

Physical Therapy Intervention for People With Huntington Disease

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Background and Purpose. The clinical symptoms of Huntington disease (HD) include progressive movement disorders, cognitive deficits, and behavioral changes, all of which affect an individual's ability to participate in activities of daily living. To date, very few quantitative or qualitative studies have been conducted to guide physical therapists working with people with HD. The objective of this study was to characterize current physical therapist practice for people with HD, thus informing the development of standardized clinical care and future research studies.

Subjects and Methods. Consultation with physical therapists working with people with HD was undertaken in the form of mailed questionnaires (n=49) and semistructured interviews (n=8). The development of the interview schedule was aided by consideration of the data obtained from the questionnaires. Themes identified from the interviews were considered in light of published literature and questionnaire responses.

Results. The main issues that emerged from the interviews were classified into 3 subthemes: (1) there is insufficient use of routine physical therapy-related outcome measures at different stages of HD, (2) there is underutilization of physical therapy services in managing HD (particularly in the early stages), and (3) the management of falls and mobility deficit progression is a key treatment aim for people with HD.

Discussion and Conclusion. A conceptual framework for physical therapy intervention in HD was developed on the basis of the themes that emerged from the data in this study. Such a framework has utility for complex, progressive conditions such as HD and may facilitate clinical decision making and standardization of practice and affect the development of future physical therapy trials.

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Huntington disease (HD) is an inherited, neurodegenerative condition resulting in clinical symptoms of progressive movement disorders, cognitive deficits, and behavioral changes that may affect an individual's ability to participate in activities of daily living, work, and community. The current management of HD is focused on symptoms because there is no curative treatment.¹ Physical therapy may play a role in assisting people with HD to maintain their independence in activities of daily living by addressing both impairments and limitations in functional activities.

Despite the potential for physical therapy to assist people with HD, evidence suggests that people with HD are not always routinely referred for physical therapy, particularly in the early stages.²⁻⁴ This lack of referral may be attributable to the limited scientific support for the effectiveness of physical therapy interventions in HD.⁵ Although animal models and small-scale studies of physical therapy in humans have led to the belief that directed physical activity, environmental enrichment, and training may be beneficial, clinical judgment, experience, and trial and error remain the major mechanisms through which interventions are justified.^{2,6-11}

A recent study¹² investigated the benefits of participation of patients with early- and middle-stage HD in an inpatient rehabilitation program. The treatment program included respiratory exercises and speech therapy, physical therapy, and occupational therapy. Patients were admitted for 3-week periods of intensive treatment that were repeated 3 times per year for 2 years. Data from the first year indicated that 25 of the initial 40 patients who started the study completed the 3 admissions during that year; only 11 patients completed all of the 6 admissions in

the 2 years. Each 3-week period of treatment resulted in highly significant ($P<.001$) improvements in motor performance, as measured by the Physical Performance Test (PPT). No motor decline, as measured by the PPT, was detected over 2 years ($n=11$). These findings give some indication that intensive rehabilitation positively influences motor and functional performance in patients with early- and middle-stage HD. The components of the provided rehabilitation that resulted in change could not be clearly determined.

The design and implementation of randomized controlled trials for people with HD are limited by a lack of knowledge of the specific content of interventions. Obtaining knowledge about the content of programs currently being implemented is an important step in providing therapists with the information to develop more standardized methods of intervention and to initiate further research that can determine the benefits of specific physical therapy interventions in a controlled manner.

The purpose of this study was to identify the specific components of physical therapy practice for people with HD. Specific research questions were: (1) What is the nature of physical therapist practice? and (2) What are the influential limiting factors relevant to the provision of physical therapy for people with HD? Because this study was focused on developing an understanding of physical therapist practice relative to HD, a qualitative methodology was considered appropriate. Qualitative research most typically involves the development of concepts that may emerge into theory¹³ and, therefore, was the approach of choice in this study.

Method

Study Design and Approach

In order to describe the nature of physical therapy practice, consultation in the form of semistructured telephone interviews with physical therapists based in specialized HD centers was undertaken. A preliminary questionnaire study was also conducted. This questionnaire data contributed to the development of the interview questions and further facilitated triangulation during the later stages of the study. From the interviews, in-depth information was gathered from participants relating to their experiences in the assessment and treatment of people with HD. Ultimately, the data were used to develop guidelines and a theoretical framework for physical therapy practice specifically relevant to people with HD.

