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People who need people : an attachment perspective on hereditary disease

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Chapter 7

Summary and discussion

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Summary and discussion

In this thesis, a perspective of attachment theory is used to investigate relationships between adverse childhood experiences, adult attachment style, emotion regulation strategies, and distress before and after predictive testing, in persons from families with a late onset hereditary disorder. The hereditary disorders at issue are Huntington's disease (HD), Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL), Hereditary Cerebral Hemorrhage With Amyloidosis – Dutch type (HCHWA-D), or *BRCA1* or *BRCA2* Hereditary Breast and Ovarian Cancer (HBOC). These disorders have an autosomal dominant pattern of inheritance, implying that offspring of effected persons have a 50% risk of inheriting the disease causing gene mutation. HD, CADASIL, and HCHWA-D are progressive and incurable neurogenetic disorders with onset generally in mid-adulthood. For gene mutation carriers, there are currently no options to prevent these diseases. HD symptoms include motor dysfunction (e.g., chorea), psychiatric problems (e.g., depression, apathy, and changes in personality), and cognitive deterioration.^{1,2} CADASIL manifests in migraine with aura, multiple strokes, psychiatric disturbances (e.g., depression, apathy and personality changes), and cognitive decline.³ HCHWA-D is characterized by recurrent strokes and cognitive deterioration.⁴ HBOC is a hereditary cancer syndrome associated with breast cancer, ovarian cancer, and other cancers.⁵ For carriers of a pathogenic *BRCA1* or *BRCA2* gene mutation, there are preventive options that reduce their chances of getting cancer. Cancer associated with HBOC can be treated and may be cured.

The aim of the studies described in this thesis was to find out whether growing up with a parent who has one of these hereditary diseases is associated with adverse childhood experiences in offspring, whether there is a relationship between these experiences and adult psychological characteristics, especially attachment style and style of emotion regulation, and whether these characteristics are associated with the level of distress during and after predictive genetic testing.

The study results increase understanding of how growing up in a family with a hereditary disorder may affect a person's childhood and later life, and contribute to personalized counseling of individuals, couples, and families confronted with HD, CADASIL, HCHWA-D, or HBOC.

In **Chapter 2**, a study is described in which attachment representations of 32 adults at 50% risk for HD, who were raised by a parent with HD, are compared to attachment representations of a non-clinical sample. The aim was to compare the prevalence of insecure attachment representations in these groups, and to see if there were any relationships between HD-related childhood experiences and adult attachment representations. Attachment representations,

defined as the way in which a person views their personal history of attachment experiences, were assessed using the Adult Attachment Interview.⁶

A higher percentage of insecure attachment representations was found in the sample of adults who were raised in a family where a parent was affected with HD. This group showed more insecure-preoccupied attachment representations (indicating ambivalence about attachment experiences) and more unresolved/disorganized attachment representations (indicating experiences of unresolved trauma or loss). A relationship was found between insecure attachment and being younger when the parent started having symptoms of HD. A relationship was found between death of the affected parent before the participant's 18th birthday and an unresolved/disorganized attachment representation.

In **Chapter 3**, the prevalence is described of adverse childhood experiences before age 16 in 74 adults at 50% risk for HD, 82 adults at 50% risk of being predisposed for HBOC, and 101 adults in a non-clinical reference group. Of offspring of a parent with HD, 53% had been exposed to adversity in childhood or adolescence. This group reported the highest mean number of adverse childhood experiences. Compared to the reference group, a higher percentage of offspring of a parent with HD had experienced parental disease and parental dysfunction before age 16. In the group of offspring of a parent with HBOC, 45% had been exposed to adversity in childhood or adolescence. Compared to the reference group, a higher percentage of HBOC offspring had lost a parent before age 16. The odds of having experienced adverse childhood experiences before age 16 were higher in offspring of a parent with HD and in offspring of a parent with HBOC, than in the reference group.

According to this study, growing up in a family where a parent has HD or HBOC is associated with experiencing adversity in childhood or adolescence. This increases the risk for psychopathology and other problems in childhood as well as in adulthood.

In **Chapter 4**, a study is described in which adverse childhood experiences and adult psychological characteristics of 96 adults who had a parent with a neurogenetic disorder (HD, CADASIL, HCHWA-D), 70 adults who had a parent with HBOC, and 127 adults in a non-clinical reference group are compared. Associations between adverse childhood experiences and adult psychological characteristics are studied. The adult psychological characteristics comprised attachment style (attachment anxiety and attachment avoidance), assessed with the Experiences in Close Relationships – Revised (ECR-R),⁷ mental health status, measured with the Mental Health Inventory-5 (MHI-5),⁸ and current psychological symptomatology, assessed with the Brief Symptom Inventory (BSI).^{9,10} Attachment anxiety is the tendency to worry about availability and responsiveness of significant others, to fear interpersonal rejection or abandonment, and to have an excessive need for approval from others. Attachment avoidance

is the tendency to feel uncomfortable with interpersonal intimacy and dependency, to have an excessive need for self-reliance, and a reluctance to self-disclose.

