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Chapter 6

Predictive testing for neurogenetic disorders: Tailored genetic counseling from an attachment point of view

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Extended version of paper 'Predictive testing for neurogenetic disorders: Tailored genetic counseling from an attachment point of view' (to be submitted).

Introduction

Counselors involved in predictive genetic testing programs for neurogenetic disorders such as Huntington's disease (HD), Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL), and Hereditary Cerebral Hemorrhage With Amyloidosis – Dutch type (HCHWA-D), frequently encounter persons who have experienced serious parental disease in their childhood.^{1,2} Insights from attachment theory may contribute to tailored psychological counseling of these persons, based on comprehension of how such a background can affect a person's development, adult psychological characteristics, and emotional reactions to the stressful process of predictive testing.^{2,3} Attachment theory describes how experiences in childhood are related to a personal style of attachment, defining an individual's social support seeking behavior throughout life. This paper aims to illustrate how results of psychological research on attachment and related concepts may be applied in genetic counseling and predictive genetic testing for neurogenetic disorders.

Neurogenetic disorders and predictive testing

HD, CADASIL, and HCHWA-D are fully penetrant autosomal dominant hereditary disorders, with onset generally in mid adulthood and death before late adulthood, for which there is currently no treatment that can alter disease onset or progression. HD is associated with progressive motor dysfunction, cognitive deterioration, and psychiatric disturbances.⁴ Psychiatric problems include apathy, depression, irritability, obsessive/compulsive behaviors, and psychotic symptoms.⁵ CADASIL is associated with migraine with aura, multiple strokes, cognitive deterioration, and psychiatric disturbances, such as apathy, depression, and changes in personality.⁶ HCHWA-D is associated with recurrent hemorrhagic strokes and dementia.⁷

Predictive genetic testing allows individuals at risk to find out whether they carry the disease causing gene mutation.⁸ The most frequently cited reasons for testing are the desire to reduce uncertainty and the need to exert control over future life, including planning a family.⁸ In the Leiden University Medical Center (LUMC, Leiden, the Netherlands), predictive testing for HD, CADASIL, and HCHWA-D is performed according to international recommendations⁹ and comprises pre-test sessions with a clinical geneticist, a psychologist, and a neurologist, consecutively. Approximately four weeks later, after another session with the clinical geneticist and, subsequently, with the psychologist, the test applicant's blood may be taken for DNA testing. Approximately four weeks thereafter, the tested person comes back to be informed on the predictive test result by the clinical geneticist, and to have post-test psychological counseling. In all stages of this procedure, additional decisional or distress counseling by the psychologist is available.

Studies worldwide have shown that most persons who opt for predictive testing for a neurogenetic disorder benefit from learning their genetic status, with few differences between carriers and non-carriers.¹⁰ The level of post-test anxiety or depression depends mainly on the level of these symptoms before the predictive test.¹⁰ Although most quantitative studies fail to find clinically elevated levels of distress after predictive testing, qualitative studies show that individuals report shock, fear, frustration and other emotional reactions after receiving test results.¹¹ Counselors in predictive testing programs regularly witness strong emotional reactions during and after testing. The measures used in quantitative studies, focusing on depression, anxiety, and distress, may not be well suited to capture the emotional reactions of persons who are tested. After two decades of clinical experience with predictive testing, there is a need to further explore concepts that may be related to emotional reactions in test candidates, and that may help explain why some individuals adjust more adequately than others. Attachment theory can provide counselors with tools that may enhance counseling in predictive testing programs, and that may help them identify test candidates who need additional support.

Psychological study on attachment and related concepts

Between January 2008 and July 2013, a study on attachment style and predictive genetic testing was conducted in the department of Clinical Genetics of the LUMC. Persons at 50% risk for HD, CADASIL, or HCHWA-D who presented for predictive testing were asked to participate. Participants' attachment style was assessed using the Experiences in Close Relationships-Revised (ECR-R),¹² a self-report scale that measures two dimensions of adult attachment, i.e., attachment anxiety and attachment avoidance. Persons with low levels of attachment anxiety and avoidance are said to have a *secure attachment style*. Persons with a secure attachment style see themselves as being able to cope with stressful situations and perceive others as supportive. They are comfortable with both autonomy and intimacy.¹³ Higher scores on one or both dimensions indicate a more *insecure attachment style*. Persons with *attachment anxiety* tend to worry about significant others not being available and responsive when needed, have an excessive need for approval from others, and fear being rejected or abandoned.¹⁴ Persons with *attachment avoidance* tend to feel uncomfortable with interpersonal intimacy and dependency, have an excessive need for self-reliance, and are reluctant to self-disclose.¹⁴

The conducted study led to sensitivity for clinical signs of attachment anxiety and attachment avoidance, as manifested in a person's verbal or non-verbal behavior during the predictive testing process. Using an attachment point of view, especially in the psychological counseling sessions, appeared to be helpful in understanding test applicants better, and offered a useful framework for counseling and interventions. Insights from attachment theory and results from the conducted study and related studies, illustrated with examples from clinical practice, may be of use for other counselors working in predictive testing.

Attachment theory

Based on interactions with parents, an individual's attachment style is formed in childhood, constituting a blueprint for social relationships in later life.¹⁵⁻¹⁸ A child will be inclined to turn to an attachment figure (mother, father, or other) when it is distressed, in an attempt to regain a sense of security. When a child experiences a parent as sensitive to its needs, and generally responding in ways that help the child feel safe, this may contribute to the development of a secure attachment style. The child learns that it may be useful to turn to others in times of need, and becomes confident that others will be available and willing to give support. The child may grow up feeling comfortable with closeness and interdependence.¹⁹ Inadequate patterns of interactions with a caregiver are associated with an insecure attachment style, in children as well as in adults.²⁰ When a father or mother is not sufficiently sensitive to the child's needs, or does not respond (by facial expressions, tone of voice, actions) in ways that help the child feel secure and protected, the child will continue to be distressed. It will not see the caregiver as a reliable source of security, and will perceive itself as being unable to cope with stressful situations.

