From, ‘‘Caring for Persons with Huntington’’s Disease’’ Editor: Edmond Chiu, AM

Chapter 5, Physiotherapy

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INTRODUCTION

Huntington’’s Disease runs progressive inexorable course, and physiotherapy is unable to affect this. However, through a programme of general and specific exercises, the effects of muscle disuse and physical inactivity which compound the other physical and emotional problems of HD may be reduced. By minimising muscle atrophy and weakness and deterioration of body fitness it is possible to improve the HD person’’s quality of life.

CLINICAL FEATURES APPARENT IN HUNTINGTON’’S DISEASE AND RELEVANT TO PHYSIOTHERAPY TREATMENT

1. Disturbed neuromuscular Function

(a) Mobility:

Usually within normal limits, although in those people in the later stages where rigidity is the predominant feature, mobility can be reduced but contractures are rare. **(b) Strength:**

A decrease in muscle strength is evident and often asymmetrical. The postural muscle groups are primarily affected— neck and trunk extensors, scapular retractors, shoulder elevators, dorsiflexors, hamstrings and most noticeably, hip abductors and extensors. **(c)Changes in Normal Posture**

Usually in the early stage postural abnormality is only slight, but there may be early signs of one or a combination of the following:

A wide base. Feet externally rotated — often one more than the other. Uneven weight bearing. Poking chin. Head held to one side and / or rotated. Later the arms assume bizarre positions — perhaps related to balance problems Hyper-extension of hips — resting back on ligaments. Usually asymmetrical.

(D) Ambulation:

There is great variation in gait pattern, e.g. Lack of rotation. Lack of arm swing Wide base. Uneven weight bearing resulting in a list to one side, making it difficult to ‘‘step through’’.There may be flexion at the hips causing weight to fall forwards and resulting in an increase in momentum and often loss of control. Sometimes ther is hyperextension at hips with a resulting danger of falling backwards. This hip extension may cause a more swaying gait. Later there is difficulty with initiating walking. The classical ‘‘dancing” ataxic gait is also seen. However, the early stage patient can walk and crawl in all directions, negotiate steps and transfer independently, but deterioration in normal timing and fluency of movement is evident. This is particularly noticeable in sideways and backwards movement.

From the early stage there is evidence of diminished control when transferring (e.g., from a standing position to a chair to a standing position).

As the disease progresses this can be further aggravated by an inability to judge distances accurately. Some have problems standing still.

(E) Co-ordination: There is evidence of deterioration of fluency and accuracy of voluntary movements. In asking a patient to perform the finger-nose test, the finger does not travel straight to the nose, but may curve or be broken by corrections. Instead of smoothly increasing at the onset and decreasing at the end, the movement may be excessive or unduly slow. Even in the early stage of the disease, rapid alternating movements (e.g. pronation, supination) may be difficult.

(F) Dyskinesia:

Movements in the early stages tend to be of small amplitude andddd promarilyaffect the distal parts of the extremities and, to a degree affect the facial muscles. As the diseaseprogresses, the movements increase in amplitude and may affect neck, shoulder and pelvic girdles. Dyskinesia increases with stress, excitement and fatigue. It also increases as the size of the base of support decreases.

(G)Muscle Tone

In the early stages, tested by passive joint movements, tone is within normal limits. But as the disease progresses there may be changes which could be hypotonic, hypertonic (rigidity) or variable (fluctuating).

(h) Reflex Response:

Cortical

There is deterioration in equilibrium reactions in the early stage when the base of support is narrow (e.g., standing) but later it is evident even when the base is wide— sitting or four-point kneeling.

Automatic Movement Reactions

Protective extension may be delayed or absent.

Mid Brain Reflexes

Righting reactions are depressed, and particularly noticeable is loss of rotation in gait and segmental break-up in rolling.

Lower Brain Stem Reflexes

Early stage patients may display subtle indications of tonic neck reflex activity, usually in the form of the Asymmetrical Tonic Neck Reflex (ATNR). The presence of the ATNR is usually associated with problems such as decreased trunk rotation and a decreasing ability to perform tasks which require a bilateral approach.

