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ABSTRACT

This speech offers a brief description of Huntington's Disease (HD): its causes, symptoms, and incidence. It then concentrates on the psychological problems of persons one of whose parents had the disease, and the role of the counselor in helping these humans cope with their fears about contacting it themselves. A relatively detailed case study is presented of a 22-year old women whose mother had HD. The paper concludes with the following therapeutic suggestions: (1) listen to the anxieties, worries, doubts, and other problems of at-risk individuals; (2) without minimizing the gravity of their concern, offer realistic hope; (3) offer support by explaining the availability of new facilities to reduce suffering in the event the disease occurs; and (4) relieve any guilt that at-risk clients or those already suffering with HD might have toward their parents or/and children. (SE)

The Counselor and Genetic Disease: Huntington's Disease as a Model

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There are some people for whom the developmental sequences do not unfold smoothly. These are the people whose biology determines that they must struggle with crises alien to those appropriate to their developmental stage. For some the path of development has been distorted at birth due to genetic illness.

Huntington's Disease (HD) is an autosomal dominant disorder of the central nervous system usually manifesting itself in the third or fourth decade of life with progressive, uncontrollable movements, intellectual deterioration, and emotional disturbance. Each child of a parent affected by the disorder therefore has a 50 percent probability of developing the disease. HD has complete penetrance, so that if one possesses the gene for it, one invariably develops the disorder. There is no adequate treatment currently available, nor is there any test which can determine whether a child of an affected parent is carrying the gene and will develop the disorder. Diagnosis must await the manifestation of symptoms. Since the disease does not usually become evident before middle life, most victims have already had children, possibly passing on the deleterious game. Some choose to forego procreation without knowing if, in fact, they will become ill.

Huntington's Disease is a relatively rare disorder. Its prevalence ranges from 4 to 7 per 100,000. However, there are over 1,000 genetic neurological illnesses alone. Hundreds of thousands of individuals live with the spectre of inherited death or debilitation. Some have specific probabilities with which to contend, while others suffer from the more diffuse anxieties of "heart disease or cancer in the family." Whether or not a client comes for help specifically in coping with a medical problem, or whether a health concern merely forms the backdrop for other difficulties, it behooves us as counselors to recognize the signs and symptoms of that concern. The state of one's health at any point along the developmental life cycle can radically alter the normal sequence of development.

In 1973 I interviewed in depth (approximately four to six hours per subject) thirty-five Ss who were at genetic risk for developing HD. The Ss selected were all between the ages of twenty and thirty-six; none of them had been diagnosed with HD or any other neurological illness.

The Ss were questioned regarding their introduction to and understanding of HD, their family backgrounds, and their own subjective experiences of being at-risk. I was particularly interested in the predominant coping strategies of these people and in understanding how their feelings and fantasies about the illness and their "risk" status affected their daily lives. It was concluded that the presence of HD in the immediate family had a profound impact on the lives of these Ss. It was also felt that the Ss would have benefited tremendously from some brief counseling which could help them to cope with separating the reality of their situation from fantasy. Although some of the Ss had received genetic counseling, the counselors had focused on providing them with the facts of genetic inheritance, rather than helping them to work through the psychological meanings of the various choices which were open to them. Ss were often flatly advised not to have children without any recognition or discussion on the counselor's part of the more subtle implications of immortality or the promise of normalcy which childbearing can have.

Rather than abstracting the predominant themes of crisis and resolution which appeared repeatedly in the interviews, the fantasies and defenses, I prefer to let the Ss speak for themselves. The following is an edited interview with a young woman at-risk for Huntington's Disease who portrays in graphic and moving detail the concerns of many in similar circumstances. Several specific counseling suggestions will conclude the paper.

Ms. S: White middle class woman of twenty-two. Single. Secondary school teacher.

My mother is 44 now, I'm 22 and my sister is 14. I do know that, like around the time my sister was born it started. I don't know if it started before or not. I was eight. I was aware that she [mother] had to go to these doctor visits all the time and later on I found that she had been going to a psychiatrist. I don't know if the fights had really started then or it was more that she just felt inadequate...