Securely attached individuals are generally resilient in stressful circumstances, whereas insecure attachment increases the risk for maladjustment, and for pervasive social, emotional, and psychological problems, in childhood and adulthood.²¹⁻²³ In adult offspring of a parent with HD, CADASIL, or HCHWA-D, an insecure attachment style was found to be associated with poor mental health.²

Attachment theory states that early interactions with parents or other attachment figures lay the foundation for later patterns in dealing with emotions in difficult situations.²² A personal attachment style is strongly linked to the way in which a person regulates unpleasant emotions.²²

An attachment perspective on three cases

To highlight how attachment anxiety and attachment avoidance may be recognized and responded to in the process of predictive testing, three illustrative cases from clinical practice will be described (*in italics*). The case of Stella describes an individual with a secure attachment style; the cases of Anna and Robert describe persons with insecure attachment styles. Attachment style was assessed in a research context using the ECR-R,¹² and was defined in terms of the level of attachment anxiety and attachment avoidance. All personal characteristics that would allow identification of the presented individuals have been altered. Risk factors for and consequences of an insecure attachment style will be addressed. Suggestions for counselors who work in predictive testing programs will be provided.

Stella (32)

Stella presents for predictive testing for HCHWA-D. Her mother (55) had several hemorrhagic strokes over the last six years due to HCHWA-D. Stella is at 50% risk of being a carrier of the gene mutation that causes HCHWA-D.

Stella's attachment style is characterized by a low level of attachment anxiety and a low level of attachment avoidance. Her attachment style is secure.

Anna (27)

Anna is at 50% risk for HD and presents for predictive testing. Her mother died from HD 17 years ago.

Anna's attachment style is characterized by a high level of attachment anxiety, and a low level of attachment avoidance. She has an insecure, anxious attachment style.

Robert (29)

Robert presents for predictive testing for CADASIL. When Robert was five years old, his father died at age 51, after a period of deterioration and several strokes.

Robert's attachment style is characterized by a high level of attachment avoidance and a low level of attachment anxiety. He has an insecure, avoidant attachment style.

1. Childhood with parental neurogenetic disease: risk factors for insecure attachment

a. Unfavorable parent-child interactions and adverse childhood experiences

HD is known to have a negative impact on family life^{24,25} and may cause inadequate parent-child interactions. The impact and psychosocial consequences of CADASIL and HCHWA-D are considered to be comparable²⁶. More than half of offspring of a parent with HD, and approximately one third of offspring of a parent with CADASIL or HCHWA-D, have experienced their parent's disease in childhood or adolescence.² Offspring of a parent with HD, CADASIL, or HCHWA-D report relatively many adverse experiences before age 16.^{1,2} Almost 30% report parental dysfunction, defined as psychiatric problems of a parent, domestic violence, alcohol or drug abuse, and/or suicide attempt of a parent.² A substantial percentage (21%) report psychiatric problems of their parent. Parental dysfunction, especially in a context of parental mental illness, is a risk factor for insecure attachment and psychological problems in offspring.²⁷⁻³⁰ Parental loss, defined as losing a parent through death or divorce before age 16, is reported by 24% of offspring of a parent with HD, CADASIL, or HCHWA-D.² Adversity in childhood is a risk factor for various mental health problems, in childhood and in later life.³¹ Growing up with a seriously ill parent or experiencing early loss of a parent may negatively

influence attachment style formation. A parent who has psychiatric problems may not be sensitive and responsive to the child's needs. Depressed parents are less capable of taking their child's perspective and of interacting sensitively with their child.³² Psychiatric symptoms may lead to inconsistent and unpredictable parenting behavior, which is considered to contribute to the development of attachment anxiety in a child.¹⁵ Both the affected parent and the healthy parent may be preoccupied with the disease and its consequences, and may therefore display little or no response to their distressed child, which is considered to contribute to the development of attachment avoidance.¹⁵ Losing a parent in childhood may lead to an increased sense of independence and self-reliance³³, and, therefore, to attachment avoidance. Having experienced parental dysfunction or parental loss in childhood is associated with strong emotional reactions to stressors and is a risk factor for short- and long-term mental health problems.^{31,34} Offspring of a parent with HD, CADASIL, or HCHWA-D have poorer mental health than a reference group.² Having experienced a parent's neurogenetic disease in childhood is associated with adult attachment anxiety; childhood adversity such as experiencing parental dysfunction is associated with poorer mental health and more psychological symptomatology in adulthood.²

When a parent is unfit or unavailable as an attachment figure, interactions with the other parent are critical for the child's attachment style formation.³¹ A child is more likely to develop a secure attachment style when the healthy or remaining parent is able to provide consistent, sensitive and responsive parenting. However, adequate parenting may be difficult when this parent has to care for an affected spouse, or when a bereaved parent has emotional problems due to the loss of a spouse.

Stella (32), at risk for HCHWA-D, secure attachment style

Stella's childhood was not influenced by HCHWA-D and there were no major adverse events in her childhood. Stella's mother was in good health until her first stroke six years ago. Stella says her parents have always been there for her when she needed them. Stella first learned about being at risk when she was an adult. Her parents were not aware of HCHWA-D in mother's family; mother's father died at 59 from a heart attack, mother's mother is 83 and in good health.

With regard to the neurogenetic disorder, there were no risk factors for insecure attachment in Stella's childhood. Both parents were available and healthy, and Stella has grown up feeling she could rely on her parents when necessary. This will have shaped her view of others as possible sources of support and of herself as being able to cope with difficulties.

Anna (27), at risk for HD, anxious attachment style

Anna's mother was affected with HD as long as Anna can remember. After Anna's younger brother was born, mother was unfit to care for her children, due to her motor, cognitive, and psychiatric problems. Anna remembers her mother as being very irritable at times, but mostly passive and ignoring her children's needs. Mother suffered from depression, for which she was once hospitalized for months. Father had a fulltime job; many different persons took care of the children and the household. Mother was permanently hospitalized in a nursing home for HD patients when Anna was 8, and died when Anna was 10 years old.

