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Effectiveness of Physiotherapy, Occupational Therapy, and Speech Pathology for People with Huntington's Disease: A Systematic Review

Belinda Bilney, Meg E. Morris, and Alison Perry

This review provides a summary of the current literature examining the outcomes of physiotherapy, occupational therapy, and speech pathology interventions for people with Huntington's disease. The literature was retrieved via a systematic search using a combination of key words that included Huntington's disease, physiotherapy, occupational therapy, and speech pathology. The electronic databases for Medline, Embase, CINAHL, Cochrane Controlled Trials Register, and PEDro were searched up to May 2002. Articles meeting the review criteria were graded for study type and rated for quality using checklists to assess study validity and methodology. The majority of articles that examined therapy outcomes for people with Huntington's disease were derived from observational studies of low methodological quality. A low level of evidence exists to support the use of physiotherapy for addressing impairments of balance, muscle strength, and flexibility. There was a small amount of evidence to support the use of speech pathology for the management of eating and swallowing disorders. The current evidence is insufficient to make strong recommendations regarding the usefulness of physiotherapy, occupational therapy, or speech pathology for people with Huntington's disease. There is further need for therapy outcomes research in Huntington's disease so that clinicians may use evidence-based practice to assist clinical decision making.

Key Words: Huntington's disease—Speech-language pathology—Occupational therapy—Physiotherapy

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The aim of this article is to assist physiotherapists, occupational therapists, speech pathologists, and rehabilitation physicians to effectively treat people with Huntington's disease by providing a review and critical evaluation of the evidence on therapy outcomes. A second aim is to identify the types of interventions or treatments most frequently used by therapists in the management of people with this debilitating neurological condition. Recommendations for therapy are made based on the evidence presented.

BACKGROUND

Huntington's disease (HD) is an inherited neurodegenerative condition that occurs owing to a mutation of the gene (IT15) located on chromosome 4.^{1,2} The condition is typified by progressive degeneration of the medium spiny neurons within the basal ganglia, primarily the caudate and putamen.^{3,4} As the disease progresses, neuronal loss occurs in the white matter, cerebral cortex, and thalamus.^{5,6} Although the age of disease onset cannot be accurately predicted,^{7,8} adult-onset HD usually occurs around 35 to 45 years of age.^{9,10} HD is progressive until death 5 to 20 years after onset.^{10,11} A juvenile form of the disease, known as Westphal variant, is associated with an earlier age at onset and is more aggressively progressive.¹² Clinical symptoms can occur in physical, cognitive, and emotional domains.

Huntington's disease is characterized by a disorder of voluntary movement as well as involuntary movements. Voluntary movement can be adversely affected by bradykinesia and akinesia, resulting in reduced gait speed and difficulty initiating movements such as walking. Postural stability may also be impaired,^{13–15} as exemplified by loss of balance

during movement and frequent falls.¹⁶⁻¹⁸ According to Folstein, "falls become common and life threatening with a risk of death due to subdural hematoma."^{18,p27} Involuntary movements in Huntington's disease include chorea, dystonia, athetosis, and tics. There is some evidence to suggest that involuntary movements such as chorea affect motor function, so that movements are less accurate or clumsy.^{19,20}

Cognitive deficits are associated with degeneration of the striatum and subsequent disruption to the frontal-subcortical neural circuits.²¹ Disorders of executive function and memory retrieval²² are reflected by difficulty in maintaining attention to a task,²³ planning, organizing information, and initiating activity.²¹ Cognitive changes associated with HD can compromise ability to complete everyday tasks such as dressing, shopping, and preparing meals. Deficits in arithmetic ability can also make money transactions more difficult, and high-level deficits in comprehension impede communication in situations where information is ambiguous or implied.²⁴ Deficits in cognitive function can have a considerable impact on family members by increasing the burden of care.

Behavioral symptoms of HD can include depression and irritability.^{21,25-27} Although these symptoms have a significant effect on the person's quality of life, and may negatively affect relationships with family and friends, they usually respond well to drug therapies and can be managed effectively once diagnosed.^{25,27} Depression may be managed with prescription of tricyclic antidepressants.²⁷ Irritability may respond to counseling and behavior management.²⁷

As the disease progresses, a combination of physical, cognitive, and emotional deficits impact the person's ability to continue in paid employment. In the later stages of HD, people frequently require assistance to complete basic bathing, grooming, dressing, mobility, and eating tasks. Communication becomes increasingly difficult owing to a combination of dysarthria and cognitive deficits,^{24,28} and there is an increased risk of aspiration due to dysphagia.²⁹⁻³¹

The management of Huntington's disease currently remains symptomatic because there are no known treatments to prevent, cure, or slow down disease progression. Physiotherapy, occupational therapy, and speech pathology address impairments and limitations to functional activity and participation in society (Table 1). The overall aim of therapy is to facilitate independence in activities of daily life and optimize participation in family,

work, leisure, and community activities. Some allied health interventions also aim to reduce participation restrictions, enabling the person to fulfill a wider range of societal roles. These aims may be achieved through a process of education, the delivery of specific advice on safety and risk management, application of specific therapeutic treatments, modification of activities or environments, or the provision of assistive devices such as adaptive eating utensils or wheelchairs.

