Huntington's Disease Service Model Requirement

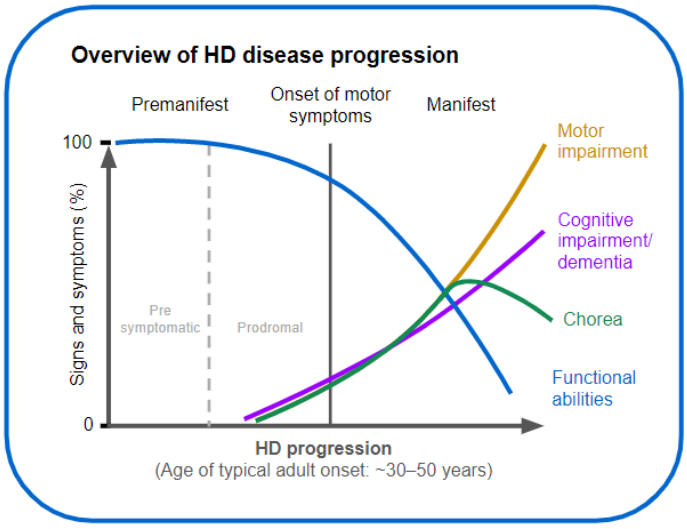
Population Needs

Huntington's disease (HD) is a rare, inherited, neurodegenerative disease, with onset usually in mid-adult life. HD progressively affects a person's everyday functions with particular impact on movement, thinking, personality and behaviour HD has a devastating impact across generations, due to the inherited nature of the disease but also due to the effect on family members and carers living with affected individuals. Whilst there are medications and interventions that can help control some of the effects of the disease, there is currently no way to prevent, slow or stop disease progression. HD is ultimately fatal and progresses relentlessly from appearance of symptoms to loss of functional capacity and disability, with a median survival of 15 years from onset of motor symptoms.

For every parent with HD, there is a 50% chance for each of their children of inheriting the condition. HD can show genetic anticipation with an earlier age of onset of symptoms in subsequent generations. This leads to significant worry and stress for those with HD, and their relatives who are seeing the effect HD has on their loved one and deciding whether to test for the gene themselves.

HD symptoms typically start between age 30- to 50-year-old, with prodromal (pre-diagnosis) subtle changes in thinking, mood and behaviour. The early stage of the disease (current point of clinical diagnosis) commonly presents with chorea (jerky involuntary movements throughout the body) which may start to interfere with balance, walking and eating. It can become difficult to plan and carry out day-to-day home and work tasks.

This then progresses to the moderate stage of the disease, with affected individuals experiencing increasing difficulty in carrying out day-to-day tasks and progressive motor and mental health symptoms. In the advanced stages and end of life care, patients will require 24/7 support, usually from 24 hour care or from a professional care home. Basic abilities, such as swallowing and maintaining body temperature, become increasingly difficult.



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While research is ongoing, there are currently no approved therapies that can slow, stop or reverse disease progression in HD. The treatments currently available focus on providing symptom relief, maximising function and optimising quality of life. Multidisciplinary management is key and symptomatic management is beneficial for many of the features in line with International Guidance (Frontiers International Guidelines for the Treatment of Huntington's Disease 2019[1])

National Context

NHS Long Term Plan (January 2019)[2]

As medicine advances, health needs change and society develops, so the NHS has to continually move forward so that in 10 years' time we have a service fit for the future. The NHS Long Term Plan will do just that. Drawn up by those who know the NHS best – frontline health and care staff, patients and their families and other experts – the Long Term Plan is ambitious but realistic.

The plan includes:

- Moving to a new service model in which patients get more options, better support, and properly joined-up care at the right time in the optimal care setting
- Setting out new, funded, action the NHS will take to strengthen its contribution to prevention and health inequalities

UK Strategy for Rare Diseases[3]

In the UK there are many examples of excellent and world-class services for people with rare diseases. However there needs to be further improvement across the whole 'patient journey'; the patient's experience from their first contact with the NHS through to reaching a diagnosis and onto managing their condition and on-going care for example empowering those affected by rarer disease and co-ordination of care.

Local Context

A study undertaken nationally which found that the prevalence per 100,000 population in the South East was 10.9 (6.8 to 16.4), which in Surrey would translate to approximately 131 patients with this condition, but this could be a low as 82 or as high as 197. Approximately 5 times this number are at risk of developing HD.

