

## OVERVIEW

Huntington's Disease (HD) is a neurodegenerative condition that progresses from the very earliest stages over a period of 10-30 years. Onset is most commonly in mid-life, but may commence at any stage of life, and starts insidiously. It results in progressive loss of mobility and is also associated with progressive impairment of swallowing and speech, with slurring early in the disease progressing to difficulty with speech initiation and word-finding. By late stage disease, most patients have severely impaired communication or are mute. Subtle intellectual deterioration can be seen for years prior to motor onset in many individuals, and these symptoms deteriorate as the condition progresses.

HD is accompanied by personality change, which can include loss of empathy, apathy, impulsivity, perseveration, depression and psychosis. Generally, patients in the late stage of the disease need help with most activities of daily living, are mute and may require PEG feeding.

There are approximately 9000 affected people in the UK. Mean annual costs of care per person early stage are approx. £2250, rising to £89 760 per year in the later stages. The average cost across all disease stages is approx. £21 605 per year. This equates to £195 million per year in the UK<sup>1</sup>.

## THE CLINICAL SERVICE IN SOUTH WALES

The South Wales Neurology services, provided through Cardiff & Vale UHB, includes as part of its service delivery a regional Huntington's Disease Research and Management Clinic. This service has been heavily underpinned since 2003 through strong links with Cardiff University who currently provide all staff for the service apart from a part-time clinical nurse and a part-time admin assistant. In this form, it has been at the forefront of both research and service delivery to the South Wales population, avoiding numerous and costly hospital readmissions and facilitating ongoing care in the community. The service offers an essential lifeline to patients from across South Wales in increasing numbers (Table 1, below) and has represented cost-effectiveness for many years for patients who would have potentially not had access to the correct expertise in a timely manner.

*Table 1: Clinic attendance figures over the last five years (April to April)*

	2013/14	2014/15	2015/16	2016/7	2017/8
Cardiff & Vale	18	35	46	58	62
Aneurin Bevan	27	58	62	55	55
Cwm Taf	18	24	28	22	27
ABMU	20	35	57	64	48
Hywel Dda	10	14	21	24	21
Powys	2	3	6	6	4
<b>Total</b>	<b>95</b>	<b>169</b>	<b>220</b>	<b>229</b>	<b>218</b>

As a service that has its heart within the community, it has developed increasing patient numbers without a corresponding increase in resources. The current patient volume is well over four times that experienced in most other HD centres across the UK and has reached a point where the service can no longer continue to provide a quality service in the manner that it currently operates.

<sup>1</sup> Jones, C., Busse, M., Quinn, L., Dawes, H., Drew, C., Kelson, M., Hood, K., Rosser, A. & Edwards, R. (2016) "[The societal cost of Huntington's disease: are we underestimating the burden?](#)", *European Journal of Neurology*, vol. 23, no. 10, pp. 1588-1590.

## THE SCOTTISH MODEL

In 2016, the Scottish Government and the Scottish Huntington's Association (SHA) developed a National Care Framework for Huntington's Disease<sup>2</sup> to help ensure families affected by the condition are given the best possible care, information and support regardless of where they live in Scotland. The Framework does not advocate a "single pathway" or "one-size-fits-all" approach; instead, individual care and support packages are created in partnership with families. This requires four key elements:

- 1) Care coordination should be provided by a single named specialist as determined by each Health & Social Care Partnership<sup>3</sup> (ideally, but not necessarily, an HD Specialist) who has a key role in assisting families to navigate their way through their HD journey;
- 2) Care should be provided by a clearly defined multi-disciplinary team consisting of core members whose roles are essential in managing health and social care needs;
- 3) Access is required to a wider and clearly mapped network of services where well-developed referral and liaison arrangements are in place;
- 4) Specialist staff play a central role in providing training and education to the wider support network. Each HSCP area should have arrangements to deliver training, or work with other HSCTPs to deliver training, appropriate to its own workforce.

It is currently too early to assess the impact of the Scottish Framework, although anecdotally it has improved provision by Health Boards.

There is no such integrated model in Wales despite the Welsh Rare Diseases Implementation Plan<sup>4</sup> mandating the highest standard of care for everyone with a rare disease.

## RECOMMENDATIONS

- The Health, Social Care & Sport Committee should hold an inquiry into services and support available to Huntington's Disease patients in Wales, with an aspiration that this should lead to a wider, more sustainable co-ordinated model of care for Huntington's Disease.
- Wales should consider developing a National Care Framework for Huntington's Disease similar to that developed in Scotland in 2016. The Scottish Framework seeks to ensure that the care and support provided to individuals and their families living with Huntington's disease takes account of their specific health & social care needs throughout their experience.

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<sup>2</sup> The full Framework can be seen at [care.hdscotland.org](http://care.hdscotland.org)

<sup>3</sup> Health and Social Care Partnerships, (HSCPs) are the organisations formed as part of the integration of services provided by Health Boards and Councils in Scotland. Each partnership is jointly run by the NHS and local authority.

<sup>4</sup> Welsh Government (2017) [Welsh Rare Diseases Implementation Plan: highest standard of care for everyone with a rare disease](#), Wales: Welsh Government / NHS Wales / Welsh Rare Disease Implementation Group.