As deteriorate occurs ATNR may become more pronounced. There may also emerge a Symmetrical Tonic Neck Reflex — extension of the head beyond vertical may produce a strong generalised extensor spasm whilst a flexion in front of vertical produces a generalised flexion pattern.

Associated reactions are often evident fairly early and become stronger as the disease progresses. These reaction may be evident even when there are no other signs of tonic reflex activity.

2 Respiratory Dysfunction

On assessment, HD persons have been found to have:

> Poor lung capacity (VC) and forced expiratory volume (FEV1) Poor ability to blow— they often force air out in the form of a short ''huff'' Reduced control of expiration— they tend to exhale by passive relaxation or recoil of the chest and lungs. Poor cough particularly voluntary cough, and even when choking, cough is poor and unproductive. Reduced ability to breathe through the nose. Irregular, uncoordinated breathing pattern. Often inhaling air only when needed, they may gulp or sniff in and may use accessory muscles excessively. Reduced control of respiratory muscles, often due to involuntary movements. Abnormal movements in the form of a ''fluttering'' of the abdominals or a jerkiness of the intercostals may also be observed.

Thus, in practical terms, persons with HD are more susceptible to:

> Chest infections. As documented, the greatest cause of death is respiratory failure, pneumonia, etc. Dysphagia contributes to this.
>
> Reduced endurance.
>
> Difficulty with speech, as it requires controlled expiration.
>
> Difficulty with swallowing and eating, as we usually breathe through our nose when doing so.
>
> Difficulty with coughing, because to do so effectively we need to be able to (i) hold breath and (ii) exhale forcefully.

3 Sensation

Assessment results indicate that some HD persons may experience disturbance of sensory function— loss of discriminative ability.

Hot/Cold

Generally the abilllllllity to discriminate between the two remains intact. However, there are instances where:

(a) Sensation is dulled e.g., reduced ability to discriminate between, say, hot and very hot.

(b)It takes longer for the information to reach the cortex. The correct response may be given, but the test tube had to be held against the skin for several seconds longer than normal.

Two Point:

Discriminative ability is generally diminished and becomes more so as the disease progresses

Touch Localisation

Particularly in latter stage sufferers the ability to localise touch is very poor.

Steroegnosis (the ability to recognise familiar objects by touch with vision occluded):

Overall this was within normal limits in the early stages of the disease. However, in the later stages problems with stereognosis were recognised, due either to reduced manipulative skills or the inability to put a name to the object, i.e., the problem was primarily a word finding difficulty.

Anaesthesia and Pain Tolerance

Localised areas of anaesthesia are not common, and are usually only found in persons with additional problems, e.g., stroke or alcoholism. However, staff at Arthur Preston Centre have noted high levels of pain tolerance in many residents. For instance, smokers can burn their fingers without complaining of pain. Similarly, patients have been known to lie on open sores with no apparent discomfort.

4 Sphincter Function

Incontinence:

As deterioration occurs, persons with HD find a sudden ‘‘urgency’’ in the need to empty the bladder. Poor control of sphincter musculature can result in some leakage of urine or, in the advanced stages of the disease, may result in total bladder emptying. Regular emptying of the bladder often helps resolve or control this problem (e.g., toileting before and after meals, or hourly visits.

Total bowel incontinence is rare, but is more prevalent in the laterstages of HD, though it is difficult to determine to what extent this is due to inability to communicate.

5 Dysphagia

Difficulty swallowing fluids and /or solids and even saliva is a common feature of the advanced stages of the disease. It can lead to choking, asphyxia and subsequent death. As noted earlier dysphagia also contributes to respiratory infection and inhalation pneumonia.

6 Higher Cortical Function

Memory

Research has indicated the persons with HD commonly experience memory disorders, and thatthese disorders are apparent prior to the emergence of motor dysfunction. This has been found to be the case with our population. Memory dysfunction is often severe enough to cause significant problems with work and family routines, even in the early stages. As the disease progresses, further deterioration is likely to occur.

Memory dysfunction commonly takes the form of:

Poor retrieval and word-finding difficulties.

Poor encoding (inability to process several pieces of information at once.

Poor short-term memory

Long-term memory usually stays relatively intact, although toward the latter stages of the disease this may be affected.