Despite the potential for allied health professionals to assist people with HD to maintain independence, functional capacity, and participation in society, recent research suggests that therapy is not always routinely provided.^{32,33} A survey of families caring for people with HD reported that a small proportion of people (24%) had been assessed by an occupational therapist and only 8% had been seen by a physiotherapist. None of the respondents had been contacted by a speech pathologist.³² A more recent survey of 25 community dwelling people with HD and their carers reported that a lack of speech pathology services was the most common area of unmet need for people with HD.³³ Sixty-four percent of the survey respondents were assessed as requiring speech pathology intervention, although none had been previously seen by a speech pathologist.³³ Previous research also suggested that for people living in the community, contact with an allied health professional usually occurred for a single assessment,³² without the follow-up over time normally provided to people with progressive neurological disease.^{34,35}

A common perception of people with HD is that therapy services are beneficial.^{32,33} This is despite a low referral rate to physiotherapy, occupational therapy, and speech pathology practitioners.^{32,33} The low referral rate may be due to a reluctance to recommend therapy when there is limited scientific evidence demonstrating effectiveness. It might also be due to a limited knowledge about the roles and services provided by allied health clinicians. Alternatively, there may be resistance from service providers to accept people with progressive neurological conditions into rehabilitation programs based on an expectation of improvement, given that HD leads to progressive deterioration in movement and cognition. It is possible that some allied health professionals are unsure how to effectively treat people with HD because a strong theoretical framework to guide selection of treatments for adults with progressive neurological conditions has not yet been established.³⁶ This suggestion is supported by Peacock's³⁷ findings from a survey of 585

Table 1. Summary of Areas of Impairments and Activity Limitations Treated by Physiotherapists, Occupational Therapists, and Speech Pathologists

Physiotherapy	Occupational Therapy	Speech Therapy
<ul style="list-style-type: none"> • Gait and balance • Falls • Aerobic capacity • Muscle strength • Joint range of movement • Respiratory function • Transfers • Wheelchair prescription and training • Task-specific reach, grasp, and manipulation • Relaxation • Dystonia management • Prevention of contractures • Prescription of safety equipment, e.g., hip protectors • Family/carer education on mobility and transfers 	<ul style="list-style-type: none"> • Personal hygiene • Grooming • Dressing • Eating/drinking • Toileting • Transfers/mobility • Work restructuring • Driving assessment • Promotion of safe home environment • Provision of equipment and adaptive devices • Memory training • Task planning • Task execution • Problem solving • Fatigue 	<ul style="list-style-type: none"> • Speech intelligibility—rate and volume • Intonation • Rhythm and prosody • Respiration • Confrontational naming • Language comprehension • Eating—bolus preparation • Swallowing • Prescription and training in use of augmentative communication devices

registered physical therapists. The survey quantified physical therapists' levels of experience in working with people with HD. Although 15.5% of physical therapists had treated a person with HD, only 6.2% had treated more than one person with this progressive condition. Peacock³⁷ suggested that physical therapists might be apprehensive about treating people with HD owing to the absence of data or clinical literature to guide therapy interventions and clinical decision making.

Current clinical management for people with HD is based on clinical decision-making strategies formed for each individual.³⁶ Physiotherapy aims to reduce impairments and to improve function in motor activities.^{35,37-39} In the early phases of HD, physiotherapists are usually involved in the assessment of movement disorders, mobility, and functional activity.^{38,39} Recommendations are made for physical activity programs aimed at maintaining optimum cardiovascular fitness, muscle strength, and flexibility.^{38,39} Gait and balance are usually assessed, and where appropriate, the person is taught strategies to maintain the ability to walk and to prevent falls.^{36,38} As the disease progresses, physiotherapy is frequently directed toward maintaining or retraining the reach to grasp movement,⁴⁰ bed mobility, transfers, and walking.^{36,39} When mobility becomes further compromised, wheelchair prescription and training of wheelchair use may become a priority.^{27,39} If there is a risk of aspiration, the physiotherapist works closely with the speech pathologist to teach the person how to clear sputum and cough effectively.⁴¹

An occupational therapist may be asked to conduct a workplace assessment and advise the person with HD and his or her employer about work restructuring. As the progression of the disease impacts cognitive or functional abilities, occupational therapists can provide assistance with the transition from paid employment to a societal role that is structured and meaningful.⁴² The occupational therapist may also be asked to participate in an assessment of the person's automobile driving skill and make recommendations about future driving behavior.⁴³ Another role of occupational therapy is to conduct home visits to assess functional abilities and the home environment. Recommendations for equipment or modifications are made to optimize independence and safety.³⁹ A person with HD can be taught strategies to maintain independence in tasks such as grooming, dressing, and eating.³⁹ Educating the person and his or her family about changes to cognitive ability and the consequent impact on function is another important role of the occupational therapist.³⁹

