Journal of Genetic Counseling, Vol. 2, No. 3, 1993

Offering Predictive Testing for Huntington Disease in a Medical Genetics Clinic: Practical Applications

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Predictive testing for Huntington disease is presently offered in a select few medical genetics centers in the United States. This is in part due to the labor intensive counseling and psychological testing suggested by the research protocols. We discuss some specific suggestions for establishing programs for Huntington disease predictive testing within pre-existing medical genetics clinics to encourage more centers to offer presymptomatic testing. This will allow more at risk individuals the opportunity to consider predictive testing and cut down the expenses of traveling to the few predictive testing centers that currently exist. The counseling principals will remain similar to those discussed here, even following the identification of the Huntington disease mutation.

KEY WORDS: genetic counseling; Huntington disease; molecular diagnosis; predictive testing.

INTRODUCTION

Programs offering presymptomatic testing for Huntington disease (HD) have been available since approximately 1985 using the technology of linked DNA markers (Gusella *et al.*, 1983). Initially, as part of research protocols, testing was offered at no charge and involved both extensive counseling and neuropsychological testing (Brandt *et al.*, 1989; Crauford *et al.*, 1989; Folstein, 1989). In the last few years, presymptomatic testing for Huntington disease has moved from research to genetic counseling centers. Multiple medical centers across the United States now offer predictive testing and counseling for HD (Quaid, 1992). However, many

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1059-7700/93/0900-0123$07.00/1

clinical genetic centers have been wary of offering this service because of the extensive time commitment required if the research protocols for counseling and testing are followed. We believe counseling for presymptomatic testing for HD can be offered in a conscientious and cost effective manner within the structure of pre-existing medical genetics clinics. We began a predictive testing program for HD in 1989 using the regular staff of our medical genetics clinic with consultation from a psychologist within our University system. Our protocol was patterned after the previous research protocols and guidelines developed by the Pacific Northwest Regional Genetics Group Huntington's Disease Diagnosis and Counseling Task Force, the Huntington's Disease Society of America and the World Federation of Neurology (Bennett, 1989; Brandt *et al.*, 1989; Huntington's Disease Society of America, 1989; Went, 1990; World Federation of Neurology, 1990). Based on our recent experience we present a practical approach to use in general medical genetics clinics for offering predictive testing for HD.

The concept for this paper was formulated before the discovery of the Huntington disease mutation (Huntington's Disease Collaborative Research Group, 1973). Although the counseling protocols will certainly be modified, counseling will and should remain an essential component of Huntington disease predictive testing programs. Direct mutational analysis will be cheaper than linkage analysis and the client seeking predictive testing will have more privacy with regards to their choice to pursue predictive testing since DNA samples will not be required from multiple family members. The reduced costs and ease in access to the testing will likely increase the demand for services related to Huntington disease predictive testing and increase the need for more testing centers.

ESTABLISHING A PREDICTIVE TESTING PROGRAM

Personnel

We suggest the minimum support team include a board certified/eligible genetic counselor (or similarly qualified individual), a medical geneticist and access to consultation from a neurologist, and psychologist familiar with HD, particularly the early manifestations of the disease. Expertise from a panel of laboratory, medical and ethical experts for consultation when psychosocial, ethical, and legal problems arise is also helpful. The genetic support team should decide if a team member will be on call to handle program participants in crisis or whether clients will be instructed to contact their existing therapist or the emergency room.

Our clinical staff includes a board certified medical geneticist and neurologist, a board certified genetic counselor and a doctoral clinical psychologist. After each client's visit, the case is anonymously discussed from the laboratory, medical, counseling, and ethical standpoints as part of our regularly scheduled medical genetics clinic conference.

Costs

The program staff should decide if costs will cover the entire counseling program or if charges will be made for each visit. If the DNA diagnostic laboratory is not on site then the staff should determine if they will charge a fee for coordinating and sending blood samples. Programs which have one fee for counseling, psychological, and neurological testing may have a better client return rate, particularly for follow-up visits after the result session, since additional visits are at no added expense. A problem with a comprehensive fee is that clients may feel committed to testing since they have made a financial investment which may affect their decision making.