Care for HD should begin prior to the onset of symptoms in those 'at risk' and spans primary care, clinical genetics, community mental health teams, neurology, neuropsychiatry and social care networks. These health services are commissioned by both CCG's and NHS Specialist Commissioning but support includes multi partnership models within Primary Care services and Adult Community Services. A key element of this model includes voluntary sector support and in particular the Huntington's Disease Association.

[1] Frontiers | International Guidelines for the Treatment of Huntington's Disease | Neurology (frontiersin.org)
[2] NHS Long Term Plan v1.2 August 2019
[3] UK_Strategy_for_Rare_Diseases.pdf (publishing.service.gov.uk)

Due to the nature of HD and the combination of neurological features, cognitive features and neuropsychiatric features, affected individuals and their families need to navigate a complex combination of health care professionals and are at risk of falling between the gaps in commissioned services.

Outcomes

The vision for the NHS in England is to secure better outcomes for patients as defined by the five domains of the NHS Outcomes Framework and uphold the rights and pledges in the NHS Constitution. Surrey Heartlands CCG will deliver the outcomes for its population in conjunction with a range of stakeholders from the health economy as defined through the quality, performance and financial metrics in:

Domains	Overarching outcomes	
1. Preventing people from dying prematurely *This domain captures how successful the NHS is in reducing the number of avoidable deaths*	● Each patient living with HD is referred to appropriate mental health services for supporting and monitoring ● Each patient living with HD has the appropriate support provided around nutrition, hydration and weight monitoring	✓
2. Enhancing quality of life for people with long-term conditions *This domain captures how successful the NHS is in supporting people with long-term conditions to live as normal a life as possible*	● Each patient's care is managed through an MDT ● Each patient with HD has an individualised assessment, agreed person-centred goals and care plan goals achieved ● Individual care plans are reassessed at regular intervals to take into account the ever changing needs of the individual ● Demonstrate improvement in health outcomes for people by supporting the patients, families and carers to manage their long term condition in the community ● Enable individuals to be supported and receive as much care as possible closer to home ● Training accessible to HCPs to support HD patients with complex needs	✓
3. Helping people to recover from episodes of ill-health or following injury *This domain captures how people recover from ill-health or injury and wherever possible how can it be prevented*	● There is equality in access to residential and nursing home care/respite care with staff appropriately trained to manage patients with HD ● Reduction in hospital admissions, through improved clinical management and supported self-care, however it is important to note due to the nature of HD symptoms (cognitive challenges / unawareness) self care can be challenging	✓

4. Ensuring people have a positive experience of care *This domain looks at the importance of providing a positive experience of care for patients, service users and carers*	• Each patient with HD has a named care-coordinator to support the individual, their family and carers to understand their healthcare needs and coordinate care • Ensuring patients experience seamless care between all healthcare service providers and are signposted to appropriate social care or other support • There is access to psychological support for HD patients and their carers across all stages of the disease • Carers and family members are supported to actively participate in decision making, and care planning for the person they care for • Work with commissioners to refine the model of care and service delivery, utilising evolving best practice	✓
5. Treating and caring for people in safe environment and protecting them from avoidable harm *This domain explores safety and its importance in terms of quality of care to deliver better health outcomes*	• Patients will be actively monitored to improve therapeutic intervention in line with national guidance to improve quality of life • Support for patients and carers with advance care planning conversations to allow considerations to be made for suitable care	✓

Scope

Aims

The aim of the service is to provide HD patients, family members and carers with co-ordinated multidisciplinary (MDT) holistic care as they progress along the stages of this disease.

Objectives

HD is a complex condition that requires a “Care Coordinator” role (please see Appendix A HD Care Coordinator job description) be established to facilitate the MDT approach of care provision to address the constantly changing needs of patients and family members living with this degenerative disease.

The service will deliver these aims by establishing defined models of care with robust shared/network care arrangements where appropriate.

Service description/care pathway

This disease is complex and progressive so the patients’ needs are very individual and will require continual assessment as the disease progresses. Ideally this care should be provided by a multidisciplinary team which includes doctors and specialist nurses from psychiatry, clinical genetics and neurology, physiotherapists, occupational therapists, dietitians, speech and language therapists and dentists. A long-standing problem for those affected by HD is that their disease crosses the boundaries of medical, psychiatric and social problems. This complex care should be supported by a care coordinator who can help and support the patient and careers to navigate the care they need.