Speech pathologists provide treatment strategies for people with dysphagia (swallowing disorders), aimed at reducing their risk of aspiration.³⁰ Aspiration may be related to eating and can be either dangerously silent or associated with coughing. Modification of food consistency, reeducation of eating behaviors, and teaching techniques to assist a safer swallow may be used to increase independence in eating.⁴⁴ Strategies to aid communication problems due to dysarthria (slurred speech) or impaired comprehension of language

may also be used.⁴⁵ The speech pathologist also educates the person with HD and his or her family regarding eating and safe swallowing as well as teaching strategies to enhance communication.⁴⁴

Given that HD is an autosomal dominant disease, therapists frequently have a role in educating family members who are at risk of developing the disease. The inheritable nature of HD may significantly impact families with feelings of anxiety about the future and guilt that their children may also develop the disease. Financial hardships and social isolation are also frequently associated with chronic illness.⁴⁶ Therapists who work with people with HD should therefore be skilled in counseling and be aware of the broader issues that affect people with HD and their family.³⁸

Defining the Search

This systematic review was designed to identify journal articles that examined treatment for subjects over 18 years of age with a confirmed diagnosis of HD, evidenced by a positive genetic test or a family history of HD with signs of chorea. The aim was to capture all treatments and interventions implemented by physiotherapists, occupational therapists, and speech pathologists. Interventions were included if they were provided for impairments of movement, cognition, or emotional status. Therapeutic interventions that aimed to improve the performance of activities or participation in society were also included. Interventions that were not specifically documented in the reviewed articles as being delivered by a physiotherapist, occupational therapist, or speech pathologist were excluded.

A preliminary search of the HD literature failed to identify intervention studies specific to physiotherapy, occupational therapy, or speech pathology that were of experimental or quasi-experimental type. Experimental studies allocate participants to different groups in order to determine the effect of a particular intervention. In experimental designs, the allocation of participants to groups is concealed to avoid selection bias.⁴⁷ In contrast, quasi-experimental designs do not have true randomization of participants into treatment groups or group allocation concealment.⁴⁷ Because of the limited scope of the literature, observational studies were included in this systematic review process. Observational studies do not try to control allocation to intervention groups and include before and after studies, cohort studies, and case control studies.⁴⁷ Articles containing only expert opinion were identified but not

included in the data analysis. Only treatment outcomes reported with quantitative or qualitative data were included in the systematic review.

Journal articles published in English and indexed on electronic databases were included in the systematic review. Book chapters or sections not accessible via the electronic databases were excluded. Literature published prior to 1966 was not accessible via the electronic databases and was therefore excluded. The review included journal articles published up to week 2 May 2002. In addition, reference lists from key journal articles were hand searched to identify relevant papers not located by the electronic search.

Databases

Electronic databases were searched during the period commencing week 2 of October 2001 until week 1 November 2001. The following databases were examined: CINAHL (1982 to week 1 November 2001), Embase (1984 to week 4 October 2001), Medline (1966 to week 5 October 2001), Cochrane Controlled Trials Register (November 2001), Allied and Complementary Medicine (1985 to November 2001), and PEDro database for physiotherapy trials, (November 2001). To update the search, the electronic databases for Medline, Embase, and CINAHL were searched again during May 2002.

Search Terms

The search strategy was divided into components in order to collect data on study types, study samples, intervention types, and therapy outcomes. Study designs were identified using the exploded terms *meta-analysis*, *case-control studies*, *prospective studies*, *retrospective studies*, *case study*, *record review*, and *review literature*. To identify the study sample, the terms *Huntington's disease* and *chorea* were exploded. In addition, the terms *Huntington* and *Huntington chorea* were used. Intervention types were searched using a combination of phrases: *physiotherapy*, *physical therapy*, *occupational therapy*, *speech therapy*, and *rehabilitation*. The terms *communicative disorders*, *speech disorders*, *impaired swallowing*, *deglutition disorder*, and *swallowing therapy* were exploded. The terms *physical activity*, *exercise therapy*, *muscle strength*, *physical mobility*, *stretching*, and *respiratory therapy* were also exploded. To identify papers that contained additional information about occupational

therapy interventions, the exploded terms *activities of daily living*, *functional status*, and *assistive technological devices* were used.

The initial search of the electronic database retrieved 87 papers using a broad application of the systematic review inclusion criteria. The abstracts of the retrieved articles were read and then eliminated if they failed to meet the review inclusion criteria. The full texts of the remaining 24 articles were then read and rated by 2 reviewers (NS, BB).

