Some struggled with juggling the task of preventing adverse experiences from one parent and the task of providing support to their other caregiver, feeling torn between their parents, and often trying to repair distressing situations. Persistent feelings of distress and the unavailability of support profoundly affected the well-being of several participants. Adversity was described as always present, and they were left with no positive impressions of their childhood or of the adults in charge of their care. To cope with the situation, some withdrew completely. They felt hopeless and became silent, invisible children in an effort to avoid any potential conflict. Others ran away or planned to run away from home to escape from their dysfunctional households. Most participants without support described their childhood as overshadowed by adversity, using expressions such as 'living a nightmare,' 'filled with chaos', and 'a lack of normality'. One participant described: 'It leaves scars when you have such a difficult childhood, and when there are so many factors that play their part. Illness… Traumatic fights, bullying at school, no one to support you, no one to see you' (Hannah; young adult female, father with HD).

### *Having a lightning rod*

In contrast to the group of participants who felt that the caregiver without HD was unavailable for support and protection, some participants grew up with one or more adult caregivers without HD whom they saw as a source of support. All these participants gave positive descriptions of sacrificing, warm, and nurturing caregivers. They talked about positive caregiving relationships, such as having a parent who was the most important person in their lives, and expressed admiration for how their caregiver managed his or her multiple roles in the family. The type of support provided by the adult caregiver without HD varied. Some participants felt they received mostly instrumental support, such as making dinners and facilitating their participation in activities. Others described emotional support, reporting that their caregiver was someone they could talk to and a source of comfort. A few participants had a caregiver who provided both forms of support. Having a close relationship with a supportive adult appeared to have a profoundly positive impact on the frequency and severity of their adverse experiences as well as on their interpretation of these experiences. The adverse experiences were related mainly to changes in their parent with HD, such as the presence of unpredictability and loss. A few participants with support also described major or potentially traumatic events, such as having a parent who attempted suicide or witnessing violence between their parents. Like those without support, these participants felt afraid and distressed. However, their distress was mostly described as isolated to the specific events that occurred and not as constant feelings that followed them through their upbringing. In contrast to those without enough support, they felt their caregiver was a buffer or mediator in the conflicts and adversity that occurred: 'All honour to my mom. She is quite tolerant. A lot falls on her, and she stands there in the middle of the storm. I call her a lightning rod' (Marilyn; adult female, father with HD). They reported feeling that they had a protector in the midst of their adversity. Having support that helped them make sense or develop an understanding of the expressions and behaviours of the parent with HD appeared to reduce the feelings of stress from adverse experiences. They described being validated and understood. Some described how this support helped them develop strategies to cope with and interpret their experiences. One participant talked about her mother's unpredictable behaviour and the constant conflict she experienced in their relationship. In this context, she explained how closeness with her father had helped her deal with conflicts between her and her mother when they occurred:

> I felt that I could talk with him a lot and that he supported me. When we [participant and mother with HD] got in a fight, he would take our side… If I did not have the courage… It is not always you dare to talk back at your mom… But he would take our side and tell her she was unreasonable. And that, in turn, gave me the courage to tell her when she was unreasonable too (Isabella; female teenager, mother with HD).

Despite the adverse experiences related to growing up with a parent with HD that most participants described, those who felt they had supportive and protective caregiving relationships generally gave positive descriptions of their childhood. These included describing their upbringing as 'normal', reporting that they had 'good family relations', and reporting that they 'felt a sense of stability' growing up.

## Discussion

This study explored ACEs of individuals who grew up with a parent with HD. Previous studies suggest that these young people can be exposed to several negative life events and challenging tasks (Kjoelaas et al., 2020; van der Meer et al., 2012). However, this is the first study to provide an in-depth exploration of this group's exposure to ACEs and the relationship between caregiver support and adversity. Our findings therefore contribute valuable knowledge that can guide the prevention of childhood stress and adversity in families affected by HD.

Participants in this study were exposed to a range of adverse events that frequently occurred throughout their childhoods. Their adverse experiences seemed to reflect unpredictability at home, complex feelings of loss, a lack of care, and experiences that compromised participants sense of safety; including being exposed to domestic violence, rejection, embarrassment, or substance abuse, or having a parent who attempted suicide. These experiences can easily be grouped into categories that are applied in standardised measures of ACEs and that have been used for decades in research and clinics (Boullier & Blair, 2018), such as different forms of household dysfunction, abuse, and neglect. Many of our participants experienced some degree of adversity on a regular or even daily basis. The forms of adversity experienced by a given participant also often fell within several or all categories of ACEs. The frequency and co-occurrence of adversity described by our participants is particularly important, because research has clearly demonstrated that the more adverse experiences someone endures, the higher the risk of having to struggle with a range of health challenges throughout life, such as diabetes, respiratory disease, low life satisfaction, mental illness, or problematic drug use (Hughes et al., 2017). Given these lifelong consequences of childhood adversity, it is particularly important to identify groups that are at high risk of experiencing adversity growing up, as we have done in this study.

Another key finding involved how the presence or lack of support from a caregiver without HD appeared to influence participants' descriptions of adversity. Participants without support from either caregiver seemed to have grown up with constant underlying feelings of distress, as they often recalled poor caregiving relationships, low levels of well-being, and childhoods overshadowed by adversity. In contrast, participants with support seemed to give different, more positive, accounts of their caregiving relationships, well-being, and childhoods. This finding is important because it suggests that having a supportive caregiver without HD can act as protection against negative experiences associated HD. Research highlights several factors that could promote this type of protection, or resilience, in children who experience adversity (e.g. Bellis et al., 2017; Franke, 2014). Our findings are in line with research pointing out that having support in adverse or traumatic childhood experiences is one of the main predictors of the impact these experiences will have later in life (Bellis et al., 2017; Shonkoff et al., 2012). Without close relationships with supportive adults, stress resulting from severe, continuous, or frequent exposure to adversity can become harmful or toxic and consequently impact health and well-being. On the other hand, having supportive adult caregivers helps the stress-response system return to baseline after distressing events. The stress they experience becomes more tolerable

instead of toxic, which will in turn have a positive or neutral (instead of negative) impact on well-being and future health.

Having support protects against the consequences of a stressful event by helping children apply effective coping strategies, such as active engagement in reducing stress and managing emotions, and by promoting a sense of control (Evans et al., 2013; Littleton et al., 2007; Shonkoff et al., 2012). Although we cannot make any conclusions about the exact mechanisms by which support acted as a buffer against adversity in our study, support from a caregiver without HD seemed to help participants cope better in adverse situations. All types of support from a caregiver were described as positive. Having a caregiver from whom participants could receive emotional support, seek information, and who provided an explanation and interpretation of their experiences seemed to be important. Help with interpretation appeared to be particularly important in terms of understanding and conceptualising the HD-related changes in their parent, so that they did not blame themselves for or feel guilty about adverse events. When faced with difficulties, children search for an explanation or reason for their occurrence. This is often an attempt to restore a sense of control or predictability in life (Meiser-Stedman et al., 2019). Therefore, we suggest that by helping participants understand the adversity they experienced, their caregiver facilitated beneficial coping, enhancing their sense of control and helped them manage their emotions.

Adverse experiences can also profoundly change the way children see themselves and the world. Unfortunately, for most of our participants, it looked as if their caregiver without HD had been unable to provide the support they needed, and they seemed to have been left to find ways to cope with adversity on their own. If adults do not help children interpret the situation, children may develop inaccurate and dysfunctional beliefs about their own responsibility (de Haan et al., 2017). This study's participants felt that they lacked protection and lacked control over their lives. Having no one to turn to when experiencing adversity appeared to lead to a lack of understanding of what they were going through, resulting in constant and intolerable stress. It seemed like those without caregiver support used coping strategies such as attempting to avoid stressful situations, withdrawing, trying to accommodate their parent's behaviours, fixing distressing situations, or acting out. These ways of coping, instead of reducing harmful levels of stress, may contribute to maintaining higher levels of stress for longer periods (Meiser-Stedman et al., 2019). Based on these findings, we argue that caregiver support may help children experience stress as more tolerable and may reduce the frequency and severity of adverse events. In contrast, the lack of a close relationship with a supportive adult may lead to the development of inadequate coping strategies, which may in turn severely compromise the child's ability to tolerate distress.

For many caregivers without HD, their partner's disease seemed to lead to mental strain that explained their lack of availability and reduced their caregiving abilities. The worst-case scenarios involved participants who grew up with more than one caregiver suffering from mental illness or instability or participants who grew up in single-parent households, having only their parent with HD for support. Available research supports that the burden of having a partner with HD can be overwhelming and that neither partners nor patients with HD get the help they need (Røthing et al.,

2015; Van Walsem et al., 2017). This burden can, in turn, be expected to influence parenting styles and the availability of both caregivers in families with HD. Interestingly, one study compared parenting styles in families in which one caregiver had HD, families without any illness, and families in which one caregiver suffered from depression; it found that compared with the other groups, in families in which a parent had HD, both parents used more dysfunctional parenting styles (Vamos et al., 2007). Our findings suggest that the strain associated with the presence of HD in any family can be so overwhelming that without appropriate help, there will be a risk that all caregivers' ability to provide adequate support to their children is compromised. Moreover, in families in which a caregiver without HD also suffers from physical or psychological illness, high household dysfunction may contribute to a severely adverse upbringing for children.

To our knowledge, no previous study has described how the dynamics between caregivers may affect children in families with HD. These findings raise questions that need further investigation. For instance, we could ask whether families in which a parent has HD is at a higher risk of household dysfunction, abuse, and neglect, due not only to the disease characteristics of HD but also to the dysfunctional dynamics between the caregivers.

### *Implications for practice*

In view of the present study's findings, healthcare and support providers should pay particular attention to children in families with HD. All members of the family should be provided with support throughout the progression of the disease and should be questioned about the possible occurrence of abuse, household dysfunction, and neglect. Because the disease will affect individuals and their families differently and at different developmental stages, emphasis must be placed on each family's specific needs. A thorough investigation of resources and support available to the children from caregivers without HD, as well as of the dynamics between caregivers, needs to be included. Healthcare and support providers should be particularly aware of extra support needs when the caregiver without HD is suffering from illness or experiences severe strain. It is likely that young people or adults who grew up with a parent with HD may need psychological help to deal with the negative experiences and adversity they have experienced while growing up. They may also experience physical and psychological difficulties later in life and may need to seek out psychological support as adults. Clinicians offering support to these adults should be aware of the possibility that they have endured a great deal of adversity, which should therefore be taken into consideration in any treatment or support given to this group.

We cannot eliminate the adversity that is inherent in growing up with a parent with a severe illness, such as HD. We can, however, work actively to build a support system that helps children cope with the distressing experiences they may encounter in order to counteract their negative effects on health and well-being. Our findings suggest that we can help children cope with the stress of having a parent with HD by ensuring that they grow up with caregiver(s) without HD who have capacity to support and provide resources to their children. Understanding the actual and

complex experiences of these children is essential. We believe parent–child support programmes that educate parents about how to foster supportive adult relationships could be beneficial and need to be developed.

### *Strengths and limitations*

This study included a relatively large sample of individuals who grew up with a parent with HD and presented their perceptions of ACEs and caregiver support. Participants were recruited nationally and are believed to be representative of the population under investigation; nevertheless, the qualitative nature of the study may limit the generalisability of the results. We included both teenagers with current experiences and adults remembering experiences from the past. Experiences recounted by the older participants did not reflect current support services. However, our participants' experiences did not appear to differ significantly according to age or available support services; rather, they differed according to the provision of support from their caregiver(s). It is also possible that participants who wished to be a part of this study on children's experiences with HD had more difficult childhoods, whereas people with few negative experiences may not have felt the same need to participate and share their stories. However, the study recruited participants broadly and from several locations to reach participants with a range of experiences. We also consulted with user representatives to verify our findings. We therefore believe our results accurately represent the childhood experiences of many who grew up with a parent with HD.

Support is one of several factors that may act as protection in adversity in childhood. Research also suggest that other factors, such as personal characteristics (e.g. temperament and self-efficacy) and additional support (e.g. extended family members and teachers), could have impacted our participant's perceptions of adversity (e.g. Franke, 2014). Future research should elaborate on our findings, looking at other factors that could help build resilience in children who grow up with a parent with HD.

## Conclusion

Our findings provide new knowledge about adverse experiences of children who grew up in families with HD and their caregiver support. Participants shared an overwhelming number of descriptions of frequent and severe adversity, providing evidence that this group is at risk of negative health consequences during childhood and later in life. Participants without caregiver support often felt unable to cope and described overwhelming feelings of distress. In contrast, having a caregiver who provided support seemed to help participants tolerate stress and adversity. The findings therefore suggest that having available caregiving support can act as a buffer or protect against negative childhood experiences related to growing up with a parent with HD. Our study illustrates the importance of providing support, resources, and education to caregivers without HD, because doing so can prevent or mitigate the negative consequences of HD for the whole family, particularly the children.

## Acknowledgements

We thank those who participated in the study, and their families. We also thank the Norwegian Association for Huntington's Disease, user representatives, and health professionals at the Centre for rare disorders in Norway, in particular Gunvor A. Ruud, for their collaboration and assistance in the development of the study, the recruitment of participants, and encouragement throughout the research process.

## Disclosure statement

The authors report no conflict of interest.

## Funding

This work was supported by Stiftelsen Dam [grant number 2019/FO247779].

## ORCID

Siri Kjoelaas http://orcid.org/0000-0003-3119-703X

## Data availability statement

Due to the nature of this research, participants in this study did not agree for their data to be shared publicly, therefore, supporting data is not available.

## References

Bellis, M. A., Hardcastle, K., Ford, K., Hughes, K., Ashton, K., Quigg, Z., & Butler, N. (2017). Does continuous trusted adult support in childhood impart life-course resilience against adverse childhood experiences – A retrospective study on adult health-harming behaviours and mental well-being. *BMC Psychiatry*, *17*(1), 1–12. https://doi.org/10.1186/s12888-017-1260-z

Boullier, M., & Blair, M. (2018). Adverse childhood experiences. *Paediatrics and Child Health*, *28*(3), 132–137. https://doi.org/10.1016/j.paed.2017.12.008

de Haan, A., Ganser, H. G., Münzer, A., Witt, A., & Goldbeck, L. (2017). Dysfunctional maltreatment-related cognitions in children and adolescents. *Child and Adolescent Psychiatry and Mental Health*, *11*(1), 1–11. https://doi.org/10.1186/s13034-017-0168-1

Dondanville, D. S., Hanson-Kahn, A. K., Kavanaugh, M. S., Siskind, C. E., & Fanos, J. H. (2019). "This could be me": Exploring the impact of genetic risk for Huntington's disease young caregivers. *Journal of Community Genetics*, *10*(2), 291–302. https://doi.org/10.1007/s12687-018-0395-z

Duff, K., Paulsen, J. S., Beglinger, L. J., Langbehn, D. R., & Stout, J. C. (2007). Psychiatric symptoms in Huntington's disease before diagnosis: the predict-HD study. *Biological Psychiatry*, *62*(12), 1341–1346. https://doi.org/10.1016/j.biopsych.2006.11.034

Evans, S. E., Steel, A. L., & DiLillo, D. (2013). Child maltreatment severity and adult trauma symptoms: Does perceived social support play a buffering role?*Child Abuse & Neglect*, *37*(11), 934–943. https://doi.org/10.1016/j.chiabu.2013.03.005

Felitti, V. J., Anda, R. F., Nordenberg, D., Williamson, D. F., Spitz, A. M., Edwards, V., Koss, M. P., & Marks, J. S. (1998). Relationship of childhood abuse and household dysfunction to many of the leading causes of death in adults: The Adverse Childhood Experiences (ACE) Study. *American Journal of Preventive Medicine*, *14*(4), 245–258. https://doi.org/10.1016/j.amepre.2019.04.001

Franke, H. A. (2014). Toxic stress: Effects, prevention and treatment. *Children (Basel, Switzerland)*, *1*(3), 390–402. https://doi.org/10.3390/children1030390

Hill, C. E., Thompson, B. J., Hess, S. A., Knox, S., Williams, E. N., & Ladany, N. (2005). Consensual qualitative research: An update. *Journal of Counseling Psychology*, *52*(2), 196–205. https://doi.org/10.1037/0022-0167.52.2.196

Hill, C. E., Thompson, B. J., & Williams, E. N. (1997). A guide to conducting consensual qualitative research. *The Counseling Psychologist*, *25*(4), 517–572. https://doi.org/10.1177/0011000097254001

Hughes, K., Bellis, M. A., Hardcastle, K. A., Sethi, D., Butchart, A., Mikton, C., Jones, L., & Dunne, M. P. (2017). The effect of multiple adverse childhood experiences on health: A systematic review and meta-analysis. *The Lancet Public Health*, *2*(8), e356–366. https://doi.org/10.1016/S2468-2667(17)30118-4

Kalmakis, K. A., & Chandler, G. E. (2014). Adverse childhood experiences: Towards a clear conceptual meaning. *Journal of Advanced Nursing*, *70*(7), 1489–1501. https://doi.org/10.1111/jan.12329

Kjoelaas, S., Tillerås, K. H., & Feragen, K. B. (2020). The ripple effect: A qualitative overview of challenges when growing up in families affected by Huntington's disease. *Journal of Huntington's Disease*, *9*(2), 129–141. https://doi.org/10.3233/JHD-190377 32065801

Lewit-Mendes, M. F., Lowe, G. C., Lewis, S., Corben, L. A., & Delatycki, M. B. (2018). Young people living at risk of Huntington's Disease: The lived experience. *Journal of Huntington's Disease*, *7*(4), 391–402. https://doi.org/10.3233/JHD-180308

Littleton, H., Horsley, S., John, S., & Nelson, D. V. (2007). Trauma coping strategies and psychological distress: A meta-analysis. *Journal of Traumatic Stress*, *20*(6), 977–988. https://doi.org/10.1002/jts.20276

Meiser-Stedman, R., McKinnon, A., Dixon, C., Boyle, A., Smith, P., & Dalgleish, T. (2019). A core role for cognitive processes in the acute onset and maintenance of post-traumatic stress in children and adolescents. *Journal of Child Psychology and Psychiatry, and Allied Disciplines*, *60*(8), 844–875. https://doi.org/10.1111/jcpp.13054

Roos, R. A. C. (2010). Huntington's disease: A clinical review. *Orphanet Journal of Rare Diseases*, *5*(1), 40–48. https://doi.org/10.1186/1750-1172-5-40

Røthing, M., Malterud, K., & Frich, J. C. (2015). Balancing needs as a family caregiver in Huntington's disease: A qualitative interview study. *Health & Social Care in the Community*, *23*(5), 569–576. https://doi.org/10.1111/hsc.12174

Shonkoff, J. P., Garner, A. S., Siegel, B. S., Dobbins, M. I., Earls, M. F., McGuinn, L., Pascoe, J., Wood, D. L., High, P. C., Donoghue, E., Fussell, J. J., Gleason, M. M., Jaudes, P. K., Jones, V. F., Rubin, D. M., Schulte, E. E., Macias, M. M., Bridgemohan, C., Fussell, J., … Wegner, L. M. (2012). The lifelong effects of early childhood adversity and toxic stress. *Pediatrics*, *129*(1), e232–e246. https://doi.org/10.1542/peds.2011-2663

Smith, J. A., Larkin, M. H., & Flowers, P. (2009). *Interpretative phenomenological analysis: Theory, method and research*. Sage.