Q: Did you wonder if there was something the matter with her?

A: Yeah. Around when I was 12 or so she started getting into a lot of car accidents, like hitting fenders and stuff. I just sort of remember vaguely before my father had really come to grips with admitting that she had it. I remember seeing the nervous hands and stuttering and stuff. I sort of had to come to the conclusion that she had it before he verbalized it.

Q: Your father knew about HD when he married your mother?

A: Yeah, he knew. He tells me that he talked it over with his relatives and they all said, "She's such a wonderful girl, go ahead and marry her." He also says he just didn't think it would really happen ... He kept hoping to get a different diagnosis.

I think (mother) was really gentle before. Then she started getting bitchy with my sister. She couldn't tolerate a normal child's antics. She'd start screaming and getting upset. And then, also, she and my father would get in fights in public, and she'd start crying or screaming or hitting him. That really upset me a lot. The stuff in private I could handle, but the stuff in public, that was just a disgrace.

Ever since all this started happening, I never really felt very good about having friends over, having people sleep overnight. I really feel like I've been deprived. (Laugh) ... I feel embarrassed. It's embarrassing to go out in public with her, she staggers so much.

I guess, uh, the thing I feel bad about on my part, the part I really dread, is having boyfriends see her, 'cause I am very open about what it is and the fact that its genetic and everything. And I think, wow, if they see how bad it is, it's just gonna be, "Forget it, baby." So, in the relationships where I haven't been really close to the guy, I just haven't had the nerve to bring him home. And my father gets really angry. "Why don't we see any of these guys that you're dating?" I sort of told him why, but I don't tell my mother.

I was about 14 when it hit me, hey, it's genetic, I know I have a chance and that was when it just hit me like a ton of rocks, you know. That was a whole new thing to go through. Just the fact that your mother's got a disease, big deal, a lot of people have sick parents (nervous laugh) ... I have never really gotten that upset or worried about the fact that, hey, I'm gonna have to go through this whole thing of going through the disease. Like sometimes it upsets me when someone at one of these HD meetings talked about having one of their relatives tied down to the bed, that was pretty vivid for me to take. Other than that, it's not being terrified about having it sometime way in the future, like at the age of 35, it's always even when I was in high school, the thought of, oh, I'll never get married and, oh, I'll never be able to, like, it's never bothered me too much the idea of not being able to bear my own children, like I wouldn't mind adopting, but the thing of no one will marry me and the whole thing of being an old maid type of thing. It's really scared me more than just thinking about having the disease.

Q: Do you feel that your father regretted the decision that he made?

A: Yeah. And I think it's really hard for me because there's been so much turmoil in the family and I've seen how awful it's been for him and how awful it's been for her and stuff. One time I went to this youth meeting for at-risks or whatever and ... it was really amazing to me to see that some of these people had gotten married...But on that score, too, maybe I'm even (sigh) decreasing my chances even more of getting married because I've made up my mind not to have kids as long as there is a genetic risk, 'cause I really feel strongly about that. Why perpetuate the disease?

Also, I know that I'd always feel guilty if I had kids. I'd always worry a whole lot if they were gonna get it and if I got it I'd really!!! feel sorry that I had them and stuff. And I don't want to have to go through that whole trip. Consequently, like I've met a lot of guys who think, "Oh well, so what about the disease," just like my father and want to have kids. They can't realize it could happen. Plus, like one guy I was really serious about said everybody's gotta die some time. But this guy wanted to have kids anyway. And you know, me having made the decision, no, I'm not going to, a lot of guys really have a trip about wanting to have kids. So, you know, that's kind of a bad thing.