Acceptance of Clients

Our eligibility requirements are that the person be at 50% risk to develop HD (25% if the at risk parent is deceased) and of legal age to be able to make an informed decision about testing. Arguments against testing children for late onset genetic disorders such as Huntington disease are discussed by Bloch and Hayden (1990), Harper and Clark (1990), and Quaid (1992). We discourage testing at our center for people who live more than 300 miles away as we feel such distances prevent us from providing adequate psychological support in pre- and post-testing periods. If the client has a local professional counselor we have offered testing to individuals who live several hundred miles away assuming they agree to attend the sessions required. We also encourage postponement of testing for individuals who are in psychological crisis (e.g., divorce, substance abuse).

Client Support

Each consultant is required to have a support person, such as a friend, relative, partner, or spouse, accompany them to each visit. It is best that this support person not be an at risk family member such as a sibling since he or she may not be able to provide subjective support (Quaid, 1992). The client should be encouraged to establish a relationship with a local professional support person, preferably a psychologist or psychiatrist who

can be available on an ongoing basis. Support in the form of a clergy member, family therapist, or social worker is also encouraged. Many clients have reached their decision to pursue predictive testing as a result of such a therapeutic relationship. Permission is obtained from the client to provide general information about Huntington disease to the professional and personal support persons.

COUNSELING PROTOCOL

Number of Counseling Visits

In our experience, four visits seems to be the minimum for a person given a decreased risk and five for a person given an increased risk. Although this may seem extensive within the constraints of a regular genetic counseling clinic, this is about the average number of visits required when providing counseling and molecular diagnostic linkage analysis for other complex genetic disorders such as polycystic kidney disease and neurofibromatosis.

Structure of Visits and Counseling

Pre-Disclosure Visits

We recommend two to three visits before the result session. In our experience it has been helpful to keep the structure of the visits flexible and to cater the discussions to the needs of the individual client. Clients who enter the program have varying degrees of prior knowledge about Huntington disease and linkage analysis as well as different levels of insight as to their anticipated adjustment to their test results. Visits are adjusted accordingly.

The family history and pedigree are obtained by phone or during the first visit. Arrangements should be made to obtain documentation of the diagnosis (i.e., autopsy, brain scans, medical records) since confirmation of the diagnosis is critical to provide accurate predictive diagnosis. Costs for DNA testing, genetic, and psychological counseling should be discussed at this time. In our program the initial visit is with a genetic counselor who assesses the client's knowledge of HD, obtains or confirms the family history, determines suitability of the family structure for linkage analysis, discusses DNA banking, and explores the motivations for testing. In future visits the genetic counselor serves as the primary professional support person and co-counsels with the medical geneticist and psychologist. A counseling approach which challenges the client's rationale for seeking predictive testing is a useful technique although this is

counter to the usual nondirective school of thought in genetic counseling (Hayes, 1992). The counselor should explore the client's motivations for seeking testing since once the DNA results are given the client cannot turn back. Many clients have not fully examined how an increased, decreased or uninformative result will affect their partner or spouse, friends, colleagues at work, and affected and at risk family members (Hayes, 1992). Table I provides a list of questions which we find useful for the genetic counselor and other team members to discuss with the client, and Table II lists questions that are helpful to address to the support person. Quaid (1992) provides a detailed review of specific recommendations for the counseling process which can be used for clients in presymptomatic testing programs.

We use an informal checklist which the genetic counselor reviews with the client at the initial visit to assure that relevant topics have been discussed (Table III). Written information for the client is useful and the Huntington Disease Society of America is an excellent resource for materials. It may be difficult for the client to imagine fully how the predictive test results will affect his or her life. We suggest the client and support person view (at home) a videotape produced by the Canadian Predictive Testing Program entitled "Predictive testing for Huntington's disease" which consists of testimonials from four individuals who have pursued presymptomatic testing and discussion of the impact the results had on their lives (Hayden, 1991). Though our clients have expressed a positive reaction to viewing this videotape, it is an emotional experience and we suggest that it be viewed with another person at a time when discussion can occur afterward or when the program staff is available by phone. Role playing is an excellent way for clients to experience how the test results may affect their lives (Crauford, 1992; Quaid, 1992).