Vamos, M., Hambridge, J., Edwards, M., & Conaghan, J. (2007). The impact of Huntington's disease on family life. *Psychosomatics*, *48*(5), 400–404. https://doi.org/10.1176/appi.psy.48.5.400

van der Meer, L. B., van Duijn, E., Wolterbeek, R., & Tibben, A. (2012). Adverse childhood experiences of persons at risk for Huntington's disease or BRCA1/2 hereditary breast/ovarian cancer. *Clinical Genetics*, *81*(1), 18–23. https://doi.org/10.1111/j.1399-0004.2011.01778.x

Van Walsem, M. R., Howe, E. I., Ruud, G. A., Frich, J. C., & Andelic, N. (2017). Health-related quality of life and unmet healthcare needs in Huntington's disease. *Health and Quality of Life Outcomes*, *15*(1), 1–10. https://doi.org/10.1186/s12955-016-0575-7

# II

Received: 29 September 2021 | Revised: 31 May 2022 | Accepted: 12 June 2022

DOI: 10.1002/jgc4.1610

ORIGINAL ARTICLE

National Society of Genetic Counselors WILEY

# Dilemmas when talking about Huntington's disease: A qualitative study of offspring and caregiver experiences in Norway

Siri Kjoelaas[1,2] | Tine K. Jensen[2,3] | Kristin B. Feragen[1]

[1]Centre for Rare Disorders, Oslo University Hospital HF Rikshospitalet, Oslo, Norway

[2]Department of Psychology, University of Oslo, Oslo, Norway

[3]Norwegian Centre for Violence and Traumatic Stress Studies, Oslo, Norway

**Correspondence**
Siri Kjoelaas, Centre for Rare Disorders, Oslo University Hospital HF, Rikshospitalet, P.O. Box 4950, Nydalen, 0424 Oslo, Norway.
Email: hagsir@ous-hf.no

**Funding information**
Ekstrastiftelsen Helse og Rehabilitering (Stiftelsen Dam), Grant/Award Number: 2019/FO247779

## Abstract

Research provides a compelling list of reasons why offspring should be included in honest conversations about disease when the disease affects their caregivers. Despite this, we lack in-depth knowledge about how families affected by the severe and complex genetic condition Huntington's disease (HD) experience talking about the many aspects of how this disease affects their lives. This study aimed to provide an in-depth exploration of how offspring with a caregiver with HD and caregivers with a partner with HD experienced talking about disease throughout childhood. Thematic analysis was conducted with semistructured interviews of both caregivers ($n$ = 14) and offspring ($n$ = 36) from families affected by HD, reflecting both current and past experiences. In addition to highlighting the many needs offspring have for knowledge and conversation about the disease with their caregivers, our findings also show that a variety of dilemmas can follow these conversations, including when to talk, what to say, how often HD should be talked about on a day-to-day basis, and whether to share disease-related information with others outside the family. The findings show the complexity of talking with offspring about HD. A difficult task for both offspring and caregivers seemed to be finding out how to balance the many dilemmas that arise in conversations and how to use dialogue to best help offspring adapt and cope with the many challenges that can come with HD. The findings can assist health care professionals, such as genetic counselors, prepare, and guide families affected by HD in the many and complex conversations that arise about the disease, in turn helping offspring adjust and cope with their current lives or future lives affected by HD.

KEYWORDS
caregivers, communication, family, Huntington's disease, offspring

## 1 | INTRODUCTION

Severe genetic disease does not only affect one individual; it can affect a whole family, particularly the offspring. To help offspring adapt to and cope with having a caregiver who suffers from severe disease, research strongly suggests that they should be included in honest conversations about their life situation (Dalton et al., 2019). However, caregivers often find it hard to talk about the disease with

their children and struggle with whether, how, and when to have these talks, and with managing the emotional responses that follow (e.g., Dalton et al., 2019; Metcalfe et al., 2008). This also seems true when talking with offspring about genetic risk in families affected by severe and complex heritable conditions, such as Huntington's disease (HD; e.g., Gaff et al., 2007; Metcalfe et al., 2008; Rowland & Metcalfe, 2013). Although caregivers may need help from health care professionals, such as genetic counselors, to successfully manage conversations with their offspring, we lack in-depth knowledge about how both caregivers and offspring experience the many aspects of the conversations that can come with having a caregiver with HD and how talking about the disease helps or hinders offspring's adaptation and coping.

### What is known about this topic

- Huntington's disease is a neurodegenerative genetic disease that, in affected families, presents major challenges for offspring.
- The many benefits that come with informing and including offspring in honest conversations about the disease when it affects their caregivers are well-established.

### What this paper adds to the topic

- This paper adds a unique and in-depth presentation of both the caregiver's and offspring's perspectives regarding the many aspects of talking about the complex genetic condition of Huntington's disease with offspring.
- The findings suggest that talking with offspring about Huntington's disease extends beyond informing them about genetic risk, and that many dilemmas arise through these conversations.
- Caregivers may need help to navigate through the many dilemmas that arise in conversations with offspring to ensure that offspring in families with Huntington's disease have the best opportunity to adapt and cope.

## 1.1 | Huntington's disease

A neurodegenerative genetic disease, HD causes a progressive and fundamental deterioration of an individual's physical, cognitive, and psychological functioning (Roos, 2010). Because the typical age of onset for HD is between 30 and 50 years, it is also a time in life when many have caretaking responsibilities for offspring when their symptoms first occur (Roos, 2010). HD has a 50% risk of genetic transmission, meaning that offspring are at risk of having inherited the gene and developing the disease themselves. The slow and long-lasting progression also means that both the individual with HD, and their family will live with the disease for an extended time, an average of 17–20 years, before it ultimately leads to death (Roos, 2010). As the disease progresses through multiple stages, the impact on offspring can be even longer. One of these stages includes having a caregiver who is at risk or presymptomatic, with progressing changes that develop slowly and symptoms that are often present years before a diagnosis is given (Duff et al., 2007). Such symptoms include, but are not limited to, involuntary movements, personality changes, aggression, and apathy (Roos, 2010). Many of these symptoms change the functions needed for caregivers to appropriately understand and respond to their offspring. For instance, cognitive symptoms interfere with the individual's emotional recognition, which may hinder a caregiver's abilities to appropriately read, recognize, and respond to their child's emotions (Watson et al., 2021).

## 1.2 | Growing up with a caregiver with HD

A relatively small but significant body of research has reported on the lived experiences of young people in families with HD, showing that they can grow up with many challenges and stressors as a result of their caregiver's disease. Although growing up with a caregiver with any disease can be a major challenge, HD may present an additional range of unique and complex issues. For instance, HD can break down family systems and compromise the support young people get from both caregivers, effectively leaving many isolated (Forrest Keenan et al., 2007; Kjoelaas et al., 2021; Vamos et al., 2007). One study found that compared with young people who have a caregiver with another inheritable condition—BRCA1/2 breast/ovarian cancer—those with a caregiver with HD reported significantly more negative life events (van der Meer et al., 2012). The profound challenges posed in the lives of young people in families with HD include, but are not limited to, overwhelming responsibilities as carers (Kavanaugh, 2014; Williams et al., 2009), insecurities about their own future because of the inheritability of the disease (Lewit-Mendes et al., 2018), and disrupted family functioning (Forrest Keenan et al., 2007), including parental dysfunction (Vamos et al., 2007) and exposure to a range of adverse experiences, such as complicated grief, unpredictability, violence, aggression, substance abuse, and suicide (Kjoelaas et al., 2021; van der Meer et al., 2012).

## 1.3 | Talking about Huntington's disease

Although there are available resources for support for family members of someone with HD, such as the Huntington's Disease Youth Organization (HDYO), informing and talking with offspring about the disease is usually seen as the responsibility of family members (Forrest et al., 2003). However, past research looking at families' experiences with informing offspring about genetic risk has indicated that talking about HD may not be an easy or straightforward task. Therefore, those at risk of inheriting HD learn about their family's history of disease in different ways and at different times in their lives (Etchegary, 2006; Forrest et al., 2009; Metcalfe et al., 2011). When it comes to telling or not telling offspring about HD, caregivers seem

to be faced with many challenges, including whether to tell, who should tell, when to tell, and how to tell (e.g., Forrest et al., 2003; Stuttgen et al., 2021). This could partly be because of an inherent dilemma caregivers face: They must choose between the obligation to convey valuable information about genetic risk and their wish to protect their children from information that could cause them to harm or worry (Gaff et al., 2007). Other studies have suggested that offspring generally want to learn about HD and its genetic risk at a young age (Forrest Keenan et al., 2007; Holt, 2006; Stuttgen et al., 2021) and have emphasized the importance of having honest conversations about HD (Sparbel et al., 2008). However, research also suggests that receiving information about HD and its associated risks can be difficult for offspring, leaving them feeling overwhelmed by the information, or feeling like they were told the wrong way (Forrest et al., 2009). These and similar studies have highlighted the distressing and often conflicting processes involved in conversations about the genetic risk that families with HD go through. So far, the main focus of research on talking with offspring about HD has been on informing or being informed about genetic risk and genetic testing. Therefore, we continue to lack in-depth knowledge about the many conversations families could have related to all other aspects of the consequences of the disease and how these conversations are experienced by both the offspring and caregivers.

### 1.4 | The importance of conversations

The many stressors and challenges young people in families with HD experience provide strong reasons to believe that there is a range of aspects regarding the disease that offspring may want to know about and discuss with their caregivers. The conversations caregivers have with their children will play a major role in any child's development but are perhaps particularly important when trying to help children make meaning out of challenging situations (e.g., Ellis et al., 2017; Morris et al., 2016). For instance, children in any family will develop an understanding of themselves and others based on their social experiences (Feldman, 1992). Through verbal and nonverbal interactions with those who are close to them, children build ideas about how the world works by actively trying to make sense of situations, events, and their surroundings (Bruner, 1990). Understanding their surroundings is perhaps particularly important for children who are faced with changes and difficulties, such as their caregiver having HD, as they try to restore a sense of control and predictability by attempting to understand the changes that are occurring. In these situations, conversations within the family help children form an understanding of existential concepts, such as life and death (Keeley, 2016). Research seems to support this notion, providing compelling evidence for why offspring should be included in continuous conversations when their caregivers are affected by the severe disease (Dalton et al., 2019; Rowland & Metcalfe, 2013). For instance, one meta-synthesis of research on family communication about genetic risk concluded that for coping, offspring are helped by having conversations that will check their understanding, provide explanations, and help with ongoing feelings and concerns (Metcalfe et al., 2008). For other types of disease, such as cancer, conversations between caregivers and offspring have been established as a particularly important tool for support and coping in everyday life (Ellis et al., 2017; Morris et al., 2016; Stone et al., 2012) and for decreasing their risk of a range of negative psychological outcomes, including anxiety and depression (Dalton et al., 2019).

### 1.5 | Study aim

Given the importance placed on talking with children about their caregiver's disease in general, there is a need to understand more about conversations between caregivers and offspring when it comes to HD, along with how these conversations help or hinder offspring's coping and adaptation. Because the nature of HD is complex and presents a variety of unique and prolonged stressors for offspring, talking about the disease could be particularly challenging for both the offspring and caregivers. Therefore, families might need help and guidance from health care professionals to effectively prepare for and navigate through the many topics of conversations about the disease so that the offspring will adapt and cope. To accomplish this, those in contact with families affected by HD, such as genetic counselors, need more knowledge about the range and complexity of conversations families possibly have with offspring about HD. To address this, the current study aimed to provide an in-depth exploration of both caregivers' and offspring's perspectives on talking about the many aspects of HD throughout childhood.

## 2 | METHODS

### 2.1 | Study participants and recruitment

Qualitative data were collected as part of a larger study that has the overall aim of exploring several topics related to experiences of growing up with a parent with HD, here in a Norwegian context. In the present study, we analyzed interviews of offspring with a caregiver affected by HD (collected from 04/2018–09/2018) and caregivers who have children with a partner with HD (collected from 07/2019–08/2020). Anyone in Norway over the age of 12 years who had current or previous experiences of growing up with a caregiver with HD and anyone who has/had children with a partner with HD were invited to participate. Information was distributed verbally and through information sheets in different settings where individuals in families affected by HD could be reached. These locations included educational courses for families affected by HD, counseling services at Oslo and Haukeland University Hospitals and at St. Olavs Hospital, the Norwegian Association for Huntington's Disease, along with online settings. Information sheets outlined the study's purpose, provided information about the interview topics, and included the main researcher's name and contact information. Table 1 summarizes the participants' demographic information. In response to the formal and

informal invitations, 36 offspring and 14 caregivers gave their written consent to participate and were included in the analysis ($N = 50$). Offspring ranged in age from 13 to 65 years ($M_{age} = 36.6$ years) and caregivers from 42 to 67 years ($M_{age} = 54.9$ years). The participants reflect a broad spectrum of family life stages, patterns of progression of HD, and formal health care and support systems.

## 2.2 | Data collection

Separate interview guides for the offspring and caregivers were created based on similar topics, relevant literature, and feedback from clinical experts and a group of user representatives. Individual semi-structured interviews were conducted, focusing on both the challenges and protective factors. Broad topics included offspring and caregiver's experiences of growing up in a family with HD, openness about the disease, and experiences of support. For the purpose of the current study, we defined childhood as the phase of life between 1 and 12 years and adolescence as the phase between 13 and 19 years. Table 2 displays the interview topics and sample questions. On average, the interviews lasted 60 min (range: 27–90 min). Face-to-face interviews were generally preferred ($n = 46$); however, a few participants preferred telephone interviews ($n = 4$). The interviews were conducted at the Centre for Rare Disorders at Oslo University Hospital, in other locations outside the home, or in the homes of a few participants. The location of the interviews was based on the preference of our participants. Because many of the participants had challenging home lives with partners or caregivers with HD or felt it was easier to talk in the privacy of our outpatient hospital department, most of the participants preferred to be interviewed outside the setting of their home. Counselors from the Centre for Rare Disorders were involved in planning the study, but they were not involved in data analyses because of their involvement with potential participants. Several researchers with formal training in addressing sensitive topics conducted the interviews, including postgraduate clinical psychology students, a clinical psychologist, and a genetic counselor. The students had no previous experience with HD, whereas the health professionals had previous research experience with the disease. All the interviewers had or had received formal training in qualitative methods before conducting the interviews. The project manager performed sample tests on the correspondence between the recorded interviews and transcripts to ensure accuracy between the transcripts produced by different members of the research team. The interviewers had no previous familiarity with the study participants. Supervision regarding interview techniques was provided, if needed. The project manager (an experienced licensed clinical psychologist) participated in at least two interviews conducted by novice researchers to ensure the reliability and consistency of all the interviewers' practices.

**TABLE 1** Demographic characteristics of the participants ($N = 50$)

| | |
|---|---|
| Offspring ($N = 36$) | |
| Age Group of Offspring | |
| Teenager (13–18 years) | 7 |
| Young adult (19–35 years) | 10 |
| Adult (36–65 years) | 19 |
| Gender | |
| Female | 26 |
| Male | 10 |
| Caregiver with HD | |
| Mother | 19 |
| Father | 17 |
| Caregivers ($N = 14$) | |
| Age Group of Children | |
| School-aged children (0–12 years) | 3 |
| Teenagers (13–18 years) | 4 |
| Young adults (19–35 years) | 5 |
| Adults 36–65 years | 2 |
| Gender | |
| Female | 11 |
| Male | 3 |

## 2.3 | Data analysis

The last author, who is also the project manager, performed sample tests on the correspondence between the recorded interviews and transcripts to ensure accuracy between the transcripts produced by different members of the research team. The analysis was guided by Braun and Clarke's thematic analysis (Braun & Clarke, 2006, 2019). During the analysis process, all authors became familiar with the data by reading and rereading the interview transcripts. The first and second authors generated initial codes by isolating phrases, sentences, and paragraphs, generating a list of codes representing every transcript. The lists of codes were collated to search for themes according to the similarities between them. During this step, we noted how the participants consistently spoke about how they had prepared, communicated, and experienced the topic of talking about HD; this became the focus of the analysis. As we continued to analyze the topic, we found that the participants described a multitude of difficult choices or *dilemmas* that did not necessarily have an obvious or preferable solution. These dilemmas were categorized and used as the overarching framework. It may be important to note that although several participants reflected on the actual dilemmas presented in their interviews, this was not the case for all the caregivers or offspring. Rather, the dilemmas used as the framework in the analysis were found within and between the participants. Themes were reviewed against the data and discussed between the three authors until full agreement had been reached. Table 3 summarizes the main themes and corresponding subthemes.

In the presentation of the findings, the frequency labels *general*, *typical* and *variant*, as suggested by Hill et al. (2005), are used to

**TABLE 2** Main topics and sample items applied in the semistructured interview guide

| Interview topic | Sample question offspring | Sample question caregiver |
|---|---|---|
| Background information | Please describe the family you grew up in? | Please describe the family your children grew up in? |
| Childhood experience | How did your parents' disease affect your family? | How did your children experience the disease? |
| | How has growing up with a parent with HD affected you? | How do you think growing up with a parent with HD affected your child/ children? |
| Disease- and self-disclosure | How did you first learn about HD? | How did your children first learn about HD? |
| | What do you feel are the benefits and disadvantages of disclosing information about HD? | What do you feel are the benefits and disadvantages of disclosing information about HD to children? |
| | What were your experiences with asking about HD if you had questions or concerns? | What information and conversations about HD did you feel like your children needed? |
| Resources and support | What sources of support did you have growing up? | What sources of support did your children have growing up? |
| | What support could you have needed that you did not get? | What support could your children have needed that they did not get? |

**TABLE 3** Overview of themes and subthemes

| Main theme | Subtheme |
|---|---|
| Too soon or too late? | Maturity or adaptability? |
| | My timing or your timing? |
| Too much or too little? | Honesty or caution? |
| | Too often or not enough? |
| To share or not to share? | Help or stigma? |
| | My choice or your choice? |

indicate the degree of representativeness across individual cases. The topics or themes that were general in the sense that they applied to all but one participant are referred to in the text as *all participants*. Topics were considered typical if they applied to more than half of the cases and are referred to as *most participants*. Topics were variant if they were represented in less than half of the sample but in more than two cases; they are referred to in the text as *some participants*. Quotes that represent the themes and subthemes were selected and translated from the original language into English. The participants were given pseudonyms to preserve their anonymity.

## 2.4 | Reflexivity

Reflexivity was important throughout the research process. The first and last authors have experience with HD as researchers. Theoretical knowledge about HD was seen as beneficial during data collection and analysis because it helped to recognize features in the participants' experiences specific to HD. However, this knowledge could also create biased interpretations of the interviews. To enhance validity and counteract group thinking and researcher bias, the first and last authors formed the primary analytic team. The second author, who has extensive experience working with developmental psychology in clinical and research settings but no previous experience or knowledge of HD, analyzed the interviews independently and served as a discussant. Therefore, we were able to benefit from the different positions we had and question any theoretical and professional interpretations made. We encouraged reflection on our different positions and experiences. Consensus was obtained after repeated rounds of independent reading, sharing of notes and open discussions, and then rereading and rediscussing of the interviews.

## 2.5 | Ethical considerations

The participants were informed about the study and their right to withdraw at any time before giving their written consent. The confidentiality of participant information was ensured throughout the research process. Caregiver consent was obtained from the participants under the legal age for health consent (16 years in Norway). Because of the sensitivity of the topics discussed during the interviews, relevant referrals or subsequent follow-ups were arranged, if necessary. Five participants wished to receive follow-up after the interviews and were referred to a clinical psychologist or counselor working with families affected by HD.

# 3 | RESULTS

Despite variations in how and when HD affected the families of the participants when it came to talking about the disease, there seemed to be a large overlap in the main topics addressed by both the offspring and caregivers. In general, having conversations with offspring about HD seems to be a complex task that could last throughout childhood. Common to most families was the lack of available guidance from health care or support services when navigating the dilemmas raised by their conversations. The dilemmas

that the families had been faced with were categorized and used as the overarching framework to present the participants' experiences of the different aspects of talking with offspring about HD. These categories included three main themes: *Too soon or too late? Too much or too little?* and *To share or not to share?* and their corresponding subthemes.

## 3.1 | Too soon or too late?

Taken together, both the caregivers and offspring brought forth several dilemmas related to *when* offspring should be told about HD.

### 3.1.1 | Maturity or adaptability?

A central dilemma relating to *when* offspring should be told about HD is whether they would benefit from waiting to learn about the disease until they were mature enough to understand its complexity or if it was best to talk to offspring at a young age so that they could adapt to and integrate the knowledge as they grew older.