I went with this guy for one and one-half years and he knew that my mother had it and that it was genetic and stuff and we never talked that much about the future, you know, and I was getting really serious about him and I started asking him more pointed questions. I asked him, I said that, "I'm not telling you that I want to get married now, I'm not telling you you have to marry me, but I want to know if you'd ever consider marrying me," and at that point he said, "No. As long as you're at-risk I'd never consider marrying you." And I felt, really like, wow, you never told me that before. And so, at that point, also because the relationship was pretty screwed, like he'd get violent and he'd throw tantrums and stuff, he was really immature, I decided, wow, this is nowhere for two reasons. So, I broke things off and then he started trying to get me back by saying, well, "If I said I'd marry you would you come back to me," and stuff. I thought it was just a ploy, plus I wasn't interested anymore ... And then finally after we decided we were gonna break up and stuff, I started talking to him more about the whole thing and this is what really got me. He said when he'd found out ... well, I should explain something to you. At that point we had both been virgins. And then we got involved -- sexually as well and he was twenty-three and had never had a sexual relationship with a girl, the reason being he had never wanted to get committed to a girl, and he thought that getting involved sexually was going to commit himself. He said that the reason he'd gotten involved with me was because he knew he'd never marry me so he knew he wouldn't be trapped. And I felt like, WOW, this is some game we've been playing all around and I haven't known the rules. And so, I've always really resented him. I just think he's such a schmuck!!

I have thought that it would have been better if I hadn't been born. You know, I have thought about that. Like, those fools, why did they ever decide to have children. That was a real egotistical, selfish thing to do, type of thing. But now that I'm here, I'm going to make the best of it.

One time my mother said something about, "You're so unsympathetic. Just wait until you have it someday." And, wow! I really laid into her for that. What a natty thing to say, especially in front of my little sister. That really hurt. I'm really bitter at her for that. But, you know, you have to kinda make allowances for her when she's in that condition.

One time I was going with a med student and I was really scared to tell him. I thought that he would know the manifestations so much more. And I guess I thought that if anyone knew the manifestations they wouldn't want to get involved. A lot of people that hear about it without having seen it so vividly, I don't think they worry about it as much ... I guess the thing I worry about, too, is oh my God, should I ever turn into what she's turned into, a bitch, an alienated vegetable, that really is scary, you know. And I guess I really have a lot of drive to get everything done before I'm 35, type of thing. Make my mark on the world, or whatever.

Q: Do you think that maybe you turn away some of the men who are interested in you?

A: Well, I have had this feeling. When I meet somebody who's super intelligent, super good-looking -- this guy should have kids; this guy should propagate his kind. This guy wouldn't want to adopt. I do think about that. And having the guy smarter than me is an absolute requirement and that's something that I'm not willing to compromise. But having the guy not too good looking is one thing I even search for. Number one, because then he won't feel like, oh, I'm so good looking I've got to have a whole bunch of me's running all over the world. And also, I feel like I'd be able to hold him easier, be able to trap him easier. Which is kind of a whole bad syndrome that I'm in ... I think, wow, since I'm at-risk, I'm less attractive as a possible mate, so I've got to compromise somewhere, give up something of my desires for a mate. So, I'm not willing to give up personality or strength or the intellectual capacity, so the one thing that's left is looks. So I'm sort of hunting for some ugly guy (laugh).

I think the worst part is that no one will marry me and I'll have to be alone. I guess it's the fear of having HD and being alone at that point, and being abandoned by all my so-called friends. But that could happen with a spouse, too. I guess that's why having a sister is a real comfort, because I don't think she'd abandon me.

But I think really, the thing that really scares me the most is I know, despite all my determination to hold myself together and be a pleasant person to be around, I fear that having the disease will make me lose that control and I'll turn into a shrew like my mother. And that will be what will alienate me from people. And that will be against my control, even though I won't want to do it. Like I see she does things that she doesn't want to do, but she can't help herself. It's gonna be interesting to listen to this tape after I change!

The daydreams I used to have often, maybe I still have them sometimes, you know, the martyr thing of "Look, she's got this terrible disease btu she's handling it so well and she's so brave. Look how well she's coping and we really admire her. Rally around her," and all that garbage. But I really don't want to be a martyr, I want to express what I feel and still have people be understanding and accept me.

