

# Statewide and Mental Health Services

## Huntington's Disease Service Model of Care



## Table of Contents

<b>Introduction and Purpose</b> .....	4
<b>Health Policy Context</b> .....	5
Relevant National Policy and Priorities.....	5
National Service Principles .....	5
<b>State and Service Directions</b> .....	5
Relevant State and/or Service Policy and Priorities .....	5
Recommendations from relevant Review/Inquiries .....	5
<b>Relevant Legislative Framework</b> .....	5
<b>Current Context</b> .....	6
Definition of the Service/Population Group.....	6
Prevalence Rates .....	6
Scope of Current Care Provided .....	6
<b>Future Service Directions</b> .....	7
Relevant Service Evidence and Research Findings .....	7
Future Goals and Targets .....	7
<b>Components</b> .....	8
Philosophies and Approaches Underpinning Care.....	8
Customer Focused.....	8
Family Friendly Practices .....	8
Access .....	8
Issues of Confidentiality.....	8
Early Engagement .....	9
<b>Target Groups</b> .....	11
Case Management.....	11
Predictive Testing .....	11
<b>Service Description</b> .....	12
Location .....	12
Hours of Operation .....	12
Staffing Profile.....	12
Linkages with other Services .....	12

<b>Types of Services Delivered</b> .....	<b>13</b>
Roles and Function of the Case Manager.....	13
Case Management Core Functions and Clinical Activities .....	13
Areas of Activity .....	14
Roles and Function of Predictive Genetic Testing.....	14
To Constantly Monitor and Review Services to Ensure Continuous Improvement By:.....	15
Triage/Intake by HDS.....	15
Assertive Case Management.....	16
Individual Service Plan (ISP) .....	16
Consultation and Liaison .....	16
Performance and Outcome Monitoring.....	16
Client Registration .....	16
 <b>Client Service Pathway</b> .....	 <b>17</b>
Entry to Service .....	17
Assessment.....	17
Ongoing Care/Shared Care .....	17
Clinical Review .....	17
Separation from Service .....	17
 <b>Service Evaluation</b> .....	 <b>18</b>
Client Outcome Measurement.....	18

## **Introduction and Purpose**

A review of Huntington's Disease Service in Tasmania was carried out in September 2007. One of the recommendations from the review related to the development of a model of care for Huntington's Disease Service (HDS) which would provide the framework for a Tasmanian-relevant pathway for Huntington's Disease outreach and Case Management services.

A working group involving all HDS staff was convened to build on the extensive work already done by HDS staff to develop the Model of Care. This draft Model of Care is the outcome of this working group.

Draft for Consultation

## Health Policy Context

### National Directions

#### Relevant National Policy and Priorities

- International Guidelines: Standards of Care for Huntington's disease. Standards of Care Working Group: European Huntington's Disease Network (EHDN) Plenary Meeting Prague 2010
- International Guidelines: Guidelines for the molecular genetics predictive test in Huntington's Disease 1994
- National Mental Health Standards 2011
- Australian Council of Health Standards Accreditation

#### National Service Principles

- Australian Law Reform Committee: The Protection of Human Genetic Information in Australia, Essentially Yours Report, Volume I Report 96, March 2003

## State and Service Directions

#### Relevant State and/or Service Policy and Priorities:

- Tasmanian Health Plan 2009
- Mental Health Services Strategic Plan 2006-2011

#### Recommendations from relevant Review/Inquiries:

- Huntington's Disease Service Review Report, Part I  
Prepared for the Department of Health and Human Services September 2007.

## Relevant Legislative Framework

- *Anti-Discrimination Act 1998*
- *Disability Discrimination Act 1992*
- *Human Tissues Act 1985*
- *Right to Information Act 2009*
- *Personal Information and Protection Act 2004*
- *Federal Privacy Legislation Amendment Act 2006*

## Current Context

### Definition of the Service/Population Group

Huntington's Disease (HD) is an inherited neurological disorder that is inherited in an autosomal dominant manner such that each child of an affected parent has a 50-50 chance of having inherited the HD gene. Men and women are equally likely to be affected. Penetrance, the likelihood of showing symptoms of the disease if the associated genetic mutation is present, is virtually 100 per cent. The characteristic symptoms of HD, (abnormal movements, intellectual deterioration, emotional instability and some psychiatric disturbances, most notably depression), usually begin around the age of 40 although the disease onset has been seen in children as young as two and adults as old as 80. Death usually occurs between 15 and 20 years after onset.

At the present time, treatment for HD is entirely symptomatic, with no treatment or cure that affects or slows the underlying disease process. Research, especially since the discovery of the HD gene, is progressing.

### Prevalence Rates

The most recent prevalence study of Huntington's Disease in Tasmania was conducted in 1990 by Professor Saxby Pridmore and published in the Medical Journal of Australia Vol. 153 August 6, 1990.