In the current study, the caregivers argued for both options. When making this decision, all the caregivers seemingly weighed the risks and benefits of the different options before choosing the option they felt would best protect their child. Some wanted to wait until their children were older because they were concerned that talking with them about HD at an early age would lead to fear and worry about the future and leave their children in a situation that could be difficult to cope with. One father, who had not yet told his school-aged son that the disease affecting his mother was HD, described why:

> It comes down to the fact that it's a disease he might start thinking he could have. I want him to wait to have to do that until he is done being a kid. But then again, he could figure it out on his own. I can't control that.
>
> (Jeffrey)

As this father also suggested, the other option caregivers had chosen was to tell as early as possible, based on the fear that by waiting too long, their children could get the information elsewhere and possibly be misinformed and confused.

Many of the offspring seemed to understand the dilemma their caregivers faced when choosing between telling earlier or later. One teenager, who had known that his father had HD for as long as he could remember, reflected on the choice his mother had made:

> It's a really difficult balance. You either wait until they are old enough to understand, but then, you have to wait for a long time. Or you tell them right away, but then, they might not understand and might worry unnecessarily.
>
> (Michael, teenager with a father with HD)

As Michael has indicated, there was not necessarily a 'good' time to learn that his caregiver had HD. Nevertheless, most offspring seemed to strongly favor being told as early as possible:

> Children will adapt to things; that's the way it is. You learn how the world works and that this is a part of your world. (…) So yes, I believe it was right that I was told as early as I was.
>
> (Nicole, adult with a father with HD)

The benefits of being told in early childhood, as Nicole suggested, include adapting to this knowledge as one grows older. Learning about HD at an early age also seems to have spared the offspring much of the shock that those who were told in adolescence and adulthood often described:

> I would have wanted to grow up knowing that my mom was going to get sick instead of them lying and it coming as a shock.
>
> (Mia, teenager with a mother with HD)

Not being told until adolescence or adulthood, but only if the disease had been known to their caregivers, often made the offspring feel like they had been lied to and had the possibility of severely disrupting their relationship with and trust in their caregivers. In some instances, the offspring said that they had accepted and understood why they had not learned about the disease earlier. For instance, HD had not been known in some families, and the caregivers did not have this information to share with their children at a younger age. Also, knowing that their caregivers had withheld information from them in an effort to protect them seemed to help the offspring accept why they had not been told sooner. However, learning about HD in adolescence and adulthood came with a range of other complications, regardless of the reasons why their caregivers had not talked about HD at an earlier age. For instance, the new information seemed to collide with already challenging tasks at the time they were told, such as planning a future, engaging in romantic relationships, or having children (especially regarding the possibility of having passed the disease on to their offspring). For some, it also seemed to have been the catalyst for severe reactions, including depression and suicidal thoughts.

### 3.1.2 | My timing or your timing?

Another central dilemma related to *when* offspring are told about HD is the decision that nonaffected caregivers have to make between the needs of their children and those of their partner with HD.

A few caregivers had anticipated that this might become a dilemma, often in collaboration with their partner with HD before symptoms occurred, and had planned decisions about the time

and event before talking with their children about the disease. However, putting their children's needs first was not always an easy task. Instead, because of a lack of insight into the progression of the disease, their partners with HD had not wanted or did not feel the need to tell their children. Therefore, the nonaffected caregivers had been caught in the dilemma of choosing between their own wish to tell their children and their partner's need to deny the subject. For many, this included long negotiations between caregivers, often at the expense of telling their children, as described by this father:

> I knew for years that she [partner with HD] would get the disease, but I couldn't tell the kids because that would go against what she wanted. Of course, I could have done that, but I didn't feel it was right. (…) And what's right then? I don't know … It will be a challenge as long as the one involved doesn't want to tell.
>
> (Gregory, father of young adult children)

The question of whose needs should be prioritized when it comes to children learning about HD did not seem to be experienced as a dilemma for most of the offspring. Instead, they consistently emphasized how they needed or would have needed to learn about HD before symptoms occurred. They were sensitive to changes brought on by the disease and shared how not understanding the slow and subtle alterations had negatively affected them long before their caregivers thought they were noticeable and needed to be explained. Lacking an explanation, many had blamed themselves and were consequently ill-equipped to mitigate the impact HD could have on their future. The offspring also highlighted the importance of being told in planned, rather than spontaneous, events and the importance of caregivers coming forth as emotionally stable and consistent. However, for many, this had not been the case, and they had experiences of seemingly normal events that transitioned into being told about the disease without forewarning. Common to the spontaneous events was that seemed to leave the offspring with the impression that it was their caregiver's needs, and not their own, that had been the priority. Conversations that did not come across as planned seemed to increase feelings of shock, as in the case of this teenager, who spoke about the recent event in which he learned that his father had HD:

> It was so strange how he told us about it; he was probably out of it himself. He just walked up to my mom and said that he got the test results and that he had HD. Smiling and crying, like he had just won a football game. That really pissed me off.
>
> (Jason)

Other offspring described how they had only learned about the disease as a premise for the next steps they were to take in life, such as choosing their education or entering serious romantic relationships.

### 3.2 | Too much or too little?

In addition to *when* offspring should be told about HD, there also seemed to be a range of dilemmas related to *what* to say and *how often* they needed to talk about the disease on a day-to-day basis.

#### 3.2.1 | Honesty or caution?

A central dilemma in terms of *what* to say when talking with offspring about HD was whether they should be told about the disease with complete honesty or whether caregivers should be more careful when conveying this message.

Being left with the power to decide what information was important for their children to have had not been an easy task for caregivers. On the one hand, some feared the consequences of being completely honest and had been cautious about any information they provided:

> I think it's ok to tell the kids about the disease so that they will know what it is, but I feel that you should be really careful about saying too much. Because my son started thinking about it a lot, it really made him worry.
>
> (Barbara, mother to adult children)

On the other hand, other the caregivers wanted to be as honest as possible because they feared the consequences of providing too little information. However, despite wanting to be honest, it still seemed difficult to determine what to say to children at different ages and how to ensure that their children understood the information they received:

> I do wonder how informed they are. At the same time, we have all these magazines available so that they can read when they want and figure it out. I think it's difficult to force-feed them with information when they are not asking for it. But what Huntington's looks like in their minds? I really don't know.
>
> (Jill, mother of teenage children)

Other caregivers had been left with questions about who should be in charge of asking for more information if needed, fearing that they would create worry by informing too much, but finding it difficult to ask their children what they understood and what information they needed.

Taken together, the offspring also seemed conflicted between honesty and caution:

> I want to know, but at the same time, I really don't want to know. Very difficult. I told my dad that unless

it's incredibly relevant, I don't want to hear about it at all.

(Kyle, teenager with a father with HD)

Having been shocked when learning about the disease, some were unsure if children should be told about the more detrimental aspects of HD from a young age:

When I was a teenager, I was told everything at once and that it was inheritable. It was a lot; it really was. So I am delighted to hear that people are getting better at talking with their children because I didn't really have that.

(Tina, adult with a mother with HD)

At the same time, not being sufficiently informed, receiving inaccurate or misleading information, or feeling rejected by caregivers when asking for information seemed to leave the offspring without an understanding of the disease and its consequences. Others felt that they had been provided with either too much information or insufficient information at a young age, without any follow-up to information as they grew older. Although the offspring seemed unsure about what information they wanted, they provided clear suggestions on what they did not want. For instance, they consistently stressed how important it was for children to not be lied to: 'Don't lie to children about HD. Nothing good will come of that. Both options will be difficult for them, but it's better to just be honest' (Kourtney, adult with a mother with HD). To accommodate the seemingly conflicting needs of the offspring for honesty and caution, being informed via a continuous process rather than a surprising, one-time event seemed particularly important.

### 3.2.2 | Too often or not enough?

After the offspring had been told about HD, another dilemma seemed to arise: How often do they need to talk about the disease on a day-to-day basis?

The caregivers had seemingly been faced with the dilemma of finding out how much their children needed to talk and how to support those struggling to cope with the disease. After initially telling their children, some had avoided addressing the subject because they feared what emotions the conversations could trigger and worried that if they mentioned HD too often, their children would not be able to escape from the disease and live a normal life without excessive worry:

It can't be all about Huntington's every day. I could see in my daughter how talking about it bought her down and all the time we spent trying to get her back up …. Many people in our family have committed suicide because of this, and her thoughts were there, too …. So you should tread carefully. That's my opinion.

(Barbara, mother to adult children)

In contrast, other caregivers wanted to ensure that their children were not repressing feelings and aimed to model openness by addressing the day-to-day difficulties:

When something happens, I will talk with the kids right away. I tell them they know how their dad is …. And usually they will answer, 'Yes Mom, we know it's Huntington's. You don't have to talk about it right now, Mom. It's enough, we know'.

(Christina, mother to school-aged children)

As this mother suggests, when it comes to talking about the disease, the caregiver's and offspring's needs do not always match. Some were genuinely worried about how their children were coping with HD and the difficulties they could see that it was creating in their developing lives; they had wanted to use conversation as a tool to help their children but had felt rejected in their attempts because their children felt that any mention of the subject was too much. Feeling powerless over their lack of ability to reach out and provide support, these caregivers had to go to great lengths to ensure that their children felt they had someone available to them if they needed to talk about their situation. To accomplish this, they had to be creative, use indirect ways of communication, and often endure difficult conversations with their children, particularly during the teenage years.

The offspring seemed to have a variety of individual needs when it came to talking about HD on a day-to-day basis. On the one hand, several said that they consistently rejected their caregiver's attempts to talk about the disease; they struggled with expressing and understanding their emotions related to the disease, and wanted to refrain from the subject to maintain a sense of normalcy in their lives or wanted to protect their caregivers from their own thoughts and worries about HD:

My dad and I do talk quite a lot. But there are things I don't want to tell him because I feel that he has a lot on him already. So I kind of limit how much I say because of that.

(Penelope, teenager with a mother with HD)

However, not wanting or feeling the need to talk about HD seemed to be reserved for those who felt that they had caregivers available to do so if they wanted. For most of the offspring, HD had not been addressed on a day-to-day basis, and they did not feel like they had a caregiver to whom they could talk. Consequently, they often longed for a caregiver who would talk to them about minor or major difficulties, finding that a lack of initiative or silence when it came to issues related to HD had severe consequences, including increased worry:

> I think talking about it [HD] and involving everyone, regardless of age, is super important. (…) I can remember all the whispering and secrets, even when I was a teenager, giving me the impression that I wasn't included. It was not a good thing. And when you sense something is said and done, you'll create this version in your head that's a lot worse than the reality. So … to communicate, that is very important.
>
> (Brenda, adult with a mother with HD)

For Brenda, not talking about HD growing up had truly impacted her childhood because it left her wondering what was happening and created a feeling of not having much-needed support.

## 3.3 | To share or not to share?

In addition to the dilemmas faced when talking with offspring about HD within the family, the participants had also faced dilemmas related to whether they should tell others outside the family about the disease.

### 3.3.1 | Help or stigma?

A central dilemma was whether sharing information about HD with others would be of help or if it would increase the burden on the offspring.

For most of the caregivers, informing others, such as schools or other health care services, was seen as an opportunity for their children to be supported, but it was followed by a potential risk of social cost. Some feared that the curiosity or judgment of others could leave their children vulnerable to others' questions and concerns, and they felt they had to prepare their children for the reactions of others. One mother, for whom HD had come as a surprise when her children were teenagers and who had chosen to fully disclose information to everyone they knew, stated the following:

> I had to go and tell my kids that if anyone asks any questions, you can choose if you want to answer them or not. Because I knew that people who wouldn't ask me might use them to get information.
>
> (Mary, mother to teenage children)

Other caregivers wished they had disclosed this information sooner, having seen that it could have had the potential to help their children.

The offspring shared both positive and negative experiences of talking with others outside their families about HD. The decision to tell others seemed to be based on certain premises of those with whom the information would be shared, such as talking with others who are trusted and understood, and/or who were in a similar situation. The benefits of talking with others included having friends who know what the family is going through, which made them feel less lonely and reduced negative social reactions from friends. However, the offspring also frequently encountered negative experiences when telling others. For instance, some had tried talking with others for support but did not feel that it had helped their situation. Others described what they perceived as stigmatizing experiences, such as having a teacher point them out in a class while lecturing about genetic diseases. Not receiving the support and understanding they needed when talking with others seemed to leave the offspring feeling rejected, hindering them from reaching out to others for help and support in the future.

Regardless of how the offspring felt about talking with others about HD, the heritability of the disease seemed to be a particular barrier. The offspring said that they had often been conscious about how this information could be interpreted and feared that their friends would shy away from them socially or that others would be looking for symptoms of HD in them:

> Because of the possibility that I could develop HD, too, I was careful with what I told others. It's a little scary; they might see things they otherwise wouldn't see, and I could be treated differently.
>
> (Gina, adult with a father with HD)

Fear of the stigma associated with disclosing information about HD also extended beyond social interactions. For instance, the offspring feared that it could potentially be harmful to their futures, such as hindering them from obtaining promotions at work or insurance.

### 3.3.2 | My choice or your choice?

Another dilemma that the participants faced when deciding whether to tell others was who had or should have the right to decide to share on behalf of the offspring.

For the caregivers, this seemed to be a dilemma of whether to tell others, with or without the blessing of their children. Although the caregivers wanted to help their children by telling others, it could also be a violation of their children's privacy. This decision had not always been static but rather evolved with their child's age and needs. For instance, information that caregivers had confidently shared with others when their children were younger could have the potential to later interfere with their children's evolving need for independence and control as they grew older:

> I am much more nuanced when it comes to sharing information than I used to be. It's something they have to be able to choose and that they are not obligated to do. And I'm terrible at it. I'll answer every time someone asks about inheritance and everything. And I think to myself, why do I have to do that? Can't they be allowed to decide if they want to answer these questions? Why do I need to hold that lecture? Not

> that there is anything they should be ashamed of but because it's their choice to make.
>
> (Jill, mother to teenage children)

Others also expressed how they did not want to overstep the boundaries set by their children and described how they had waited, 'planting the seeds' of possibilities and hoping that one day, their children would reach out for help and support.

Seen from the perspective of the offspring, being able to choose what others know about HD was related to a dilemma between maintaining control and autonomy and receiving help. Several offspring appeared to have quite strong opinions about sharing their caregiver's diagnosis with others. For instance, some offspring insisted that caregivers should not share any information on their behalf:

> I got the feeling that I didn't have that control. One thing is what I chose to share; another thing is what I can't control. So in terms of HD and children, I would never have involved the school or friends or anyone. I would have kept my mouth shut and let the children make their own decisions.
>
> (Cheryl, adult with a mother with HD)

As Cheryl suggested, the offspring's need for control over what is shared on their behalf seems at times to collide with their caregivers' idea of what would help them. In contrast, other offspring had wanted to share information about their situation but had been hindered by their caregivers' or family's desire to maintain secrecy. Some of these offspring described how they deliberately crossed their caregivers by telling others about the disease and facing the possibility of alienation from their family. The offspring were also particularly aware that choosing to tell others about HD would provide information about other family members' genetic risk. This complicating factor often hindered their decision to disclose information about the disease and seek the support of others.

## 4 | DISCUSSION

The current study provides an in-depth exploration of both the caregivers' and offspring's perspectives on the many aspects of talking about HD. First, the findings suggest that offspring in families with HD have several general needs when it comes to talking about the disease with their caregivers. These needs included wanting to learn about the disease as early as possible, to receive age-appropriate information that is followed up as they grow older, to feel like they can talk about HD on a day-to-day basis, if needed, and to feel like they are in control of the information that is shared with others on their behalf. Second, the findings also suggest that a variety of dilemmas seem to follow the many conversations caregivers have with offspring about HD, including when to talk, what to say, how often the disease should be addressed on a day-to-day basis and whom outside the family should be told.

The offspring and caregivers' perceptions of many of the topics that needed to be addressed seemed to mirror one another and reflect something basic in a parent–child relationship: the child's need for knowledge so that the child can make sense of the situation and the dilemmas that arise when parents want to protect the child from the harshness of their life. The general needs the offspring had to talk about regarding the many aspects that came with growing up with a caregiver with HD that corresponds well with findings from past research. For instance, the fact that the offspring wanted to learn about disease from an early age has been well documented when it comes to HD, genetic illness in general, and a range of other diseases, including cancer and HIV (e.g., Dalton et al., 2019; Forrest Keenan et al., 2007; Rowland & Metcalfe, 2013). The need for offspring to talk about HD as a continuous process that develops over time, rather than a one-time event, has also been reflected in past studies that highlight the incremental nature of telling offspring about genetic risk with HD and other genetic diseases (e.g., Forrest et al., 2003, 2009; Gaff et al., 2007). Our findings add to this knowledge by showing that these conversations also extended to many other aspects of growing up with a caregiver with HD, in addition to talking about genetic risk. For instance, the offspring had to be provided with explanations for the many progressing changes in the caregiver that were experienced as confusing or even frightening, particularly when the offspring did not understand why they occurred. These findings mirror research on other diseases, such as cancer, by showing that children want to be included in honest conversations about the many ways their lives are affected and that caregivers play an important role in helping offspring create an understanding of HD through a variety of different conversations (Dalton et al., 2019). The findings also show that the many consequences it can have for offspring when they do not have their needs for information and conversation met. As supported by past research, not receiving sufficient information leads to feelings of distress, hinders their ability to cope with the situation (Forrest Keenan et al., 2007), and could result in inaccurate and dysfunctional beliefs about their own responsibility for the changes that occurred (Eklund et al., 2020).

The need to talk about HD expressed by the offspring in the current study may also mirror the developmental needs of any child in any situation. For instance, the need to receive information with follow-up as they grow older and the need to have caregivers who are available for conversations can reflect how a child's understanding develops over time. With younger children, who have a combination of a growing need to predict the world but a poor understanding of the complexity of disease, early information that is not followed up on or explained in age-appropriate ways can easily lead to misunderstandings and self-blame (Schonfeld, 1993). The difficulties those who learned about HD in adolescence described, in contrast to learning about the disease when they were younger, reflect how adolescence can be a time of increased vulnerability for anyone, possibly making it a particularly difficult time to first learn about their caregiver's complex disease (Davidson et al., 2015). Also, with the developmental changes that occur from childhood to adolescence,

the need for control over information and interference from others will change; here, a need to establish autonomy and identity can easily come in conflict with an increased need for support because of their caregiver's disease (Davidson et al., 2015).

Another key finding in the current study included the variety of dilemmas that seemed to present a more nuanced and complex picture of the many conversations that offspring and their caregivers have about HD than previously presented by the literature. Whereas past research has suggested that questions about whether, what and how to inform offspring about genetic risk is a dilemma for many caregivers (e.g., Forrest et al., 2003, 2009; Gaff et al., 2007; Klitzman et al., 2007), our findings suggest that when it comes to HD, dilemmas also arise in a range of other topics of conversation and, importantly, for both the caregivers and offspring. For instance, our data show that offspring generally preferred being told as early as possible. At the same time, they were unsure about whether all information about HD should be disclosed at this time, how often the topic of the disease needed to be talked about in their everyday lives and whether it would be helpful to share information about HD with others.

Whereas past research has demonstrated how HD severely disrupts family systems and demands that family members try to cope (Brouwer-DudokdeWit et al., 2002; Forrest Keenan et al., 2007), a major issue posed by HD on family processes identified in the current study was finding the right balance in the many dilemmas they faced throughout conversations about the disease that would lead to the best outcome for the offspring. Some of the families in our study felt they had been successful at achieving a balance in their conversations about disease, that HD had not defined them, that their family had been a resource for the offspring by helping them cope and adapt and that being open to others had been a support. However, other caregivers and offspring did not feel they had achieved a balance in the dilemmas they encountered, and efforts to protect and shield the one another had instead left the offspring uninformed to make important decisions in their lives, with damaged relations within the family and with a lack of trust in those they felt should have helped them. Drawing on knowledge about other diseases and HD (Dematteo et al., 2010; Forrest et al., 2009; Klitzman et al., 2007), we suggest that the ability to successfully balance dilemmas that arise when talking with offspring could partly be tied to the concept of trust. We are not the first to suggest that trust in relationships impacts conversations about HD. The results of one study by Forrest et al. (2009) suggest that when young people first find out about their family history of HD, the trust they have in those who provide the information may impact how the message is received. In another study that looked at patterns of decision-making when disclosing genetic risk information within families, the concept of trust was described as a 'gift' that has the power to strengthen or break relationships when disclosing information more generally (Klitzman et al., 2007). The present study elaborates on these findings by suggesting that trust in relationships could also be relevant in the many conversations that arise about HD, in addition to the disclosure of genetic risk information. For instance, our findings suggest that decisions about *when* and *what* offspring were told seemed to revolve around whether the caregivers trusted their child's ability to understand and cope with information about the disease, and whether the child felt they had trusted that their caregivers' decisions were made in their best interest. Talking about HD on a day-to-day basis also triggered issues of trust in the caregiver–child relationship. For the offspring, the actual frequency of the conversations they had seemed secondary to them trusting that a caregiver was available, if needed. The caregivers, on the other hand, needed to trust their ability to handle difficult conversations involving HD and correctly read and interpret their children's evolving needs.