I don't know if I feel if I will get HD or not. It's just a big question mark. Sometimes ~~I think~~, "What if you really do have it?"

Then I go, "Aaaahh." Sometimes I think, "What if you don't have it, I mean right now you just really don't have it and you're worrying about all this for nothing," and I think, wow, that's just an entirely different feeling too. I think the state I'm in now, of not knowing, is a completely different state from either knowing or not knowing. And like with this experiment, where they took the skin biopsy, it's really scarey, I feel really weird towards this scar. It's like an A for Adultery on your chest or something. When this guy asked me what it was, I just didn't tell him. First I evaded the question. The second time he asked me, I was kind of playful about it, I said, "Well, it's a scar." He says, "A scar from what?" I said, "A scar from a wound." "What kind of wound?" "A flesh wound." And then he said, "How'd you get it?" And I said, "Self-inflicted." And he just didn't ask anymore ... It's kind of an ugly thing. When it was healing it was kind of a hard thing for me psychologically. (?) Yeah, it was like a manifestation of the disease, or something. When they first talked about having it done I didn't know it was going to be so big and have such a big scar. They said it was like a little slit, I mean, a hole punch like that is a pretty weird scar to have. You know, it doesn't look too normal.

What I'd say is that it's sort of put a pall over my whole life. I feel like it's kind of like a veil, a darkness overshadowing it. Because always in the back of my mind, is the thought, "Oh, am I gonna have it. Oh, am I ever gonna get married. Oh, am I gonna be rejected because of it?" The fear of having it, the fear of being alienated. And like, when I was going with this guy J, he kept talking about the fact that until he was 26 the draft was just a pall over him. And I thought, well, that's what HD is to me; it's always in the back of your mind. It's always there to lurch out. If I have any other problem usually HD will surface with it because it's always in the back of my mind type of thing. I don't think I try to repress it or completely put it out of my mind, like I wrote a lot of poetry and I think about it and I think I've done an awful lot of growing about it. But I don't just want to be depressed all the time. It is sort of like a terminal cancer patient, knowing: run for your life. I think I'm running to sort of cram as much as I can into 15 years or something.

Well, I remember being in Junior High and thinking to myself, I don't know whether this was before my parents started having the fights or not, because I remember thinking how happy I was, how perfectly happy I was.

* * *

Ms. S. is a strikingly attractive young woman and quite intelligent. She walks with unusual grace and has a quality in her movements and in her speech of a restive energy barely arrested and suppressed. Just below the surface of her rather exaggerated outward calm, she is volatile and sad. She speaks with the "pseudo-toughness" and bravado of adolescence but her general demeanor and understanding is far more mature and quite insightful. Ms. S's father has suffered two psychotic episodes since her mother's diagnosis of HD. During one episode he threatened to rape Ms. S and kill several other persons. Ms. S is extremely tied to her family, despite her verbally blase attitude, and has tried until very recently to be the mainstay and mediator of the household. She visits home frequently and talks to her mother on the telephone when away. In recent years she has come to realize the necessity for separating from her family to preserve her own sanity, but feels extremely guilty over individuating and "abandoning them."

Ms. S. often speaks as if she definitely will get HD but the real affectual force of her concern is toward getting married. Even though she is only 22 years of age and lives in an environment where marriage is increasingly less emphasized, Ms. S has a desperate need to "catch a man," "to trap him" and "to hold him." She is most bothered by the physical manifestations of HD and feels them to be repulsive and embarrassing. She considers herself to be "defective" because she is at-risk.

Ms. S's fantasies about HD are very much entwined with her attitudes towards herself as a person and particularly as a woman. She seeks an ideal of a man to marry and with whom to identify. Her descriptions of men are idealistic and over-romanticized. The man becomes her "well" self, her "whole" self. However, she is afraid that a really ideal man would not be interested in her. She compromises by choosing in fantasy, an ugly man. His physical ugliness becomes an external representation of what she experiences as her hidden ugliness. With an ugly man, she feels equal; with her "ideal" man she feels humiliated and ashamed of what she considers her "defects."