Concentration:

Reduction of concentration span and an impairment of cognitive and organisational skills are someof the first obvious symptoms to occur, and again, cause difficulties in the family and workplace.

Body Image

Body image (ability to perceive the location and relationship of body parts) usually remains intact until well on in the disease process. If dyspraxis develops, then it is likely that body image will also be affected.

Praxis:

Dyspraxis is the inability to carry out unfamiliar motor tasks, even though the means to do it remain intact. It may be seen in HD persons who are able to automatically carryout an action, but are unable

to do so on command (e.g., the person may be fully mobile but is unable to walk sideways on command or even on demonstration.

Spatial Orientation (form and space perception):

Dysfunction is apparent in patients who are, for example, unable to distinguish front from back, or the inside of a sweater. This problem with body image and spatial confusion makes dressing difficult, and misinterpretation of facial relationships makes eating a major problem. Also, it may be that a patient who has reasonably good motor control displays very poor ability to judge the amount of space required to fit through a gap (e.g., a doorway) or to reach or negotiate an object (e.g., sitting down in a char). The patient loses contact with his/her environment, internal state, posture and movement. In other words, there is a malfunction of the interaction between sensory input, neurological interpretation and appropriate behavioural responses.

Generally. This is seen in the latter stages of HD, but theremay be subtle indications of it early on in the disease process.

7. Behavioural Components

Depression:

Understandably, a common complaint amongst persons with HD is depression. Even if not manifest as psychotic depression, persistent feelings of despair, low energy, poor motivation, lack of alertness and general low spirits can severely affect the individual''s ability to cope with, and respond to, therapy.

Internal Regulation:

Some individuals in advanced stages of the disease suffer a breakdown in the internal mechanism concerned with regulating emotional expression. Personalities do not change as such, but there is less control over personality traits. "Difficult" personalities become even less controlled and this can cause significant patient management problems.

Interpersonal Skills

In the more advanced stages, the ability to relate effectively with others in terms of appropriatness and awareness of social norms may become disrupted (e.g., overstepping personal space boundaries or failure to make eye contact).

8.Hand Function

Generally the HD process does not affect the range of movement of the hand, but the ability to coordinate finger movement or regulate grip strength is affected. Thus, hand dexterity and manipulative skills decrease. Fine motor skills deteriorate first, and this may be evident fairly early on in the disease. Gross hand skills may remain relatively unimpaired, but if involuntary movement becomes severe then hand function also suffers.

There are instances where persons in the advanced stages of HD have developed a ''tonic grasp''. That is, where the hand is constantly held in a clenched position. Initially some voluntary release of the grasp is usually possible, but over time contractures may form and the hand becomes ''set''.

9 Visual Disorder

Eye movement controls usually unaffected in the early stages. Latter-stage persons may be unable to smoothly follow an object travelling across the visual field, or may find itdifficult to rapidly focus on an object. Controlled eye movements, independent of neck movements, may also be lost.

10. Exercise Tolerance

Poor exercise tolerance due to factors such as decreased respiratory capacity, inactivity muscle weakness and dysfunction of normal movement is evident at all stages of the disease.

PHYSIOTHERAPY TREATMENT

The broad overall aim is to maintain function and independence. For example, the ability to:

Transfer with or without assistance.

Sit safely.

Walk with minimal assistance.

Maintain some degree of hand function in order to encourage independence (e.g., dressing, toilet, feeding) for as long as possible.

Try and save themselves if or when they fall, i.e., the reinforcement of equilibrium reactions and protective

extension.

The specific aims are to:

Assess initially and at regular six-month follow-ups.

Maintain and/or improve lung function.

Teach relaxation techniques.

Maintain full range of movement and prevent long-term contractures.

Maintain/strengthen muscle groups as needed.

Reinforce and/or improve equilibrium and righting reactions.

Facilitate normal movement patterns.

Improve gait pattern — to keep patient mobile for as long as possible.

Reinforce and/or maintain perceptual skills.

Maintain/improve balance and postural control.

Improve endurance.

Assist with healing pressure areas if necessary.

Maintain some hand function for which head control, symmetry, eye/hand co-ordination, the ability to grasp and release, and sitting balance are all necessary.