Mother's parenting behavior, as described by Anna, was dysfunctional. Anna's mother may not have been sufficiently sensitive and responsive to Anna's needs, because of her complex HD symptomatology. Anna's father was insufficiently available to respond to Anna's needs and to help her feel secure when she was distressed. The early parental loss that Anna experienced, through her mother's hospitalization and death, has undoubtedly also negatively influenced the formation of Anna's anxious attachment style, as she lost one of her most important attachment figures.

Robert (29), at risk for CADASIL, avoidant attachment style

Robert was 5 years old when his father died. In the following years, Robert's mother developed a severe alcohol addiction. When Robert was 8 years old, his mother was unable to care for her children, and Robert and his sister (2 years older) were placed in foster care.

In a sense, Robert lost both parents in early childhood. His father was hardly able to serve as an attachment figure due to his disease process and untimely death. Robert's mother had great difficulty being responsive to her children's needs during her husband's disease process, and was almost completely lost as an attachment figure after her husband had died. The early loss of his father and the lack of responsiveness of his mother may have contributed to Robert's avoidant attachment style.

Suggestions for counselors:

- Consider the knowledge on risk factors for insecure attachment to understand counselees' presentation and behavior in the counseling process, and to estimate whether there is an increased risk for poor mental health.
- Explore the counselee's recollection of interactions with parents. When the person describes having felt insecure, unsupported, or anxious with their parent(s) in childhood, there may be a higher risk of insecure attachment.
- Explore adverse childhood experiences, especially those associated with parental dysfunction and parental loss. The risk of attachment anxiety is increased when the counselee has experienced the parent's disease in childhood. The risk of poor mental health

is increased when the person experienced parental dysfunction in childhood, or other adverse childhood experiences that may have negatively influenced attachment formation.

- When counseling parents, refer for psychiatric evaluation when psychiatric symptoms are observed; early diagnosis and adequate treatment of psychiatric symptoms in parents with a neurogenetic disorder may prevent inadequate parenting and adverse childhood experiences.

When counseling parents, refer for family interventions when children are at risk of being exposed to dysfunctional parenting.³⁵

b. Role reversal or parentification

When a parent has a neurogenetic disorder, a child may take on parental roles towards the affected parent, or may function as a substitute partner for the non-affected parent who lacks a supporting spouse. In such cases of parent-child role reversal, or parentification, the child's own need for attention, comfort, and support will be insufficiently met.³⁶ This interferes with the formation of a secure attachment style³⁷ and with the development of skills for mobilizing social support throughout life.

Anna (27), at risk for HD, anxious attachment style

Anna used to feel responsible for her mother's well-being. Even at a very young age, she was attuned to her mother's moods and needs and helped her whenever possible. Furthermore, she looked after her younger brother and performed many adult tasks, both when her mother was ill and after her mother's death.

Although it is not necessarily harmful for a child to take on some adult tasks, they should be age-appropriate and should not exceed the child's resources. In Anna's case, role reversal may have contributed to her attachment anxiety, due to the lack of attention, support, and comfort that she experienced in childhood. Caregiving for an affected parent by minors is very common in families with HD, which in many cases leads to emotional distress.³⁸

Suggestions for counselors:

- Check for signs of role reversal in the test applicant's personal history, to identify persons who are likely to have an insecure attachment style.
- Be aware that persons with an insecure attachment style have trouble mobilizing social support during and after testing and are more vulnerable for distress.

c. Intergenerational transmission of insecure attachment

Given the autosomal dominant hereditary nature of HD, CADASIL, and HCHWA-D, it is likely that the test applicant's father or mother also grew up with a parent affected with this disorder. The parent is therefore more likely to have an insecure attachment style. Parents' attachment styles

(secure, anxious, avoidant) positively correlate with their children's attachment styles, through a process of intergenerational transmission.³⁹ Parents with an insecure attachment style may be less able to help their child develop a sense of security, and the child may develop an insecure attachment style. In families with HD, CADASIL, or HCHWA-D, the risk for consecutive generations of developing an insecure attachment style is increased.

Anna (27), at risk for HD, anxious attachment style

Anna's grandfather also died from HD, after a long disease process. Anna knows that her mother had a difficult childhood herself, with domestic violence and a father who was severely irritable and aggressive.

Anna's mother may have had an insecure attachment style due to a lack of security in childhood. She may not have learned adaptive ways of managing distress, making it difficult for her to be sensitive and responsive to the emotional needs of her offspring. For Anna, the risk of developing an insecure attachment style was therefore not only increased by her mother's disease and psychiatric problems, but also by her mother's family background. When Anna would have children herself, they would have an elevated risk of developing an insecure attachment style as well, even when Anna would not be affected with HD.

Suggestion for counselors:

- Adopt an intergenerational perspective to gain awareness on how neurogenetic disorders may influence psychological characteristics of consecutive generations.

d. Social isolation

When one or both parents are insufficiently available or adequate as attachment figures, it is important that a child has additional or substitute attachment figures to rely on and get support from in stressful circumstances. Other persons than parents may thus help the child feel secure. Chances of finding additional attachment figures may be reduced for children in HD families, due to social isolation, which is relatively common in families with HD.⁴⁰ This may be similar in families where a parent has CADASIL or HCHWA-D. Children may feel ashamed of their parent's behavior and of the situation at home, and may be reluctant to invite friends at their home, or they may devote their time to helping their parents rather than to social interactions with peers. This may reduce the child's possibilities of learning how to engage in social interactions and how to maintain relationships.

Stella (32), at risk for HCHWA-D, secure attachment style

Stella's parents were very active in their community and had a large social network. Stella had a special relationship with her aunt (mother's sister), with whom she often stayed during school

holidays. This aunt has undergone predictive testing and does not carry the disease causing gene mutation. They discussed the test several times. Stella feels supported by this aunt, who was very close to her mother before and during mother's disease process.

The strong social network may have helped all family members to feel supported when needed. Stella's aunt has been an additional attachment figure for Stella and still serves as a support person for her in adulthood. The familial disorder provides a special bond between them, as they have both experienced the disease process of Stella's mother and the risk of being a carrier themselves.