Ms. S gives us some understanding of her unconscious elaborations of the meaning of HD when she speaks of her skin biopsy scar. The scar came to represent the disease, like a constant advertisement. She speaks of it as a "sore," a "slit," a "wound." It is like the red letter A for Adultery blazoned across Hester Prynne to announce her sexuality, her excesses, and her transgressions. It is also "self-inflicted." Unconsciously, the image of HD becomes enmeshed with her image of her female identity: to be a woman is to be like mother is to be defective is to be sick. The secret and ugliness of HD become a representation of the secret ugliness of her feminine sexual self. And Ms. S chose infantile and sadistic men who reinforced her in this devalued image of herself as an object to be "screwed" and not loved.

It is not surprising that fantasies of this disease which so affects the body should alter fantasies of the body image. Ms. S is terrified that should she develop HD she will literally become her mother, despite her best efforts. She will move as her mother, become thin as her mother, speak as her mother, and, worst of all, behave as her mother.

Despite the upheaval in her family, despite the fact that both her mother and her father are seriously physically and psychologically ill, Ms. S has a great deal of resilience and ego strength. She is someone who could probably profit a great deal from some short-term counseling aimed toward separating the realities of the disease from her fantasies and rather more frightening representations of it.

Psychotherapeutic Suggestions

Almost every subject in this sample could benefit from short-term or long-term counseling focused on coping with HD in the family. When at-risk persons first learn of their own risk or when they come for genetic counseling, there is often so much substantive information to be imparted that there is not enough time for discussing emotional reactions. There is also an initial shock which shields against problems which arise later. None of the subjects had had any counseling other than that provided sporadically and on a volunteer basis by the Committee to Combat Huntington's Disease. For most, the only contact they had had with a knowledgeable professional was in talking with their parents' physician. Genetic counselors, for the most part, focus on issues of procreation.

Despite all the diversity in background, education, employment and general life styles of these subjects, certain commonalities of concern existed. In response to these shared problems, the following counseling suggestions are offered. These suggestions are culled from my experience working with persons at-risk for HD but it is hoped that they will be relevant to many counseling situations in which a genetic illness is involved.

1. Listen. Many at-risk individuals find that their spouses, their immediate family, or their relatives are too involved, too frightened or too guilty to really listen. Most at-risk persons do not want to frighten their families with their concerns. They also especially do not want other family members to watch them for symptoms.

2. Do not minimize the gravity of their concern but offer

realistic hope. Because of their own difficulties in coping with the risk situation, family members often brush aside the at-risk person's concerns, scoff at them, or offer magical-omnipotent solutions. Spouses are notorious for such statements of denial as: "Don't worry, honey, I won't let you get it," or "It can't happen to us." Although optimism is a must, it can also be frightening to the at-risk person to feel that the spouse cannot afford to think that it could happen to them. It means that the disease is truly too terrible to think about. Frequently the spouse has a realistic appraisal of the situation but a conspiracy of silence regarding the disease grows between the couple because each does not want to frighten the other. A counselor can be extremely helpful in guiding the individual or the couple toward a realistic appraisal of the disorder, acknowledging the reality of their concerns, giving hope, and thereby demonstrating that the illness can be reasonably discussed without anybody coming to grief.

3. "Don't worry about it, you could step off the curb and get hit by a truck." True, but not truly helpful. Most at-risks are concerned about the process of dying, not with death itself. It is more valuable to stress the quality of life, both in health and sickness. In this case getting HD is only a 50 percent risk, but many at-risks make themselves 100 percent miserable worrying while they are healthy. Much of their apprehension concerns the treatment they will receive should they become ill. Many have retained childhood visions of a "lunatic" parent, strapped in bed, with no medication. The counselor should emphasize the new drugs

which are now available, new health care insurance which is pending, better nursing facilities, increased awareness of HD in the medical community, and the efficacy of physical and psychological therapy in staving off some of the most frightening symptomatology. The slow development of the disorder is a boon in the sense that medical science is experimenting with new projects daily and the brain is capable of a certain amount of restitution even after some damage is suffered. If the state of being ill is seen as less frightening, anxiety will decrease. Remind the individual that a 50/50 probability means as great a chance that they will not get HD as that they will. Odds also decrease after the forties.