The result indicated that as of January 1990, the prevalence of HD in Tasmania was 12.1 per 100 000. Professor Saxby Pridmore indicated that there may be some under-reporting.

It is noted that the report completed in 1990 is significantly dated and does not appear to reflect the current population. Current service provision suggests prevalence may be approximately 14 + per 100 000.

### Scope of the Current Care Provided

#### Resource Allocation – Physical, Human, Financial

- North West: 0.5 Case Management/20 hours fortnight
- North: 0.8 Case Management
- North/NorthWest: 0.2 Predictive Testing
- South: 1.0 Case Management and Predictive Testing

## Future Service Directions

### Relevant Service Evidence and Research Findings

- Draft Standards of Care for Huntington's which should inform practice over time.
- A Physician's Guide to the Management of Huntington's Disease, 3<sup>rd</sup> ed 2012
- Lifting the Veil of Huntington's Disease Recommendations to the Field from the Huntington's Disease Peer Workgroup.
- Essentially Yours Report, The Protection of Human Genetic Information in Australia, Volume I Report 96, March 2003
- Huntington's Disease Third Edition, Edited by Gillian Bates, Peter Harper, Lesley Jones, Oxford Monographs on Medical Genetics 45
- Caring for Persons with Huntington's disease; A handbook for Health Care Professional, Second Edition, Edmond Chiu, A.M.
- European Huntington's Disease Network – Working on Standards of Care Draft, Draft Care Pathway, January 2006
- Case management should be consistently applied in line with the Case Management Society of Australia's 'National Standards of Practice for Case Management.

### Future Goals and Targets

- Development of a Huntington's Disease Resource Manual.
- Updated Huntington's Disease Prevalence Study.
- Statewide Huntington's Tasmania liaison meeting with Mental Health Services.
- Review and monitor opportunities for carers/significant others to become involved in the HDS.
- Review and monitor the opportunities for group programs as part of the HDS.

## Components

### **Philosophies and Approaches Underpinning Care**

The Huntington's Disease Service (HDS) is committed to promoting and supporting the health and well-being of people from families in which Huntington's Disease is known to exist. It is a specialist service that works collaboratively with other services in the extension of comprehensive treatment and care. The aim of the services is to provide the best quality of clinical service and support to enhance quality of life to families affected by this chronic degenerative genetic disease.

The HDS work in conjunction with Huntington's Tasmania.

### **Customer Focused**

The HDS enables choice and flexibility, offering the maximum choice possible in the Tasmanian context for individual clients and their family members including out of area referrals in accordance with National Mental Health Standards. The service recognises that not all people want or need the same things and is able to respond to people's changing needs over time.

### **Family Friendly Practices**

The service is family and social networks focused, recognising that family members are impacted by the illness and are potential clients of the service, along with the individual with Huntington's Disease.

### **Access**

The service simplifies access to the health system for clients, provides for continuity and integration of care for clients and delivers services equitably statewide. Throughout the client journey, clients at different times will come and go from the HDS. To make this as easy as possible, referrals to the service can occur directly through to the HDS. Referrals will be accepted from clients, their carers/significant others, GP's or other services involved in the clients care.

### **Issues of Confidentiality**

HD is a hereditary or genetic disease. Children of a parent with HD have a 50 per cent chance of inheritance. There is no cure or treatment for HD at this time.

In this context, issues of confidentiality for clients of the HD Service, including storage of their information and data, have particular sensitivities and risks for individuals and their relatives. Information about a person's health and future health and that of their family can be correctly or incorrectly inferred by the person having contact with the HD Service or from family history. This can potentially lead to discrimination, stigmatisation and harm to the families involved. Consideration needs to be given to confidentiality for the individual person within the family context.



Discrimination can take many forms, for example social, financial, employment, and insurance, and can be perpetrated by families, friends, employers, institutions, doctors, health services and in public sector settings. Discrimination can harm a person's psychosocial wellbeing, their life opportunities, employment or commercial opportunities. It can occur incorrectly when assumptions are made about a family where the future health of any individual may not include HD. Fear of discrimination may lead people not to make contact with the services they require. Genetic discrimination can lead to a genetic underclass existing within our community.

Taking a predictive genetic test can be useful to an individual for a range of reasons, however it carries high risk of discrimination if an individual is found to be gene positive and that information is misused. Test results are therefore given to the individual and only to a third party with client consent.

A number of papers over the last decade have expressed concern about the emergence of genetic discrimination. Two recent papers (1,2) have highlighted the reality of genetic discrimination for individuals and their relatives. Researchers indicate that up to 40 per cent of individuals with a genetic risk have experienced discrimination (1).