In our program if the client wishes to pursue predictive testing, the next visit is with the neurologist/medical geneticist, genetic counselor, and psychologist to discuss testing in more detail, review medical records and family history to document the diagnosis of HD and give a neurological examination. The likelihood of informativeness and a review of which family members must be sampled are given. The proband's *a priori* risk is determined. Often we combine the initial genetic counseling visit with the neurological evaluation particularly for clients who, during the phone interview, seem well informed about testing or for clients who believe they are symptomatic. Clients who are symptomatic and seek predictive testing may be in denial and it may be difficult for the counselor to ignore the symptoms and remain unbiased until after the neurological exam.

Table I. Discussion Questions for Clients Considering Predictive Testing for Huntington Disease

What has been your experience living with people in your family with Huntington disease?
What is the single most important reason for wanting to know if you have inherited the HD allele?
Why have you chosen to consider predictive testing at this time vs. earlier or later than now?
What decisions in your life have you made that you would have made differently if you had not been at risk for HD?
Do you believe you have inherited the HD allele? What does your spouse/partner think?
What does your family think?
What effects do you think being given an increased risk would have on your: Personal life? Significant other? Decision to have children/more children? Children if you have any? Parent(s) including your affected parent if living? Siblings who are at risk/siblings who are affected? Friends? Other relatives including those affected or at risk? Career plans/present and future? Colleagues at work? Financial planning? Medical and life insurance? Choices in how you would spend the next 10–20 years?
What about a decreased risk? What about an un-informative risk?
If the DNA testing discloses non-paternity, do you want to know this information if it does not alter your final test results?
Who will you tell that you are participating in predictive testing? Who will you tell of your results (e.g. family members, friends, colleagues at work)?
How have you coped at difficult points in your life in the past? Who do you turn to for support? How do you ask for help when you need it?
Have you ever been depressed? How did you get yourself out of depression? Have you ever considered suicide? If so, what or who helped prevent you from taking this action?
Is there anyone pressuring you to find out your status? Explain.
What will you do the day you are given your results if you are given an increased risk? A decreased risk? An uninformative result? What will you do the next week? The next month? The next year? (e.g., take the day off work, go out to dinner, stay home).

Table II. Questions to Ask Client's Support Person in Presymptomatic Testing for Huntington Disease

What is your relationship to the client? How long have you known each other?
Have you been through difficult times with this person before? Explain.
How have you provided this person with support in the past?
Do you have a pre-conceived notion about whether or not the client has inherited the HD allele?
How do you anticipate this person will react to being given an increased risk? A decreased risk? An uninformative result?
How will you provide this person with support during the process and afterward?
Is this person able to ask for your support? How do you recognize when this person needs support (e.g., they ask you, they get quiet, they get angry, etc.)?
Who will you turn to for support in this process?
How do you anticipate you will react to the client being given an increased risk? A decreased risk? An uninformative result?

The involvement of in-house psychological support personnel is crucial but the extent of involvement depends on whether such a support person is a permanent part of the medical genetics clinic staff. If this professional support is only available on a consultative basis, we recommend that the client identify a local psychological professional who is available before and after results are given.

The decision of whether to use a battery of psychological tests or rely on a clinical interview is up to the individual testing center. Our center has used clinical interviews although we have recommended formalized testing for selected clients. Other testing programs have used psychological tests such as the Beck Depression Inventory, Beck Hopelessness Scale, Symptom Checklist 90, Ways of Coping, State-Trait Anxiety Inventory and the Emotional Support System Scale (Brandt *et al.*, 1989; Crauford *et al.*, 1989; Crauford *et al.*, 1992; Evers-Kiebooms *et al.*, 1991; Folstein, 1989; Wiggins *et al.*, 1992).