General Rehabilitation Treatment Programme

The most comprehensive treatment programme is undertaken with persons in the early stage of HD. Work is done on an individual basis and in small groups which offer the opportunity for the socialisation, support and stimulation which may be lacking in the outside community where many may feel and be isolated.

1.Individual Muscle Re-education

Where necessary.

2.Mat Work

Rolling, bridging prone on elbows, four foot kneeling, crawling, kneel standing,etc.

Using compression, elastic bands, tapping, rhythmic stabilisations, resistance, etc.

During these activities, rotation, strengthening of weak muscle groups, balance, equilibrium and righting reactions, as well as maintenance of function and independence, are emphasized (e.g., teaching the patient to get up from the floor after a fall).

In the more advanced stages of HD, the emphasis is on the basic changes of body position with some passive/assisted active movements where possible.

3.Balance and Gait Training

Each patient has an individual gait pattern and one or more of a number of techniques may be used in treatment:

Sailor''s gait, canes, etc., to emphasise rotation and normal rhythm and flow.

Clasping of his/her hands together in front or behind body if involuntary movements are upsetting balance.

Practise sidewards and backwards walking for weak hip abductors/extensors— with resistance or assistance.

Compression and emphasis on a ''heavy'' (stamping) gait pattern.

Transference practice and training. Reduced rotation, perceptual problems and involuntary movements cause problems in this area.

Standing — encouragement of balance reactions and weight bearing.

Even if persons are not ambulant, they are all assisted and encouraged to bear their weightand even walk with maximum assistance. The policy at the Arthur Preston Centre is to keep all patients mobile with or without assistance for as long as possible.

4.General Activities

A normal exercise programme is often substituted with general activitiesfor variety and fun (in many ways they fulfill much the same aims as an exercise programme).

For example:

Ball games — tunnel ball, cross ball.

Work with hoops.

Table tennis (ping pong).

Snooker.

Soccer.

Obstacle course.

Outside walks.

Quoits.

5. Perceptual Training

The treatment program has been directed at attempting to improve:

Body image.

Shape recognition.

Spatial relationships.

Object negotiation.

The following methods/activities are used:

Puzzles, jigsaws.

Felt face with removable parts.

Hoops — to reinforce up, down, in, out, through, left, right. Whilst performing this task, the person is encouraged to name the part of the body he/she is using at that time.

Maze.

6. Hand Function

Specific exercises.

Activities — puzzles, stacks, etc.

Important Points in Any Treatment

Skills must be broken up into simple parts.

Repetition of exercise — with verbal reinforcement.

One-to-one supervision if necessary and possible.

Explain reasons behind activities (e.g., hoop work will help with dressing, maze will help with doorways, obstacles, etc.).

Equipment Used in General Rehabilitation Treatment Programme

Hand equipment.

Weighted cuffs and belts.

Large 120cm diameter exercise ball (for vestibular stimulation).

Octagonal roller 86cm diameter (for strengthening activities and encouraging equilibrium and righting reactions).

Large mats.

Stools — solid, height of chair seat approx. 40cms.

Obstacle course (for balance and perceptual training).

Rods, hoops.

Balls (varying weights).

Balance board.

Elastic bands.

Pulleys.

Stairs (to encourage weight transference and reciprocal leg movement).

Games and puzzles (for hand activities and perceptual training).

SPECIAL REHABILITATION PROGRAMMES

Chest Physiotherapy

Treatment

Where there is infection — postural drainage with vibrations and percussion, coughing and deep breathing. In the later stages, persons with HD are unable to cough or deep breathe on command and so vibration and percussion often help stimulate a cough.

Preventative Treatment

a.Breathing exercises — usually incorporated into the general exercise class.

b. Coughing.

c. Breathing activities:

Blowing balloon off the palm of the hand.

Blowing ping pong balls across a table — races, etc.

Blowing a tissue (held by one corner) at different distances from mouth.

Counting, vocalising during expiration.

Breath holding.

Encouragement of correct lip positioning during exercises.

General mouth and tongue exercises.