4. "Symptom Searching." Every at-risk individual is continually on the alert for any suspicious signs of the disorder. Even if they deny that they check themselves if asked directly, many will give examples of such self-diagnoses in the course of general conversation. Every time an at-risk person trips, stumbles, mumbles, falls, forgets, has a car accident, gets enraged, gets divorced, etc. the spectre of HD is aroused for themselves and for others. Many are so hyper-alert that they make themselves uncoordinated, frightening themselves even more. There are at-risk individuals who practice walking on lines, walking on curbs, controlling their handwriting, controlling their speech, touching their fingers to their noses, and even rehearsing Serial Sevens! There are others who practice how it would be to have HD. Occasionally they frighten themselves by not knowing when the practice stops

and the real thing begins. They try to master the disease through activity in the same way that people who very much fear "going crazy" play "being crazy." Some at-risks will imitate mannerisms of the affected parent as an identification with that parent. It can be extremely reassuring to explain that all at-risks "symptom-seek" and that most feel convinced that they will develop the disease. The therapist should teach the individual about psychological defenses and how they may be operating, as well as about normal muscular tics and twitches such as myoclonic bursts and normal psychological lapses, including especially normal forgetfulness.

5. Differentiate HD from the rest of the environment. Freud once said that if you cordon off one portion of a city and tell the police they cannot enter, you can be sure where to find all the criminals. The prospect of having HD can feed into every conflict; and each problem can be interpreted in terms of HD, rendering it relatively hopeless in the eyes of the individual. For some it may be easier to lay the blame with HD rather than face vulnerabilities, failures, weaknesses, which have nothing to do with the disease. Work with the individual to differentiate realistic concerns regarding the illness from fantasied concerns and from conflicts which are unrelated. Most problems stem from the usually disrupted environments in which these people have been raised.

6. Relieve guilt. The counselor should be attuned to any expressions of conscious or unconscious guilt on the part of the client. Often the guilt is pervasive and extends both toward the parents and

and toward the children. Guilt may be over anger toward and neglect of the parent, over envy of an obviously well sibling, over the desire to bear one's own children, etc. In particular, the counselor should try to make explicit the common belief that good or bad behavior will have an influencing effect on whether or not the individual develops the disease. Often the environment conspires with this belief: a twelve-year-old at-risk girl was told by a police matron that she had "better behave or she would get what her mother had."

7. "Unnecessary and never sufficient." Just as prediction is usually foremost in the minds of the at-risk individual, it is often foremost in the mind of the counselor: will this person develop HD?

In the opinion of this investigator, most HD patients do not get diagnosed until approximately three to ten years or more after the initial manifestations of the disorder. This does not mean that they are unaware of the disease prior to the diagnosis. A well-trained observer may be able to detect subtle neurological, cognitive, and psychological cues long before the HD patient feels it necessary (or is pushed) to be diagnosed. What have been thought in the past to be socio-psychological indicators of neurological pathology may be, in fact, indicative of a psychological reaction to a perceived change in performance, but should never be taken as sufficient indications of the disease in and of themselves. Extreme pain, anger, and/or an ill-advised decision to have children can result from an inaccurate prediction of future events and counselors should avoid speculations, even if they are optimistic. If, on the other hand, the counselor feels convinced that the individual is not manifesting any signs of the illness <u>at that moment in time</u>, it can be very encouraging for the client to hear this opinion. Above all, the counselor should be empathic and respond to the concerns of the client as they are expressed.