Table III. Client Check-List for Presymptomatic Testing for Huntington Disease

I choose to participate in predictive testing for the presence of the Huntington disease (HD) gene. Although the gene for HD has not been found, a DNA marker (pattern) has been located near the gene on chromosome 4. In some families it is possible to trace this marker through the family to predict who has and has not inherited the Huntington disease gene. I understand it is the marker that will be tested for, not the gene itself.

I understand that my participation in this program is wholly voluntary and I can terminate at any time without jeopardy to my medical care. I also understand that if at any time my continued participation in this program is considered injurious to my health or disadvantageous to me, the testing staff may recommend postponement of the testing.

I understand that DNA marker testing is not 100% accurate. For some individuals the testing can give a result of a 99% probability of an increased or decreased risk of developing HD. For others the results will be less accurate (e.g., 84% increased risk).

I will be responsible for the costs of the laboratory testing and counseling regardless of the outcome of my testing. The total cost ranges from $2000 to over $5000 depending on the number of blood samples and visits.

I understand there can be four outcomes to the test:

1. **Uninformative.** The pattern of the marker in my family might not be interpretable. This might be because of my family structure (a limited number of relatives) or the genetic information may be such that I cannot learn anything further.
2. **Decreased risk.** I will be told that there is a reduced chance I am carrying the HD gene.
3. **Increased risk.** I will be told that there is an increased chance that I am carrying the HD gene.
4. **Non-paternity in the case of an affected father.**

If I am given an increased risk to develop HD, I am aware that the testing cannot predict when I first will show signs of HD nor can it predict the severity or rate of progression of the disorder.

Due to the nature of DNA testing, I am aware that the testing may identify non-paternity (e.g., my father might not be my biological father).

I understand that blood samples must be collected from relatives both affected and unaffected with HD. The specific samples needed will depend upon my family pedigree and will be discussed with me.

It is my responsibility, working with the staff of the testing site, to arrange for the necessary blood samples to be collected. How rapidly or slowly this collection process takes place will determine in large measure the length of the interval between my initial visit for counseling and the availability of my test results.

(Continued)

Table III. *Continued*

The results of the analysis of my relatives' blood samples will be strictly confidential. I will receive only information about my own probability of inheriting HD.

I agree to have a companion of my choice accompany me through the entire program or parts of it as agreed upon with the testing staff. Further, I am aware that I am encouraged to identify a counselor in my community with whom I can meet and discuss my decision to have predictive testing. This individual should be prepared to provide psychosocial support and ongoing counseling for me, if necessary.

I agree to participate in the minimum of four counseling sessions required for the testing. There will be at least two sessions prior to the test result session and a minimum of one session (two for individuals given an increased risk) following it. Sessions will last 1–3 hours. I understand that during this time I will receive neurological and psychological examinations and evaluations including in-depth interviews regarding my attitudes toward predictive testing, my reactions to various test outcomes including who I plan to tell of my results, my personal relationships, and other aspects of psychological functioning which have a bearing on my adjustment to the test results.

I agree to have a neurological exam. I am aware this examination may disclose that I have clinical signs of Huntington disease and I will be told of my results.

The risks of such testing are primarily of a psychological nature. A non-informative result can be frustrating and can intensify the ambiguity of the risk situation or can provide relief. A decreased risk can produce feelings of guilt as well as joy. An increased risk could lead to serious psychological consequences including feelings of depression, futility, despair, and severe stress. Counseling provided during the test is designed to help me adjust as best as possible to non-informative, positive, and negative information. Counselors will discuss with me other possible risks such as difficulties with employment or insurance. Counselors will assist me in arranging for any longer term counseling and support I may need.

Physical risks include the discomfort of a needle prick and the possibility that a black and blue mark may form as a result of blood being drawn, a mark which will fade in a few days.

Client Date Counselor Date

[a] Adapted from Huntington's Disease Society of America 1989.