Anna (27), at risk for HD, anxious attachment style

Anna was ashamed of her mother's behavior and never invited any friends. She went home immediately after school to look after her brother and to help her mother. The family was socially isolated. Anna describes how her parents have lost their friends when her mother became progressively affected. She is very afraid that this may happen to her too when she would become affected with HD.

Due to the family's social isolation, there were no substitute attachment figures that Anna could rely on. Anna had limited opportunities to practice social skills with peers and she has not learned to see others as reliable sources of support. She has experienced that friends may be present in good times, but may disappear when circumstances become difficult. This may have contributed to her fear of rejection and abandonment, which is one of the characteristics of attachment anxiety.

Robert (29), at risk for CADASIL, avoidant attachment style

As a child, Robert sometimes found shelter with a neighbor when there was no one at his home who could look after him. She was one of the very few people his parents socialized with. He thinks this neighbor knew what his family situation was like, although Robert never talked to her about it, as he was ashamed of his mother's drinking behavior. Recently, Robert has contacted this neighbor, hoping she could help him remember who he was as a child. Unfortunately, the neighbor hardly remembered anything about him.

Robert had very little opportunity to find substitute support figures. He never experienced what it is like to rely on others in times of distress, and has not learned the social skills necessary to appeal to others for support. This may have contributed to the development of his attachment avoidance. When others are not perceived as sources of support, individuals develop ways of coping that do not involve other people. The resulting independence may lead to feelings of loneliness and may have negative consequences for partner relationships.⁴¹

Suggestions for counselors:

- Ask about the family of origin’s social situation and about possible attachment figures other than parents to understand how a person’s attachment style may have developed.
- Be aware that past experiences with social interactions have shaped current expectations of others, including counselors. Due to a lack of practice with social situations, persons from socially isolated families may either depend strongly on counselors’ support during predictive testing, or be too independent to accept support.

2. Adulthood: Attachment style

Attachment anxiety and avoidance can be recognized in clinical practice, without formal assessment. A test candidate’s social support seeking behavior throughout the testing procedure and his/her degree of autonomy are suggestive of the levels of attachment anxiety and attachment avoidance. Persons with an insecure attachment style experience more distress during and after predictive testing,³ and may therefore require additional psychological counseling.

a. Social support seeking and autonomy

Insecure attachment styles are found more frequently in persons at 50% risk for HD, CADASIL, or HCHWA-D than in the general population.^{2,42} Whether or not a person brings a companion for predictive testing may give important information on support seeking behavior in stressful situations and, therefore, on attachment style. The predictive test guidelines and recommendations for HD⁹ (also applicable in predictive testing for CADASIL and HCHWA-D) state: “The participant should be encouraged to select a companion to accompany him/her throughout all the different stages: the pre-test, the taking of the test, the delivery of the results and the post-test stage.” (Recommendation 3). Most people who have a partner will come for predictive testing together. When there is no partner or when the partner is unable to be present, another security-providing person (relative, friend, or other) may be selected by the test applicant. Test applicants usually value the support of a companion during the process of decision making and throughout the test procedure, to help them feel secure enough to deal with arising dilemmas and emotions.

Securely attached individuals tend to seek support in stressful situations such as predictive testing, because they expect others to be accepting and responsive to their needs,¹³ which allows them to feel emotionally supported.⁴³

Insecurely attached individuals may be more inclined to present for testing without a companion. Persons with attachment anxiety will worry about other persons' availability and responsiveness and they fear being rejected or abandoned. This is reflected in thoughts like "my partner does not really love me, won't care about me as much as I care about him/her", "my partner will not want to stay with me, will abandon me", or "my partner may become interested in someone else".¹² Persons with attachment avoidance feel uncomfortable with interpersonal intimacy and dependency. They do not expect or need support from others in stressful circumstances, and see themselves as being able to cope with difficulties alone. They do not usually discuss their concerns and problems with others²² and may be inclined not to share their feelings about testing with their partner or anyone else. Attachment avoidance is reflected in thoughts like "I prefer not to be too close to romantic partners", "I find it difficult to allow myself to depend on a partner", or "I prefer not to show a partner how I feel deep down".¹²

Stella (32), at risk for HCHWA-D, secure attachment style

Stella is accompanied by her husband David throughout the test. Stella and David are planning on starting a family and they wish to prevent transmission of the genetic predisposition for HCHWA-D to the next generation.

Stella has thought about testing ever since she learned about HCHWA-D and her 50% risk of being a carrier. She and David have talked about it several times. During counseling, Stella is capable of clarifying her motivations for testing and of exploring different scenarios of her life after testing. She is confident that either test result (favorable or unfavorable) will help her and David plan their future. Stella discussed the test with her sisters (28, 27), and although her sisters suggested they go for testing together, Stella feels that it is important she decides independently about testing.

A secure attachment style provides a secure base for decision making and for confronting difficult situations.^{16,44} Securely attached individuals like Stella will generally be able to discuss their reasons for testing clearly and concisely, and are not overwhelmed by their emotions when thinking about a future in which they will develop symptoms of a neurogenetic disorder. Stella trusts she will be able to cope with the perspective of future disease, with the support of David, their relatives and friends.

Persons with a secure attachment style, although seeking and valuing support of others, are generally autonomous enough to make independent decisions,⁴⁵ and to feel they will be able to cope with stressful situations, like undergoing predictive testing.

Anna (27), at risk for HD, anxious attachment style

Anna has a boyfriend, Ben, but she comes to the appointment alone. She is reluctant to discuss the test and its consequences with Ben, fearing he will abandon her in case of an unfavorable result.

Anna considered predictive testing two years ago, but did not proceed with it at the time. Her father had expressed a fear that she would not be able to cope and had advised her not to take the test.

During the first psychological counseling session, Anna is overwhelmed by her emotions. She describes considerable instability in many domains of her life and cannot decide about testing. Additional psychological counseling is offered.

Due to a lack of confidence in their partner's availability and responsiveness and a fear of rejection or abandonment, persons with attachment anxiety are likely to worry excessively about their partner's reaction to the test and to receiving test results. They may therefore prefer to come for testing unaccompanied.