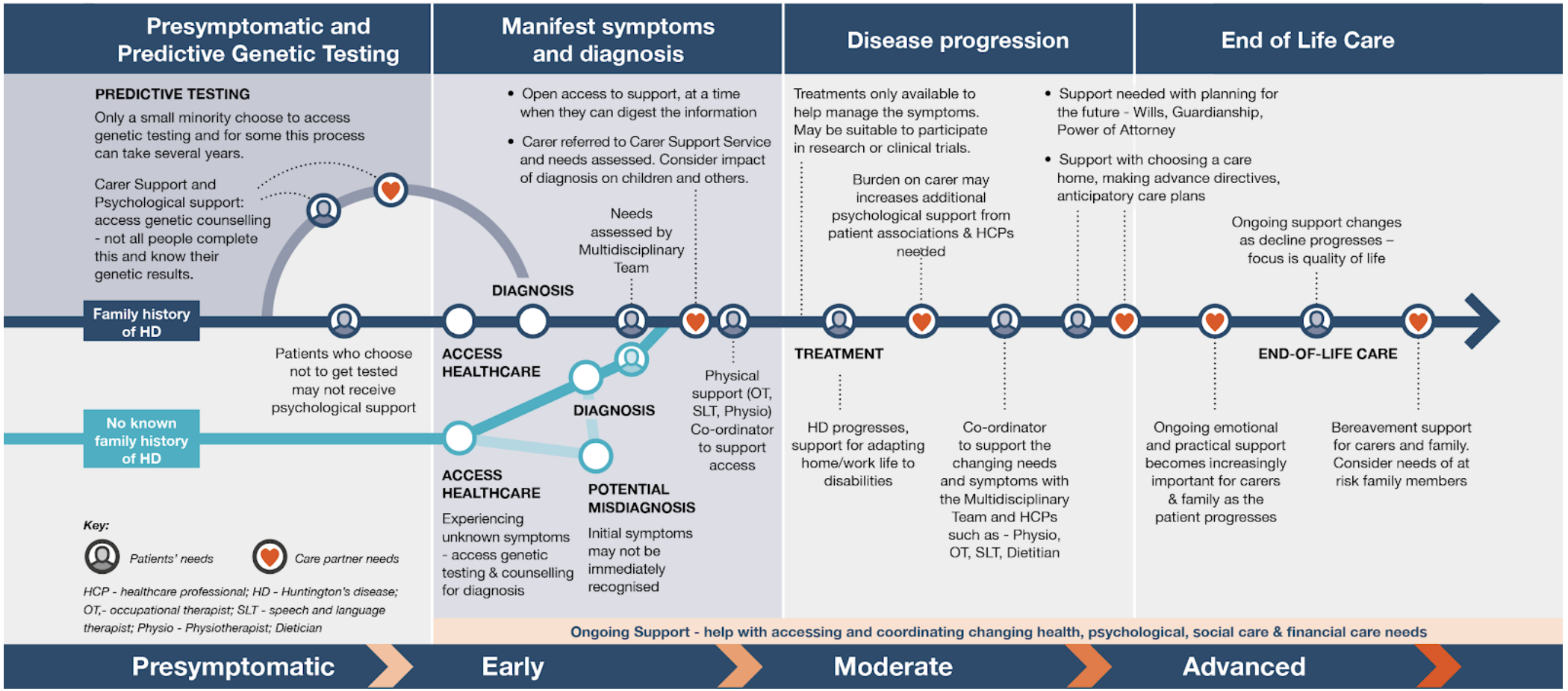
HD is a family disease. Indeed, the impact of HD goes beyond the immediate symptoms experienced by the person who is ill. It affects the whole family: the carer and the person living at risk, the person in receipt of an unfavourable test result as well as the symptomatic patient and those with a favourable test result. HD is a complex disease and requires a multidisciplinary approach, involving a range of services that are required at each, differing stage of a person’s life with the disease. These other specialties include neurology, genetics, psychiatry, neuropsychiatry, neuropsychology and specialists in movement disorders.

The diagram below describes the HD Care Pathway across the four disease stages and the support required during these stages for patients and carers.