The semistructured telephone interview, which was the primary method of data collection in this study, allowed exploration of the issues of interest with the use of open-ended questions without the constraint of predetermined categories of analysis.¹⁴ In accordance with this method, a "grounded theory" approach was used to influence data gathering and analysis.¹⁵ This has been described as an approach that "starts from the ground and works up in an inductive fashion to make sense of what people say about their experiences, and then to convert those statements into theoretical propositions."^{15(p102)}

Study Procedures

Preliminary questionnaires. Physical therapists with a special interest in neurology ($n=200$) were selected randomly from the United Kingdom Association of Chartered Physiotherapists Interested in Neurology research database to receive a mailed questionnaire. Stamped, self-addressed envelopes were provided for the return of the completed ques-

Table 1.
Participant Experience in Working With People With Huntington Disease (HD)

Participant	Place of Current Work	Specific Experience With HD
1	Neurological rehabilitation unit	Average of 1 or 2 adults with HD per year over the last 10 y.
2	Specialized HD unit	Many adults with HD and mainly psychiatric problems. Average of 15–20 people per year over the last 4 y in a private setting.
3	Specialized HD unit	Average of 11–15 people per year over the last 18 y in an HD day therapy unit and community outreach service.
4	General hospital	One case of juvenile HD.
5	Specialized HD unit	Average of 10 people per year over the last 5 y (adults and older people with HD in an HD day therapy unit).
6	General hospital	Average of 1 or 2 people per year over the last 18 y.
7	Mental health team	One case of adult HD; referral for anxiety.
8	Community physical disability team	Average of 15–20 people per year.

tionnaires. The questionnaire comprised 3 sections. The first section collected respondents' demographic details relevant to their treatment of people with HD. The second section considered referral for physical therapy and the treatments provided. Open-ended questions were used to obtain qualitative information about issues such as falls and the effects of behavior and dementia on physical therapy treatments. The third section included questions about guidelines for care and outcome measures routinely used in the treatment of people with HD.

Semistructured interviews. In order for the interviews to yield sufficient amounts of descriptive data, we used a purposive sampling strategy¹⁴ that involved recruiting physical therapists from a broad geographical area and with a range of experiences to be interviewed. Participants were specialist physical therapists who had experience working with people with HD. Table 1 provides a detailed description of the participants.

There are no established rules to aid in sample size determination in qualitative research.¹⁶ The decision as to when sufficient participants had been interviewed was related to the point at which no new themes emerged; that

is, when theoretical saturation was achieved.¹³ Eight participants were recruited for this study, and the initial analysis of transcribed data indicated that the saturation point had been achieved. Interviewing was discontinued at this stage, and further analysis of the conducted interviews confirmed that recurrent themes appeared to be emerging in all of the interviews.

The participants were recruited via advertisement on the Chartered Society of Physiotherapy discussion board. All of the telephone interviews were conducted by one researcher and audio recorded. Field notes were made by an additional researcher, who also was present during all of the interviews. Special attention was given within the interview process to reframing, repetition, and expansion of the questions as required, thus enhancing the credibility of the information that was obtained. The interviewer used a preplanned schedule (Appendix) that served as a checklist for the questions covered during the course of each interview and as a guide toward the topic areas for discussion, thereby facilitating a consistent and systematic method of data collection. The development of the interview schedule was aided by consideration of the data ob-

tained from the preliminary questionnaires. The schedule aimed to cover 3 basic topic areas during each interview: (1) assessment and treatment planning according to the stage of the condition; (2) the effects of specific issues, such as behavior and dementia problems, on the management of HD; and (3) departmental policies with respect to people with HD and, specifically, with respect to the potential for developing care guidelines.

Responses to open-ended questions were analyzed with a thematic approach. Closed questions were coded and then entered into and analyzed descriptively with the Statistical Package for the Social Sciences (version 14).*

Interview Data Analysis

The interviews were transcribed in full into text. This process of full transcription helped to strengthen the trustworthiness of the data by avoiding selective recording of the information.¹⁴ Ensuring the truth value of the study, the accuracy of the transcripts was confirmed by both the interviewer and the independent researcher as well as by consultation with the participants

* SPSS Inc, 233 S Wacker Dr, Chicago, IL 60606.

Table 2.

Summary of Questionnaire Results Relating to Referrals and Evaluation Procedures

Question	Response	Valid Percentage of Respondents Giving That Response (N=49)
Most common no. of people treated per year?	0	5.1
	1	46.2
	2	38.5
	>5	10.2
First referral?	Presymptomatic	0.0
	Early stage	8.9
	Middle stage	53.3
	Late stage	37.8
Most frequent referral reason?	Mobility, balance, falls	57.1
	Deteriorating function	20.0
	Assessment only	8.5
	Advice and support	5.7
	Moving and handling	2.9
	Respiratory	2.9
	Seating	2.6
Are falls an issue?	Yes	95.3
Is behavior a limitation?	Yes	58.5
Is motivation a limitation?	Yes	56.4
Is dementia a limitation?	Yes	74.3
Is a physical therapy guideline available?	No	95.4
What outcome measures should be used in treating people with Huntington disease?	None	44.4
	Rating scales and timed tests	35.6
	Rating scales only	11.1
	Timed tests only	6.7
	Goal setting	2.2

who were interviewed.¹⁷ Copies of the transcripts were sent, along with self-addressed, stamped return envelopes, to the interviewed participants, inviting them to comment on whether the transcripts fully represented their ideas or not. Final themes also were sent to the participants for comment.