Anna needs others to be able to cope with distress, and she would not take the test without her father's support. Autonomous decision making on important matters like predictive testing may be difficult for persons with attachment anxiety, as they perceive themselves as being unable to cope with difficulties and are dependent on others' support.

Anna (27), at risk for HD, anxious attachment style

In a series of additional psychological counseling sessions, Anna describes a pattern of difficult relationships. She is afraid of being rejected and is easily hurt. The relationship with Ben has been unstable from the beginning and has actually ended three months ago. She cannot accept that the relationship is over and contacts Ben regularly.

Anna's social interaction patterns are explored, in relationship to her earlier attachment experiences. She is encouraged to seek opportunities for social interactions; her anxiety in these situations is discussed and she gradually gets more confidence in herself and in others.

Anna's clinging behavior and lack of autonomy towards Ben are signs of attachment anxiety. In stressful circumstances, persons with attachment anxiety tend to seek proximity of a partner, friend or relative by expressing high amounts of distress.²² They feel that the only way to get affection or support is to show extremely emotional behavior. When the other person is insufficiently supportive, feelings of insecurity and anxiety may result, leading to even higher amounts of distress.

Robert (29), at risk for CADASIL, avoidant attachment style

Robert comes to the appointment alone. He has not informed his partner Louise (25) about his intention to undergo predictive testing. He considers the test to be his own decision, and does not need her support. Louise has talked about starting a family, but Robert has decided he will never have any children.

Robert did not inform his sister (31) either. He has never been close to her and feels no need to talk with her about the test. He says it may be possible his sister already had the test herself, without informing him. As Robert was reluctant to contact his family members before and during the predictive test, he was unable to provide any details on the family history regarding CADASIL. Robert says he has no friends.

Robert's tendency to decide about testing independently and without informing anyone is a sign of attachment avoidance. He relies mainly on himself and seeks no support from others. Attachment avoidance is associated with seeing a partner as untrustworthy and unsupportive.⁴¹ Individuals with attachment avoidance may have a very limited number of social relationships. They may not perceive this as problematic, because they feel autonomous enough to deal with difficulties without the support of other people. Robert's behavior may be difficult for Louise if she has a desire to share thoughts and feelings on important aspects of life. Partners of persons with an avoidant attachment style (who are not avoidant themselves) may seek additional psychological counseling to discuss the consequences of a predictive test. The presence of the avoidant spouse is preferred during such sessions, to enhance communication and mutual support seeking. In some cases, the partner may wish to come alone.

Suggestions for counselors:

- Be aware of test candidates' support seeking behavior to understand how they cope with the emotional consequences of testing and receiving test results.
- If a person presents for predictive testing without a companion, other than for practical reasons, explore the reasons for this.
- Be aware that a lack of autonomy (in decision making, in dealing with distress) may be a sign of attachment anxiety, whereas being overly autonomous (not needing others during or after testing) may indicate attachment avoidance.
- Offer additional psychological counseling to partners of persons with an avoidant attachment style.

b. Dealing with predictive testing

In emotional and stressful circumstances, such as being involved in predictive testing for a neurogenetic disorder and waiting for test results, attachment styles will be activated.¹⁹ During

the process of predictive testing, experiences with parental disease will often be remembered and discussed, which may contribute to the activation of a person's attachment style. Securely attached individuals feel secure about receiving support as needed, based on a stable sense of self-esteem and self-efficacy.¹⁹ They will seek adequate social support before and after testing and are generally capable of regulating their emotions to the extent that they feel able to cope with the distress of testing and to adapt to the test result. Securely attached persons will be cooperative in genetic counseling and receptive to any suggested psychological interventions. Undergoing predictive testing and coping with test results may be more difficult for persons with an insecure attachment style, who often struggle with support seeking, are more likely to interpret and evaluate events negatively,⁴⁶ and will find it harder to regulate their emotions. Persons with attachment anxiety experience and express intense and strongly fluctuating emotions, whereas persons with attachment avoidance experience and express little emotionality.¹⁹ Adult attachment anxiety was found to be associated with distress during and after predictive testing.³

Stella (32), at risk for HCHWA-D, secure attachment style

Stella opts for predictive testing because she feels the need to know whether there is a risk of transmitting the gene mutation to the next generation. To reduce nervousness in the time before receiving results, she schedules some extra sports and social activities.

Stella is confident that she and David will be able to cope with any result. They lead a satisfying life and trust they will continue to do so after an unfavorable result. They feel supported by their friends and by Stella's aunt who underwent testing as well.

Stella's trust in herself and others allows her to adequately deal with the distress of testing. The strategy she uses to deal with the emotions of waiting for test results is an example of positive refocusing, i.e., thinking about other, pleasant matters instead of the event in question. Adaptive emotion regulation strategies are associated with secure attachment.²²

Anna (27), at risk for HD, anxious attachment style

Anna feels that the uncertainty about her genetic status leads to ongoing distress and blocks her in her decisions. Talking about a future with HD upsets her; she fears becoming as lonely as her mother. Anna's motivations for predictive testing are further explored, as well as her expectations on how the results may affect her life. After some emotional sessions, she is able to talk more freely about what may lie ahead and gradually feels less distressed. Eventually, she feels she would benefit from both a favorable and an unfavorable test result and she takes the predictive test.

Anna experiences distress from not knowing her genetic status, but also fears what will happen once she does know her test result. She sees herself as unable to deal with negative emotions,

and has an excessive need for approval and support from others. However, based on her past attachment experiences, she worries that no one will respond to her needs. This combination of a lack of trust in herself and in others contributes to her strong emotional reactions and makes her vulnerable for distress.

Individuals with attachment anxiety may need and accept additional psychological counseling and support.⁴⁷

Robert (29), at risk for CADASIL, avoidant attachment style

Robert is a student in mechanical engineering and a part-time computer repairer. He has been unable to concentrate on his study lately, and minimizes contact with his fellow-students. He postpones starting an internship.

Robert is afraid of developing dementia at a young age. He feels obsessed with testing his cognitive abilities and spends several hours a day playing competitive games on the internet. Losing a game makes him think his deterioration has already started, and provokes a state of panic. His partner Louise does not know how he has been feeling lately.