Few studies have explicitly focused on how families affected by complex heritable conditions, such as HD, talk with offspring about the situations they face in their daily lives. The new knowledge brought forth by the current study, which highlights the many dilemmas that can follow conversations about disease and how it impacts the lives of offspring in detail, provides a better understanding of caregivers' and offspring's perspectives on living with HD and directions for areas where these families may need support from health care professionals in the future.

### 4.1 | Implications for practice

One of the major objectives for health care professionals, such as genetic counselors, when meeting caregivers affected by disease is to facilitate family communication. However, recent research suggests that there is uncertainty among genetic counselors about how to guide parents to talk with their children (Keenan et al., 2020). The in-depth insights of the present study can be used to provide clarity and understanding about the many aspects of talking with offspring about HD. The findings demonstrate the need for families to be helped to meet the individual and general needs offspring have for talking about HD and, equally important, help families prepare for and navigate the many and complex dilemmas they may face during these conversations. We suggest that the caregivers in families with HD should be helped to strengthen their ability to make conscious choices regarding current and future conversations with their offspring. We also suggest that families should be helped develop trust in their caregiver–child relationships to feel confident when exploring and addressing their child's individual and developmental needs for conversation about disease. Also, given that offspring in this and past studies indicate that they do not get the emotional support they need from their caregivers, referring offspring to available patient organizations for support could be beneficial and should be considered when assessing the family's needs.

### 4.2 | Strengths and limitations

A major strength of the present study is the inclusion of the different perspectives of both caregivers and offspring, reflecting both current and past experiences. Although the experiences recounted by the older participants did not necessarily reflect contemporary views on

openness and communication about disease, our participants consistently addressed similar topics, regardless of age. Research has suggested that attitudes and reluctance towards informing offspring in families with HD about the genetic risk have remained stable, despite current possibilities to test for the genetic mutation before symptoms occur (Pierron et al., 2020). As such, we believe this inclusion provided a unique benefit of investigating participants' experiences of conversations about HD throughout childhood and across time. The nature of the sampling in the current study may present some limitations. For instance, it is possible that the offspring who wished to be part of the present study had more difficult experiences they wanted to share, whereas those offspring with few negative experiences may not have felt the same need to participate. In contrast, it is also possible that caregivers who wanted to share their experiences could be those who felt comfortable and confident in their actions and choices regarding their children. Although the current study focused on the perspectives of offspring with a caregiver with HD and caregivers with a partner with HD, we did not include the perspectives of caregivers at risk, presymptomatic, or diagnosed with HD. Because this group is also an integral part of family experiences, future studies could benefit from including this perspective.

## 5 | CONCLUSION

The current study has provided a detailed and in-depth exploration of both caregivers' and offspring's perspectives on talking with offspring about HD. We found that the offspring generally wanted age-appropriate conversations about the disease as early as possible, which are followed up as they grow older, to have caregivers who are available to talk to on a day-to-day basis and to have control over the information that is shared with others about the disease. However, we also found that talking with the offspring about HD included a range of dilemmas that seemed to arise throughout a variety of conversations about the disease, including when to tell, what to say, how often the topic should be brought up in day-to-day life and who to tell outside the family. A difficult task for many families appeared to be finding a balance in these dilemmas that would lead the offspring to adapt to and cope with the many challenges presented by having HD in their lives. Our study illustrates the importance of genetic counselors and other health care professionals to help families with HD prepare for and navigate these dilemmas so that knowledge and conversations can be used to strengthen the offspring's ability to adapt to and cope with their caregiver's disease.

### AUTHOR CONTRIBUTIONS

**Siri Kjoelaas:** Conceptualization; data curation; formal analysis; investigation; methodology; project administration; validation; writing – original draft; writing – review and editing. **Tine Jensen:** Formal analysis; methodology; supervision; validation; visualization; writing – review and editing. **Kristin Feragen:** Conceptualization; data curation; formal analysis; funding acquisition; investigation; methodology; project administration; resources; supervision; validation; visualization; writing – review and editing. Authors SK, TKJ and KBF confirm that they had full access to all the data in the study and take responsibility for the integrity of the data and accuracy of the data analysis. All of the authors gave final approval of this version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work were appropriately investigated and resolved.

### ACKNOWLEDGEMENTS

We thank those who participated in the study and their families. We thank Stiftelsen Dam for funding the study. We also thank the Norwegian Association for Huntington's Disease, user representatives, and health professionals at the Centre for Rare Disorders in Norway, in particular Gunvor A. Ruud, for their collaboration and assistance in the development of the study, the recruitment of participants and encouragement throughout the research process. The work was conducted to fulfill a PhD degree requirement. We thank Stiftelsen Dam for funding the study.

### COMPLIANCE WITH ETHICAL STANDARDS

### CONFLICT OF INTEREST

The authors SK, TKJ, and KBF declare that they have no conflicts of interest.

### HUMAN STUDIES AND INFORMED CONSENT

Approval to conduct this human subjects research was obtained by the Regional Committee for Medical Research Ethics [Health region East, Norway, reference number: 2017/864347].

### ANIMAL STUDIES

No non-human animal studies were carried out by the authors of this article.

### DATA SHARING AND DATA ACCESSIBILITY

Because of the nature of this research, the participants in this study did not agree that their data would be shared publicly; therefore, supporting data are not available.

### ORCID

*Siri Kjoelaas* https://orcid.org/0000-0003-3119-703X
*Tine K. Jensen* https://orcid.org/0000-0002-0751-0228
*Kristin B. Feragen* https://orcid.org/0000-0002-0329-5671

### REFERENCES

Braun, V., & Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative Research in Psychology*, *3*(2), 77–101. https://doi.org/10.1191/1478088706qp063oa

Braun, V., & Clarke, V. (2019). Reflecting on reflexive thematic analysis. *Qualitative Research in Sport, Exercise and Health*, *11*(4), 589–597. https://doi.org/10.1080/2159676X.2019.1628806

Brouwer-DudokdeWit, A. C., Savenije, A., Zoeteweij, M. W., Maat-Kievit, A., & Tibben, A. (2002). A hereditary

disorder in the family and the family life cycle: Huntington disease as a paradigm. *Family Process*, *41*(4), 677–692. https://doi.org/10.1111/j.1545-5300.2002.00677.x

Bruner, J. S. (1990). *Acts of meaning*. Harvard University Press.

Dalton, L., Rapa, E., Ziebland, S., Rochat, T., Kelly, B., Hanington, L., Bland, R., Yousafzai, A., Stein, A., Betancourt, T., Bluebond-Langner, M., D'Souza, C., Fazel, M., Fredman-Stein, K., Harrop, E., Hochhauser, D., Kolucki, B., Lowney, A. C., Netsi, E., & Richter, L. (2019). Communication with children and adolescents about the diagnosis of a life-threatening condition in their parent. *The Lancet*, *393*(10176), 1164–1176. https://doi.org/10.1016/S0140-6736(18)33202-1

Davidson, L. L., Grigorenko, E. L., Boivin, M. J., Rapa, E., & Stein, A. (2015). A focus on adolescence to reduce neurological, mental health and substance-use disability. *Nature*, *527*(7578), 161–166. https://doi.org/10.1038/nature16030

Dematteo, D., Harrison, C., Arneson, C., Salter Goldie, R., & Lefebvre, A. (2010). Disclosing HIV/AIDS to children: The paths families take to truthtelling. *Psychology, Health & Medicine*, *7*(3), 339–356. https://doi.org/10.1080/13548500220139395

Duff, K., Paulsen, J. S., Beglinger, L. J., Langbehn, D. R., Stout, J. C., & Predict-HD Investigators of the Huntington Study Group. (2007). Psychiatric symptoms in Huntington's disease before diagnosis: The predict-HD study. *Biological Psychiatry*, *62*(12), 1341–1346. https://doi.org/10.1016/j.biopsych.2006.11.034

Eklund, R., Kreicbergs, U., Alvariza, A., & Lövgren, M. (2020). Children's self-reports about illness- related information and family communication when a parent has a life-threatning illness. *Journal of Family Nursing*, *26*(2), 102–110. https://doi.org/10.1177/1074840719898192

Ellis, S. J., Wakefield, C. E., Antill, G., Burns, M., & Patterson, P. (2017). Supporting children facing a parent's cancer diagnosis: A systematic review of children's psychosocial needs and existing interventions. *European Journal of Cancer Care*, *26*(1), e12432. https://doi.org/10.1111/ECC.12432

Etchegary, H. (2006). Discovering the family history of Huntington disease (HD). *Journal of Genetic Counseling*, *15*(2), 105–117. https://doi.org/10.1007/s10897-006-9018-7

Feldman, C. F. (1992). The new theory of theory of mind. *Human Development*, *35*(2), 107–117. https://doi.org/10.1159/000277138

Forrest Keenan, K., Miedzybrodzka, Z., Van Teijlingen, E., McKee, L., & Simpson, S. (2007). Young people's experiences of growing up in a family affected by Huntington's disease. *Clinical Genetics*, *71*(2), 120–129. https://doi.org/10.1111/j.1399-0004.2006.00702.x

Forrest, K., Simpson, S. A., Wilson, B. J., Van Teijlingen, E. R., McKee, L., Haites, N., & Matthews, E. (2003). To tell or not to tell: Barriers and facilitators in family communication about genetic risk. *Clinical Genetics*, *64*(4), 317–326. https://doi.org/10.1034/j.1399-0004.2003.00142.x

Forrest, K., van Teijlingen, E., McKee, L., Miedzybrodzka, Z., & Simpson, S. A. (2009). How young people find out about their family history of Huntington's disease. *Social Science and Medicine*, *68*(10), 1892–1900. https://doi.org/10.1016/j.socscimed.2009.02.049

Gaff, C. L., Clarke, A. J., Atkinson, P., Sivell, S., Elwyn, G., Iredale, R., Thornton, H., Dundon, J., Shaw, C., & Edwards, A. (2007). Process and outcome in communication of genetic information within families: A systematic review. *European Journal of Human Genetics*, *15*(10), 999–1011. https://doi.org/10.1038/sj.ejhg.5201883

Hill, C. E., Thompson, B. J., Hess, S. A., Knox, S., Williams, E. N., & Ladany, N. (2005). Consensual qualitative research: An update. *Journal of Counseling Psychology*, *52*(2), 196–205. https://doi.org/10.1037/0022-0167.52.2.196

Holt, K. (2006). What do we tell the children? Contrasting the disclosure choices of two HD families regarding risk Status and predictive genetic testing. *Journal of Genetic Counseling*, *15*(4), 253–265. https://doi.org/10.1007/s10897-006-9021-z

Kavanaugh, M. S. (2014). Children and adolescents providing care to a parent with Huntington's disease: Disease symptoms, caregiving tasks and young carer well-being. *Child & Youth Care Forum*, *43*(6), 675–690. https://doi.org/10.1007/s10566-014-9258-x

Keeley, M. P. (2016). Family communication at the end of life. *Journal of Family Communication*, *16*(3), 189–197. https://doi.org/10.1080/15267431.2016.1181070

Keenan, K. F., McKee, L., & Miedzybrodzka, Z. (2020). Genetics professionals' experiences of facilitating parent/child communication through the genetic clinic. *Journal of Genetic Counseling*, *29*(1), 44–55. https://doi.org/10.1002/jgc4.1179

Klitzman, R., Thorne, D., Williamson, J., Chung, W., & Marder, K. (2007). Disclosures of Huntington disease risk within families: Patterns of decision-making and implications. *American Journal of Medical Genetics Part A*, *143*(16), 1835–1849. https://doi.org/10.1002/ajmg.a.31864

Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2021). 'I knew it wasn't normal, I just didn't know what to do about it': Adversity and caregiver support when growing up in a family with Huntington's disease. *Psychology & Health*, *1-19*, 211–229. https://doi.org/10.1080/08870446.2021.1907387

Lewit-Mendes, M. F., Lowe, G. C., Lewis, S., Corben, L. A., & Delatycki, M. B. (2018). Young people living at risk of Huntington's disease: The lived experience. *Journal of Huntington's Disease*, *7*(4), 391–402. https://doi.org/10.3233/JHD-180308

Metcalfe, A., Coad, J., Plumridge, G. M., Gill, P., & Farndon, P. (2008). Family communication between children and their parents about inherited genetic conditions: A meta-synthesis of the research. *European Journal of Human Genetics*, *16*(10), 1193–1200. https://doi.org/10.1038/ejhg.2008.84

Metcalfe, A., Plumridge, G., Coad, J., Shanks, A., & Gill, P. (2011). Parents and children's communication about genetic risk: A qualitative study, learning from families experiences. *European Journal of Human Genetics*, *19*(6), 640–646. https://doi.org/10.1038/ejhg.2010.258

Morris, J. N., Martini, A., & Preen, D. (2016). The well-being of children impacted by a parent with cancer: An integrative review. *Supportive Care in Cancer*, *24*(7), 3235–3251. https://doi.org/10.1007/S00520-016-3214-2

Pierron, L., Hennessy, J., Tezenas du Montcel, S., Coarelli, G., Heinzmann, A., Schaerer, E., Herson, A., Petit, E., Gargiulo, M., & Durr, A. (2020). Informing about genetic risk in families with Huntington disease: Comparison of attitudes across two decades. *European Journal of Human Genetics*, *29*(4), 672–679. https://doi.org/10.1038/s41431-020-00776-8

Roos, R. A. C. (2010). Huntington's disease: A clinical review. *Orphanet Journal of Rare Diseases*, *5*(1), 1–8. https://doi.org/10.1186/1750-1172-5-40

Rowland, E., & Metcalfe, A. (2013). Communicating inherited genetic risk between parent and child: A meta-thematic synthesis. *International Journal of Nursing Studies*, *50*(6), 870–880. https://doi.org/10.1016/j.ijnurstu.2012.09.002

Schonfeld, D. J. (1993). Talking with children about death. *Journal of Pediatric Health Care*, *7*(6), 269–274. https://doi.org/10.1016/s0891-5245(06)80008-8

Sparbel, K. J., Driessnack, M., Williams, J. K., Schutte, D. L., Tripp-Reimer, T., McGonigal-Kenney, M., Jarmon, L., & Paulsen, J. S. (2008). Experiences of teens living in the shadow of Huntington disease. *Journal of Genetic Counseling*, *17*(4), 327–335. https://doi.org/10.1007/s10897-008-9151-6

Stone, A. M., Mikucki-Enyart, S., Middleton, A., Caughlin, J. P., & Brown, L. E. (2012). Caring for a parent with lung cancer: caregivers' perspectives on the role of communication. *Qualitative Health Research*, *22*(7), 957–970. https://doi.org/10.1177/1049732312443428

Stuttgen, K., McCague, A., Bollinger, J., Dvoskin, R., & Mathews, D. (2021). Whether, when, and how to communicate genetic risk to minors: 'I wanted more information but I think they were scared I couldn't handle it. *Journal of Genetic Counseling*, *30*(1), 237–245. https://doi.org/10.1002/jgc4.1314

Vamos, M., Hambridge, J., Edwards, M., & Conaghan, J. (2007). The impact of Huntington's disease on family life. *Psychosomatics*, *48*(5), 400–404. https://doi.org/10.1176/appi.psy.48.5.400

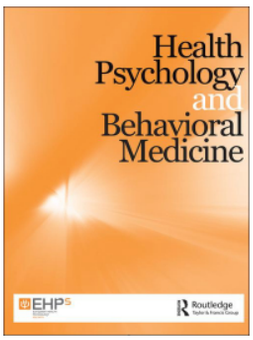
van der Meer, L. B., van Duijn, E., Wolterbeek, R., & Tibben, A. (2012). Adverse childhood experiences of persons at risk for Huntington's disease or BRCA1/2 hereditary breast/ovarian cancer. *Clinical Genetics*, *81*(1), 18–23. https://doi.org/10.1111/j.1399-0004.2011.01778.x

Watson, K. H., Ciriegio, A. E., Pfalzer, A. C., Hale, L., Jones, M. T., Brown, B., Grice, V., Moroz, S., McDonell, K., Claassen, D. O., & Compas, B. E. (2021). Neurobiological and psychosocial correlates of communication between Huntington's disease patients and their offspring. *The Journal of Neuropsychiatry and Clinical Neurosciences*, *33*(4), 321–327. https://doi.org/10.1176/appi.neuropsych.20120309

Williams, J. K., Ayres, L., Specht, J., Sparbel, K., & Klimek, M. L. (2009). Caregiving by teens for family members with Huntington disease. *Journal of Family Nursing*, *15*(3), 273–294. https://doi.org/10.1177/1074840709337126

**How to cite this article:** Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2022). Dilemmas when talking about Huntington's disease: A qualitative study of offspring and caregiver experiences in Norway. *Journal of Genetic Counseling*, *00*, 1–14. https://doi.org/10.1002/jgc4.1610

# III



Routledge
Taylor & Francis Group

Health Psychology and Behavioral Medicine

An Open Access Journal

ISSN: (Print) (Online) Journal homepage: https://www.tandfonline.com/loi/rhpb20

# Social support experiences when growing up with a parent with Huntington's disease

Siri Kjoelaas, Kristin B. Feragen & Tine K. Jensen

**To cite this article:** Siri Kjoelaas, Kristin B. Feragen & Tine K. Jensen (2022) Social support experiences when growing up with a parent with Huntington's disease, Health Psychology and Behavioral Medicine, 10:1, 655-675, DOI: 10.1080/21642850.2022.2104286

**To link to this article:** https://doi.org/10.1080/21642850.2022.2104286



Published online: 29 Jul 2022.

HEALTH PSYCHOLOGY AND BEHAVIORAL MEDICINE
2022, VOL. 10, NO. 1, 655–675
https://doi.org/10.1080/21642850.2022.2104286

Taylor & Francis
Taylor & Francis Group

RESEARCH ARTICLE

OPEN ACCESS Check for updates

# Social support experiences when growing up with a parent with Huntington's disease

Siri Kjoelaas [a,b], Kristin B. Feragen [a] and Tine K. Jensen[b,c]

[a]Centre for Rare Disorders, Oslo University Hospital HF, Oslo, Norway; [b]Department of Psychology, University of Oslo, Oslo, Norway; [c]Norwegian Centre for Violence and Traumatic Stress Studies, Oslo, Norway

**ABSTRACT**

**Background:** Social support is a strong protector factor against the many negative effects stress and adversity in childhood can have on short- and long-term health. However, for young people who are exposed to adversity because their parent suffers from severe neurodegenerative disease, such as Huntington's disease (HD), support from close caregiving relationships can be compromised. This study aimed to investigate what current and past experiences young people who grow up with a parent with HD have with social support outside the parent–child context.

**Methods:** A total of 36 semi-structured qualitative interviews with individuals who had current and past experiences growing up with a parent with HD were analysed using thematic analysis.

**Findings:** Relationships were experienced as supportive when they provided a sense of love, care, or belonging; when they provided coping skills; and when they reduced or alleviated stressors at home. Barriers to receiving and accepting support included their parent's and others' lack of acknowledgement and understanding about their situation and the young people's own need to protect themselves or their family from support they feared could cause harm.

**Conclusion:** Our findings highlight the many important roles persons other than caregivers can have in helping young people who grow up with the distress and adversity of having a parent with a severe disease, such as HD. The findings suggest that by sustaining positive and adaptive emotions and/ or changing distressing emotions, social support help and can compensate for a lack of support in their caregiving relationships. In order for others to be experienced as supportive, the many barriers this vulnerable group may encounter must be addressed and overcome. Most importantly, support providers must understand how HD affects young people.