A qualitative data analysis package (QSR Nvivo, version 2.0.163[†]) was

[†] QSR International Pty Ltd, 225-R Concord Ave, Cambridge, MA 02138.

used to aid in the analysis process for the interview data. All codes were saved within the NVivo database as nodes. As a result of highlighting areas that were unclear and performing further coding and refinement, the initial nodes were reordered, duplicated, or merged. Following identification of the themes, labeling and layering of connections between ideas and concepts were performed with the modeling option in NVivo. Final themes were agreed on by the research team and were sent to the participants for checking. The

themes were further cross-checked and triangulated with the preliminary questionnaire data as well as with information obtained from a critical review of available literature. This process allowed us to confirm the content and the results obtained from the interviews.¹⁸

Results

Preliminary Questionnaires

Of 200 questionnaires sent, 118 (59%) were returned and included in the analysis. Of the 118 respondents, 49 had treated an individual with HD during the course of their career. All of these 49 respondents had obtained their training in the United Kingdom. The respondents had an average of 15.3 years of experience (range=2.5-40). Therapists ranged in position from staff physical therapist to senior therapist/clinical specialist. Therapists practiced in a range of environments, including hospitals, outpatient settings, and specialized long-term-care HD units. The majority of the therapists treated 1 or 2 patients per annum (46.2% and 38.5% of total questionnaire responses, respectively). The therapists most frequently saw patients in the middle and late stages of the disease (47.3% of the respondents for both). Only 5.3% of the respondents saw patients in the early stage.

Table 2 shows a summary of the questionnaire results related to referral for physical therapy and evaluation procedures. Table 3 summarizes the respondents' answers about types of interventions that they used for people at different stages of HD. In the early to middle stages, the focus was generally on balance, coordination, and general physical conditioning; in the late stages, therapists reported focusing more on functional activities, equipment provision, mobility, and prevention of falls.

Semistructured Interviews

All 8 recruited participants had obtained their undergraduate training in the United Kingdom. All were senior physical therapists with varied work experience in the field of neurological rehabilitation (Tab. 1).

The questions used for the semistructured interviews are shown in the Appendix. The main issues that emerged from the interviews were classified into 3 subthemes: (1) there is insufficient use of routine physical therapy-related outcome measures at different disease stages, (2) there is underutilization of physical therapy services in managing HD (particularly in the early stages), and (3) the management of falls and mobility deficit progression is a key treatment aim for people with HD. The identification of these 3 subthemes led to the development of an overriding master theme: The management of HD must be modified according to an individual's problems and to the stage of disease (Fig. 1).

Subtheme 1: Insufficient use of physical therapy-related outcome measures that address HD-specific problems. Functional assessments were considered the most appropriate method of evaluation. Participants reported using general mobility assessments that were part of the assessment procedures at their hospital or facility. These included observational items, such as movement from a sitting position to a standing position, sitting and standing balance, bed mobility, walking, and use of stairs. The participants were in agreement that functional assessments yielded the most information for them to make appropriate clinical decisions. Participant 6 said:

I think that much more important really is to know what they can do. For example, if they can get to the toilet, get up or down stairs, make themselves a meal, etc, that is much more

Table 3.

Types of Interventions Most Frequently Used by Therapists for People at Different Stages of Huntington Disease (HD)^a

Intervention	Valid Percentage of Respondents Using That Intervention for HD at Following Stage:			
	Presymptomatic	Early	Middle	Late
No treatment	86.7	62.2	26.7	33.3
Balance, mobility, and prevention of falls	2.2			2.2
Core stability	2.2		2.2	
Provision of equipment and postural support				2.2
Physical conditioning				2.3
Problem-oriented approach and functional activities	6.7	33.3	55.5	15.6
Respiratory therapy	2.2	4.5	13.3	44.4
Patient education			2.3	

^a No entry in a cell indicates no response to that particular intervention for that stage.

important than them being able to walk 10 m outside.

However, participants did express a desire for more functionally relevant, validated outcome measures. Although several participants realized the potential for standardized tests for the purposes of obtaining information about an individual's current functional status (eg, 10-m walk test) and highlighting the nature of any impairments (eg, Berg Balance Scale, Timed "Up & Go" Test [TUG]), none of the therapists was currently using any tools as outcome measures to assess the effectiveness of specific physical therapy interventions. Participants agreed that it would be feasible to use the types of measures that are used for people with other neurological conditions. Data from the questionnaires revealed that 57% of the respondents who did report using outcome measures routinely used a combination of rating scales and timed tests (such as the Berg Balance Scale, the TUG, and the 10-m walk test).