When asked to reflect on the consequences of predictive testing, Robert becomes restless and gives short answers. Eventually, he expresses considerable doubt as to whether the test result will be of any benefit for him. A new appointment is made to further discuss the test, against the background of his psychological situation.

A personal history with unavailable or irresponsible attachment figures may be reflected in a tendency not to confide in others or to expect others to be supportive in stressful situations. Individuals with attachment avoidance will try to deal with distress themselves, rather than expressing their feelings and sharing their experiences. They may be inclined to avoid social situations, especially when feeling distressed. In genetic counseling, persons with an avoidant attachment style may be short in their responses to questions and may not be willing or able to reflect on their thoughts and feelings.

Robert (29), at risk for CADASIL, avoidant attachment style

At the beginning of the next appointment, Robert says he will not proceed with predictive testing. He has noticed that talking about his personal situation and reflecting on the possibility of a future with CADASIL leads to an increase of intrusive and negative thoughts.

Robert expresses considerable pessimism for his future. He thinks he will not be able to get a degree and find a job, because of his tendency to avoid social situations.

Additional psychological counseling is offered, but Robert does not want to “take up someone else’s time”.

Robert is referred to the psychiatrist who is part of the multidisciplinary team, for psychiatric evaluation and diagnostics, but does not show up at the appointment. His GP is informed; she will contact Robert to discuss his situation.

Persons with an avoidant attachment style may be reluctant to accept psychological counseling and support,⁴⁷ and may not be willing to discuss the implications of testing, or to express their thoughts and feelings. In situations that are stressful for most people, like predictive testing, they may not experience distress, or may suppress their feelings. Avoidant persons may not be inclined to provide much detail on past experiences, which makes it harder for counselors to understand the influence of the hereditary disorder on their lives. It may take more effort for counselors to understand an avoidant person's motivations for testing and to estimate how receiving test results may affect them.

Suggestions for counselors:

- Be aware of how early adverse experiences and a personal attachment style may influence a person's social interactions patterns, to enhance understanding of a counselee's behavior during and after predictive testing.
- Allow more time for counselees with attachment anxiety to feel secure enough to explore and express their feelings.
- Interpret strong emotional reactions and a tendency to show clinging behavior towards a counselor as possible signs of attachment anxiety.
- Accept that counselees with attachment avoidance may have limited ability or willingness to reflect on themselves or their situation.
- Be aware that avoidant individuals may be reluctant to mention their psychological problems, or to seek adequate treatment. If there are indications of psychological or psychiatric problems, offer additional psychological counseling or refer to other mental health care providers before proceeding with testing.

c. Post-test adaptation and counseling

The result of the predictive test – favorable or unfavorable – does not predict a person's distress after the test.⁴⁸ Non-carriers as well as carriers may find it difficult to adapt to the new situation. Since insecure attachment is associated with inadequate emotion regulation and with personal and social difficulties,⁴⁹ dealing with test results will be more difficult for persons with an insecure attachment style.³

Stella (32), at risk for HCHWA-D, secure attachment style

Stella receives an unfavorable test result. In the post-test psychological counseling session, Stella and David express their feelings of sadness and seek comfort with each other. They are

able to discuss their new situation coherently, and to reflect on how to proceed today and the next couple of days. They have decided to go for a walk along the beach together, and will inform some relatives and friends after that.

Two weeks later, Stella and David describe how they felt in the days after hearing the test result. They have felt sad, but they experience a strong bond between them, and feel supported by relatives and friends. Stella and David would like to talk about ways to start a family without transmitting the gene mutation. Their thoughts and feelings on all available reproductive options are explored.

Three weeks later again, they feel quite well and are able to accept their new situation. They will opt for a natural pregnancy and prenatal diagnosis to determine whether the fetus has the genetic predisposition for HCHWA-D. They are invited to come back when they need more psychological support. Seven months later, Stella informs the psychologist that she is pregnant, and that a prenatal test showed that the gene mutation was absent in the unborn child. She and David are doing well.

Securely attached persons like Stella may show an emotional reaction upon hearing test results, but, based on their confidence in themselves and others, will generally adapt well.

Anna (27), at risk for HD, anxious attachment style

Anna receives a favorable test result. She feels relief, but also sorrow and confusion. She has always feared she would develop HD, and finds it difficult to adjust to her new status. Anna has two post-test counseling sessions to explore her mixed feelings, after which she says she feels better. Three months later, Anna comes back for additional psychological counseling, wondering if it is normal to feel so distressed after a favorable result. She often thinks about the negative influence HD has had on her life, both in childhood and during the years in which she feared she would develop HD. She feels others cannot understand what she has experienced, and she is upset when someone says she has to be happy with the favorable result. Her psychological situation has not improved.

Anna is vulnerable for distress and mental health problems after the emotional event of predictive testing, which may be based on her childhood with inadequate parenting and parental loss,³⁴ and her insecure attachment style. Anna's tendency to ruminate (i.e., excessive thinking about the feelings or thoughts associated with a negative event) and to catastrophize (i.e., explicitly emphasizing how terrible an experience has been) are examples of maladaptive emotion regulation strategies, which insecurely attached persons are more inclined to deploy. Maladaptive emotion regulation strategies such as rumination and catastrophizing predict emotional problems and psychopathology after stressful events.⁵⁰ Therefore, these strategies increase the risk of maladjustment after predictive testing.

Anna (27), proven non-carrier of the HD mutation, anxious attachment style

Anna finds it beneficial to talk about the impact of HD on her past and present. More adaptive ways of emotion regulation are explored. She understands that being at risk for HD has been part of her identity, and she is invited to explore how her identity has changed now that she is no longer at risk for HD. She shows several emotional outbursts, after which she feels ashamed as well as relieved. She gradually experiences that expressing her feelings does not lead to rejection.

For the last of four sessions, she asks her father to come with her. She feels supported by his presence, and is happy that he agrees to talk about their shared experiences with HD.

Anna is now able to focus on other aspects of her life and has decided to apply for a new job.

