

The role of the physiotherapist in Huntington's disease

People with Huntington's disease (HD) present with a variety of physical symptoms owing to the complex degeneration that occurs in the basal ganglia, the centre within the brain for controlling voluntary movement and establishing postures. Physiotherapists can help people with HD at various stages of the disease with certain aspects of these physical changes and are essential members of the multidisciplinary team (MDT).

In the Harper Huntington's disease unit at St Andrews Hospital there are people at both ends of the physical spectrum, some who are independently mobile whilst others who are dependent on a wheel chair. This provides a great challenge for staff to cater for people with such a broad range of physical disability. I propose to outline some of the physiotherapy interventions that are adopted in the early, middle and late stages of the disease. Due to the nature of the progression of the disease physiotherapy intervention will need to be constantly reevaluated. I have for the purposes of this article discussed physiotherapy in isolation but in reality MDT work is essential and physiotherapy is only one important but often forgotten cog in the wheel.

Physiotherapy in the earlier stages

It is important to encourage people with HD to remain as active as possible, maintaining cardiovascular fitness should be encouraged, this can involve a range of activities from outdoor walks to more challenging activities such as Tai Chi, which is exceptionally good at challenging balance. In this stage people with HD are dealing with the onset of their symptoms, it can be very stressful and depression is not uncommon. There is a vast amount of literature advocating exercise, as a treatment of depression so keeping people active is an important first port of call. There may well be physical limitations, which restrict the person partaking in some generic exercise classes. The physiotherapist can assess appropriateness of different forms of exercise and make suitable recommendations.

At a first meeting the physiotherapist should complete a full assessment; particular attention should be drawn to any muscle weakness, muscle shortening, balance deficits and analysis of gait. It is not uncommon for the postural neck and trunk muscles to be weakened in the earlier stages; these people tend to walk lurching forwards at the hips. Subtle changes in posture can occur, the physiotherapist should pick up on this and postural re-education can help in the long run. Any specific alterations in musculature, balance and gait identified should benefit from a specific exercise regime, which will be developed to minimise the impact of these factors.

The physiotherapist can teach the person deep breathing exercises; this is useful for relaxation purposes but also to help maintain lung capacity. A lung function test can be completed, the result being used as a baseline and as a useful reference point for later stages of the disease when lung functioning may become impaired.

Fine hand movements and gripping can begin to become more difficult, exercises to prevent this should form part of the treatment plan. Simple strategies such as taking part in cards and board games can be enjoyable and challenge these skills and hopefully maintain hand function.

The middle stages

With the progression of HD the movements typically worsen (in particular chorea), this can make walking increasingly difficult and so falls become more of an occurrence. Falling is a concern for both the person with HD and the carer. In one sense it would be easier when falls start to occur to just rely on a wheel chair for transport. However it is important that person remains active and 'use it or lose it'. People with HD are ingenious in developing their own gait patterns as the disease progresses; it is important however that person is mobilising in a safe as possible manner. The physiotherapist can identify possible specific physical causes of falls such as poor balance, muscle weakness or shortening and devise specific exercises to help combat this. Walking aids can be used for those people whose movements and handgrip allows them to do so. To help minimise the impact of any falls people can wear protective padding on vulnerable bony prominences including hip protectors, knee and elbow pads and reinforced hats.

The person will spend an increasing amount of time sitting down, therefore the seating should be considered carefully. It is important the seat provides sufficient support so the patient can sit upright, is comfortable and in a functional position. The chair should not provide too much support otherwise the patient will rely on the contours of the chair to remain upright and not use the trunk stabilising muscles. Poor postural habits for long periods of time will result in negative soft tissue changes occurring (Two common occurrences are shortening of hamstrings or the feet not resting flat on the floor and turning inwards). The person should be able to support his or her own body in the chair. Rounded shoulders and slouching will impair chest expansion and therefore lung functioning. It is important the seat provides some support but still ensures the patients are utilising their postural stability muscles in the trunk. The person's trunk stabilising muscles will benefit from a daily period of unsupported seating.

Transfer practice should be ongoing. People with HD have decreased awareness of their body position, decreased spatial awareness and are often impulsive. This can result in rather precarious transfers. Practicing completing safe transfers should be extended from the physiotherapy session into the daily routine with particular emphasis on slowing them down!

People with HD cardiovascular fitness will inevitably decline during this stage; activity should continue to be encouraged as much as possible but always ensuring the patient is safe.

Hand exercises should continue to help maintain fine motor coordination. Many people have great difficulty with fine hand movements in the middle and later stages, this can be particularly problematic to those who smoke and can be distressing for both patients and carers alike. Burns can then become an issue.

Exercises to activate the small muscles in the feet should form part of the treatment plan, as these are essential in maintaining balance.

Later stages

In the later stages the person may become dependent on a wheel chair and posture and seating becomes the main focus of physiotherapy input. A daily posture plan should be developed; this should involve the person, carers, nursing staff, speech and language therapist (SALT) and occupational therapy staff (OT). The emphasis should be on

comfort, maximising functional ability and to minimise secondary complications such as muscle contractures, pressure sores, chest infections etc. It is a good idea to develop simple photographic illustrations of the different postures a patient may adopt in the day. Feedback from the person, family and carers and the people who know the patient best is essential. Specialist wheelchairs will be required as people with HD have great difficulty fitting into standard prototype wheel chairs. Sleeping posture needs to be considered but is often forgotten. The person with HD will spend an increasingly large amount of time in bed; poor postural habits are often acquired in the bed undoing the good postural work achieved in the daytime.

Any contractions that may have developed should be stretched regularly to try and reverse this and prevent further development. Muscle contractions can become painful and make personal hygiene tasks become difficult. The physiotherapist can help with muscle rigidity commonly experienced in the later stages. Hydrotherapy is a good tool to help the patients muscles relax so that more effective stretching may then take place. Standing and walking practice can take place in the pool when the support provided by the buoyancy of the water makes this easier.

For those people still able to weight bear safely independent or assisted wheelchair transfers should be continued, this can provide an important feeling of some independence. If the person is able to stand with staff they should do so regularly throughout the day, this will help maintain muscle strength, muscle length, bone density, assist with bladder and bowel functioning and decrease the risk of pressure sores. If the person is able to only partially weight bear then standing can take place in a standing frame or walking hoist.

In the later stages the physiotherapist will have an increasingly important role in monitoring the chest status as the person with HD may become more susceptible to chest infections. The physiotherapist should be working closely with the SALT to develop ideal posture at meal times to minimise the risk of choking and aspiration of the fluid into the lungs and ultimately chest infections. The physiotherapist can monitor chest status whilst the SALT can consider the swallowing.

Conclusion

People with HD will experience many physical changes; a physiotherapist is an essential member of the MDT to help manage this aspect of the triad of symptoms. Specialised physiotherapy input is essential at each stage of the disease as it can help to maintain optimum physical function and minimise the complications during the progression of the disease.

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