However, it was suggested that although these measures might be used as baseline measures, they might not be good indicators for predicting the probability of the risk of falls in people with HD. Participant 2 said:

I sometimes use the Berg Balance Scale, but it is far from the ideal when you've got patients who got 25, 26 on Berg Balance Scale and are pretty much largely independently mobile with the occasional fall. Most people with other conditions and such scores would not be able to walk and have recurrent falls. So, from that point of view, it is not a good indicator except as a baseline to see how they are getting on.

None of the interviewed participants reported access to scores on the Unified Huntington's Disease Rating Scale (UHDRS), even though the test was designed specifically for HD and includes a Motor Assessment and a Functional Assessment. The UHDRS is an observer-rated and clinic-based outcome measure that may not re-

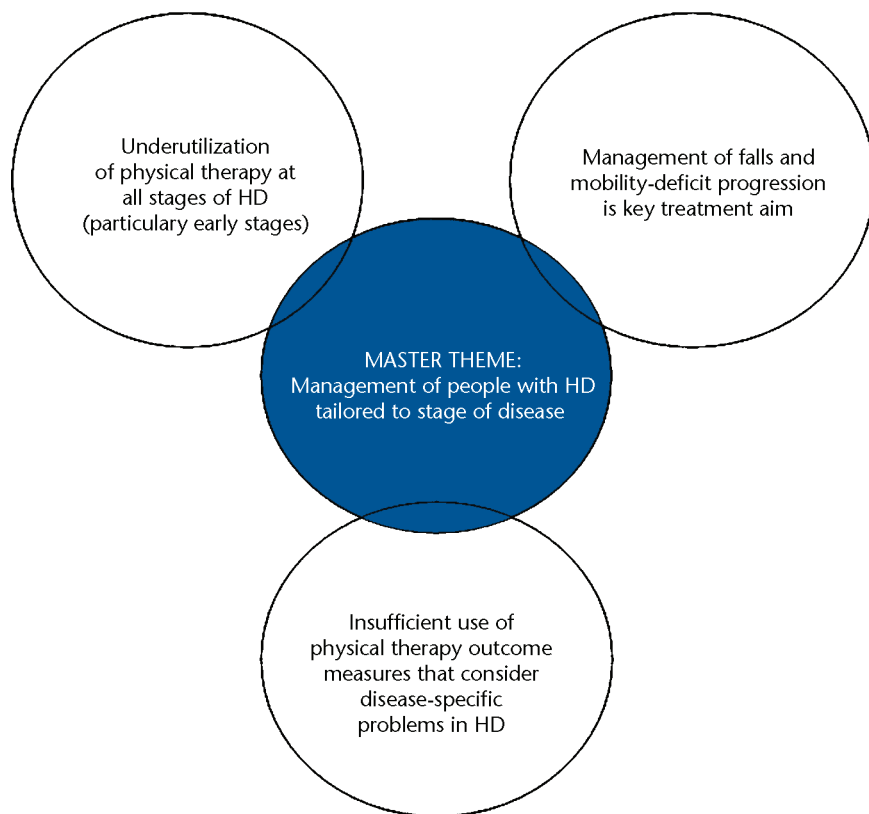


Figure 1. Representation of the 3 subthemes and the master theme for Huntington disease (HD).

flect what people are actually able to do in their own environments. Participant 6 said:

It is so difficult, because there are so many outcome measures out there. I deal a lot with multiple sclerosis (MS), and there is not one that suits everybody, and I really get upset because the clinic uses the EDSS [Expanded Disability Status Scale], which is similar to the UHDRS for HD; I found that does not mean anything. I mean there are a lot of variables. Time of day, place of assessment, people are nervous, and it does not necessarily relate to what the patient can do at home.

In summary, there was agreement that functional assessments provided the most information for therapists to make appropriate clinical decisions and to guide their interven-

tions. Although therapists recognized the need for using outcome measures, they were unsure of which tools were most appropriate to use.

Subtheme 2: Underutilization of physical therapy services in managing HD. A second subtheme that emerged from the interviews was that physical therapy services were underutilized for people with HD, particularly in the early stages. The main issues discussed were policies for the referral of people with HD for physical therapy services, the number of people referred per year, and the stages at which referral occurred. The responses regarding the number of people referred for physical therapy services differed according to the areas in which the people were referred. Interviewed partici-

pants working in nonspecialist areas such as general hospitals (participants 4 and 6) treated 1 or 2 people per year. In contrast, participants working in specialist areas such as specialized HD units (participants 2, 3, and 5) reported treating larger numbers of people (10–20) per year. Most of the interviewed participants reported that referrals occurred in the middle to late stages of the disease, although there was agreement that physical therapy would be most beneficial in the early stages.