Offspring of a parent with a neurogenetic disorder were found to have experienced more parental dysfunction in childhood, and showed higher levels of adult attachment anxiety and poorer mental health, compared to the reference group. Offspring of a parent with HBOC had experienced more parental loss in childhood and showed poorer mental health, compared to the reference group. In both clinical groups, a higher number of adverse childhood experiences was associated with poorer mental health and more psychological symptomatology in adulthood. A higher level of attachment anxiety was found in persons who were younger than 16 when their parent started having symptoms of the genetic disease, compared to persons who experienced their parent's disease later in life.

This study indicates that adult offspring of a parent with genetic disease may have a more insecure attachment style and poorer mental health than persons without such a background, and that this may be related to adverse childhood experiences.

In **Chapter 5**, a study is presented that investigates whether adult attachment style and strategies for emotion regulation are related to distress, in 98 adults at 50% risk for a neurogenetic disorder (HD, CADASIL, HCHWA-D) who presented for predictive testing. Two dimensions of attachment style, i.e., attachment anxiety and attachment avoidance, were assessed with the Experiences in Close Relationships – Revised (ECR-R).⁷ Three emotion regulation strategies known to be maladaptive in stressful circumstances were assessed with the Cognitive Emotion Regulation Questionnaire (CERQ): self-blame, rumination (i.e. excessive thinking about the feelings or thoughts associated with a negative event), and catastrophizing (i.e., explicitly emphasizing the terror of an experience).¹¹ Distress before and after predictive testing was measured with the Brief Symptom Inventory (BSI).^{9,10}

The level of attachment anxiety was found to be associated with being distressed before the predictive test. Persons who had a tendency to catastrophize were more distressed before testing. The level of attachment anxiety predicted the level of distress up to two months after testing.

The findings suggest that it may be useful for counselors involved in predictive testing programs to look for signs of attachment anxiety and catastrophizing in their counselees, to identify persons who may be vulnerable for distress during and after testing. Persons with such signs of vulnerability might benefit from additional psychological counseling, in which a sense of security and the use of more adequate emotion regulation strategies should be promoted.

In **Chapter 6**, it is explained how the findings from the described studies may be applied in genetic counseling and psychological care in procedures of predictive testing for a neurogenetic disorder.

Three cases from clinical practice describe persons with varying levels of attachment anxiety and attachment avoidance. Adopting an attachment point of view in counseling for predictive testing is advocated, with an open eye for counselees' childhood experiences with a parent who had a neurogenetic disorder and the effect of these experiences for later psychological make-up. It is explained how attachment insecurity may be recognized in persons who present for predictive testing for HD, CADASIL, or HCHWA-D, and how such an attachment style may be related to emotional and psychological problems.

Persons with a high level of attachment anxiety may need additional psychological counseling before and after predictive testing, because of their tendency to worry about a lack of support of a partner or others, and the use of inadequate emotion regulation strategies. Persons with a high level of attachment avoidance may be more likely to decline psychological counseling, because of their need for independence and self-reliance.

Knowledge on risk factors for an insecure attachment style and the psychological consequences thereof can enhance counselors' understanding of the behavior and emotional reactions of counselees, and can contribute to more personalized care.

Moving forward in clinical care and research

Being born into a family with a serious neurogenetic disorder or a hereditary cancer syndrome, and growing up with a parent who is affected with such a disease, engenders a number of inevitable medical and psychological risks. The medical consequences of neurogenetic disorders or cancer syndromes are in many cases severe and life threatening. The psychological impact for affected individuals and their families is often substantial, especially when several family members are affected, in consecutive generations. There may be negative changes in family dynamics and parent-child relationships.¹²⁻¹⁷

This thesis shows the importance for clinicians to be aware that adversity in childhood, associated with parental hereditary disease, may affect the lives of offspring to a considerable extent throughout life, and that some of the risk factors for developing an insecure attachment style may be prevented. More personalized genetic counseling can be achieved when counselors understand how childhood experiences may affect a person's behavior during and

after predictive testing, and may determine important aspects of the relationship between counselor and counselee.