Methodological Evaluation

The reviewers identified the study type using a hierarchy of levels of evidence adapted from the United States Department of Health and Human Services.⁴⁸ The classification ranks study evidence as (I) systematic review, meta-analyses of randomized controlled trials; (II) randomized controlled studies; (III) nonrandomized intervention studies; (IV) observational studies; (V) nonexperimental studies; and (VI) expert opinion. Observational studies were subdivided and ranked according to the method suggested by Khan et al.;⁴⁷ thus, (1) cohort study, (2) case-control study, (3) cross-sectional study, (4) before-and-after study, and (5) case series. The study methodology was systematically evaluated using a checklist of items relevant to the study type. Four separate checklists were used to evaluate randomized control studies, cohort, case control, and case series studies. The checklists for the cohort studies, case control, and case series were drawn from a list of suggested quality criteria for the assessment of observational studies by Khan et al.⁴⁷ The checklists enable evaluation of the presence or absence of quality elements considered important for each study type. The assessment included items to determine subject selection bias, reliability and blinding of outcome assessment, and appropriateness of follow-up period.⁴⁷ The quality checklists were independently scored by 2 raters (NS, BB). The maximum possible score for each of the study types was as follows: randomized control studies (10), cohort study (10), case control (9), and case series (6). Disagreements between the 2 raters for the checklist scoring were settled by discussion.

Data Extraction

Relevant data were extracted and recorded on a standardized form by 2 independent raters (NS,

BB). Data of interest included subject information: sex, age, confirmation of diagnosis, duration of illness, severity of symptoms, cognition, depression, and medication status. Also documented were descriptions of therapy intervention, frequency, and duration. The validity and reliability of the study outcome measures and study results were also recorded.

Data Synthesis

The data extracted from the literature will be presented as a descriptive summary and synthesis in a table format. It was not possible to determine the treatment effect size or the confidence intervals around the treatment effect owing to insufficient statistical analysis or reporting of descriptive data within the extracted journal articles. Tables 2, 3, and 4 summarize the range of physiotherapy, occupational therapy, and speech pathology interventions provided to people with HD that were identified by the literature search but were not necessarily supported by outcome evidence (e.g., expert opinion).

RESULTS

The search failed to identify any studies ranked as level I, II, or III evidence.⁴⁸ There were no randomized clinical trials or nonrandomized controlled clinical trials on the effects of physiotherapy, occupational therapy, or speech pathology for people with HD. A small number of observational (level IV) and nonexperimental studies (level V) were identified. In addition, 6 journal articles rated as expert opinion (level VI) were retrieved via the electronic search. The following section summarizes the evidence of treatment outcomes for people with HD obtained from the level IV and V studies. The studies are presented in rank order for level of evidence and methodological quality, with the most robust studies presented first. In addition, the scope of therapy interventions is summarized in table format (Tables 2-4).

Physiotherapy for People with HD

Peacock³⁷ evaluated the effects of an outpatient exercise program designed for people with HD. The study measured therapy outcomes for 10 participants diagnosed with HD who were living in the

Table 2. Physiotherapy Interventions Identified in the Literature Review

Impairment	Treatment/Intervention	Authors	Level of Evidence*
Reduced coordination Reduced strength Poor posture	Mat exercise	Peacock ³⁷	Level 4
		Imbriglio and Peacock ³⁸	Level 6
	Chair exercise Weighted cuffs Strengthening	Lavers ⁵⁰	Level 6
		Lavers ⁵⁰	Level 6
		Lavers ⁵⁰	Level 6
		Peacock ³⁷	Level 4
		Imbriglio and Peacock ³⁸	Level 6
		Churchyard et al. ³⁶	Level 6
		Imbriglio ³⁸	Level 6
		Binswanger ⁴⁹	Level 4
Decreased range of movement	Home exercise program Neurophysiological techniques	Binswanger ⁴⁹	Level 4
		Sheaff ⁵¹	Level 5
	Splinting Positioning Provision of supportive devices	Lavers ⁵⁰	Level 6
		Lavers ⁵⁰	Level 6
		Lavers ⁵⁰	Level 6
Bradykinesia in gait	Mat exercise Walking with an external cue	Peacock ³⁷	Level 4
		Thaut et al. ⁷⁸	Level 4
Postural instability	Attentional strategies when walking Assisted walking	Churchyard ³⁶	Level 6
		Lavers ⁵⁰	Level 6
	Avoidance of dual tasks Environmental modification Falls prevention strategies	Churchyard et al. ³⁶	Level 6
		Churchyard et al. ³⁶	Level 6
		Churchyard et al. ³⁶	Level 6
Bradykinesia in reach and grasp	Task-specific practice Movement transitions Movement initiation	Quinn et al. ⁴⁰	Level 6
		Quinn et al. ⁴⁰	Level 6
		Quinn et al. ⁴⁰	Level 6
		Quinn et al. ⁴⁰	Level 6
Force variability Chorea	Increase weight of object to be lifted Hydrotherapy	Quinn et al. ⁴⁰	Level 6
		Sheaff ⁵¹	Level 5
Reduced cardiovascular fitness	Relaxation therapy Cardiovascular fitness program Ball games	Imbriglio and Peacock ³⁸	Level 6
		Churchyard et al. ³⁶	Level 6
		Imbriglio and Peacock ³⁸	Level 6
Reduced breath control and volume	Breath control exercises	Lavers ⁵⁰	Level 6
		Peacock ³⁷	Level 4
		Imbriglio and Peacock ³⁸	Level 6