Participant 1 was particularly concerned about the lack of early intervention and indicated that because early-stage referral was achievable for people with Parkinson disease, it also should be available for those with HD. This participant indicated that the benefits of early-stage intervention would include helping the therapist to establish a baseline, establishing a relationship with the patient and the caregivers at an early stage, and assisting the patient in maintaining mobility and functional independence for as long as possible. This participant said:

We do not see them early enough. We do not see them as soon as diagnosed in the same way as we see MS patients, very close to diagnosis. It used to be a problem with Parkinson disease, we used to see them at the very late stages, but we see them now much, much earlier, so with HD, if we see people earlier, then we can have baselines and establish relationships with people and their carers from that early stage, and then give them advice on activities to maintain mobility.

Other factors related to people with HD themselves were considered to influence the initial referral for physical therapy services. Participant 3, working in a day therapy unit and a community outreach service, commented that people in the presymptomatic stage rarely were seen in the clinic and instead were seen in the

community setting. Participant 3 felt strongly that fear of the future may act as a barrier to coming to a clinic, where they may see what the future would be like, saying:

A lot of patients that start as outreach are perhaps the children of patients who are actually coming into the unit. They may be presymptomatic, and they do not want to come to the unit, where they may see what the future holds.

Participant 6 added that a patient's refusal to confront or acknowledge the problem might be another factor, saying:

I would say that I have not seen pre. I've seen 2 or 3 patients at the early diagnosis side of it, just after they've been diagnosed. Most of those who were diagnosed ignore it unless the families were keen on our involvement.

Thus, the problem of early-stage referral may be multifaceted. Although most of the interviewed participants received referrals in the middle to late stages of the disease, the more experienced therapists felt strongly that they did not see people with HD early enough. They agreed that physical therapy would be most beneficial in the early stages of the condition.

Subtheme 3: Management of falls and mobility deficit progression in HD is a key treatment aim. All participants identified falls and risk of falls as some of the primary problems that need to be addressed in people with HD. The progressive nature of the disease results in alterations in mobility that place people in a high-risk category for falls and injuries. Accordingly, prevention of falls was suggested as an important element in a physical therapy program implemented for people with HD.

Facilitation of the development of coping strategies was one of the

methods recommended for reducing the risk of falls in people with HD. Despite the fact that people with HD in the late stages look unsafe and uncontrolled when they move, many of the participants believed that gait should not be corrected or changed. It may be preferable to allow people to develop their own coping strategies. Participant 1 said:

I have not felt that I could intervene in such a way to make a significant change in that gait, and I have been afraid that by trying to do something, that could actually interfere with the way that these individuals have developed their gait. I have not tried to modify the gait but to modify the environment and the carer.

The fact that safety and mobility may become severely compromised in the late stages of the disease was commented on by participant 1, who said:

I think for anybody observing someone at the late stage of HD with multiple choreic movements occurring during the day, it is frightening to watch, it just looks very dangerous and very uncontrolled even though the person can actually move from point A to B safely; it does look very unsafe and then we need to introduce that to the home environment where there are a lot more things to go wrong, to trip over.

With regard to what is believed to be safe performance of an activity (such as ambulation), participants believed that there were frequent disagreements between family members or caregivers and people with HD. Therapists working with people with HD should acknowledge and accept the need of these people for functional independence. It seems that this is part of living with the disease, although it may mean an increased risk of injuries from falls. Participant 3 said:

We had one patient, a very young man . . . and had great difficulties

with him when he wanted to mobilize. The unit staff were really very concerned because he was falling so much when he got out of his chair, and they were rushing to prevent him getting up, which he found frustrating. In the end, we agreed on a compromise, where he would be allowed to mobilise freely and the staff would not get too close . . . that was the way he was less likely to fall.

On the basis of the data from the interviews, walking aids for safer mobility can be prescribed for people with HD. However, their use should be individually assessed and treated with caution. It seemed that most therapists believed that walking aids such as walkers and canes were more of a hindrance. Chorea can mean that a walking aid becomes more of a hazard for people with HD. In addition, some people with HD have difficulty with persistent motor activity and are therefore likely to let go of the aid while using it. With regard to cognition-related issues, some people may have difficulties navigating the environment with the aid, an issue that may increase the risk of falls. These aspects relating to the use of walking aids for people with HD were commented on by participants 1, 6, and 8. Participant 1 said:

We will provide walking aids as they need them. We tend to find that they are resistant to using walking aids, and sometimes it is difficult. I think with choreic movement that would cause them more trouble. They can trip over them or harm themselves.

Participant 8 summarized:

Sometimes providing walking aids seems to make them worse, especially if they get festinate in their gait, it seems to hinder them. If they are falling and use side steps or step to save, then the aids get in the way. It is more trial and error with walking aids as to what they receive, when and how effective they are.