Striving for the prevention of parental dysfunction

This thesis shows that parental dysfunction is fairly common in families with a neurogenetic disorder, in particular HD (Chapter 3 & 4). This may be explained by the psychiatric and cognitive symptoms of these disorders, and by the psychological consequences of being at risk for or being affected with such a disorder. The personality changes that often occur in HD, CADASIL, or HCHWA-D may contribute to parental dysfunction, when a parent becomes unpredictable, aggressive, or insensitive to their child's needs. Psychiatric problems in a parent affected with a neurogenetic disorder may lead to insensitivity and irresponsive behavior towards children. Psychiatric problems are highly prevalent in HD patients, and may occur well before neurological diagnosis of HD and the onset of motor symptoms.² Psychiatric symptoms occur in all stages of HD, and comprise apathy, depression, irritability/aggression, obsessive/compulsive behaviors, and sometimes psychosis.² Depression and other psychiatric symptoms are probably undertreated in HD patients.² Clinical experience suggests that psychiatric symptoms like depression or apathy often occur in CADASIL patients years before clinical diagnosis, and are probably insufficiently treated. Cognitive problems in parents with a neurogenetic disorder may also negatively affect parent-child interactions, when parents are unable to comprehend what their child needs and how to respond in ways that help the child feel secure. HD, CADASIL, and HCHWA-D entail many unfavorable changes in family dynamics, even before the onset of the disease. Parents may fear the becoming symptomatic when they are aware of being at risk, and they may feel guilty for possibly having transmitted the disease causing gene mutation to their children.

The study results described in this thesis suggest that parental dysfunction is partly preventable with timely and adequate psychological or psychiatric treatment. Such treatment may improve a parent's ability for sensitive and responsive interactions with their children. Parents who are at risk for HD, CADASIL, or HCHWA-D, who are verified gene mutation carriers, or who are affected with these disorders should have full access to psychological and psychiatric services, to diminish the risk of adverse childhood experiences and insecure attachment in offspring where possible. Clinicians working with persons at risk for or diagnosed with HD, CADASIL or HCHWA-D, in any setting, should look for signs of psychiatric problems in their patients, and refer them for psychiatric treatment if needed. Support groups for persons at risk and for patients may allow parents to learn from others in similar situations and to develop constructive ways of coping. Furthermore, psychological interventions for parents affected with a neurogenetic disorder should be developed, aimed at enhancing parental sensitivity and responsivity and at

the prevention of parental dysfunction. In the absence of such specialized interventions, access to more generic interventions with a similar aim should be facilitated.

The psychological consequences of CADASIL and HCHWA-D require further study, preferably from a family perspective. The study samples of CADASIL and HCHWA-D described in this thesis are small, and larger studies on individuals at risk for these disorders and their families should be conducted to understand the impact of these disorders on individuals, couples, and families. Dysfunctional parenting could be measured using the Measure of Parenting Style (MOPS),¹⁸ in order to capture aspects of the parent (parental indifference and over-control, parental abuse) that may contribute to the development of an insecure attachment style in offspring.

Striving for the prevention of insecure attachment styles

This thesis indicates that there is an increased risk for insecure attachment in persons who grew up with a parent affected with a neurogenetic disorder (Chapter 4). The evidence is strongest for offspring of a parent with HD (Chapter 2 & 4).

In this thesis, attachment styles were assessed in adulthood. Further studies are needed to investigate attachment styles in children currently growing up in families with HD, CADASIL, or HCHWA-D, to see how parent-child interactions and adverse childhood experiences affect children's attachment style formation. Knowledge on relationships between early experiences, attachment styles in childhood and adult attachment styles suggests that preventing parental dysfunction and psychiatric problems of a parent helps to prevent insecure attachment in childhood and in adulthood. Interventions aimed at the prevention of insecure attachment should ideally start as early as possible, when attachment style formation is ongoing and insecurity can be prevented.

Insecure attachment in children of a parent with HD, CADASIL, or HCHWA-D may also be prevented by promoting circumstances in which the healthy parent can be as available, sensitive, and responsive to children as possible. Spouses of affected persons should have access to professional support in child rearing when desired, as well as to psychological support in coping with the consequences of hereditary disease. Support from the social environment of persons with a neurogenetic disease is also important to prevent insecure attachment in children. A personal social network may help parents cope during the disease process, and may present children with opportunities for finding additional attachment figures.

During the process of decision making on reproductive options, couples who consider having a child often reflect on possibilities to prevent transmission of the genetic disorder to the next generation. Clinical experience suggests that couples find it more difficult to reflect on the consequences of the hereditary disease for family dynamics and parenting. However,

prospective parents might benefit from psychological counseling on how a neurogenetic disorder may affect the upbringing of their child or children. Discussing risks and protective factors for adversity and insecure attachment in offspring, and possibilities for psychological and psychiatric interventions, may contribute to the prevention of problems in the next generation. Parents who are aware of these aspects before parenting problems occur may be more inclined to seek help when needed.