A third visit is usually offered the week before the result session is given. The week before the final results is a time of high stress. Prior to this time the client has been involved with details such as obtaining blood samples and worrying about financing the testing. Once the laboratory testing has begun there can be feelings of not being able to turn back. It is important to re-emphasize that the program is voluntary and testing may be declined (Quaid, 1992).

Special Considerations

Prenatal Diagnosis

Each program should decide if they will offer prenatal testing including prenatal exclusion testing. In prenatal exclusion testing, linkage studies are done to determine if the fetus has inherited the linked markers from the affected or non-affected grandparent, but the parent's risk status is not altered. For example the result may be that the fetus is at 50% risk to have inherited the allele for Huntington disease. Because currently there is unanimous agreement that predictive testing should not be done for children, prenatal testing is not offered unless the couple agrees to terminate a pregnancy at increased risk because otherwise a child will have been tested. Such directive counseling is controversial and at odds with the traditional nondirective training of medical genetic professionals. We suggest genetic counseling for these couples include at least two visits before the results are given and a follow-up visit for those given an increased risk. Couples should be encouraged to begin the testing process pre-conceptually or very early in the pregnancy. We recommend that pregnant women at risk for HD should not find out their own risk and the fetal status simultaneously since they have the potential to have double grief if both they and the fetus are at increased risk (Crauford *et al.*, 1992).

Non-Paternity

Establishing guidelines to handle disputed paternity discovered through DNA testing is important. For example, if the at risk individual has an affected mother and the social father is found not to be the biological father the informativeness of the DNA test results may not be altered but the information of non-paternity may be devastating for the client. Also, in this scenario, would the non-paternity result be given to the adult client or to the client's mother who may be demented or even deceased? The issues of non-paternity should be addressed in the consent forms to obtain blood samples from family members as well as for the client

seeking testing. We involve the client in a discussion of how they wish to handle non-paternity results. If the client wishes to be informed, it may be helpful to give this information in person or over the phone separately from the final result session so that the client does not have the emotional issues involved with non-paternity as well as the emotions that may be raised by the predictive testing results.

Confidentiality of Results

The predictive testing team will need to decide where the final results from the DNA analysis will be placed. Will results be placed in the client's permanent record or in the personal office files of team members? Does the report contain information about other family members or just the proband? Our center places a report in the permanent medical record that contains only the client's risk information with the markers that are used but not the actual linkage data since this includes information about other family members. The linkage data remains in the team members' office files. During the initial contacts with the client it is important to discuss what type of record is kept of their testing.

Result Session

Some predictive testing programs choose to perform DNA informativeness studies before the proband's sample is analyzed (Crauford *et al.*, 1992; Quaid *et al.*, 1992). Although we discuss this option with our clients, most have chosen to have all family samples analyzed simultaneously. This reduces the number of pre-visit counseling sessions and generally decreases the time until the client is given results. Our clients also have stated that they feel being given a non-informative result would be just as traumatic as being given an increased risk result and they prefer to be told this result in person.

The date the results will be given is established well in advance so that the client will not have to experience the devastation of being told that the laboratory results are not completed on the day they were expected: If the results are available sooner than expected, the result appointment day is not changed. If the genetic counselor is the primary professional support person, he or she may choose to delay obtaining laboratory results until immediately before the result session since the ability to remain an unbiased support person may be hampered. A client may decline to know his or her results at the last moment and if the counseling

staff already has test information, unbiased counseling in the future will be virtually impossible.

The day of the results can be an acutely stressful one for the client and support person. The client is prepared in advance as to how and where the results will be given and by whom. In our program the medical geneticist gives the actual test results and the genetic counselor and psychologist are usually both present. The client is scheduled early in the day and escorted to the room quickly. The client may interpret every movement to be indicative of the test results. If a member of the genetic support staff normally escorts the client into the room, on that day it may be best to have other available staff place the client in the room since the client will try to read the face and actions of the team member.