Physical Therapy in Huntington Disease

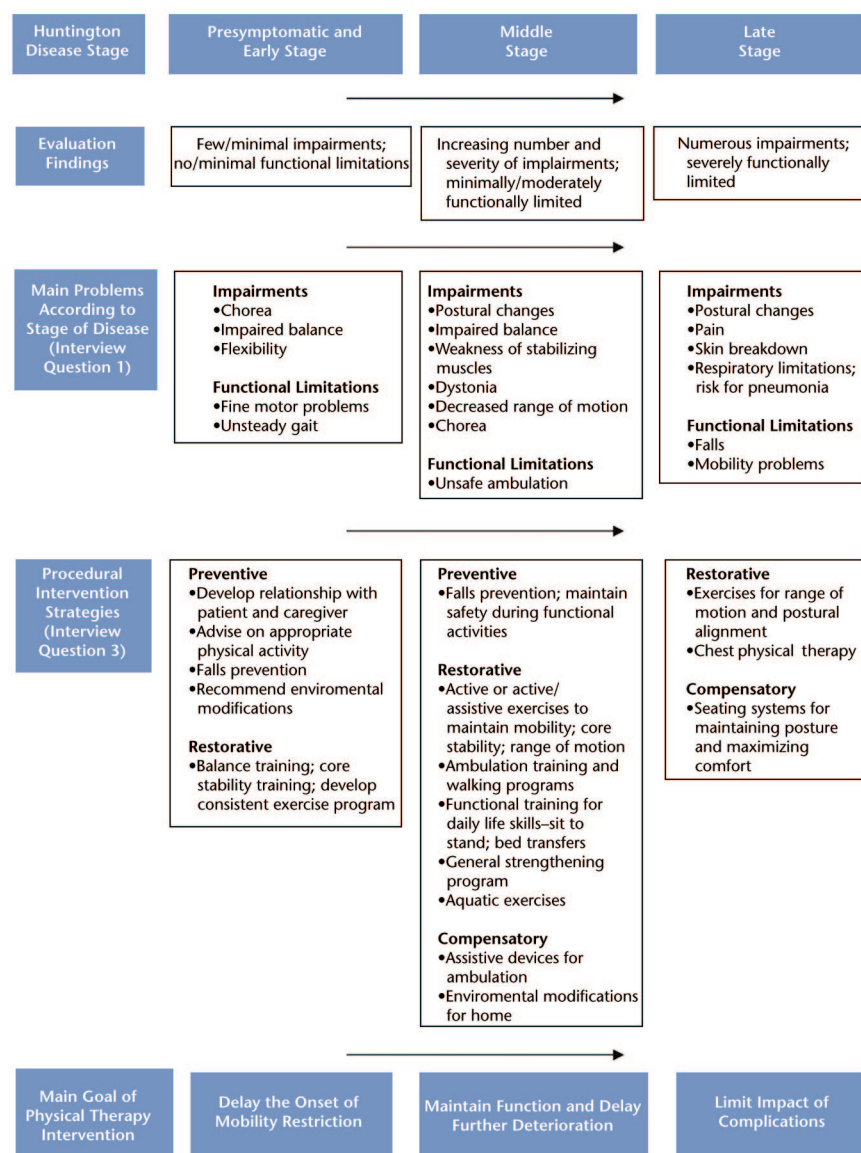


Figure 2. Conceptual framework for physical therapy intervention in Huntington disease.

Participant 6 agreed with these 2 participants and said:

From an aids point of view, I think it is very difficult, because I do not think we have ever been successful supplying anybody with a walking aid because of the arm movement. Sometimes if you can get them to concentrate long enough, they can do it, but of course towards the end, their concentration is poor. I mean there are so many cognitive problems as well.

Participants believed that teaching people to develop compensatory strategies would be most effective in aiding in the prevention of falls. These strategies included developing coping mechanisms, modifying the environment, modifying caregiver behavior, and providing aids in certain situations.

Master theme. The 3 subthemes all related to a master theme that

emerged from our analysis: Physical therapy management of HD must be modified according to an individual's problems and to the stage of the disease. To illustrate this master theme, we extended a framework (Fig. 2) originally developed by Dal Bello-Haas¹⁹ for people with neurodegenerative diseases. When the interview responses from main questions 1 and 3 (Appendix) are applied, this framework relates specifically to people with HD and is supported by statements made by the participants.

The respondents identified functional deterioration as being correlated with increases in the number and severity of motor impairments, as indicated by participant 8, who said:

At the early stage . . . more balance and flexibility problems, coordination, and sometimes loss of muscle strength. Mild to moderate [mid-stage] usually includes problems with walking and falls and sometimes posture problems and provision of equipment. Severe [late stage] tends to be more maintaining range of movement, posture, and flexibility.

Therapists reported that at different stages of the disease, different problems emerged. Problems with behavior, cognition, and motivation were reported to be some of the main issues throughout the disease stages. As the motor disorders progress, cognitive impairments may interfere with the learning of new skills. However, some simple strategies can be implemented by family members or caregivers and by therapists to compensate for this situation. Participant 8 commented on one patient:

We did try to involve him and use cueing references. We tried to keep instructions quite simple and straight. We gave him pictures of exercises to do, and we did not change the program. We kept on repeating it in the hope that he would learn that and involved his wife quite heavily with

that as well to try to reinforce the pattern of what we're trying to get him to learn.