*Levels of evidence are based on the classification system adapted from the United States Department of Health and Human Services.⁴⁸

Table 3. Occupational Therapy Interventions Identified in the Literature Review

Activity	Treatment/Intervention	Authors	Level of Evidence*
Grooming	Reeducation	Mason et al. ⁵²	Level 4
Showering/bathing	Provision of aids; reeducation	Mason et al. ⁵²	Level 4
Dressing	Reeducation	Mason et al. ⁵²	Level 4
Feeding	Provision of aids	Lavers ⁵⁰	Level 6
Mobility	Provision of aids, wheelchair education	Mason et al. ⁵²	Level 4
	Rood techniques	Di Scipio and Hannesson ⁵³	Level 4

*Levels of evidence are based on the classification system adapted from the United States Department of Health and Human Services.⁴⁸

community and were able to travel to attend an exercise group. A 5-level disease severity rating scale was used to screen for people not severely affected by HD (level 1 and 2 severity). Cognition, depression, and medication status were not documented. Outcome measures were briefly described and included tests of flexibility, range of movement, standing balance, balance in kneeling, coordination, and breathing control. Physiotherapy intervention consisted of 12 weekly sessions of

relaxation training followed by 45 minutes of active exercise directed toward improving flexibility, coordination, balance, strength, and breathing control. Specific exercises within the session were not operationally defined. Although exercises were delivered in a group setting, they were individualized for each person. Following the 12-week program, participants attended 3 additional booster sessions held monthly. In addition, they were encouraged to complete a home exercise program.

Table 4. Speech Pathology Interventions Identified by the Literature Review

Impairment	Treatment/Intervention	Authors	Level of Evidence*	
Dysphagia	Modify head posture	Kagel and Leopold ⁵⁵	Level 4	
		Leopold and Kagel ⁵⁶	Level 4	
		Logemann ²⁹	Level 6	
	Modify body posture	Hunt and Walker ³¹	Level 4	
		Kagel and Leopold ⁵⁵	Level 4	
		Leopold and Kagel ⁵⁶	Level 4	
	Modified food/bolus consistency	Logemann ²⁹	Level 6	
		Kagel and Leopold ⁵⁵	Level 4	
		Leopold and Kagel ⁵⁶	Level 4	
	Thermal/gustatory stimuli	Kagel and Leopold ⁵⁵	Level 4	
		Verbal cueing	Kagel and Leopold ⁵⁵	Level 4
			Leopold and Kagel ⁵⁶	Level 4
	Modified swallow sequence	Logemann ²⁹	Level 6	
		Hunt and Walker ³¹	Level 4	
		Kagel and Leopold ⁵⁵	Level 4	
Modified utensils	Leopold and Kagel ⁵⁶	Level 4		
	Hunt and Walker ³¹	Level 4		
	Kagel and Leopold ⁵⁵	Level 4		
	Leopold and Kagel ⁵⁶	Level 4		
	Reduce rapid eating	Miller and Langmore ³⁰	Level 6	
	Family education regarding choking	Leopold and Kagel ⁵⁶	Level 4	
Dysarthria	Treat respiratory chorea	Logemann ²⁹	Level 6	
		Prescription of lightwriter/communication aids	Klasner and Yorkston ⁵⁷	Level 4
	Chalmers ⁸⁰	Level 6		
Productive syntax impairments	Compensatory strategies	Murray ^{24,28}	Level 6	
Impairment in language comprehension	Teach strategies to improve language comprehension	Murray ²⁴	Level 6	

*Levels of evidence are based on the classification system adapted from the United States Department of Health and Human Services.⁴⁸

The details of the exercise content, duration, frequency of completion, or compliance to the home exercise program were not reported. Peacock reported that “at the end of six months in the exercise program, all 10 patients had improved to some degree on seven or more of the 10 functional tests.”^{37,p34} Peacock reported that participants improved on the tests of flexibility, coordination, breath volume, and breath control during vocalization. Nine people improved on tests of static standing balance and strength. For the tests of reciprocal movement, 2 participants remained unable to complete the test and 4 showed only minimal improvement. This study provides limited evidence that exercises may be helpful for some people with mild impairment in the early stages of HD. However, the study was limited by low methodological quality (4/8 checklist elements) because of small sample size and the absence of data pre- and postintervention. The exercises completed during the group session and home exercise program were not sufficiently described to enable replication. The inclusion of a home exercise program that was not monitored for compliance limits the ability to draw firm conclusions about how fre-

quently exercises should be performed to gain therapeutic effects. In addition, the multifaceted nature of the program makes it difficult to draw direct links between specific types of exercises performed and therapy outcomes.