With newly emerging genetic technology, many countries including Australia are moving to legislate special protection for the genetic information of individuals and their families and to update Health Policies to respect the particular sensitivities of a person's genetic makeup. In 2002 the Australian Government began this process, launching a major national enquiry into the protection of human genetic information. This has resulted in the March 2003 report *Essentially Yours; The Protection of Human Genetic Information in Australia*. National Health and Medical Research Council, Australian Health Ethics Committee.

These ethical, personal and legal concerns underpin the provision of services by the HDS. Factors that must be considered include the location of the service, how clients access the service and the storage of data and information needed to meet the particular sensitivities of genetic disease.

Because HD is a genetic disease, it is a family disease. The effect on the entire family can be profound. A single diagnosis has implications for immediate and extended family members. Some families are very open and share information across generations. Other families do not share information or knowledge and strictly adhere to individual privacy or autonomy. This familial nature of HD and its inherent complexities need to be given special ethical attention by the HDS.

Particular attention needs to be paid to the storage of information, awareness about the need to keep individual family members information/appointments separate. Also the awareness of the rights of the individual "not to know" need to be taken into consideration.

	<p>Although all members of a family will not get HD, almost all are affected emotionally, socially, financially and by stigma. Some symptoms of HD, particularly denial, mood swings, irritability, sleep disturbance and apparent changes to personality can make life difficult for all family members.</p> <p>Some family members will be part of caring for affected members of the family and the caring role can be complex. It can lead to social isolation, changed roles for individuals within the family, including the affected individuals role and can result in a profound impact on the carer's and the individual's emotional well-being. Depression and incidences of suicide have been shown to be as high in HD families as in any identifiable group. Some families separate under the strain of living with HD which can result in further isolation for family members.</p> <p>There is a profound impact on a family as a result of a genetic disease like HD, because of the severe and progressively disabling nature of the condition. There is symptomatic treatment but no known cure, and limited health professional knowledge of this rare condition. The provision of timely, knowledgeable, professional services is critical. Developing a relationship with a family at an early stage can assist the family in a range of ways and can provide a contact point where individuals or families can receive a service which is comprehensive and competent.</p>
<p><b>Early Engagement</b></p>	<p>Early engagement for people with HD may be beneficial in a number of ways. It enables the professional to establish a therapeutic relationship with the client and care giver and ensures early intervention to maintain function and quality of life for as long as possible.</p> <p>For individuals with early symptoms, developing a professional, therapeutic relationship prior to the cognitive and emotional changes that accompany the disease is vital to working together as the disease progresses. Connecting people in the early stages of the disease to health professionals such as physiotherapists and speech therapists is also critical. Early engagement with families can promote communication and education, exploring the options of predictive genetic testing, family planning, preparation of power of attorney and living will documents, financial planning and employment issues. This can assist with a future that will include difficulty with functions such as memory, speech and cognitive ability.</p> <p>Early engagement assures families that specialised clinical services are available when needed and that broader services such as groups, carer respite referrals and referrals to specialised health professionals can be arranged.</p>

## Target Groups

There are two components of Huntington's service delivery:

### Case Management

Registered clients:

- People who are gene positive and experiencing Huntington's symptoms that require psychiatric treatment.

Non registered clients:

- People who are at 50 per cent risk of HD
- People who are experiencing symptoms of HD
- People undertaking diagnostic genetic testing
- People who have had the genetic test and are living with the knowledge that they have the HD genetic mutation and will develop HD.
- Carers family members/significant others experiencing issues that come with living in a family in which HD is known to exist
- Children or adolescents coping with the complex issues that emerge from the trans-generational impact of having HD in the family
- People who have taken the genetic test and do not have the HD mutation, but need help with other Huntington's related issues
- People considering testing.

Note: throughout the document clients receiving on-going psychiatric treatment will be referred to as "registered clients", that is, they are registered clients within the Mental Health Service system. "Non-registered" clients are clients who are actively receiving service from HDS staff without receiving ongoing psychiatric services. This differentiation relates only to the way data relating to HDS clients is reported at a state and national level.

### Predictive Testing (also known as presymptomatic testing)

- People considering testing
- People undergoing pre-test counselling
- People undergoing post-test counselling.