**ARTICLE HISTORY**
Received 17 February 2022
Accepted 17 July 2022

**KEYWORDS**
Social support; Huntington's disease; adversity; stress; young people

## Introduction

Social support is one of the most significant determinants of psychological health (Thompson, Flood, & Goodvin, 2015). For children and adolescents, studies consistently show that social support can protect against the many damaging effects adversity and

**CONTACT** Siri Kjoelaas hagsir@ous-hf.no Centre for Rare Disorders, Oslo University Hospital HF, Rikshospitalet, Oslo, Norway; Department of Psychology, University of Oslo, Oslo, Norway

distress can have on short- and long-term health (Hughes et al., 2017; Thompson et al., 2015). Young people can be exposed to adversity and distress in many situations, one of which is when their parents suffer an illness or injury that disrupts their ability to provide appropriate care for them. However, we continue to lack in-depth knowledge about how young people who grow up with a caregiver with severe disease, such as Huntington's disease (HD), are helped by their relationships in coping with and adjusting to the many stressors they endure, as well as what potentially hinders this vulnerable group from receiving the support they need.

Support can come from any part of a young person's social network, ranging from informal relationships, such as their parents, extended family members, or peers, to formal contacts, such as teachers or healthcare workers (Taylor, 2011). Theory and research has defined and categorised social support in many ways; however, the literature generally relates to relationships that provide a variety of emotional, informational, or instrumental resources (Thoits, 2011). Emotional support occurs when others provide values, such as love or sympathy; informational support occurs when others provide facts or advice; and instrumental support occurs when others provide behavioural or material assistance. While having these types of supportive relationships is beneficial in any life stage, it is perhaps particularly important in helping to maintain mental and physical health in times of stress and adversity. In times of stress, the relationships we have with others have the power to reduce otherwise harmful psychological reactions and promote positive adjustments by acting as a resource and buffer against the effects of stress (Taylor, 2011). When children and adolescents endure significant adversity and distress, having a robust social support system has been found to be one of the most important factors in minimising the many risks these experiences pose to their short- and long-term health (Hughes et al., 2017; Shonkoff et al., 2012). Consequently, children and adolescents who experience significant ongoing adversity without having the help of important relationships have a higher risk of poor mental health, particularly if they experience this adversity at home (Thompson et al., 2015). A central point for interventions that aims to help families with adversity and distress is, therefore, to enhance existing social support and build new networks in which such children can be supported (Thompson et al., 2015).

Because the idea that social relationships are beneficial is an ingrained part of our understanding of health and well-being, it is perhaps natural to assume that it can easily be implemented for families in which children are exposed to significant adversity and distress and that social support will automatically be beneficial. However, the way in which social support actually assists young people in families with high levels of stress and adversity is, in reality, a much more complex and multifaceted process that is made difficult by a range of factors (Thompson et al., 2015). For instance, there are stressful contexts that may not allow youth to draw on the benefits of social support (Rueger, Malecki, Pyun, Aycock, & Coyle, 2016). Whether support will actually help depends on that person's individual needs and values (Thompson et al., 2015). Also, when considering support, another complicating factor is the need to distinguish between support that is received and support that is perceived. Received support refers to the actual support that is provided, whereas perceived support reflects the person's sense or experience that support would be available if needed (Taylor, 2011). Interestingly, while perceived support has repeatedly been associated with better mental and physical health, the

results are more mixed for received support, where research report weak and even contradictory results (e.g. Bolger & Amarel, 2007; del-Pino-Casado, Frías-Osuna, Palomino-Moral, Ruzafa-Martínez, & Ramos-Morcillo, 2018; Thoresen, Jensen, Wentzel-Larsen, & Dyb, 2014). However, we lack knowledge that takes into account the many ways social support can help young people who grow up with a parent who suffers from severe disease, as well as the barriers this vulnerable group may face in obtaining the support they need. In the current study, we examine social support experiences in a sample of adolescents and adults who grew up in families with a severe and complex illness, Huntington's disease (HD).

Huntington's disease is a progressive neurodegenerative disease with a 50% risk of genetic transmission (McColgan & Tabrizi, 2018). The disease has unquestionably devastating effects on those affected, their families, and the young people whose caregivers are affected in particular. The disease will cause damage to the brain that slowly and gradually affects most functional abilities, and symptoms usually appear between 30 and 50 years of age (McColgan & Tabrizi, 2018), a time in life in which many people have caretaking responsibilities for families and children. The burdens on those affected by HD often last a long time, averaging 17–20 years (McColgan & Tabrizi, 2018), and there is currently no cure and few alternatives available for symptom relief. HD is perhaps most detectable via the disease's visible symptoms, which include uncontrollable movements and changes in motor function (McColgan & Tabrizi, 2018). However, the disease will also create a range of cognitive, and psychiatric symptoms, including personality changes, aggression or apathy, psychosis, a lack of empathy and insight, and difficulties with social perspective-taking years prior to physical symptoms (Eddy & Rickards, 2015; McColgan & Tabrizi, 2018). Although these symptoms are less visible, they may present the most significant challenges to the ability to appropriately care for children.

There exists a small but significant body of research reflecting how HD can impact the lives of young people. Among the multiple and ongoing stressors they may endure are the high risk of being left with overwhelming responsibilities at home (Kavanaugh, 2014) and being exposed to a range of adverse experiences, such as chronic unpredictability, domestic violence, and suicide (Forrest Keenan, Miedzybrodzka, Van Teijlingen, McKee, & Simpson, 2007; Kjoelaas, Jensen, & Feragen, 2021; van der Meer, van Duijn, Wolterbeek, & Tibben, 2012). In addition, many struggle with the constant worry about their own possibility of one day developing the disease and whether or not to get tested as they approach adulthood. It seems many do not get the support they need during this process (Lewit-Mendes, Lowe, Lewis, Corben, & Delatycki, 2018; Forrest Keenan, McKee, & Miedzybrodzka, 2015; Tillerås, Kjoelaas, Dramstad, Feragen, & von der Lippe, 2020). In response to the many and chronic stressors they face, these young people have often been found to struggle with generally poor psychological well-being, including depression and anxiety (Ciriegio et al., 2020; Lewit-Mendes et al., 2018). However, the experiences of these young people do vary, and while some struggle, others cope more successfully with the challenges of growing up with a parent with HD (Forrest Keenan et al., 2007).

To further understand how young people with a parent with HD could be helped to cope and adapt, research from other broader categories of children at risk may be useful, such as at risk of averse childhood experiences (ACE's; Kjoelaas et al., 2021; van der Meer et al., 2012) or with young people who grow up serving as a caregiver for their parent with

physical or mental illness in general, called 'young carers' (Becker, 2000). For this group, one systematic-review shows that taking on caregiving tasks in the family can have both a positive and a negative impact on a young person's development (Chikhradze, Knecht, & Metzing, 2017). A key factor to cope and adapt for young people in families with HD, those at risk of ACEs, and young carers in general, seems to be the protection and guidance provided by having good systems of social support and strong attachments within relationships (e.g. Forrest Keenan et al., 2007; Pakenham, Chiu, Bursnall, & Cannon, 2007; Kjoelaas et al., 2021). Consequently, past research strongly advocates that this group is in need of social support systems that can help them cope and adapt (e.g. Forrest Keenan et al., 2007; Kavanaugh, 2014; Lewit-Mendes et al., 2018). However, we still know little about the ways in which others outside the parent–child context may help young people in families with HD cope with and adapt to the stressors they endure throughout childhood.

What research has suggested is that, for this group of children and adolescents, finding support from others may also not be an easy task. The close attachments they find in their relationships with their caregivers are an important source of social support for any child or adolescent (Bowlby, 1979). However, in families with HD, the disease often disrupts family systems and can compromise the resources and availability of both caregivers as a source of support for their children (Forrest Keenan et al., 2007; Kjoelaas et al., 2021; Mand et al., 2015; Vamos, Hambridge, Edwards, & Conaghan, 2007; Van der Meer et al., 2006). For instance, as one caregiver gradually loses his or her caregiving and supportive abilities due to the progression of HD, the other caregiver may also become less available due to increasing preoccupations with the tasks of caring for their partner (Mand et al., 2015). It is therefore likely that young people in families with HD must rely more heavily on social support from outside the parent–child relationship for help in coping. However, this particular group has also been found to have a range of unmet needs regarding support from others, including help with complex caregiving tasks, and many experience social isolation, receive little support from peers or adults, and want more emotional support (Kavanaugh, 2014; Kavanaugh, Noh, & Studer, 2015; Forrest Keenan et al., 2007).

Given the major risks on short- and long-term health that can come with the enduring stress and adversity many young people with a parent with HD experience with the addition of having support from their caregivers compromised by the disease, understanding how social support can help their coping and adjustment is important for interventions to provide actual help. What possibly hinders this vulnerable group from obtaining the support they need may also be a particularly important piece of this puzzle. Therefore, this study aimed to provide an in-depth understanding of social support for this group. Specifically, we addressed the research question: What are the current and past experiences of social support outside the parent–child context for young people with a parent with HD?

## Methods

### *Sample and setting*

As a part of a larger national study with the overall aim of exploring the experiences of growing up in a family affected by HD [Regional Committee for Medical Research Ethics

Health region East, Norway, reference number: 2017/1613], we analysed interviews collected in 2018. Anyone in Norway over the age of 12 years who had current or previous experiences of growing up with a parent with HD was invited to participate in the study. Information about the study was distributed verbally and through information sheets in several settings where individuals who had grown up in a family affected by HD could be reached. Information about the study was received directly (e.g. via counsellors or other health care professionals) and indirectly (e.g. via the offspring's caregivers or peers). The main locations where information was distributed included educational courses for families affected by HD, genetic counselling services at Oslo and Haukeland University Hospitals and St. Olavs Hospital, the Norwegian Association for Huntington's Disease, and Facebook. The information sheets outlined the study's purpose, provided information about the interview topics, and included the main researcher's name and contact information and a consent form that could be returned by mail. In response to the formal and informal invitations, 42 people provided initial written consent to participate via mail or at the time of the interviews. Of these, six people could not be reached when subsequently contacted for an interview. A total of 36 participants (26 females, 10 males) were interviewed and included in the analysis. The participants were adolescents, young adults, and adults reflecting on their current and past experiences (age range = 13–65 years; $M_{age} = 36.6$ years). Table 1 summarises the participant's demographic information.

### *Data collection procedures*

An interview guide was created based on the relevant literature and feedback from clinical experts and a group of user representatives. The clinical expert references that helped to develop the interview guide included four counsellors from the Centre for Rare disorders at Oslo University Hospital, with extensive experience from working with families with HD in a clinical setting. The user representative group consisted of three individuals with experiences of either growing up with a parent with HD or having children with a

**Table 1.** Participant demographic characteristics ($N = 36$).

| Variable | | Label | *N* |
|---|---|---|---|
| Age | 13–18 years | Teenager | 7 |
| | 19–35 years | Young adult | 10 |
| | 36–65 years | Adult | 19 |
| Gender | Female | | 26 |
| | Male | | 10 |
| Parent with HD | Mother | | 19 |
| | Father | | 17 |
| Occupational status | Student | | 8 |
| | Full-time employment | | 14 |
| | Part-time employment | | 3 |
| | No employment | | 8 |
| | Unknown | | 3 |
| Marital status | In a relationship/married | | 18 |
| | Single | | 12 |
| | Unknown | | 6 |
| Family status | One or more child | | 16 |
| | No children | | 17 |
| | Unknown | | 3 |

partner with HD. Individual semi-structured interviews were conducted, focusing on childhood experiences, family relations, and experiences of support. Table 2 displays interview topics and sample questions. Face-to-face interviews were generally preferred ($n = 33$); however, a few participants preferred telephone interviews ($n = 3$). The interviews were conducted at the Centre for Rare Disorders at Oslo University Hospital, in other locations outside the home, or in the homes of a few participants. The locations of the interviews were based on the preferences of our participants. Because many participants had challenging home lives with partners or caregivers with HD or felt it would be easier to talk in the privacy of our out-patient hospital department, most participants preferred to be interviewed outside their homes. Counsellors from the Centre for Rare Disorders with many years of clinical experience with HD were involved in planning the study, but they were not involved in the data analyses, due to their involvement with potential participants. Five researchers conducted the interviews, and these included one clinical psychologist, three postgraduate psychology students, and two trained health professionals at Oslo University Hospital. The students had no previous experience with HD, whereas the health professionals had previous research experience with the disease. All the interviewers had or received formal training in qualitative methods before conducting the interviews and supervision regarding interview techniques was provided if needed. The interviewers had no previous familiarity with the study participants. The project manager, an experienced licenced clinical psychologist, participated in at least two interviews conducted by novice researchers to ensure the reliability and consistency of all interviewers' practices.

### *Data analysis*

The interviews were recorded and transcribed verbatim. We followed the guidance of Braun and Clarke's reflexive (2006, 2019) thematic analysis. During the first and second steps, all the authors became familiar with the data by reading and re-reading the interview transcripts and noted any text related to support. During this step, we noted that lack of support often was spoken of as barriers to support. That is, the participants experiences of what may have stood in the way of receiving support. In the further analysis we therefore separated experiences of support and barriers in two separate superordinate themes. In the following steps, the first and second authors generated initial codes by isolating phrases, sentences, and paragraphs and generated a list of

**Table 2.** Main topics and sample items applied in the semi-structured interview guide.

| Interview topic | Sample question |
|---|---|
| Background information | *What is your motivation to participate in this study?* |
| | *Please describe the family you grew up in?* |
| Childhood experience | *Tell me about your childhood?* |
| | *How did your parents' disease affect your family?* |
| | *What was your relationship with your mother and father like?* |
| | *How has growing up with a parent with HD affected you?* |
| Disease- and self-disclosure | *What is your experience of disclosing information about HD growing up?* |
| | *What are your thoughts on how parents should inform their children about HD?* |
| Resources and support | *What relationships were your sources of support growing up?* |
| | *How did you/ do you feel about having friends over to visit?* |
| | *How did relationships help you understand and cope with your situation?* |

codes representing every transcript for both superordinate themes separately, i.e. support and barriers to support. The lists of codes were collated to search for themes according to the similarities between them. Themes were chosen for their prevalence in relation to the research question(s). These themes were then reviewed against the data and discussed between the three authors until full agreement was reached; the final themes were then determined and named. Finally, the report was produced.

Reflexivity was emphasised throughout the analysis. Two of the authors (the first and the second) have previous research experience with HD. Knowledge about HD was helpful during the analytic phase. However, this experience also had the potential to create biased interpretations of the interviews. Inspired by the consensual qualitative research model (Hill, Thompson, & Williams, 1997), to enhance the study's validity and counteract group thinking and researcher bias, the first and second author formed the primary analytic team. The third author, who had no previous experience or knowledge of HD, read most interviews independently and served as a discussant during the analyses. Consensus on topics was obtained after repeated rounds of independent reading, sharing notes, discussion, and, finally, rereading and re-discussing the interviews. Last, all cases were analyzed, checking for consistency and soundness, both case by case and across cases. Some participants were young when their parents developed symptoms of HD, whereas others reported memories from late adolescence. The interviewers asked participants to provide the approximate age at which they experienced these events. Given this study's focus, only experiences that had occurred in childhood or adolescence were included in the analysis.

In the presentation of the findings, the frequency labels *general*, *typical*, and *variant*, as suggested by Hill et al. (2005), are used to indicate the degree of representativeness across individual cases. The themes that were general in the sense that they applied to all but one participant are referred to in the text as *all participants*. Topics were considered typical if they applied to more than half of the cases and are referred to as *most participants*. Topics were variant if they were represented in less than half of the sample but appeared in more than two cases; they are referred to in the text as *some participants.* Quotes that illustrate themes and subthemes were selected and translated from the original language into English. Participants were given pseudonyms and identifying information has been omitted.

## Ethical considerations

Ethical approval for the study was obtained from the Regional Committee for Medical Research Ethics (Health region South-East, Norway, reference number: 2017/1613). Participants were informed about the study, provided written consent, and were informed of their right to withdraw at any time. In accordance with ethical regulations, parental consent was obtained for those under the legal age for health consent (16 years in Norway). Due to the sensitivity of the topics discussed during the interviews, relevant referrals or subsequent follow-ups were arranged by a clinical psychologist and project manager if necessary. All the participants received a follow-up call within two weeks after the interview to assess their need for referral to a clinical psychologist and obtain their feedback regarding how they experienced being interviewed about the topics in question. Three participants wished to receive follow-ups after the interviews and were referred to a clinical psychologist.

## Findings

In this study, we examined young people in families with HD's experiences of being supported by others outside the parent–child relationship. The participants' accounts differed in many ways, including when their parents developed symptoms, the severity of the disease, and the disease's impact on them. Despite these differences, they still seemed to share similar descriptions of how the support of others had helped them cope with and adapt to stressors, as well as what barriers they experienced. Their experiences are presented within two superordinate themes, *'How support helped'* and *'Support barriers'*. Table 3 summarises the superordinate themes and corresponding main themes and subthemes.

## How support helped

The first superordinate theme, '*How support helped,*' reflects the different ways others had supported our participants in coping with and adapting to the variety of stressors they experienced during childhood and adolescence. While some participants described having dynamic networks of family, friends, or professionals that had been or could be a form of support when needed, this seemed not to be the case for most participants. Instead, the presence of one significant person or group were the ones providing support. Three main themes were derived reflecting the different ways in which relationships with others were described as supportive: '*I felt connected to someone,*' '*I learned how to cope,*' and *'I got a break from reality.'*

### *I felt connected to someone*

Feeling connected to others was an important source of support that seemed to reflect two emotional functions: 'I *felt loved and cared for*' and '*I felt less alone*'.

**I felt loved and cared for:** Relationships had helped participants when these connections made the young people feel loved and cared for. These were relationships that subtly and continuously helped in the background, such as displaying love and care by staying close and being involved in minor or major events that occur on an everyday basis:

> I have always been very connected to my grandfather. If he had not been there, I wouldn't be … here … We didn't talk all that much, but at the same time, I always knew he was there and

**Table 3.** Superordinate themes and corresponding main themes and subthemes.

| Superordinate themes | Main themes | Subthemes |
|---|---|---|
| How support helped | I felt connected to someone | I felt loved and cared for |
| | | I felt less alone |
| | I learned how to cope | I developed skills |
| | | I developed an understanding |
| | I got a break from reality | I had an escape |
| | | My responsibilities were reduced |
| Support barriers | My needs for support were not understood | Parents are gatekeepers to support |
| | | Others lack of knowledge about my situation |
| | I did not know if involving others would help | Protecting my family |
| | | Protecting myself |

> that he was my protector … . He showed me that he loved me just by being there and that I had someone who cared. It was probably what I had been missing the most, but I had him … who made me who I am and gave me the strength to pull through the way I did. (Tina; adult female, father with HD)

Tina highlights several important functions of such supportive relationships. Through these relationships they had someone who they were close to, who worked to keep them safe and secure, who took notice of them and their situation, and who was on their side. Their home environments were more often than not characterised by chaos and conflict and parents who did not have the capacity to show their children the love and care they needed. In this sense, having close ties with others who made them feel loved and cared for provided a 'buffer' that helped participants cope with and adapt to the distressing emotions they had felt at home: '*My aunt was always the buffer I had. She could be there when my mom and dad were not there, and she would acknowledge me*' (Tammy; adult female, mother with HD).

**I felt less alone:** Relationships with others with similar or comparable experiences had also helped because this connection made the young people feel less alone. These ranged from short encounters to lifelong and profound relationships. One commonality among these relationships was that others with similar experiences seemed to convey an implicit understanding of the young people's true experiences, which helped by making them feel less alone:

> In the beginning, I felt all alone. I felt like I was the only one in the world who had these feelings and these experiences … But when I got to this camp for young people in families with HD, I was really happy to see that I was not the only one and to feel that, finally, I had someone I could talk to. I felt that everyone there was just like me and that I didn't stick out. Everyone was the same and spoke about the same things. Everyone could share their feelings in the same way, and everyone would understand everything that was said. (Danielle; young adult female, father with HD)

As Danielle demonstrates, a mutual or shared understanding helped by providing a sense of belonging, companionship, and normalcy and compensated for or 'buffered' feelings of being alone that many had experienced in other relationships. A sense of trust was also established through these relationships, which provided the young people with the opportunity to have a confidante who could validate their feelings and concerns.