Motivation also was considered to have an impact on the physical therapy program, with an individual's feelings of helplessness, depression, and lack of understanding of the need for physical therapy being the main reasons identified for nonadherence. Some strategies for encouraging an individual's participation in a rehabilitation program and continuing home exercises were mentioned by participant 5, who said:

Family members have a big role really, and it is largely that. If a patient goes home, they won't do the exercises unless somebody is there trying to motivate and encourage them to do them. We get family and carers on board and use diaries that they tick when they have done exercises. That helps. . . .

The uncontrolled behavior of people with HD was also considered to have a potential impact on the physical therapy treatment. Participant 2, who had extensive experience working with people with HD and psychiatric problems, suggested strategies to manage uncontrolled behavior. Participant 2 said:

We have patients with behavioral problems that can have a massive impact on physiotherapy. Clearly, we have quite a lot of experience with dealing with very difficult, aggressive patients, so I normally manage that. I try strategies to find ways around and get them involved and doing something. If they are physically aggressive, then I will just try to gently hold the patient and try to facilitate them through and just do a lot of positive praise. . . . If they continue to be very aggressive and resistant, then I try another day.

The noticeable shift in the management strategy from preventive in the early stages to supportive in the late

stages was mentioned by participant 2, who said:

In terms of physiotherapy, I think they should be more involved before they become symptomatic to work on things like core stability and the other things that they could benefit from in the long term. Right the way through, it is spending more time on thinking down the line, maintaining their posture, prevent muscle shortening and when to keep them standing and weight bearing, making sure posture is good at the meal time to prevent aspiration and then to make sure they are weight bearing evenly when they are sitting down or lying to prevent any pressure sores or these sorts of things.

It is clear from the results presented that each of the participants felt strongly that, at the different stages of HD, therapists needed different management strategies, assessments, and goals. Although preventive strategies should be implemented in the early stages of the disease, with the aim of delaying a further deterioration in an individual's condition, current management is mostly supportive in the late stages of the condition. Late-stage management aims to reduce the impact of complications.

Discussion

Physical therapy may play a role in facilitating independence in activities of daily living for people with HD. However, evidence for the efficacy of physical therapy treatment in people with HD is almost nonexistent, with little systematic research aimed at developing strategies for managing the mobility deficits present in this condition. Currently, for physical therapists working with people with HD, there is little documented information to guide their intervention strategies. The results of the present study have provided a framework that may help to better direct physical therapists in the provision of services for people with HD

and to facilitate the development of efficacy studies for people with HD.

Three subthemes and one master theme emerged from the interviews. The first subtheme involved physical therapy assessment procedures for people with HD. The UHDRS was not reported as being used, although it has a high degree of validity and reliability for motor scores, meaning that it can be used for tracking HD clinical features over time.²⁰ Functional assessments were considered the most appropriate methods of evaluation. Measures such as the Berg Balance Scale and the TUG, which are used for people with other conditions, were recommended for people with HD. There is a need to develop appropriate outcome measures that are sensitive to physical therapy-related changes and that consider both impairments and associated functional limitations for people with HD.

The impact of cognition may be a limitation with any of the suggested outcome measures. Although the TUG has been recommended as a screening tool for identifying older people who are at risk for falling,²¹ poor performance on the TUG may be attributed to cognitive impairments. For this particular test (as for most functional tests that are timed), an understanding of the instructions and interactions among the patient, the assessor, and the environmental setting are required.²² Given the deterioration of cerebral integration and processing of sensory (visual, vestibular, and proprioceptive) information in the presence of cognitive impairments,²³ mobility functions such as walking can be negatively influenced. It has been found that slower gait speed, decreased step length, and increased step-to-step variability are more common in people with cognitive impairments than in older adults who are healthy.^{24,25}

For the second subtheme, the present study demonstrated the underutilization of physical therapy services in managing HD, particularly in the early stages of the condition. Referrals for physical therapy services seem to be occurring in the middle and late stages, with insufficient referrals occurring in the presymptomatic and early stages.

Despite the underutilization of physical therapy services in the early stages of the disease, the present study highlighted the benefits of physical therapy in the early stages. A physical therapy program implemented in the early stages and directed toward improving impairments such as reduced flexibility, coordination, balance, and muscle strength (force-generating capacity) has the potential to reduce disease manifestations and improve an individual's quality of life.² The importance of providing physical therapy in the early stages of the disease has a solid scientific foundation and theoretical support in recent mouse model studies.^{8,9,26,27} These studies suggested that environmental stimulation in the early stages delayed the degenerative loss of cerebral volume in mice. This remarkable effect of environmental enrichment in delaying the onset of the disease in affected mice inferred a positive impact of physical therapy programs in humans.