Based on the finding in this thesis that the risk for insecure attachment is higher in offspring who experienced their parent's disease early in life, prospective parents might want to start their family sooner rather than later. Thus, the child has the best chances of benefiting from a family life and parent-child interactions that are not negatively influenced by a parent's symptoms. Furthermore, they should try to establish and maintain supportive personal relationships with friends and relatives before onset of the disease, to increase chances for optimal support during the disease process. This may be for the benefit of patients, their partners and their children.

Striving for the prevention of psychological problems

Mental health was found to be poorer in offspring of a parent with a neurogenetic disorder and in offspring of a parent with HBOC (Chapter 4), and a relationship was found between adversity in childhood and psychological problems in adulthood. Childhood adversity and inconsistent or insensitive parenting are known to make individuals vulnerable for psychological problems throughout life.¹⁹ Attachment insecurity is associated with a lack of resilience in coping with stressful life events, and with various mental health problems, including depression, anxiety, PTSD, suicidal ideation, and personality disorders.²⁰ It is important for clinicians working with individuals from families with HD, CADASIL, HCHWA-D, or HBOC to be aware of the adversity in these persons' background, and to look for signs of insecure attachment. Persons with a history of childhood adversity and persons who have an insecure attachment style may require more care than others, to prevent psychological problems, especially in stressful circumstances.

Attachment anxiety and catastrophizing were found to be associated with distress during and after the predictive test for a neurogenetic disorder (HD, CADASIL, HCHWA-D) (Chapter 5). As illustrated in this thesis (Chapter 6), using an attachment perspective in counseling may help develop a way of counseling that can contribute to the prevention of psychological problems in persons undergoing predictive testing. The described case stories and suggestions for counselors may lead to a sensitivity for characteristics of insecure attachment. More detailed information on how to use insights from attachment theory in a clinical or therapeutic relationship can be obtained from publications on this subject.^{21,22} It is recommended to incorporate knowledge on risk factors for and consequences of insecure attachment styles in the training programs for clinicians working in clinical genetics, such as clinical geneticists, genetic counselors,

psychologists or social workers. Moreover, a specialized training program might be developed in which clinicians can learn how to recognize characteristics of an insecure attachment style in their counselees or patients.

Clinicians may try to provide an environment and a relationship in which insecurely attached individuals can build on feelings of security. In particular, a clinician's sensitivity and supportiveness may promote a sense of security and may improve counselees' mental health.²⁰ Attachment styles are relatively stable throughout life, but research suggests that attachment insecurity is subject to some change, based on favorable experiences with relationships after the initial formation of an individual's attachment style.²⁰ Therefore, building on a supportive social network throughout life should be promoted.

Interventions promoting adequate emotion regulation strategies may help prevent psychological problems. Future studies are needed to investigate whether improvements in emotion regulation reduce psychological problems in persons confronted with HD, CADASIL, HCHWA-D, or HBOC. Such studies might inform intervention programs in which persons can be assisted in learning more effective ways of regulating their emotions, and in finding new life goals when some existing goals may no longer be attained. Similar coping-effectiveness training programs have been developed and studied in other clinical populations,²³⁻²⁸ and may be useful in the populations studied in this thesis.

Couples confronted with a hereditary disorder like HD, CADASIL, HCHWA-D, or HBOC, in which one of the spouses has an insecure attachment style, may benefit from Emotionally Focused Therapy (EFT).²⁹ EFT is based on the principles of Bowlby's attachment theory and has been shown to effectively reduce distress in couples living with chronic disease.²⁹ Improving relationships may be of major importance for individuals, couples, and families confronted with a serious hereditary disorder like HD, CADASIL, HCHWA-D, or HBOC, and can be of particular benefit for children who grow up in these families.

Little is known about the psychological characteristics of persons at risk for HD, CADASIL, HCHWA-D, or HBOC who do not present for predictive testing. Future studies should investigate whether these persons differ from persons who choose to be tested, in attachment style, the use of emotion regulation strategies, and psychological well-being. One of the reasons for not wanting to know whether one will develop a serious hereditary disorder in the future may be a fear of not being able to cope with such a perspective. This thesis shows that insecure attachment styles and maladaptive emotion regulation strategies are associated with psychological vulnerability. Therefore, a higher prevalence of insecure attachment and maladaptive emotion regulation strategies might be found in persons who do not opt for predictive genetic testing.

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