Binswanger⁴⁹ described the effects of a physiotherapy program for 5 people diagnosed with HD. The description was provided in a letter to the editor and therefore was limited in detail. Participants lived at home with support from carers. They were mentally alert and appeared to be motivated to change their level of ability. Neither disease severity nor functional status were described. Each person with HD received home physiotherapy for 1 h, twice a week for 4 weeks. Treatments were not specifically described, although they were reported to utilize neurophysiological techniques and therapy directed at specific areas of deficit such as muscle weakness, reduced range of movement, gait, and breathing patterns. Although objective outcome measurements were not reported, Binswanger noted subjective improvements in alertness and balance that appeared to result in safer ambulation. The methodological quality of this study was low (2/8) owing to bias in the selection of subjects

and failure to report the reliability and validity of the measurement tools. Although it was known that the participants had different levels of functional ability, neither disease severity nor functional status were quantified. The failure to include a follow-up period to assess maintenance of gains prevents any conclusion being drawn regarding the long-term effectiveness of physiotherapy techniques utilized in this study.

The effects of a weekly group intervention program conducted over a 12-month period for 6 residents of a long-term care psychiatric ward was described by Lavers.⁵⁰ Group therapy aimed to maintain mobility, balance, and coordination; prevent contractures; increase social interaction; and reduce passive behavior. The physiotherapy interventions used in the program are summarized in Table 2 and included formal mat exercises in prone-lying, kneeling, bridging, weighted cuff exercises, and ball games. The author did not report objective therapy outcomes, so it is not possible to draw conclusions regarding the effectiveness of treatment.

A single case study by Sheaff⁵¹ reported the effects of a hydrotherapy program for a severely disabled male with HD. The intervention involved gentle exercises in water supervised by a physiotherapist for an unknown period. Outcomes of the intervention were not quantified, although the author reported that physiotherapy was beneficial. The failure to provide any objective outcome measurements precludes conclusions being drawn regarding the usefulness of hydrotherapy for people with HD.

There are a small number of observational studies indicating that exercise for strengthening, range of movement, mobility, and balance may reduce the severity of impairment.^{37,49} This evidence is generally of low quality and is limited by selection biases, failure to use valid and reliable measurement tools, and small participant numbers.

Occupational Therapy for People with HD

The systematic review process identified 3 journal articles that described interventions provided by occupational therapists for adults with HD (Table 3). These included 2 observational level studies of single-case study design.

Mason et al.⁵² described the effects of an occupational therapy program aimed at enabling people to carry out personal activities of daily living. The

article reported results for 4 participants using single-case, A-B-A experimental design. An 8-week period of baseline assessment was followed by a 16-week intervention phase and then a further 8 weeks of observational assessment. The participants were selected from a group of 11 people diagnosed with HD who were residents of a long-term care facility. Participants were excluded if they had severe cognitive impairment or were unable to comply with a rehabilitation program. Disease severity was not documented, although all participants were reported to require nursing assistance to complete daily living tasks. Disease duration ranged from 11 to 20 years. Occupational therapy involved reeducating each person in personal care tasks such as face washing, drinking from a cup, teeth cleaning, or mobility. Treatment outcome was measured on a 9-point scale used to rate the level of external assistance required to complete the activity. Nonblinded raters assessed the participants at several points during the baseline periods and at 2 weekly intervals during the intervention phase. Performance ratings revealed considerable fluctuations in ability to complete the assessed activities during the baseline and intervention periods, making interpretation of the data difficult. Visual analysis of the data suggested that no gains in function occurred as a result of occupational therapy. Nevertheless, it cannot be discounted that occupational therapy intervention prevented deterioration in the functional task.

The fluctuations of performance over time, in the study by Mason et al.,⁵² may have occurred because of real changes in the person's ability to successfully complete the motor task. Alternatively, the measurement tool used in this study may not have been reliable and may have contributed to the inconsistency of the results. The validity of this study is further compromised by the poorly defined inclusion and exclusion criteria. The 4 participants were not clearly described in terms of disease severity, cognitive status, or willingness to comply with a rehabilitation program, limiting the ability to generalize the results. Adverse events such as deterioration in medical status were not documented during the preintervention, intervention, and postintervention phases. The frequency, duration, and type of treatment technique utilized in the study were not described, preventing any conclusions being drawn regarding the effectiveness of the interventions.