A controlled study was undertaken by physiotherapists at the Arthur Preston Centre in an attempt to evaluate the short-term effects of physiotherapy on the respiratory function of Huntington''s Disease persons. A significant improvement was noted in a number of respiratory function tests in an experimental group of five patients treated with chest physiotherapy over a two-week period. It was also shown in Huntington''s Disease persons that various respiratory functions were reduced when compared with normal values.

It was interesting to note that during this respiratory study persons with Huntington''s Disease who smoked did considerably better in the lung function tests that the non-smokers. They smoked on average 15 to 20 cigarettes a day, inhaling deeply and thus supposedly exercising their chest and lungs. However, to balance this, the only ones who showed signs of chest infection when examined medically were these smokers.

2. Oral Control

The main problems are:

Difficulty in swallowing

Dribbling

Open mouth

Lack of tongue control

Treatment includes:

Facial massage, including stretching of facial muscles (direction of stretch is toward the lips).

Voluntary holding of lips together after stretches, while encouraging nose breathing and swallowing. Correct position of head and neck is necessary as extension of the head may result in re-opening of the mouth.

Ice and swipe to sternomastoid muscles and below the cricoid cartilage, particularly before all drinks and meals.

3. Hydrotherapy

A high staff/patient ratio is necessary (one to one is preferable) for safety as much as for quality of treatment.

The main benefits of hydrotherapy are:

Relaxation.

Recreation, fun and enjoyment.

Psychological — can do more in the water than on land, can still ''swim'' and enjoy water.

Increase or maintenance or range of movement and muscle strength.

Physical contact with water and with helper.

Sensory stimulation and awareness.

A variety of exercises and activities are performed using vertical, prone and supine positions. The water properties of warmth, viscosity and buoyancy aid in relaxation, assistance and resistance during exercises.

Hydrotherapy Equipment

Rubber rings and tyres of various sizes.

Life jackets.

Balls.

Hoist.

Floaties.

Bars/rails around edge of pool.

4. Relaxation Therapy

This is a very important part of the treatment programme as it provides many obvious benefits to the Huntington''s Disease person as well as being a most enjoyable experience. Sessions are best conducted individually or in groups with a high staff/patient ratio. General physical relaxation is accompanied by rhythmical massage and, where appropriate, rhythmical passive and/or assisted techniques.

The aims of the relaxation group are to:

Provide an atmosphere which is conducive to relaxation, e.g. rugs and pillows on the floor, subdued lighting, a quiet environment without conversation and an accompaniment of slow, non-obtrusive music.

Reduce both mental and physical tension.

Reduce involuntary movements. (This becomes very obvious as the person with HD relaxes and is often able to sleep).

Improve breathing patterns — a more co-ordinated breathing pattern resulting in greater air entry with less effort (also becomes obvious as the patient relaxes).

Increase body awareness and improve self-image through the tactile sense.

Decrease involuntary movements around the face and tension around the temporomandibular joint by facial massage.

Enable the Huntington''s Disease person to gain pure pleasure through the tactile sense rather than experiencing touch only when he/she is dependent on staff for being washed, dressed, showered etc.

Gain fuller joint range through relaxation.

Gain pleasure from being a member of a group (even if active group participation is no longer possible).

Involve volunteers and staff from all areas in a positive and beneficial experience with their Huntington''s Disease residents.

Enable nursing and other staff to see how relaxation can be used at other times for the benefit of persons with Huntington''s Disease, e.g., when having difficulty sleeping or when distressed.

Enhance the sensitivity of both staff and Huntington''s Disease persons to the power of non-verbal communication.

5. Creative Movement

a. The Physical Aims are to:

Improve body awareness and achieve a realistic body image.

Improve joint flexibility, muscular strength and exercise tolerance.

Improve weight transference and balance.

Improve postural awareness.

Gain muscular relaxation.

Improve gross motor skills such as rolling, crawling and walking.

Increase physical stamina and fitness.

Improve co-ordination and control of movement.

Encourage enjoyment of physical activity.

Improve breathing patterns.

Enhance space perception.

Increase sensory input and awareness to all the senses (kinesthetic, tactile, vestibular, proprioceptive, visual and auditory).

Improve rhythm in movement.

b. The Emotional and Self-Expressive Aims are to:

Develop greater levels of self-confidence, self-image and self-esteem.