### *I learned how to cope*

Relationships had also consistently helped when they provided the young people with information or advice. The supportive ingredient of this help seemed to be the provision of different tools to cope: '*I developed skills*' and '*I developed an understanding*'.

**I developed skills:** Some participants described how acquiring skills to cope with their feelings and reactions to stressors was helpful. Relationships with others were viewed as supportive when they helped to alter destructive thoughts and reactions to stress and develop strategies they could continue to use to work through problems:

> What has helped is that we have someone who actually steps into our home on a day-to-day basis. She is here a few times a week and talks with my mom and dad, talks with me, talks with my brother. Mostly with me … It has helped because we almost always talk about how

> to handle the conflicts, and as you know, conflicts are one of those things that will intensify when someone in the family gets Huntington's … . (Zachary; teenage male, father with HD)

As Zachary suggests, not all challenges related to HD can be eliminated. Instead, others help by teaching young people to develop tools via which to work around and adapt to obstacles. Different skills taught by support providers had strengthened the young people's coping abilities. Other participants described how such tools gave them the self-efficacy necessary to handle ongoing feelings and concerns and communicate their thoughts and feelings to others using effective strategies.

**I developed an understanding:** Some participants' had also found support in those who specifically increased their understanding of HD. Some talked about those who helped them broaden their perspectives on the disease and develop a positive outlook and hope, despite the possibility that they could also inherit the disease one day:

> I have a lot of good experiences from the stays we had at these healthcare facilities where the whole family was included. I processed a lot there, changed how I viewed things. The more information I got, the safer I felt. In terms of the possibility that I could become ill one day too, I learned that everyone with HD is not the same. That was really good to know. (Dawn; adult female, mother with HD)

As Dawn highlights, information and understanding was a support because it helped the participants view negative experiences or thoughts about their futures in a more adaptive light. An increased understanding gave them room to process distressing experiences from the past and helped them prepare for the progression of their parents' disease in the future with less distress.

### *I got a break from reality*

Relationships also consistently helped participants' cope by providing assistance. Support that came with assistance reflected two types of relief from stress: '*I had an escape' and 'My responsibilities were reduced.'*

**I had an escape.** Several participants described how relationships were supportive because these relationships provided a sense of distance or escape from the distress the participants were experiencing at home. For some, these relationships were with their friends and family, who provided them with the benefit of having somewhere to be outside their home. Dawn highlighted how organised activities had been of help to her:

> I did sports, played instruments, did art, and went hiking. I was given time to be just me but also to get out and to have someone who would see that I was good at something. It was a little bit of a kick, doing sports and activities and having someone notice … . Someone who would acknowledge me during those years. (Dawn; adult female, mother with HD).

In this context, the actions and behaviours of others were helpful because they provided a direct sense of distance from home and an emotional distraction from distressing experiences. Through the eyes of someone outside their family, the young people could also have a break from their distressing thoughts and feelings and build self-esteem, a sense of self, and friendships without constant thoughts about the disease.

**My responsibilities were reduced:** Relationships also helped when others' actions lessened the overwhelming responsibilities young people felt at home. In contrast to the

previous sub-theme reflecting the supportive element of relationships that made young people feeling like they had somewhere to escape, these experiences reflected tangible measures others provided at home and how they helped. For some, support providers alleviated growing responsibilities by providing separate housing for the young people and their parents who could not live at home, or by taking over care tasks such as cleaning the house or personal care for their parent with a disease. For other participants, the measures employed by support providers did not alleviate participants' responsibilities, but they were helpful because they changed their sense of responsibility for the better:

> After we got a puppy, everything changed, or at least a lot. It was like therapy because the dog and my dad are best friends. The dog quickly learned that we need to look out for Dad. He will notice everything and is really good at it … . When we are somewhere else in the house, the dog will come running and bark to let us know when there is something wrong with Dad and we need to come and help … . (Danielle; young adult female, father with HD)

Although the support provided to this participant came with fur and four legs, Danielle's account still highlights the importance of support that changed young people's sense of responsibility. Because young people often worry about and feel responsible for the safety and care of their parents, measures that help them care for their parents also decrease the burden of responsibility, resulting in their home lives feeling less overwhelming.

## Why support was hindered

While reflecting on how others had supported them during their childhood and adolescence, participants generally placed more emphasis on how they felt they had lacked support from others and the many barriers to support they experienced. The second superordinate theme, *Why support was hindered,* therefore covers the participants' descriptions of obstacles to social support, illustrated by two themes: '*My needs for support were not understood*' and '*I did not know if involving others would help',* and reflects barriers that had the potential to hinder all 'types' of support for our participants.

### *My needs for support were not understood*

A general lack of understanding of HD, how the disease truly affected their lives and their needs was described as a major barrier to receiving support. These experiences were captured by two sub-themes; '*Parents are gatekeepers to support'* and '*Others lack of knowledge about my situation'*.

**Parents are gatekeepers to support:** Almost all of the participants described how their parent with HD's lack of insight and awareness of their disease was a barrier to receiving support: '*It's the most difficult part about this whole thing when you are a young carer who wants help but you are not getting anywhere because your parent is denying that they have a disease*' (Ian; young adult male, mother with HD). Some parents were described as unaware of how their own disease developed and oblivious to or in denial about the changes and challenges it was creating in their children's lives. In many instances, their caregivers with the disease were also the gatekeepers regarding whether others could gain insight into the true nature of their home lives and thus make the changes needed for the children. However, because their caregivers'

disease often progressed without being noticed by others outside the home, obtaining help and support was difficult. For instance, when in contact with others, parents could disguise symptoms, often creating an illusion that everything was all right. In addition, some consequently refused or ignored any help or medical attention because they did not find it necessary:

> She did not understand that I needed someone to talk to or why someone came from the hospital to inform us and our friends about what this disease is … . Even to this day, she claims that she is not sick … . (Morgan, teenage female, mother with HD)

Numerous descriptions similar to morgan's account further highlight that the need for support went undetected by others and conflicts between the needs of young people and the needs of parents with HD were generally resolved in favour of the parents.

**Others lack of knowledge about my situation:** When others lacked knowledge about the true nature of HD, this created a range of obstacles that prevented young people from finding support. For some, this meant that their need for support went undetected by potential support providers. This played into parents' denial of the disease because the perception of the needs of the young people was often based on the information provided by parents with HD. As a result, their children's need for support was not taken seriously. Some participants highlighted the fact that this meant they were not provided with continuous support, as is necessary considering the progressive nature of the disease. Others brought forth the idea that the need for mandatory or automatic support systems and measures was not recognised, as it normally would have been for children whose parents had illness or disease with better understood support needs. A general lack of knowledge also seemed to leave others not knowing what to do to help the young people:

> I stood all alone as a teenager … . I later realized that others knew more than they let on, and I can't really understand why they didn't go in to check if things were working at all. Perhaps, they didn't know how to tackle it. Perhaps, it was just easier to do nothing when they didn't know what to do. (Lori; adult female, mother with HD)

As this Lori suggests, believing that they did not have the appropriate tools to help others, potential support providers shied away from intervening in their lives. These experiences could also be related to the stigma, misconceptions, and fear attached to the disease, leaving young people isolated from potential support.

When others lacked knowledge about HD, the support that was actually provided often did not align with young people's support needs:

> I don't think they understand that they don't have the full picture and that I don't feel like I can talk to them at all. Even though I know they are only trying to be supportive [and] they want me to talk to them and want to help me, it's like … it only makes it worse. (Brooke; teenage female, mother with HD)

As Brooke suggests, when potential support providers did not understand the true nature of the young peoples' situation, any support received could feel wrong or misplaced and, in turn, increase young people's distress instead of decreasing it. Generally, meeting others who did not understand seemed to come at a great cost; these situations caused them to feel rejected and helpless in trying to find someone to turn to, potentially deterring them from seeking the help and support of others in the future.

### *I did not know if involving others would help*

Relying on others outside the parent–child relationship for support would, in many instances, also mean allowing others access to their families, home lives or personal thoughts and feelings. Fear of the consequences of this exposure led young people or other family members to avoid the potential support of others, as captured by two sub-themes: '*Protecting my family*' and '*Protecting myself*'.

**Protecting my family:** When young people or other family members were scared of the potential repercussions that could come with others gaining insight into their families, support was hindered. Some young people attempted to protect their families from exposure by hiding their need for support or not reaching out to others. The young people had considered several reasons to do so. Some were scared that others would disrupt their family cohesion, such as separating them from their parent(s) or siblings, or that the help of others would trigger adverse reactions in their parents. Others expressed how they had felt guilty knowing that their need for support would be in conflict with the needs of their parents with HD; for instance, they feared that their parents or themselves would have to leave the home and wondered who would take care of them if the family did not:

> When I look back on the situation today, I think it would have been a very good thing if I had allowed someone to see and take me away, but with children and loyalty, I don't know if that would have worked … (Steven; adult male, father with HD)

As Steven demonstrates, some participants felt an innate loyalty to and need to take care of and protect their parents, even at the cost of finding help themselves. Other family members need to protect their families and themselves could also result in them rejecting and overlooking young people's support needs. Many of the extended families of our participants had been overwhelmed by the responsibilities that have followed generations with HD. These social contacts, which could offer support because they were familiar with the young people's situation and had had similar experiences themselves, had therefore, instead, been caught up in conflicting responsibilities, loyalties, and desires to care for those who had developed the disease, and they protected their own agendas over the support needs of the young people: '*Everybody had enough on their plate already, and they probably wanted to keep the issues within the four walls of our house*' (Steven; adult male, father with HD). Consequently, when it came to social support from extended family members, young people could be left without any neutral party who was acting in these young people's best interest.

**Protecting myself:** When young people were too overwhelmed by the issues they were facing, this could become a barrier to receiving support. Getting help could also mean that the young people would have to give others access to their distressing personal experiences and be forced to address difficult feelings and topics that not all have the strength to face:

> It's not like this 'no one sees me' thing. I have plenty of people who have tried to follow up, tried to provide support. I have been bombarded with counselling sessions, so that's not the problem. I just haven't wanted to deal with it, haven't been able to handle it. (Zachary; teenage male, father with HD)

As Zachary describes, some participants had avoided the help of others or did not reach out to others out of a sense of self-preservation or attempts to cope with ongoing and overwhelming stressful experiences to the best of their abilities. Accepting help or support from others could also mean giving others access to their lives and being exposed to potentially negative judgements or different treatment as a result:

> I didn't want them to treat me differently. I just wanted to be the same as everyone else. I felt they treated me like, 'Oh, we have to take very good care and watch out for this one,' and they didn't do that with the other children, right? (Danielle; young adult female, father with HD)

As Danielle highlights, accepting support could jeopardise participants' need to feel or appear normal. This fear consequently hindered or limited the help and support participants received from close friends, as young people often feared embarrassment or exposure due to bringing someone else into their lives.

## Discussion

This study aimed to investigate how relationships outside the parent–child context help young people who grow up with a parent with HD and explain why this group may not feel that they have the supportive relationships they need. The findings presents a number of new insights into the multifaceted and important roles others outside the parent–child context have in helping young people in families with high levels of stress and adversity, such as families affected by HD. The findings also detail the many barriers that must be overcome for this vulnerable group to receive the support they need.

Past research has shown the distressing circumstances young people in families with HD may endure, as well as showing that support from their caregivers can be severely compromised (e.g. Forrest Keenan et al., 2007; Kavanaugh et al., 2015; Kjoelaas et al., 2021; Vamos et al., 2007; van der Meer et al., 2012). The first main finding in this study builds on this knowledge by identifying the many important roles others' or groups' outside the parent–child context can serve in helping young people cope with and adapt to stressful life circumstances. On a general level, these findings mirror past theory and research by showing that relationships that help young people in families with high levels of stress and adversity can come in many forms, ranging from formal contacts such as healthcare professionals and teachers, to informal relationships, including relatives, neighbours, peers, and even devoted pets (Taylor, 2011). As with past research specifically examining the support needs of young people in families with HD regarding their caregiving duties (Kavanaugh et al., 2015), our participants had been helped through various types social support, including emotional, informational, and instrumental support. However, because research on the role social support plays under stressful circumstances often fails to explain how these relationships actually help someone (Thoits, 2011), our findings are unique due to the in-depth knowledge they provide.

First, we found that emotional support helped young people in families with HD by creating connections that made them feel loved, cared for, and less alone. It is theorised that one way social support can help with stress is when relationship indirectly reduce or

'buffer' the physical and emotional arousal related to stressors, helping sustain someone's positive and adaptive emotions. This is referred to as 'emotional sustenance' (Thoits, 2011). This seems applicable to how participants in this study described the helpful elements of emotional support. For our participants, relationships seemed to have the capacity to significantly help them cope and adapt through the sheer connectedness and value others had transferred to them. As demonstrated by this and other studies, young people with a parent with HD can experience enduring emotional strain and feelings of isolation and loneliness, and at the same time have caregiving relationships where emotional availability is compromised (e.g. Forrest Keenan et al., 2007; Vamos et al., 2007), finding ways to strengthen or build connections that provide these types of emotional support may be particularly important. Our findings suggest that emotional support from others outside the parent–child context can protect young people by sustaining positive or adaptive feelings, such as safety, and by reducing and buffering emotional distress related to a lack of love, a lack of security, or feelings of loneliness. Support providers are encouraged to help young people in families with HD strengthen or build relationships that provide these types of emotional support, such as close family members or others with similar experiences. For instance, young people in families with HD can easily be connected to available arenas for peer support, such as the Huntington's Disease Youth Organisation (HDYO), Young People Affected by HD (YPAHD), and the Huntington's Disease Society of America's National Youth Alliance (NYA).

Second, in addition to relationships that sustained positive emotions through adversity and distress, our participants had also been helped by relationships that actively changed the way they handled stressors and distressing emotions. These types of changes seemed to stem from informational support. The way our participants described informational support seems to reflect what is theorised to be stress buffering by reducing someone's psychological or physiological stress-related arousal, which is called 'active coping assistance' (Thoits, 2011). For our participants, relationships had helped because they taught them the skills needed to handle ongoing issues, which helped them think about, approach, and tackle stressors. In turn, they developed skills such as self-efficacy and effective communication that buffered or reduced emotional distress through building the capacity to handle ongoing challenges. Past research has also focused on the importance of coping strategies when families are affected by HD (Ciriegio et al., 2020; Forrest Keenan et al., 2007). Because studies have shown that these young people often lack information and communication about the disease in their caregiving relationships (e.g. Forrest Keenan et al., 2003; Stuttgen, McCague, Bollinger, Dvoskin, & Mathews, 2021), finding ways to strengthen or build relationships that they can rely on for information and help to understand their situation and HD may be particularly important. Our findings suggest that relationships outside the parent–child context can play a central role in providing information and teaching coping strategies that can help young people in families with HD cope and adapt. Support providers, such as family members, genetic counsellors, or other health care workers are encouraged to ensure that young people have relationships that give this type of informational support, such as providing help to re-structure unhelpful thoughts and worries about their own risk of having HD and whether or not to get tested.

Relationships also helped by providing instrumental support that actively changed the way the participant's handled stressors by reducing or alleviating the many stressors they

encountered at home. Building and strengthening instrumental support may be particularly important for this group because they are often overwhelmed by responsibilities at home and caregiving tasks and may find only limited time for social activities (e.g. Kavanaugh, 2014; Lewit-Mendes et al., 2018). According to our findings, instrumental support can help by transforming young people's feelings of being overwhelmed and can distract them from distressing emotions. In turn, these types of support can also build self-esteem, close connections, and identities that will help protect these young people against current and future stressors. Support providers are therefore encouraged to provide this type of instrumental support. Such support can include financial and practical aid to the families or the young people and appropriate help and support to the parent with HD to alleviate some of the responsibilities and caregiving tasks they could be left with.

The second main finding in this study was the many barriers that hindered the participant's to feel supported. On the one hand barriers were created by a lack of acknowledgement of the young people's situation or a lack of recognition of their needs. On the other hand, they related to the way in which young people and their family members themselves did not allow others to see their needs for support. On a general level, these findings are in line with past research indicating that children in families with HD feel as if they have limited opportunities to find the necessary support (e.g. Kavanaugh et al., 2015; Lewit-Mendes et al., 2018). Other studies have also found that high levels of social support barriers is related to poor mental health trajectories after adversities (e.g. Arnberg, Hultman, Michel, & Lundin, 2013; Thoresen et al., 2014). Importantly, in these studies, experiences of being hindered in obtaining social support were highly associated with psychological distress.

Many of the barriers young people with a parent with HD experienced were related to the nature of their parents' disease and the fact that those who could have helped did not have the necessary knowledge to interpret the young people's support needs correctly. The development of symptoms associated with HD, such as a lack of self-insight and symptom awareness, had severely complicated young people's access to the necessary support. While this is one of the first studies to provide an in-depth understanding of the many ways symptoms of HD can hinder social support within affected families, the neuropsychological and neurocognitive symptoms creating these barriers have been well documented (McColgan & Tabrizi, 2018). Even in the early phases of HD, before the development of visible motor disturbances and a set diagnosis, HD has been found to create fundamental challenges to someone's ability to understand his or her own and other's mental states and needs, as well as to create deficits in the ability to identify inappropriate behaviours (Eddy & Rickards, 2015). The results of our study clearly demonstrates how the early and developing cognitive symptoms of HD may not only affect the individual with the disease, but also, when left undetected, severely limit their children's access to support. For support providers it is important to be aware of these barriers and address them directly.

As is evident in our findings, the heritability of diseases such as HD also means that entire families and generations can be indirectly affected. In this study, this could severely compromise relatives' provision of support for young people. While extended family members normally have the potential to be of great support, their fear, loyalty, and protectiveness could also lead them to prioritise the affected parent's needs above the child's.

According to the social support theory literature, close family ties may be the most accessible and important sources of support because of their closeness to the distressed individual (Thoits, 2011). However, past research has suggested that family norms of secrecy or fear of exposure follow many families with a history of HD, making it difficult for young people to gain support from these relationships (Forrest Keenan et al., 2015). As such, family support can be compromised because they can be 'too close to the situation' to be of help. Because they are often upset about the same situations or behaviours, relatives may minimise the child's experiences, distance themselves from the issues, attempt to solve the problem as quickly as possible, or force a positive outlook that does not actually address a young person's true needs (Thoits, 2011; Thompson et al., 2015). Therefore, support providers are encouraged to also connect young people to arenas for social support outside their families.

Our findings also suggest that, because many of the symptoms of HD reflect impairments in thinking and are not directly observable, young people's needs for support can easily go unnoticed or be misunderstood if helpers lack knowledge about the disease and its consequences on young people. Social support literature also suggests that helpers can be 'too distant or unfamiliar' with an issue to effectively provide support (Thoits, 2011). For our participants, this had come at a great cost, leaving many without the necessary support throughout their upbringing. The general lack of knowledge young people experienced as a barrier had three main consequences. First, when others lacked knowledge about and an understanding of the distress young people were coping with at home, children's support needs could easily be neglected. For instance, the needs of the parent with a disease may be addressed, while the needs of their children are not. Second, when others lacked knowledge about the young people's situation, provided support was not in line with the children's true needs. In line with research that also suggesting that support can yield a range of negative outcomes (Maisel & Gable, 2009), wrong types of support ended up exacerbating instead of reducing stress. Third, when others lacked knowledge, they did not know what to do in order to help, and some participants had even experienced rejection when reaching out for support. Feeling rejected or feeling that the support they receive is negative is perhaps particularly damaging to vulnerable groups. Such experiences may cause someone to feel resentment and betrayal, and seriously compromise their ideas about available support (Thoits, 2011). In turn, as suggested by research on groups with other severely distressing or traumatising experiences, negative social constraints that comes with support is associated with mental health difficulties, and can lead a person to not utilise potential support that might otherwise be within reach, creating a sense of learned helplessness (Andrews, Brewin, & Rose, 2003; Kaniasty & Norris, 2008). Based on these findings, we encourage future studies to investigate ways to improve the public's understanding of the impact HD has on families.