It may take more time for persons with an insecure attachment style to adapt to their new status after predictive testing. They may need additional psychological counseling, regardless of the nature of the test result, and may require help in successfully regulating their emotions. To establish a fruitful working relationship with insecurely attached individuals, counselors should be responsive to the special characteristics of these persons, and to their clinical needs.⁵¹

Suggestions for counselors:

- Use insights on relationships between attachment style, emotion regulation and psychological problems to understand why some persons have difficulty adjusting to their test result.
- Attempt to improve counsees' emotion regulation where necessary, by exploring possibilities for adaptive strategies such as positive refocusing.

Discussion & conclusions

Adverse childhood experiences, like dysfunctional parenting or parental loss, may be seen as risk factors for an insecure attachment style. Such experiences are more likely to have occurred in persons who grew up with a parent who had HD, CADASIL, or HCHWA-D. An insecure attachment style is a risk factor for emotional and psychological problems during and after predictive testing. Individuals with a *secure attachment style* will, in general, seek support in their own social network, and will be able to adequately cope with the test and its consequences. Persons with *attachment anxiety* may worry about the support and responsiveness of their partner and other persons, may experience high amounts of distress, and tend to ruminate and catastrophize. They may need additional counseling to be able to cope with predictive testing and test results. Persons with *attachment avoidance* may not need others' support, and may not experience distress or acknowledge being distressed during and after testing. They may not feel they need psychological counseling.

Adopting an attachment perspective may contribute to tailored genetic counseling of individuals and couples confronted with HD, CADASIL, or HCHWA-D, in a context of predictive testing. Counselors may explore childhood experiences, adult attachment style, and emotion regulation strategies of counselees to understand their behavior and psychological reactions during and after the predictive testing process.

References

1. Van der Meer LB, Van Duijn E, Wolterbeek R, Tibben A. Adverse childhood experiences of persons at risk for Huntington's disease or BRCA1/2 hereditary breast/ovarian cancer. *Clin Genet* 2012; **81**(1):18-23.
2. Van der Meer L, Van Duijn E, Wolterbeek R, Tibben A. Offspring of a parent with genetic disease: Childhood experiences and adult psychological characteristics. *Health Psychol* 2014; **33**(12):1445-53.
3. Van der Meer L, Van Duijn E, Giltay E, Tibben A. Do attachment style and emotion regulation strategies indicate distress in predictive testing? *J Genet Couns* 2015 Feb 3. [Epub ahead of print]
4. Ross CA, Tabrizi SJ. Huntington's disease: from molecular pathogenesis to clinical treatment. *Lancet Neurol* 2011; **10**:83-98.
5. Van Duijn E, Craufurd D, Hubers AA, Giltay EJ, Bonelli R, Rickards H, et al. Neuropsychiatric symptoms in a European Huntington's disease cohort (REGISTRY). *J Neurol Neurosurg Psychiatry* 2014; **85**(12):1411-8.
6. Lesnik Oberstein SAJ, Boon EMJ, Terwindt GM. CADASIL [GeneReviews™, Internet, 28-6-2012]. <http://www.ncbi.nlm.nih.gov/books/NBK1500/>
7. Maat-Schieman M, Roos R, Van Duinen S. Hereditary cerebral hemorrhage with amyloidosis-Dutch type. *Neuropathology* 2005, **25**:288-97.
8. Tibben A. Predictive testing for Huntington's disease. *Brain Res Bull* 2007; **72**(2-3):165-71.
9. MacLeod R, Tibben A, Frontali M, Evers-Kiebooms G, Jones A, Martinez-Descales A et al. Recommendations for the predictive genetic test in Huntington's disease. *Clin Genet* 2013; **83**:221-31.
10. Paulsen JS, Nance M, Kim JI, Carlozzi NE, Panegyres PK, Erwin C, et al. A review of quality of life after predictive testing for and earlier identification of neurodegenerative diseases. *Prog Neurobiol* 2013; **110**:2-28.
11. Crozier S, Robertson N, Dale M. The Psychological Impact of Predictive Genetic Testing for Huntington's Disease: A Systematic Review of the Literature. *J Genet Couns* 2015; **24**(1):29-39.
12. Fraley RC, Waller NG, Brennan KA. An item response theory analysis of self-report measures of adult attachment. *J Pers Soc Psychol* 2000; **78**:350-65.
13. Bartholomew K, Horowitz LM. Attachment styles among young adults: A test of a four-category model. *J Pers Soc Psychol* 1991 Aug; **61**(2):226-44.
14. Brennan KA, Clark CL, Shaver PR (1998). Self-report measurement of adult attachment: An integrative overview. In JA Simpson, WS Rholes, JA Simpson, WS Rholes (Eds.), *Attachment theory and close relationships* (pp. 46-76). New York, NY US: Guilford Press.
15. Ainsworth MDS, Blehar MC, Waters E, Wall S. *Patterns of attachment: A psychological study of the strange situation*. Hillsdale, NJ: Erlbaum; 1978.
16. Bowlby J. *Attachment and loss. Vol II: Separation: anxiety and anger*. London: Hogarth Press; 1973.
17. Fraley RC, Shaver PR. Adult romantic attachment: Theoretical developments, emerging controversies, and unanswered questions. *Review of General Psychology* 2000; **4**:132-54.
18. Mikulincer M, Shaver PR. The Attachment Behavioral System in Adulthood: Activation, Psychodynamics, and Interpersonal Processes. In MP Zanna (Ed.), *Advances in experimental social psychology, Vol. 35* (pp. 53-152). San Diego, CA US: Elsevier Academic Press; 2003.
19. Pietromonaco P, Feldman Barrett L, Powers SI. Adult attachment theory and affective reactivity and regulation. In: Snyder DK, Simpson JA, Hughes JN (Eds.), *Emotion regulation in couples and families: pathways to dysfunction and health*. 1st ed. Washington: American Psychological Association; 2006. p. 57-74.
20. De Wolff MS, van IJzendoorn MH. Sensitivity and attachment: a meta-analysis on parental antecedents of infant attachment. *Child Dev* 1997; **68**(4):571-91.
21. Fries AB, Ziegler TE, Kurian JR, Jacoris S, Pollak SD. Early experience in humans is associated with changes in neuropeptides critical for regulating social behavior. *Proc Natl Acad Sci USA* 2005; **102**(47):17237-40.