Di Scipio and Hannesson⁵³ described the outcomes of an occupational therapy program provided to a single female participant who had been a

hospital in-patient for the previous 5 years. The participant had a history of depression, cognitive deterioration, and loss of ability to communicate. Before the commencement of occupational therapy, the participant was nursed on the floor during the day, due to an inability to maintain a sitting position. She required assistance to stand but could walk a short distance with frequent falls. Occupational therapy was provided daily for 20 to 30 min over a 10-week period. Treatment involved applying "Rood" techniques⁵⁴ such as brushing, rubbing, icing, and positioning in a flexor withdrawal posture. Objective outcome measurements were not reported; however, the authors noted improvement in the subject's function, including an ability to maintain a sitting posture, to lift her head and shoulders from a prone position, and to ambulate. The validity of this single case example is limited by the possible change of medical status during the intervention phase. In this case example, the patient had been withdrawn from all medications including tranquilizers 1 month prior to the commencement of the study. In addition, the absence of objective, valid, and reliable outcome measurements limits the interpretation of this single case study.

It is difficult to draw any occupational therapy treatment recommendations on the basis of the 3 outcome studies identified. The studies involve a small number of patients, all of whom were long-term residents of high-level nursing care facilities, restricting the generalizability to people living in institutional care.

Speech Pathology for People with HD

The systematic examination of the literature identified 3 papers that addressed the effects of speech pathology for people with HD. Two papers specifically examined treatment effectiveness for dysphagia (swallowing disorders).^{55,56} The third examined the effect of augmentative and alternate communication techniques⁵⁷ (Table 4). The literature contains many reports of the impairments present in HD such as dysphagia, dysarthria, and cognitive and comprehension difficulties^{24,28,58-65} that would traditionally be treated by a speech pathologist. The failure to identify journal papers specifically addressing treatment efficacy for these deficits in HD may reflect the organization of the speech pathology literature, which is generally structured by terms relating to presenting impairment, rather than by etiology or diagnoses.

Leopold and Kagel⁵⁶ reported the results of a study with 12 people diagnosed with HD, 11 with moderately advanced disease. Objective assessment included rating the participants on a 0- to 5-point scale for severity of dysphagia. Barium cineradiographic studies were used to rate liquid and solid bolus swallows on a nominal scale. All 12 participants were rated as having dysarthria, an impaired swallow and cough. Every participant received speech pathology intervention. One patient was managed with diet modification, and the others with intensive speech pathology treatment.⁵⁶ The authors did not provide the frequency, duration, or detailed description of the intervention, making it difficult to determine the specifics of the interventions that each patient received. The type of interventions described included "standard and modified techniques such as altering diet selection and utensil type, individualizing swallowing sequences (chew-swallow-cough-swallow), performing a modified Valsalva maneuver, and correcting the head/body positioning."^{56,p58} The results of the intervention were not subject to statistical analysis. However, on reassessment using barium cineradiographic studies, the authors reported improvements in the preparation, food transfer, and swallow phases of eating, with a decreased incidence of aspiration and reflux. Furthermore, the authors reported that gains in swallowing ability were maintained by some patients for up to 3 years after the intensive treatment phase. The methodological quality of the paper was rated as poor owing to the absence of inclusion and exclusion criteria, unreported reliability, and validity of the outcome measurement tools and lack of statistical analysis. In addition, the absence of a specific description of the interventions and the frequency that they were provided limits the conclusions that can be drawn from this study. It is interesting to note that in the opinion of Leopold and Kagel,⁵⁶ the participants with more severe dementia "required more sessions to habituate the cognitive and motor sequencing required to prevent aspiration" (p. 60). Eleven of the participants were rated as having a moderately advanced disease status, suggesting that disease status may not be a useful indicator to differentiate the participants most likely to respond to treatment. However, the addition of an objective assessment of the participant's cognitive status may assist in identifying people who will respond more quickly to therapy or make greater gains.

A paper published by the same authors in 1992⁵⁵ described 35 patients, 34 of whom underwent extensive clinical examination including videofluo-

roscopic investigation of their swallow and an assessment of self-feeding. Patients were classified according to whether rigidity or chorea was the primary presenting involuntary movement disorder. After the provision of adaptive eating equipment and modified body positioning, 29 of the 30 participants with hyperkinetic movements improved their ability to independently feed and reduced the incidence of tachyphagia, that is, excessively fast eating. Three of the 5 participants assessed as having rigidity also reduced their dependence for feeding by using adaptive equipment such as modified cutlery and non-skid mats, and by altering body posture using foam wedges.

Klasner and Yorkston⁵⁷ described a single case study of a male diagnosed with HD who was having difficulty communicating with his wife and had reduced his participation in active decision making for household management. Speech pathology was provided for 1 h per week over a 6-month period. The communication impairment was addressed by developing and implementing scripted conversations and by use of key words to prompt verbal description. Participation in household duties was addressed by developing lists of tasks to be completed and by using an alarm to prompt attendance to the task. The authors reported that both strategies were successful, resulting in improved communication and increased participation in household management.⁵⁷

There is some evidence that intervention provided by speech pathologists improves the ability of people with HD to independently feed.⁵⁵ Furthermore, there is limited evidence that dysphagia can be improved with intervention.⁵⁶ The methodological flaws in the 2 studies addressing eating and swallowing, including the failure to detail the exact intervention, its duration, and frequency, limit the conclusions that may be drawn regarding treatment effectiveness. It is unknown as to whether a number of treatment techniques are required to gain an effect or if the same result can be achieved using a single strategy. Further research is required to determine the impact of speech pathology on the ability of people with HD to communicate.