Presymptomatic	Those with family history must live with knowledge of the condition, possibly whilst caring for a family member. They may consider genetic testing and family planning options and require support through these decisions There may be subtle changes in thinking, mood and behaviour.
Early	Point of clinical diagnosis Individuals may not have insight into their symptoms at this stage of the disease and many are able to continue working and functioning normally in the early stages with minimal support. However, there are usually changes in

	thinking, mood and behaviour. chorea (jerky involuntary movements throughout the body) may start to interfere with walking and swallow. It can become difficult to carry out day to-day home and work tasks.
Moderate	Increasing difficulty carrying out day-to-day tasks. Progressing symptoms eg. **Motor** – balance, chorea, speech, fatigue, incontinence, swallowing, weight loss **Mental Health**- depression, suicidal thoughts, anxiety, irritability, lack of motivation and **Cognitive** - concentration, memory, reduced speed of processing
Advanced	Full time support is often needed in a professional care home. Basic abilities, such as communicating, swallowing and maintaining body temperature, become increasingly difficult. Palliative Care input is required.

Huntington's Disease Care Pathway



March 2022
M-GB-00006729

Provision of Clinical Advice & Support

One of the primary functions of the service is to provide specialist clinical advice and support to health and care professionals in both primary care and the wider out of hospital setting. This advice can take the form of telephone or email contact with other professionals in line.

The HD service will encompass a specialist HD multidisciplinary clinic based at a tertiary neuroscience centre (i.e St. George's) with development of regional HD MDT meetings via teams for complex case discussion, outreach and training.

Requests for advice should be easily accessible through a single email address and telephone number. Advising professionals should have access to the patient's care plan held by the GP and/or relevant locality, ideally through direct interoperability of clinical systems.

Advice and recommended interventions should be captured as part of the care plan and communicated to the GP. It is anticipated that the majority of requests for advice will originate from primary care and core community service professionals, however advice may also be requested from other care settings e.g. care homes, hospices, mental health teams.

All requests to the service should be prioritised and addressed in accordance with clinical need. Resources should be deployed appropriately, and a preventative approach taken where possible. This should be done in co-ordination with services across health and social care.

Promotion and support self-care

The service provides self-management and empowerment of the patient to self-care through promotion and education. However it is important to note due to the nature of HD symptoms (cognitive challenges / unawareness) self care can be challenging.

Carer support

The service will provide support to carers to include:

- Ensuring carer assessments are offered
- Provide information to carers about local respite services for carers (such as lunch clubs, day centres etc) and ensure this information is cascaded to GP surgeries, Huntington's Disease Association etc
- Establishing a local carer support group/network

Information provided to patients and carers

Patients will receive a personalised care plan. Those affected by Huntington's Disease, including carers will also be given information about their condition and any relevant services that may be of benefit to them.

Additional Services

The service provider should also undertake research programmes, including the conduct of clinical trials and to establish and participate in suitable regional and national programmes of clinical audit.

Signposting arrangements between services

This service refers to others such as social services, housing, health services, mental health services and charitable organisations as and when appropriate.

Population covered

The service is accessible to service users who are either registered with a GP practice within Surrey Heartlands CCG or usually resident in the area covered by Surrey Heartlands CCG but not registered with any GP, subject to Responsible Commissioner guidance.

Referrals are accepted for patients that are over 18 years of age and with a familial history of HD, or with a confirmed diagnosis of HD by a Consultant. Those individuals who have juvenile HD will be excluded from this service as they will be cared for by specialist paediatric services in a national tertiary hospital.

Interdependence with other services/providers

This disease is complex and progressive so the patients' needs are very individual and will require continual assessment as the disease progresses. As previously outlined there are a number of general services with varying levels of interdependency with HD patients, family members and carers:

- Clinical Genetics – SW Thames Centre for Genomics at St. George's Hospital with local outreach clinics in Surrey Hospitals
- Consultant Neurologists at Surrey Hospitals
- Consultant Neurologists at St George's Hospital
- Clinical Nurse Specialists (Community & Hospital based)
- General Practitioners
- Medicines Management Teams (Community & Hospital based)
- Physiotherapists (Community & Hospital based)

- Occupational Therapists (Community & Hospital based)
- Speech & Language Therapists (Community & Hospital based)
- Dieticians (Community & Hospital based)
- Psychiatrists (Community & Hospital based)
- Psychologists (Community and Hospital based)
- Palliative Care team (Community & Hospital based)
- Commissioners (Local Authority & CCG)
- Continuing Health Care Contract Manager
- Charitable Organisations/Voluntary Sector (e.g. Huntington' Disease Association, Crossroads, Brigitta Trust)