In the third subtheme, falls were considered a major issue, particularly toward the late stages of the disease. Although the incidence of falls in HD has not been empirically determined, there is some evidence that falls do occur in people with HD. In a gait study of people with HD, it was reported that 11 of 13 people had a history of 2 or more falls.²⁸ This finding is in agreement with data from our questionnaires, in that 95.3% of the respondents considered their patients with HD to be

at risk for falling, with balance and loss of coordination as the main contributing factors. Although falls are considered to be one of the major physical therapy-related issues associated with HD,^{1,7,29-31} the reasons for falls occurring in people with HD are unclear. Falling in HD is a multifactorial issue, with impaired postural stability, dystonic postures, and gait disturbances considered contributing factors.³² Falls and a loss of independent ambulation³³ are often factors that precipitate admission to nursing homes.

The master theme that evolved from the analysis of the interviews was overriding and connected the 3 subthemes: Physical therapy management in people with HD, both evaluation and intervention, must be adaptable to the individual and to the stage of the disease. This theme has implications for future physical therapy trials for HD, stratifying people according to the stages of the condition and then evaluating the effectiveness of various management approaches at these stages. Clear definitions of specific interventions and consensus as to best practice in physical therapy are essential to providing the required evidence to justify physical therapy interventions^{34,35} and are even more essential in complex, changing conditions such as HD. Furthermore, a clear understanding of the cognitive, motivational, and behavioral issues that may affect treatment at different stages of the disease is required. Developing appropriate physical therapy intervention programs for HD is likely to be complex,⁵ but a clear understanding of the components of the interventions, factors that influence responses to treatment, and potential mechanisms that may influence outcomes is essential before the initiation of large-scale trials.³⁶

Conclusion

The results of this study lend support to the suggestion that physical therapist management should be modified according to the different stages of the condition. The study was limited in that it was conducted only in the United Kingdom with a relatively small purposive sample. Despite the limitations, there are implications for future physical therapy trials for HD, stratifying people according to the stages of the condition and then evaluating the effectiveness of various management approaches at these stages. Rather than relying on clinical judgment for treatment options, therapists may use the information gathered in this study to help them make better decisions based on consensus recommendations from people working in the field and justified by the currently available evidence. The information gathered in this study also may facilitate the development of standards of care and the design of future physical therapy trials.

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Ethical approval was obtained for all aspects of this study, and all research was undertaken in accordance with the Research Governance Framework of Cardiff University. All subjects signed informed consent statements prior to participation in the study.

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Physical Therapy in Huntington Disease

Appendix.

Interview Schedule

Greeting:

Thank you for agreeing to this interview. We want to explore your experiences of assessment and treating people with Huntington disease, which we will refer to as HD. The interview will last approximately 30 to 45 minutes, and it will be audio recorded using a Dictaphone. Written notes will be taken during the interview by an additional researcher. After the interview, the recording will be transcribed. The transcript will be sent to you to see if it represents your ideas.

We have sent you an information sheet and consent form. Have you read the documentation, and do you agree to participate and consent to the audio recording?

Are you happy to continue?

Introductory Questions:

1. Can you describe your physical therapy training?

FOLLOW-UP:

- How long ago did you qualify as a physical therapist?
- What was your qualification?
- From where did you get your qualification?

2. Can you tell me about your past and current work experience?

FOLLOW-UP:

- Experience in HD, including any postgraduate training.

3. How many patients with HD do you treat per year?

4. Referring to the stages of HD as pre (presymptomatic), early (mild impairment), mid (mild to moderate impairment), and late (severe impairment, dependent for all care), at what stage of the condition usually are the patients that you treat?

5. How are they usually referred into your service?

FOLLOW-UP:

- What is the most frequent source of referral?
- What are the most frequent reasons for referral?

Main Questions:

1. According to the stage of the condition, what are the main problems that patients with HD have?

2. How do you usually assess these problems?

FOLLOW-UP:

- What outcome measures do you routinely use when treating patients with HD?

3. About your treatment plan. What treatments would you normally provide when dealing with patients with HD?

FOLLOW-UP:

- How could this differ according to the stage of the condition?
- When is the best time to address the problems?
- What is the average frequency of treatment according to the stage of the condition?
- What is your strategy to reduce the risk of falls?
- What is your provision for the use of walking aids and assistive devices?
- What support or advice is given to the family as a part of the treatment plan?

4. Do you consider behavioral problems with HD to affect your treatment?

FOLLOW-UP:

- Could you explain how and why?

5. Do you consider dementia in patients with HD to affect your treatment?

FOLLOW-UP:

- Could you explain how and why?

6. What affects or limits your proposed treatment?

7. Does your department have a care pathway for dealing with HD?

FOLLOW-UP:

- If yes, could you please explain it?
- If no, what is your opinion about using a standardized care pathway set up for dealing with HD?
- What are the most important components?
- How would it be structured?