This seems, at least in part, to be reflected through the barriers described in which the young people themselves had hindered support. In some instances, the young people were so overwhelmed by their situation that they did not have the capacity to receive or reach out for support. Social support literature also suggests that experiences can be so devastating and upsetting that supporters themselves are emotionally overwhelmed and become avoidant to protect themselves from distress (Thoits, 2011). Those in contact with these young people are therefore encouraged to provide support that accounts for how distressing experiences affect someone at an individual level. Other

obstacles may include young people's feelings of responsibility to take care of their parents, as well as the loyalty they feel. The idea that children can experience internal conflict resulting from loyalty and protectiveness is not unique to those growing up with a parent with HD. In fact, young carers with a parent with chronic illness frequently feel the need to provide help and protect both parents, and importantly, suppress their own needs to do so (Chikhradze et al., 2017). Nonetheless, young carers also frequently report positive effects, including early maturity, close relationships to their parents, and a preparedness for life (Chikhradze et al., 2017). Support providers are encouraged to help by promoting healthy aspects of having responsibilities and at the same time hindering potentially harmful costs of caring. This includes talking with young people about their needs for support and being aware that family members might have insecurities about the possible risks that can come with receiving support. In adolescence, this strong alliance with parents tends to shift, and having relationships with peers and feeling normal become more important. The literature on social support suggests that some recipients of support can have negative reactions when the support is 'too visible'; adolescents in particular may not want to draw attention to the problem, feel as though they owe others, feel controlled or feel devalued (Maisel & Gable, 2009; Thoits, 2011). This may reflect why the young people in this study frequently described what appeared to be a paradox in terms of support, both needing and hindering the support of peers.

### *Strengths and limitations*

This study included participants who reflected on both current and past experiences from their childhood and adolescence. This means that actual support, perhaps particularly support provided by healthcare services, do not reflect the forms of support available today. However, despite the many differences in the availability of support, participants' descriptions of factors defining whether support was experienced or missed did not seem to vary across time, as one might expect. Therefore, we believe that our findings demonstrate the core elements of support which can be applied across settings, times, and places. Another limitation could be the nature of the sampling. For instance, it is possible that those who grew up with a parent with HD who wished to be a part of this study had more difficult experiences that they wanted to share, whereas those with few negative experiences may not have felt the same need to participate. While a major strength of the qualitative approach in this study is that it provides a greater depth of understanding of participant's experiences than other methods, the findings should still be considered transferable rather than generalisable.

## Conclusion

Past research has shown that young people in families with HD may experience enduring adversity and distress, combined with potentially compromised parent–child relationships that in other contexts provides support. We therefore examined how relationships with others outside this context could help this vulnerable group cope and adjust, as well as the barriers they encountered to feel supported by others. The findings highlight the important role of building and strengthening relationships with others outside the parent–child context, as well as the ways in which these relationships can buffer the

many stressors this group of young people face. Supportive relationships were found to help young people by sustaining positive and adaptive emotions through connections that provide love, care, and belongingness and to handle distressing emotions by teaching coping skills and relieving or alleviating stressors at home. The many barriers to support these young people had encountered also seemed to hinder their abilities to cope with and adjustto the challenges that came with the disease. Barriers included the fact that their need for support was not recognised and understood by their parent with HD and that others who could provide support lacked the knowledge needed to understand the nature of the challenges these young people were facing. Support was also hindered by avoidance and a need to protect themselves and their families from intervention and possible harm. Together these findingssuggests that support may not be experienced as helpful unless it is perceived as addressing the issues young people themselves feel they are facing. Therefore, those in contact with families with HD need to gain knowledge and understanding of the disease and young people's needs, thus helping them overcome the many obstacles they face.

## Acknowledgements

We thank those who participated in the study. We also thank the Norwegian Association for Huntington's Disease, user representatives, and health professionals at the Centre for Rare Disorders in Norway, in particular Gunvor A. Ruud, for their collaboration and assistance in the development of the study, the recruitment of participants, and encouragement throughout the research process.

## Disclosure statement

No potential conflict of interest was reported by the author(s).

## Funding

This work was funded by Stiftelsen Dam under Grant No. 2019/FO247779.

## ORCID

*Siri Kjoelaas* http://orcid.org/0000-0003-3119-703X
*Kristin B. Feragen* http://orcid.org/0000-0002-0329-5671

## References

Andrews, B., Brewin, C. R., & Rose, S. (2003). Gender, social support, and PTSD in victims of violent crime. *Journal of Traumatic Stress*, *16*(4), 421–427. doi:10.1023/A:1024478305142

Arnberg, F. K., Hultman, C. M., Michel, P. O., & Lundin, T. (2013). Fifteen years after a ferry disaster: Clinical interviews and survivors' self-assessment of their experience. *European Journal of Psychotraumatology*, *4*(1), 1–9. doi:10.3402/ejpt.v4i0.20650

Becker, S., Dearden, C., & Aldridge, J. (2000). Young carers in the UK: research, policy and practice. *Research, Policy and Planning*, *8*(2), 13–22.

Bolger, N., & Amarel, D. (2007). Effects of social support visibility on adjustment to stress: Experimental evidence. *Journal of Personality and Social Psychology*, *92*(3), 458–475. doi: 10.1037/0022-3514.92.3.458

Bowlby, J. (1979). The Bowlby-Ainsworth attachment theory. *Behavioral and Brain Sciences*, *2*(4), 637–638. doi:10.1017/S0140525X00064955

Braun, V., & Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative Research in Psychology*, *3*(2), 77–101. doi:10.1191/1478088706qp063oa

Braun, V., & Clarke, V. (2019). Reflecting on reflexive thematic analysis. *Qualitative Research in Sport, Exercise and Health*, *11*(4), 589–597. doi:10.1080/2159676X.2019.1628806

Chikhradze, N., Knecht, C., & Metzing, S. (2017). Young carers: Growing up with chronic illness in the family – A systematic review 2007–2017. *Journal of Compassionate Health Care*, *4*(1), 1–16. doi:10.1186/s40639-017-0041-3

Ciriegio, A. E., Pfalzer, A. C., Hale, L., McDonell, K. E., Claassen, D. O., & Compas, B. E. (2020). Investigating the interplay of working memory, affective symptoms, and coping with stress in offspring of parents with huntington's disease. *Neuropsychology*, *34*(7), 791–778. doi:10.1037/neu0000692

del-Pino-Casado, R., Frías-Osuna, A., Palomino-Moral, P. A., Ruzafa-Martínez, M., & Ramos-Morcillo, A. J. (2018). Social support and subjective burden in caregivers of adults and older adults: A meta-analysis. *PLoS One*, *13*(1), 1–18. doi:10.1371/journal.pone.0189874

Eddy, C. M., & Rickards, H. E. (2015). Theory of mind can be impaired prior to motor onset in huntington's disease. *Neuropsychology*, *29*(5), 792–798. doi:10.1037/neu0000190

Forrest Keenan, K., McKee, L., & Miedzybrodzka, Z. (2015). Help or hindrance: Young people's experiences of predictive testing for Huntington's disease. *Clinical Genetics*, *87*(6), 563–569. doi:10.1111/cge.12439

Forrest Keenan, K., Miedzybrodzka, Z., Van Teijlingen, E., McKee, L., & Simpson, S. (2007). Young people's experiences of growing up in a family affected by Huntington's disease. *Clinical Genetics*, *71*(2), 120–129. doi:10.1111/j.1399-0004.2006.00702.x

Forrest Keenan, K., Simpson, S. A., Wilson, B. J., Van Teijlingen, E. R., McKee, L., Haites, N., & Matthews, E. (2003). To tell or not to tell: Barriers and facilitators in family communication about genetic risk. *Clinical Genetics*, *64*(4), 317–326. doi:10.1034/j.1399-0004.2003.00142.x

Hill, C. E., Thompson, B. J., Hess, S. A., Knox, S., Williams, E. N., & Ladany, N. (2005). Consensual qualitative research: An update. *Journal of Counseling Psychology*, *52*(2), 196–205. doi:10.1037/0022-0167.52.2.196

Hill, C. E., Thompson, B. J., & Williams, E. N. (1997). A guide to conducting consensual qualitative research. *The Counseling Psychologist*, *25*(4), 517–572. doi:10.1177/0011000097254001

Hughes, K., Bellis, M. A., Hardcastle, K. A., Sethi, D., Butchart, A., Mikton, C., Jones, L., & Dunne, M. P. (2017). The effect of multiple adverse childhood experiences on health: A systematic review and meta-analysis. *The Lancet Public Health*, *2*(8), 356–366. doi:10.1016/S2468-2667(17)30118-4

Kaniasty, K., & Norris, F. H. (2008). Longitudinal linkages between perceived social support and posttraumatic stress symptoms: Sequential roles of social causation and social selection. *Journal of Traumatic Stress*, *21*(3), 274–281. doi:10.1002/jts.20334

Kavanaugh, M. S. (2014). Children and adolescents providing care to a parent with Huntington's disease: Disease symptoms, caregiving tasks and young carer well-being. *Child & Youth Care Forum*, *43*(6), 675–690. doi:10.1007/s10566-014-9258-x

Kavanaugh, M. S., Noh, H., & Studer, L. (2015). It'd be nice if someone asked me how I was doing. Like,'cause I will have an answer": Exploring support needs of young carers of a parent with Huntington's disease. *Vulnerable Children and Youth Studies*, *10*(1), 12–25. doi:10.1080/17450128.2014.980370

Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2021). I knew it wasn't normal, I just didn't know what to do about it': Adversity and caregiver support when growing up in a family with Huntington's disease. *Psychology & Health*, *37*(2), 1–19. doi:10.1080/08870446.2021.1907387

Lewit-Mendes, M. F., Lowe, G. C., Lewis, S., Corben, L. A., & Delatycki, M. B. (2018). Young people living at risk of Huntington's disease: The lived experience. *Journal of Huntington's Disease*, *7*(4), 391–402. doi:10.3233/JHD-180308

Maisel, N. C., & Gable, S. L. (2009). The paradox of received social support: The importance of responsiveness. *Psychological Science*, *20*(8), 928–932. doi:10.1111/j.1467-9280.2009.02388.x

Mand, Cara M, Gillam, Lynn, Duncan, Rony E., & Delatycki, Martin B. (2015). "I'm scared of being like mum": The Experience of Adolescents Living in Families with Huntington Disease. *Journal of Huntington's Disease*, *4*(*3*), 209–217. http://dx.doi.org/10.3233/JHD-150148

McColgan, P., & Tabrizi, S. J. (2018). Huntington's disease: A clinical review. *European Journal of Neurology*, *25*(1), 24–34. doi:10.1111/ene.13413

Pakenham, K. I., Chiu, J., Bursnall, S., & Cannon, T. (2007). Relations between social support, appraisal and coping and both positive and negative outcomes in young carers. *Journal of Health Psychology*, *12*(1), 89–102. doi:10.1177/1359105307071743

Rueger, S. Y., Malecki, C. K., Pyun, Y., Aycock, C., & Coyle, S. (2016). A meta-analytic review of the association between perceived social support and depression in childhood and adolescence. *Psychological Bulletin*, *142*(10), 1017-1067. doi:10.1037/bul0000058

Shonkoff, J. P., Garner, A. S., Siegel, B. S., Dobbins, M. I., Earls, M. F., McGuinn, L., Pascoe, J., Wood, D. L., & Wegner, L. M. (2012). The lifelong effects of early childhood adversity and toxic stress. *Pediatrics*, *129*(1), 232–246. doi:10.1542/peds.2011-2663

Stuttgen, K., McCague, A., Bollinger, J., Dvoskin, R., & Mathews, D. (2021). Whether, when, and how to communicate genetic risk to minors:'I wanted more information but I think they were scared I couldn't handle it'. *Journal of Genetic Counseling*, *30*(1), 237–245. doi:10.1002/jgc4.1314

Taylor, S. E. (2011). "Social support: A review". In Friedman, M. S. (Ed.). *The handbook of health psychology* (pp. 189–214). New York, NY: Oxford University Press. Retrieved from: https://taylorlab.psych.ucla.edu/wp-content/uploads/sites/5/2014/11/2011_Social-support_A-review.pdf

Thoits, P. A. (2011). Mechanisms linking social ties and support to physical and mental health. *Journal of Health and Social Behavior*, *52*(2), 145–161. doi:10.1177/0022146510395592

Thompson, R. A., Flood, M. F., & Goodvin, R.. (2015). Social support and developmental psychopathology. In D. Cicchetti & D.J. Cohen (Eds.), *Developmental psychopathology, risk disorder and adaptation* (pp. 1–37). Hoboken, New Jersey: John Wiley & Sons, Inc.

Thoresen, S., Jensen, T. K., Wentzel-Larsen, T., & Dyb, G. (2014). Social support barriers and mental health in terrorist attack survivors. *Journal of Affective Disorders*, *156*, 187–193. doi:10.1016/j.jad.2013.12.014

Tillerås, K. H., Kjoelaas, S. H., Dramstad, E., Feragen, K. B., & von der Lippe, C. (2020). Psychological reactions to predictive genetic testing for Huntington's disease: A qualitative study. *Journal of Genetic Counseling*, *29*(6), 1093–1105. doi:10.1002/jgc4.1245

Vamos, M., Hambridge, J., Edwards, M., & Conaghan, J. (2007). The impact of Huntington's disease on family life. *Psychosomatics*, *48*(5), 400–404. doi:10.1176/appi.psy.48.5.400

Van der Meer, L., Timman, R., Trijsburg, W., Duisterhof, M., Erdman, R., Van Elderen, T., & Tibben, A. (2006). Attachment in families with Huntington's disease: A paradigm in clinical genetics. *Patient Education and Counseling*, *63*(1–2), 246–254. doi:10.1016/j.pec.2005.11.019

van der Meer, L., van Duijn, E., Wolterbeek, R., & Tibben, A. (2012). Adverse childhood experiences of persons at risk for Huntington's disease or BRCA1/2 hereditary breast/ovarian cancer. *Clinical Genetics*, *81*(1), 18–23. doi:10.1111/j.1399-0004.2011.01778.x

# Appendix A: Semi-structured Interview Guides (in Norwegian)

# Appendix A.1: Semi-structured Interview Guide for Offspring Participants

## Del 1: Introduksjon

Generell informasjon om transkripsjon, sletting, frivillig å delta, mulighet til å trekke seg og at dataene er avidentifiserte.

*«Du har vokst opp i en familie der en av dine foreldre har eller hadde Huntington sykdom. Vi er interessert i å høre hvordan det var for deg. Vi er interessert i din historie og din opplevelse av hvordan det var og hvordan du tenker tilbake på dette nå som du er eldre»*

*«Kan du starte med å si noe om hva som er din motivasjon for å delta?»*

## Del 2: Bakgrunnsinformasjon

*«Hvordan vil du beskrive familien du vokste opp i?»*

*«Bodde du med begge foreldrene dine? Helt til hvilken alder?»*

Prober: Hvem av dine foreldre hadde/har HS, vokste du opp sammen med din syke far/mor? dine foreldres sivilstatus da du var barn, deres yrkesbakgrunn, dine søsken (alder og kjønn).

*«Før vi fortsetter med intervjuet, er det noen viktige hendelser i barndommen din du gjerne vil fortelle om eller nevne?»*

## Del 3: Barndommen din - Å tenke tilbake

*«Hvordan opplevde du barndommen din?»*

*«Var HS en kjent sykdom i din slekt? Hvis ja, hvem andre hadde sykdommen i familien?»*

*«Hvordan fikk du høre om HS første gang?*
- *Hvordan opplevde du informasjonen du fikk?»*
- *Hvordan var det for deg å spørre om sykdommen dersom du lurte på noe?»*

*«Hvordan var forholdet ditt til din far? Din mor?»*

*«Hvordan var det for deg da du første gang begynte å merke at far/mor var syk eller rar?*
- *Hva slags forklaringer fikk du om det som var i ferd med å skje og fra hvem?*
- *Hvor gammel var du da?*
- *Hva husker du som de første tegnene på at noe var galt?*
- *Hva slags tanker og følelser hadde du om det den gangen?*
- *Vold? Rusproblemer?*

*«Vet du når far/mor fikk diagnosen HS? Var det samtidig eller ikke samtidig med at du opplevde noe var rart og endret seg for din familie og deg som barn?»*

*«Kan du beskrive hvordan mors/fars sykdom preget familien? Hva merket du mest?*

- *Hvordan opplevde du at sykdommen endret seg over tid?*
- *Hva slags følelser og tanker hadde du om det?»*

Prober: Det er helt normalt i en familie der far eller mor er syk at barn tenker mange tanker og har mange ulike følelser (Evt. eksempler på vanlige følelser: Ansvarsfølelse, ensomhet, føle seg annerledes, skam, sinne, frykt, skyldfølelse). «*I noen familier snakker man mye om det, i andre familier er det mindre vanlig. Hvordan var det i din familie?»*

*«Hvordan har sykdommen preget deg?*

**Del 4: Åpenhet**

*«Hvordan opplevde du åpenheten rundt sykdommen?*

- *Har du noen tanker om hvordan foreldre burde fortelle barna sine om sykdommen og hvor åpne de bør være?*
- *Hva tenker du er fordelene og/eller ulempene med å være åpen om sykdommen?*
- *Hvordan opplevde du å være åpen om sykdommen da du var barn?»*

**Del 5: Støtte og ressurser**

*«Hva fikk du av hjelp til å forstå og takle situasjonen din?»*

*«Hvilke støttepersoner hadde du rundt deg?* (f eks andre familiemedlemmer, lærer, sosiallærer, helsesøster, psykolog, hjemmetjeneste, annet?)

*«Hvordan var det for deg å snakke med vennene dine om at din far/mor hadde HS?*

- *Hvordan opplevde du det var å ha med venner hjem?*
- *Hva med å bli med hjem til andre?»*

*«Hvilke arena eller aktiviteter i hverdagen var en støtte for deg?»* (aktiviteter, hobbyer, kjæledyr, interesser, gode opplevelser)

*«Hva tenker du kunne blitt gjort annerledes og som hadde vært til hjelp for deg som barn?»*

**Del 6: Avslutning**

- *Hva ville du sagt til et annet menneske som er i samme situasjon som deg?*
- *Er det noen andre tema du ønsker at vi skulle snakket om?*
- *Hvordan opplevde du denne samtalen?*

Vi kommer til å ringe deg 1-2 uker etter dette intervjuet for å høre om det har dukket opp andre ting du kunne tenkt deg å snakket om eller om du har noen tanker etter intervjuet som du gjerne vil ta opp med oss eller dele (eventuelt gjøre en avtale om tidspunkt).