22. Mikulincer M, Shaver PR, Pereg D. Attachment Theory and Affect Regulation: The Dynamics, Development, and Cognitive Consequences of Attachment-Related Strategies. *Motiv Emotion* 2003; **27**(2):77-102.
23. Sroufe LA. Attachment and development: a prospective, longitudinal study from birth to adulthood. *Attach Hum Dev* 2005; **7**:349-367.
24. Forrest Keenan K, Miedzybrodzka Z, van Teijlingen E, McKee L, Simpson SA. Young people's experiences of growing up in a family affected by Huntington's disease. *Clin Genet* 2007; **71**(2):120-129.
25. Vamos M, Hambridge J, Edwards M, Conaghan J. The impact of Huntington's disease on family life. *Psychosomatics* 2007; **48**(5):400-4.
26. Rolland JS, Williams JK. Toward a biopsychosocial model for 21st-century genetics. *Fam Process* 2005; **44**(1):3-24.
27. Atkinson L, Paglia A, Coolbear J, Niccols A, Parker KC, Guger S. Attachment security: a meta-analysis of maternal mental health correlates. *Clin Psychol Rev* 2000; **20**(8):1019-40.
28. Byng-Hall J. The significance of children fulfilling parental roles: Implications for family therapy. *J Fam Ther* 2008; **30**(2):147-62.
29. Perris C, Andersson P. Experiences of parental rearing and patterns of attachment in adulthood. *Clin Psychol Psychother* 2000; **7**:279-88.
30. Wan MW, Green J. The impact of maternal psychopathology on child-mother attachment. *Arch Womens Ment Health* 2009; **12**(3):123-34.
31. Luecken LJ, Roubinov DS. Pathways to lifespan health following childhood parental death. *Soc Personal Psychol Compass* 2012; **6**(3):243-57.
32. Trapolini T, Ungerer JA, McMahon CA. Maternal depression: relations with maternal caregiving representations and emotional availability during the preschool years. *Attach Hum Dev* 2008; **10**(1):73-90.
33. Maier EH, Lachman ME. Consequences of early parental loss and separation for health and well-being in midlife. *Int J Behav Dev* 2000; **24**(2):183-9.
34. Hagan MJ, Roubinov DS, Gress-Smith J, Luecken LJ, Sandler IN, Wolchik S. Positive parenting during childhood moderates the impact of recent negative events on cortisol activity in parentally bereaved youth. *Psychopharmacology* 2011; **214**(1):231-8.
35. Cicchetti D, Rogosch FA, Toth SL. Fostering secure attachment in infants in maltreating families through preventive interventions. *Dev Psychopathol* 2006; **18**(3):623-49.
36. Chase ND. An overview of theory, research, and societal issues. In: Chase ND (Ed.), *Burdened children*. New York: Guilford; 1999. p. 3-33.
37. Byng-Hall J. Relieving parentified children's burdens in families with insecure attachment patterns. *Fam Process* 2002; **41**(3):375-88.
38. Williams JK, Skirton H, Paulsen JS, Tripp-Reimer T, Jarmon L, McGonigal KM, et al. The emotional experiences of family carers in Huntington disease. *J Adv Nurs* 2009; **65**(4):789-98.
39. Soponaru C, Dirtu MC. Antheus' effect in the intergenerational transmission of attachment styles and its importance for adult education. *Procedia - Soc Behav Sci* 2014; **142**(0):564-9.
40. Carlozzi NE, Tulsy DS. Identification of health-related quality of life (HRQOL) issues relevant to individuals with Huntington disease. *J Health Psychol* 2013; **18**(2):212-25.
41. Karantzas GC, Feeney JA, Goncalves CV, McCabe MP. Towards an integrative attachment-based model of relationship functioning. *Br J Psychol* 2014; **105**(3):413-34.
42. Van der Meer L, Timman R, Trijsburg W, Duisterhof M, Erdman R, Van Elderen T, Tibben A. Attachment in families with Huntington's disease: A paradigm in clinical genetics. *Patient Educ Couns* 2006; **63**(1-2):246-54.
43. Florian V, Mikulincer M, Bucholtz I. Effects of adult attachment style on the perception and search for social support. *J Psychol* 1995; **129**(6):665-76.

44. Bowlby J. *A secure base: parent-child attachment and healthy human development*. New York: Basic Books; 1988.
45. Van Petegem S, Beyers W, Brenning K, Vansteenkiste M. Exploring the association between insecure attachment styles and adolescent autonomy in family decision making: a differentiated approach. *J Youth Adolesc* 2013; **42**(12):1837-46.
46. Pielage SB. *Adult attachment and psychosocial functioning*. Faculty of Behavioural and Social Sciences, University of Groningen; 2006.
47. Dozier M, Bates BC. Attachment state of mind and the treatment relationship. In: Atkinson L, Goldberg S (Eds.), *Attachment issues in psychopathology and intervention*. Mahwah: Lawrence Erlbaum Associates, Inc.; 2004. p. 167-80.
48. DudokdeWit AC, Tibben A, Duivenvoorden HJ, Niermeijer MF, Passchier J. Predicting adaptation to presymptomatic DNA testing for late onset disorders: who will experience distress? Rotterdam Leiden Genetics Workgroup. *J Med Genet* 1998; **35**(9):745-54.
49. Gross JJ, Richards JM, John OP. Emotion regulation in everyday life. In: Snyder DK, Simpson JA, Hughes JN (Eds.), *Emotion regulation in couples and families: Pathways to dysfunction and health*. 1st ed. Washington: American Psychological Association; 2006. p. 13-35.
50. Garnefski N, Kraaij V. Cognitive coping and psychological adjustment in different types of stressful life events. *Individ Differ Res* 2009; **7**:168-81.
51. Bernecker SL, Levy KN, Ellison WD. A meta-analysis of the relation between patient adult attachment style and the working alliance. *Psychother Res* 2014; **24**(1):12-24.