The results are given immediately without preamble. Results are not modified with adjectives such as "good" or "bad," "high" or "low," since only the client can interpret the positive or negative impact the results will have. For example, the medical geneticist might say: "Your testing shows you have an increased risk of 98% for having inherited the gene for Huntington disease. This means there is a 2% chance of error," then explain the results and ask for the client and support person's reactions. A written laboratory report is used to make the result seem more real to the client. The result session may be relatively brief or quite extended, depending on the results and the client's reactions. No matter what the results, this session is usually very emotional for all involved.

Follow-Up Sessions

Individuals given an increased risk are contacted by phone within 24 hours. A follow-up visit is scheduled within 1–2 weeks and again within 1–3 months as well as 6–12 months. The experience of the Canadian collaborative study for HD predictive testing suggests that the greatest anxiety and depression in the increased risk group occurs in the first 2 months (Bloch *et al.*, 1992). Follow-up is either by phone contact or clinic visits. Individuals with non-informative results may need as much follow-up as those with an increased risk.

Individuals with a decreased risk are contacted by phone within a week following the result session. A follow-up appointment is made within 1–2 months and a follow-up visit or phone call again 6–12 months. The experience of the Canadian collaborative study indicates the most vulnerable time for individuals given a decreased risk is from 2–12 months after results are given (Huggins *et al.*, 1992).

COPING WITH RESULTS

In our experience and the experience of other testing centers, most clients cope well with their results (Bloch *et al.*, 1992; Brandt *et al.*, 1989; Huggins *et al.*, 1992; Wiggins *et al.*, 1992). An uninformative result is very difficult since the main reason many clients seek testing is to finally know their status to make life plans (Wiggins *et al.*, 1992). It is crucial to address this possibility and the client's anticipated reactions in the pre-result sessions.

An unexpected outcome of testing is that some individuals with a decreased risk experience depression similar to a post-partum depression (Evers-Kiebooms *et al.*, 1991; Huggins *et al.*, 1991) as well as survival guilt (Ever-Kiebooms *et al.*, 1991). People who are given a decreased risk are at higher risk for anxiety and depression if they plan on making sweeping life changes based on their test results (Huggins *et al.*, 1992). After the initial elation they may realize that the testing has not cured the problems that existed before their testing nor have they gained new abilities to make decisions or solve personal problems (Quaid, 1992; Wiggins *et al.*, 1992).

Individuals with decreased or increased risks may not believe their results. Kessler (1988) has noted preselection in some families with Huntington disease (i.e., a preconceived notion of which family members will or will not develop the disease). During the initial counseling visits it is helpful to determine in what risk group the client places himself/herself and if family members have a preconceived notion of the client's risk. If the DNA testing is discordant with the individual's or family's belief the counselor can anticipate that the client may have trouble accepting the validity of the results.

SUPPORT FOR THE PREDICTIVE TESTING TEAM

Being a member of a team providing predictive testing may be personally rewarding but also emotionally taxing. It is important for the professional staff to have time to discuss their feelings after meeting with the client, particularly after the result session. Personality conflicts may arise between a professional support staff member and the client and it is important to discuss such situations early in the testing process to resolve conflicts or choose another professional support person. Opportunities for meeting with staff from other HD predictive testing programs can provide learning experiences, maintain quality counseling and personal support.

SUMMARY

It is possible to provide predictive testing programs for Huntington disease within the structure of existing medical genetics programs. Genetic counselors play an essential role in the establishment of such programs and support for participants. Effective communication and coordination of activities between the members of the counseling team, the consultant and the support person are the most important factors in determining a successful program. As more clinical genetics centers offer predictive testing programs for Huntington disease these services will become more accessible both financially and geographically to individuals at risk.

Although the counseling protocol for Huntington disease testing will be modified with the availability of direct mutational analysis, counseling remains a crucial component of any predictive testing program. Many aspects of the model for HD predictive testing and counseling programs can be applied to the delivery of genetic services for other adult onset genetic disorders for which DNA linkage analysis is becoming an option such as familial Alzheimer's disease and familial amyotrophic lateral sclerosis.

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