# Appendix A.2: Semi-structured Interview Guide Caregivers

## Del 1: INNLEDNING

- Presenter deg selv og evt. andre å tilstede under intervjuet. Gi generell informasjon om transkripsjon, sletting, frivillig å delta, mulighet til å trekke seg og at dataene er avidentifiserte.
- Fortell om hvordan du har tenkt å legge opp intervjuet.
- *«Du har barn som har vokst opp i en familie der den andre forelderen har eller hadde Huntington sykdom. Vi er interessert i din historie og din opplevelse og det er derfor ingen riktige eller gale svar på spørsmålene. Vi forstår at du sitter med mange opplevelser av hvordan sykdommen har påvirket også deg, men ønsker å presisere at i akkurat dette intervjuet vil vi hovedsakelig å ha fokus på hvordan du har opplevd dine barns situasjon under oppveksten. Dersom du skulle ha behov for samtale vil vi tilrettelegge slik at du kan ha en samtale med fagpersonen som intervjuer deg i etterkant og/ eller muligheten til oppfølging fra en av fagpersonene ved avdelingen.*
- *«Kan du starte med å si noe om hva som er din motivasjon for å delta?»*

## Del 3: BARNDOMMEN

### FORELDRENS OPPLEVLESE

- *Kan du beskrive familien barna dine vokser opp i/ har vokst opp i?*
- *Når og hvordan fikk du høre om Huntington sykdom i din partners slekt?*
- *Hva var de første endringer du la merke til hos din partner som kunne tyde på at han/ hun hadde begynt å bli syk?*
- *Hvor gamle var barna dine da?*

### BARNAS OPPLEVELSE

- *Hvordan tror du barna dine har opplevd sykdommen?*
- *Hvordan tror du dine barn vil huske sin egen barndom?*
- *Kan du beskrive hvordan du tror din partners sykdom kan ha preget barna/ barnet ditt som person?*

## Del 4: ÅPENHET/ INFORMASJON

### FORELDRENS OPPLEVELSE

- *Hva slags informasjon fikk du om sykdommen til partneren og fra hvem?*
- *Hva slags informasjon fikk du om eventuelle konsekvenser sykdommen kunne ha for barna deres?*
- *Hvordan og av hvem fikk barna høre om HS første gang?*
- *Hvordan opplever/ opplevde du/ dere å gi informasjon til barna om HS?*
- *Hva er dine tanker om hvordan foreldre burde fortelle barna sine om sykdommen og hvor åpen de bør være?*
- *Hva slags hjelp eller støtte har du fått til å gi barna informasjon barna?*

- *Hva tenker du er fordeler / ulemper ved å være åpen om sykdommen til barna?*

**Prober:** «*I noen familier snakker man mye om det, i andre familier er det mindre vanlig. Hvordan er/ var det i din familie?*»

- *Hvordan opplever du det er å skulle følge opp dette ønske om åpenhet om sykdommen til barna?*

**Prober:** *«vi ser at mange har et ønske om å være åpen om sykdommen til barna, men syns det er vanskelig og derfor ofte venter, hvordan er det for deg?»*

### BARNAS OPPLEVELSE

- *Hva har barnet/ barna dine etterspurt av informasjon og åpenhet om sykdommen?*
- *Hva har barnet ditt sagt til deg om deres tanker og følelser rundt det at de kan arve sykdommen?*

## Del 5: FAMILIEFORHOLD OG HJEMMESITUASJON

### FORELDRENS OPPLEVELSE

- *Hvordan føler du familien og familielivet endret/ endrer seg med sykdommen?*
- *Hvis du skulle oppsummert din relasjon til din partner med 3 ord i dag, hva ville det ha vært?*
- *På hvilken måte har sykdommen påvirket ansvarsfordelingen og omsorgen for barna?*

**Prober:** *Vi ser at flere av barna får flere ansvarsoppgaver i hjemmet med en syk forelder, hvordan har det vært hjemme hos dere?*

- *Hvordan tenker du sykdommen har påvirket din egen evne til ta vare på barna under deres oppvekst?*
- *Hva er dine tanker om hvor mye tid du har hatt til barna i deres oppvekst?*
- *Hvordan vil du beskrive barnas ansvar og oppgaver hjemme?*
- *Vet du om noen opplevelser knyttet til HS som har vært ubehagelige for barna?*
- *Har du opplevd noen vold eller aggresjon mot dine barn eller mot deg selv?*

**Prober:** *Vi ser at flere av barna har negative opplevelser som at foreldrene blir fort irritert eller at personligheten deres forandrer seg, hvordan har det vært hjemme hos dere?*

- *Har det vært noen rusproblemer i deres familie?*
- *Hypotetisk scenario: Hvis du hadde våknet i morgen, og alt hadde vært bra, hva ville ha vært annerledes?*

### BARNAS OPPLEVELSE

- *Og hvis du skulle tenkt deg at du skulle svart for barna/ barnet ditt/ dine på det samme hypotetiske scenarioet: Hvis de skulle våknet i morgen, og alt hadde vært bra for dem, hva tror du ville ha vært annerledes i livene deres?*
- *Hvordan har dine barn beskrevet forhold til sin syke forelder?*

## DEL 5: STØTTE OG RESSURSER

FORELDRENS OPPLEVELSE

- *Hvordan opplevde du å takle situasjonen med en syk partner/ forelder til dine barn?*
- *Hvilken støtte har ditt barn fått fra din partners og egen familie?*
- *Hvordan opplever du hjelpen barna har fått til å forstå og takle sykdommen utenfor familien?*
- *Hvordan opplever du at barna gjør det/ gjorde det på skolen eller barnehagen?*
- *Hva tenker du kunne vært gjort annerledes som hadde vært til hjelp for barna dine?*

BARNAS OPPLEVELSE

- *Hvilken støtte tror du barna dine kunne ønske/ ønsket seg?*
- *I hvilken grad føler barna dine at de har mottatt den støtten du føler det var behov for?*
- *I hvilken grad tror du sykdommen har påvirket fritidsaktivitetene til barna og/ eller dere som familie?*
- *Hvordan opplever du det er/ var for de å ha med seg venner hjem eller være på besøk hos andre?*

## DEL 6: AVSLUTNING

- *Er det noen andre temaer du ønsker at vi skulle snakke om?*
- *Hvordan opplevde du denne samtalen?*

Vi kommer til å ringe deg 1-2 uker etter dette intervjuet for å høre om det har dukket opp andre ting du kunne tenkt deg og snakket om eller om du har noen tanker etter intervjuet som du gjerne vil ta opp med oss eller dele (eventuelt gjøre en avtale om tidspunkt). Dersom du har ønske om det kan vi ta en samtale nå etter intervjuet, eller vi kan avtale en oppfølgingssamtale med en av fagpersonene ved senteret.

# Appendix B: Information Sheet and Consent Forms (in Norwegian)

## Appendix B.1: Information Sheet and Consent Form Offspring Participants

Oslo universitetssykehus

Forespørsel om deltakelse:

**En undersøkelse om det å leve opp i en familie der en av foreldrene har/hadde Huntington sykdom (HS)**

### Bakgrunn og hensikt

Dette er en invitasjon til deg som har vokst opp i en familie der en av dine foreldre har eller hadde Huntington sykdom. Vi ønsker å spørre deg om du kan tenke deg å delta i en intervjuundersøkelse i regi av Senter for sjeldne diagnoser (SSD). Vi ønsker å få mer kunnskap om hvordan det er å være barn i en familie som rammes av HS og tenker at dere som har opplevd dette sitter med viktig informasjon om hvordan det var og hva som eventuelt kan gjøres på en annen måte for å styrke oppfølgingen av barn som står i samme situasjon i dag og deres familier.

### Hva innebærer studien?

Studien baserer seg på utfylling av et spørreskjema og et intervju i form av en samtale mellom deg og en psykolog eller psykologistudent som snart er ferdig utdannet som psykolog. Intervjuene er planlagt en gang mellom august 2017 og juni 2018.
Temaer som tas opp vil være:

- Bakgrunnsinformasjon om din familie
- Hva husker du om situasjonen din da du var barn?
- Når forsto du at din far eller mor var syk?
- Når fikk du vite om HS og hvordan ble det for deg?
- Hva var gode opplevelser og hva var vanskelig for deg?
- Hvordan opplevde du å bli ivaretatt av andre (voksne eller fagpersoner)?
- Hvordan påvirket HS familielivet deres sosialt eller i hverdagen?

### Mulige fordeler og ulemper

Fordelen for deg er at du kan dele dine erfaringer om det å vokse opp i en familie med HS, samt peke på ting som fungerer og komme med forslag til ting som kan forbedres i oppfølging av barn i samme situasjon. Ulempen er at du må bruke ca 1 time av din tid til et intervju. Det kan også være at noen temaer er vanskelige for deg å snakke om, eller oppleves belastende og krevende. Dersom det skjer, vil vi på best mulig måte ivareta deg og du vil også få tilbud om en ny samtale i etterkant dersom du ønsker det.
Du bestemmer selv om du vil komme til SSD i forbindelse med en poliklinisk konsultasjon for å bli intervjuet eller om du ønsker at vi gjør en annen avtale med deg. Studentene som er involvert i prosjektet studerer i Trondheim, Bergen og Oslo. Dersom intervjuet gjennomføres i Trondheim eller Bergen tar vi ansvar for å finne et egnet sted.

### Hva skjer med informasjonen du deler?

Intervjuene vil bli tatt opp på bånd. Dette er et hjelpemiddel, slik at vi kan gjøre om samtalen til tekst etter intervjuet. På denne måten sikrer vi at det ikke går tapt viktig informasjon ved notering, og at det ikke brukes unødvendig mye tid til skriving under intervjuet. Det sikrer også at intervjuer har fullt fokus på deg og det du forteller. Informasjonen som registreres skal kun brukes slik som beskrevet i hensikten med studien. Opplysninger fra intervjuene vil bli behandlet uten at de kan knyttes til ditt/deres navn eller andre gjenkjennende opplysninger. En kodenøkkel/navneliste oppbevares separat fra intervjuene og i låst og brannsikkert arkivskap ved senteret. Alle opplysninger om deg blir avidentifisert. Resultater som publiseres presenteres slik at identiteten til de som deltar i prosjektet ikke kommer fram. Det er kun prosjektmedarbeidere som har adgang til navnelisten og til intervjuene. Alle personopplysninger vil bli slettet når prosjektet er ferdig, trolig innen fem år.

## Frivillig Deltakelse

Det er frivillig å delta. Du kan når som helst og uten å oppgi noen grunn trekke ditt samtykke til å delta og dette vil ikke få konsekvenser for deg eller din familie i forhold til videre oppfølging. Dersom du ønsker å trekke deg ved en senere anledning, kan du kontakte prosjektleder og psykolog Kristin Feragen.

## Personvern

Prosjektet er godkjent av REK og Personvernombudet ved Oslo universitetssykehus. Senter for Sjeldne Diagnoser har det overordnede ansvaret for studien. Hvis du sier ja til å delta, har du rett til innsyn i hvilke opplysninger som er registrert om deg. Barn som er med i studien har rett til å begrense foreldrenes innsynsrett. Du har rett til å korrigere eventuelle feil i opplysninger vi har registrert. Du kan trekke deg fra studien og kreve at opplysninger slettes, med mindre disse allerede er inngått i analyser eller er brukt i publikasjoner.

## Informasjon om resultatet av undersøkelsen

Informasjon om resultater vil komme fram i artikler som vil gjøres tilgjengelig via SSD. Dersom du ønsker annen informasjon om resultatet av undersøkelsen kan du få dette ved å henvende deg til Kristin Billaud Feragen, tlf. 23 07 53 57.

## Deltakelse

Dersom du ønsker å delta i dette forskningsprosjektet, gjør du/dere det ved å underskrive vedlagte samtykkeerklæring og returnere det i vedlagte konvolutt. Psykolog Kristin Billaud Feragen eller en annen rådgiver ved SSD vil deretter ta kontakt med deg og avtale hvordan og når du vil utføre intervjuet. Dersom du ønsker mer informasjon om studien eller har andre spørsmål kan du kontakte Kristin Billaud Feragen på tlf. 23 07 53 57.

Vennlig hilsen,

**Kristin Billaud Feragen**
**Prosjektansvarlig**
**PhD, Psykolog**
**Senter for sjeldne diagnoser**

Tel: 23 07 53 57
krifer@ous-hf.no
www.sjeldnediagnoser.no

~~Senter for Sjeldne Diagnoser~~
~~Posboks 4950 Nydalen~~
~~0424 Oslo~~
~~Sentralbord: 23 07 53 40~~

# Samtykke til å delta i kartleggingsundersøkelse

## En undersøkelse om det å leve opp i en familie der en av foreldrene har/hadde Huntington sykdom (HS)

Jeg har lest den vedlagte informasjonen og er villig til å delta i undersøkelsen

Navn:

_____

(Underskrift, sted, dato)

For å avtale intervju kan Kristin Billaud Feragen eller annen medarbeider i undersøkelsen kontakte meg på telefonnummer:

_____

## Appendix B.2: Information Sheet and Consent Form Caregiver Participants

**INVITASJON TIL FORELDRE OM deltakelse i forskningsprosjektet**

# Barn som pårørende ved Huntington sykdom

Bakgrunn og hensikt

Dette er en invitasjon til deg som har barn som har vokst opp i en familie der den andre forelderen har eller hadde Huntington sykdom (HS). Vi ønsker å få mer kunnskap om hvordan det er å ha barn i en familie som rammes av HS og tenker at dere som foreldre som har opplevd dette sitter med viktig informasjon om hvordan det var og hva som eventuelt kan gjøres på en annen måte for å styrke oppfølgingen av barn som står i samme situasjon i dag og deres familier.

Hva innebærer prosjektet?

Du vil bli spurt om å fylle ut et kort spørreskjema og delta i et intervju i form av en samtale mellom deg og en intervjuer som har utdannelse i psykologi (profesjonsgrad, mastergrad, eller student mot slutten av studieforløpet). Intervjuene er planlagt mellom mars og september 2019.

Vi ønsker blant annet å spørre deg om:

- Bakgrunnsinformasjon om deg og din familie
- Hvordan du tror ditt/dine barn har opplevd sykdommen
- Dine tanker og opplevelse om ditt/dine barns oppvekst
- Dine tanker om hvordan sykdommen har påvirket familielivet og hjemmesituasjonen til barna
- Dine tanker om det å snakke med barna om sykdommen
- Hvordan du har opplevd at barnet/barna har blitt ivaretatt av helsevesenet og støtteapparatet

Hva innebærer DELTAKELSE?

Dersom du svarer ja til å delta i studien vil vi be deg om å fylle ut et spørreskjema og delta i en intervjusamtale hos oss ved SSD, i Bergen eller i Trondheim. Intervjuet kan også gjøres over telefon hvis dette er lettere for deg. Dette vil ta ca. 1 time. For å være med på prosjektet må du først fylle ut et samtykkeskjema på siste side i dette dokumentet og deretter levere eller

sende det til oss. Etterpå blir du ringt av en prosjektmedarbeider på telefon for å avtale tidspunkt for intervjuet.

Mulige fordeler og ulemper

Utover tilfredsstillelsen av å bidra personlig til at hensikten nevnt ovenfor kan oppfylles, vil du eller ditt barn neppe ha spesielle fordeler her og nå av å delta. Som antydet, vil derimot ny kunnskap basert på dine og andres opplevelser kunne komme ditt barn, deg og andre i lignende situasjon til gode i framtiden. En mulig ulempe er tiden det tar å være med på intervjuet. Det kan også være at noen temaer er vanskelige for deg å snakke om, eller oppleves belastende og krevende. Det kan også være at du har behov for å snakke om din egen opplevelse og dine utfordringer utenfor dette prosjektets fokus på barn. I begge tilfeller, vil vi på best mulig måte ivareta deg, og du vil også få tilbud om en ny samtale med psykolog ved avdelingen i etterkant.

Frivillig deltakelse og mulighet for å trekke sitt samtykke

Det er frivillig å delta. Du kan når som helst og uten å oppgi noen grunn trekke ditt samtykke til å delta og dette vil ikke få konsekvenser for deg eller din familie i forhold til videre oppfølging. Dersom du ønsker å trekke deg ved en senere anledning, kan du kontakte prosjektleder og psykolog **Kristin Feragen.**

Hva skjer med informasjonen?

Intervjuene vil bli tatt opp på bånd. Dette er et hjelpemiddel, slik at vi kan gjøre om samtalen til tekst etter intervjuet. På denne måten sikrer vi at det ikke går tapt viktig informasjon ved notering, og at det ikke brukes unødvendig mye tid til skriving under intervjuet. Det sikrer også at intervjuer har fullt fokus på deg og det du forteller. Informasjonen som registreres skal kun brukes slik som beskrevet i hensikten med studien. Opptakene vil bli slettet etter de er skrevet ned. Opplysninger fra intervjuene vil bli behandlet uten at de kan knyttes til ditt/deres navn eller andre gjenkjennende opplysninger. En kodenøkkel/navneliste oppbevares separat fra intervjuene og i låst og brannsikkert arkivskap ved senteret. Alle opplysninger om deg blir avidentifisert. Resultater som publiseres presenteres slik at identiteten til de som deltar i prosjektet ikke kommer fram. Det er kun prosjektmedarbeidere som har adgang til navnelisten og til intervjuene. Alle personopplysninger vil bli slettet når prosjektet er ferdig, senest syv år etter prosjektslutt.

PERSONVERN

Regional komité for medisinsk og helsefaglig forskningsetikk og personvernombudet ved Oslo universitetssykehus har vurdert prosjektet, og har gitt forhåndsgodkjenning [REK nr. 864347, 04.10/2017] Senter for Sjeldne Diagnoser har det overordnede ansvaret for studien. Etter ny personopplysningslov har dataansvarlig og prosjektleder **Kristin Feragen** og PhD kandidat **Siri Kjølaas** et selvstendig ansvar for å sikre at behandlingen av dine opplysninger har et lovlig grunnlag. Dette prosjektet har rettslig grunnlag i EUs personvernforordning artikkel 6a og artikkel 9 nr. 2 og ditt samtykke.

Du har rett til å klage på behandlingen av dine opplysninger til Datatilsynet.

KONTAKTINFORMASJON

Dersom du har spørsmål om prosjektet kan du ta kontakt med følgende:

**Siri H. Kjølaas**
**PhD Kandidat**
**23 07 53 37**
**hagsir@ous-hf.no**

**Kristin Billaud Feragen**
**Prosjektansvarlig**
**23 07 53 42**
**krifer@ous-hf.no**

Senter for sjeldne diagnoser
Oslo Universitetssykehus HF
Rikshospitalet
Postboks 4950 Nydalen
0424 Oslo
www.sjeldnediagnoser.no

**Jeg samtykker til å delta i prosjektet og til at mine personopplysninger brukes slik det er beskrevet**

Dersom du ønsker å delta i studien signerer du under og sender denne siden til oss i vedlagt frankert konvolutt, eller leverer den til oss i person.

- ☐ **Jeg ønsker å delta i studien**

Har du krysset av over, vennligst fyll inn feltene under. Ønsker du ikke å delta, vennligst se bakside.

| | |
|---|---|
| ---------- | ---------- |
| Sted og dato | ------<br>Signatur(er) |

| | |
|---|---|
| ---------- | ---------- |
| Navn med trykte bokstaver | |

Kontaktinformasjon:
Telefon:

Adresse:

**Jeg er IKKE villig til å delta i studien**

- ☐ **Jeg ønsker ikke å delta i studien**

Det hadde vært nyttig for oss å vite hva som er grunnen til at du ikke ønsker å delta. Denne besvarelsen er anonym og frivillig.

Jeg ønsker ikke å delta på grunn av:

# Appendix C: Demographic information questionnaire caregivers (in Norwegian)

**BAKGRUNNSINFORMASJON foreldre: BARN SOM PÅRØRENDE HUNTINGTON SYKDOM (HS)**

**I denne delen av spørreskjemaet ønsker vi bakgrunnsinformasjon om deg. Vær vennlig og besvar alle spørsmål.**

---

**1.** Bakgrunnsinformasjon

a. Hva er din alder? .....

b. Kjønn

☐ Mann
☐ Kvinne

**2.** Hva er din høyeste utdanning?

☐ Grunnskole
☐ Videregående
☐ Høyere utdanning, < 4 år
☐ Høyere utdanning, > 4 år

**3.** Hva er din sivilstatus?

☐ Alene
☐ Gift
☐ Partner/Samboer
☐ Separert/Skilt
☐ Enke/Enkemann

**4.** Hvor mange barn har du med en partener med Huntington sykdom? .....

**5.** Hva er alderen på disse barna/ dette barnet?.....

**6.** Har du barn med andre enn din partner med HS?

☐ JA
☐ NEI

- *Hvis JA, hvor mange barn har du med andre enn din partner med HS?.....*

***7.*** Hvor mange barn bor det i din husholdning?..... (antall)

**8.** Hvor lenge har du visst at din partner har HS i familien? ..... år

**9.** Er du hovedomsorgspersonen for personen med HS?

☐ JA
☐ NEI

**10.** Hvor mange familiemedlemmer bor i din husholdning? .....

**11.**Hva er din yrkesmessige status?

- ☐ Yrkesaktiv heltid
- ☐ Yrkesaktiv deltid
- ☐ Arbeidsledig
- ☐ Hjemmeværende
- ☐ Student
- ☐ Pensjonist/Ufør

**12.**Har du selv noen funksjonsnedsettelser eller sykdommer? ☐ JA

(f.eks. problemer med syn, hørsel, fysisk helse) ☐ NEI

- *Hvis ja*, vær vennlig og beskriv dette:

.....

**13.**Bor personen med HS i samme bolig som deg? ☐ JA ☐ NEI