DISCUSSION

Principal Review Findings

The current evidence regarding the effectiveness of physiotherapy, occupational therapy, and speech

pathology interventions for people with HD is not strong. Therapy outcome studies of experimental or quasi-experimental design have not been reported in the literature. This review identified only a small number of journal articles that examined treatment outcome at an observational level of evidence. The studies were not sufficiently similar in the type of intervention used to combine the evidence for analysis. Furthermore, the quality ratings for study methodology were poor for the majority of studies. This was usually due to subject selection bias or failure to use valid or reliable outcome measurement tools. For the majority of the studies, the participants were not described in sufficient detail to enable replication and sample sizes were small. In addition, factors that may impact the efficacy of treatment, such as severity of disease, functional ability, cognitive status, presence of depression, or medication usage, were frequently not recorded or taken into consideration.

Despite these shortcomings, there is a small amount of evidence suggesting that exercise may be helpful for select impairments associated with HD, such as loss of range of movement or balance deficits. The use of speech pathology treatment techniques to promote safe swallowing is also supported by one study with level 4 evidence. It was not possible to determine which of the many treatment techniques used is most effective in managing dysphagia. Speech pathology may also be useful in assisting people to become more independent in self-feeding. At the time of this review, there was insufficient evidence to guide effective occupational therapy management for people with HD.

How Can the Study Methodologies Be Improved?

A notable shortcoming in this literature was the failure to use valid and reliable objective measurement tools to quantify therapy outcomes. The main tool used for objective measurement of impairment and activity in people with HD is the Unified Huntington's Disease Rating Scale (UHDRS).⁶⁶ The UHDRS is an ordinal rating scale that measures 4 domains of HD: motor, cognitive, functional ability, and behavior. The motor section is used to assess the severity of voluntary movement impairment and the severity of involuntary movement such as chorea and dystonia. Severity is measured on a 5-point ordinal scale. The cognitive assessment includes the verbal fluency test, symbol digit modalities test, and Stroop interference test. The

scale has high internal consistency and good inter-rater reliability,⁶⁶ and it is recommended that therapists use it to quantify impairments and activity limitations.

Other tests that quantify impairment in people with HD include tests of reaction and movement time,^{40,67-69} spatiotemporal parameters of gait,^{36,70,71} clinical tests of balance,¹⁵ and tests of motor speech.⁷² Abnormal involuntary movements can be measured on an ordinal scale^{66,73} or quantified with an accelerometer,⁷³ or by analysis of electromyographic muscle activity.⁷⁴ Kinematic analysis has been used in people with HD to assess reach and grasp movements,^{19,40} writing movements,^{75,76} and gait.⁷⁷ Some of these tests have been validated and found to be reliable and may be useful for measuring therapy outcomes.

Therapy outcome literature also frequently failed to quantify disease severity and functional and cognitive ability in people with HD. The inclusion of ratings of these domains may enable more homogeneous study samples to be formed. For example, cognitive impairment may reduce the effectiveness of a treatment technique that uses principles of motor skill relearning to change function. This was partially illustrated by a study by Thaut et al.⁷⁸ who used auditory cues to increase gait speed. In this study, participants who were more severely cognitively impaired did not respond as well to the technique as those who were less impaired. Identification of the factors that influence response to treatment should be an important component in future experimental designs.

The existing literature on therapy outcomes has not investigated the effect of environment on motor or cognitive performance. A recent investigation examining HD in a transgenic mouse model has found that the onset of motor signs of disease was delayed in mice who lived in an "enriched" environment.⁷⁹ By the end of the testing period, mice exposed to frequent changes of objects within their cages had significantly less loss of cerebral volume, and only 1 (14%) had developed signs of motor incoordination.⁷⁹ The concept of environmental enrichment may have implications for the development of therapy programs for people with HD and is worthy of future investigation.

SUMMARY

There is very little published evidence evaluating the efficacy of physiotherapy, occupational therapy, or speech pathology for reducing impairment

or increasing activity and participation in people with HD. There is a complete absence of randomized control studies in the published literature. The evidence in the current literature is not of sufficient strength for strong recommendations to be made regarding the usefulness of physiotherapy, occupational therapy, or speech pathology treatment for people with HD. There is, however, a small amount of evidence that exercise may be useful in addressing specific impairments in people who are not severely affected by the disease. In addition, there is preliminary evidence that speech pathology may be effective in reducing the risk of aspiration. Given the perception of people with HD and their carers that therapy is beneficial, controlled clinical trials need to be conducted as a basis for evidence-based clinical practice.

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