Experience greater feelings of success, accomplishment and achievement in a non-competitive atmosphere.

Become more independent and self-directing.

Assert self as an individual by taking initiative.

Have fun and enjoy participation.

Gain pleasure from expressing self through movement.

c. The Social Aims are to:

Experience greater degrees of acceptance and belonging as an individual respected by the group.

Experience being a leader or follower.

Increase the capacity to trust others.

Increase awareness of non-verbal communication through the kinesthetic sense and body language.

d. The Intellectual Aims are to:

Encourage memory.

Promote imagery skills and verbalisation by constantly reinforcing word with action.

Plan actions and make decisions.

Increase pattern recognition.

Improve understanding of spatial concepts and directions.

CREATIVE MOVEMENT TREATMENT

Creative movement sessions are conducted by a physiotherapist with expertise in the therapeutic use of movement and dance. The programme is based on the work of Rudolf Laban and his basic effort elements of space, time, force and flow, which enable the content of classes to be varied in many different ways whilst still focusing on the needs of the Huntington''s Disease person. The activities involve both the functional and expressive use of movement, and are enjoyable as well as reinforcing many of the overall aims of the physiotherapy programme.

Groups consist of 6-8 individuals with several volunteers and staff members, and are approximately one hour in length.

Example of a Creative Movement Session:

1. Warm-up activities — seated in circle on chairs or stools.

Bouncing hands over different body surfaces.

Light bouncing of different body parts.

Choosing own part to move in own way and sharing movement with group.

Accompaniment: music with strong rhythmic beat.

(ii) Exploring full use of the space around the body with image of painting the inside of a large balloon with different body parts, eg. hands, head, elbow, shoulders, back, feet. Emphasis is on smooth, flowing movements.

Accompaniment: music with smooth, flowing quality.

2. Walking around room meeting others with contact between different body parts, e.g. , hands, elbows, shoulders, hips, feet, backs.

In pairs, exploring ways of moving different body parts through a movement conversation.

Accompaniment: any bright music.

3. Using brightly coloured scarves and exploring possible ways of moving them through space.

Sharing of ideas with group.

Free exploration using suitable music to support the various possible dynamics of movement.

In pairs:

i) angry conversation between two scarves (quick, strong direct movements accompanied by tambour beaten by leader).

ii) friendly conversation between two scarves (slow, gentle, flowing movements accompanied by Indian bells played by leader).

In pairs:

Sensitive mirroring movements between scarves — one is the mirror image of the other — so that both must move slowly as one.

Accompaniment: music with slow, flowing quality.

Using hoops:

Hoops placed on floor. Curved floor pathways around hoops for spatial awareness using different forms of locomotion.

As above, but stopping with different body parts in centre of hoop at signal from tambour, e.g., left elbow and right knee; right hand and left foot; bottoms only.

Sharing hoop between two people — exploring different ways of moving the hoop and the body in relationship to the hoop.

Dance with hoops.

Accompaniment: bright music.

Mat work:

Exploring different positions for rocking body with different body parts taking weight. Music to accompany must have a rocking quality, e.g., "Morning has Broken", "Annie''s Song", "Kum ba Uah".

Sitting back-to-back with partner with whole of spine relaxed and upright — very gently rocking and swaying in different directions with gentle, slow, rocking music.

Relaxation — choose own comfortable position and while suitable background music is playing leader uses own voice, verbal instructions and mental imagery to increase awareness of relaxed diaphragmatic breathing.

Group sits on mats holding hands in a circle and gently rocks and sways with image of a piece of seaweed being gently washed over by waves.

Feedback from participants as to what they enjoyed, would like to explore further, how they felt during the session, etc.

Creative Movement Equipment

Cassette tape player and library of tapes with music of various dynamic qualities.

Percussion instruments: tambour, tambourine, cymbal, bells, etc.

Personal property — extremely important for providing the individual with a focus and extension beyond the body itself; for tactile and visual stimulation; and to provide a link with others.

Coloured scarves.

Material of different sizes, colours and textures.

Ribbons and ribbon sticks.

Hoops.

Elastic.

Large woollen pom-pom.

Peacock feathers.

Expendable items, such as flowers, balloons, paper streamers, bubble mixture, etc.