## Service Description

### Location

- South
- North
- North West

### Hours of Operation

- 8:45 am to 5:06 pm

### Staffing Profile

- Services are provided by a multidisciplinary team

### Linkages with other Services

Current clinical treatment provides support and symptom management. Different professionals are involved at different stages of the disease. Continuity of care should be maintained throughout all stages of the disease so that a relationship builds up between professionals, clients and their families. HDS works closely with many services and organisations including:

- Neurologist
- Speech Pathologist
- Dietician
- Psychiatrist
- Occupational Therapist
- Physiotherapist
- Acute Health
- Primary Health
- Community Services
- Clinical Genetics
- Disability Services
- Huntington's Disease Association
- General Practitioners
- Residential Facilities
- Community Sector Organisations
- ACAT
- Palliative Care Team
- Falls Prevention Team
- Centrelink
- Legal Services

## Types of Services Delivered

### Roles and Function of the Case Manager

The role and function of the Case Manager is complex and requires flexibility and judgement in determining the most appropriate service response to the client's needs and would include:

- Ongoing bio-psychosocial cultural assessments from point of referral
- Provisional assessment, goal planning, and therapeutic intervention
- Health monitoring and symptom management
- Consumer, carer and family counselling and education
- Advocacy
- Crisis assessment and intervention
- Clinical review
- Community referral to tertiary health services and/or other community and support services
- Adherence to best practice guidelines and evidence-based practice without excluding the appropriate use of innovative practice in service delivery
- File maintenance and documentation and departmental record keeping
- Outcome monitoring and recording of Health of the Nation Outcome Scales for MHS registered clients
- Service Satisfaction Survey
- Teamwork and collaboration

It is not a core function of case management to provide non-clinical support for consumers, but rather for HDS to liaise with community sector organisations and mainstream services in partnership with the consumer to ensure all their needs are met. Therefore it is common for consumers receiving clinical support from Huntington's Services to also be receiving support from other agencies. It is the role of the case manager to have a sound knowledge of services and supports available, liaise and coordinate access, and monitor and review outcomes.

Case management should be consistently applied in line with the Case Management Society of Australia's 'National Standards of Practice for Case Management.

As part of a Multidisciplinary Team, case managers also undertake the following functions in addition to their case management role:

- Health Promotion Community Education
- Community Development
- Facilitating consumer, carer and family participation
- Specific group work

**Case Management  
Core Functions and  
Clinical Activities**

- Ongoing professional development
- Supervision of students
- Adherence to service policies and procedures and all relevant codes of conduct and standards of practice including:
  - Occupational Health and Safety policies and procedures
  - Service planning, development, implementation and evaluation
  - Data collection, monitoring and reporting
  - Participating in research activities

All therapeutic interventions are provided by HDS within a chronic disease framework and include:

- Intake, Assessment, Triage and Referral
- Assertive Case Management
- Crisis Response
- Early Intervention
- Education
- Counselling
- Therapeutic programs and groups

**Areas of Activity**

- Coordination and referral of therapy and support services,
- Support programs and groups for people living with Huntington's Disease
- Coordination of Huntington's Disease Clinics
- Information and education for people affected by Huntington's Disease
- Providing continuity of care and single point accountability through assertive case management
- Providing treatment
- Advocacy
- Making links with other internal and external stakeholders to ensure an integrated service response
- Involving the consumer, families, and other support structures in the collaborative development of individual service plans.
- Proactive review of activity to explore avenues for early intervention and health promotion
- Ongoing risk assessment.

## **Roles and Function of Predictive Genetic Testing**

The practice of advocacy is an essential activity to promote best practice and maintain care and treatment over the trajectory of the HD disorder.

At all stages of the progression of HD the HDS clinicians advocate for the recognition of rights and access to mainstream treatment for the individual and families. Clinicians advocate for the recognition of the impact of symptoms on the individual and challenge discriminatory practices and stigma in health service delivery, residential and respite provision, community care services and for the provision of appropriate resources.

The principles of advocacy also apply in the complex hereditary context of competing demands when family members or carers and the individual person have conflicting needs. Advocacy is an essential activity as the progression of the disorder presents behavioural, physical, emotional and psychiatric symptoms, including the non-verbal and dependence stage that have the potential to isolate and alienate vulnerable individuals and families. Advocacy activities may also apply to situations of involuntary treatment and formal legal processes.

People at risk of Huntington's Disease may seek predictive testing for:

- Presymptomatic testing if known to be at risk for Huntington's Disease
- Testing if there are possible or suspected symptoms of Huntington's Disease
- Pre-natal testing in pregnancy
- Pre-implantation genetic diagnosis in IVF

Adults "at risk" of having inherited the extended gene responsible for the onset of Huntington's Disease can have a predictive genetic test. As HD has 100 per cent penetrance, if the individual is found to have the extended gene they will develop the disease if they live long enough. Only about 10 to 15 per cent of individuals "at risk" of having inherited the gene have chosen to have the test over the 17 years the test has been available. This small percentage of people who are asymptomatic, find knowledge of their genetic status has psychological and life planning benefits. Some people who suspect they have early symptoms choose to be tested to clarify their status. Some people chose to have a pregnancy tested or access pre-implantation genetic diagnosis (PGD) to ensure that their children are not affected by HD.

**The HDS aims to constantly monitor and review services to ensure continuous improvement by:**

- Involving consumers and carers/significant others and staff in the review, planning and development of HDS.
- Reviewing benchmarks of other services, striving to attain 'best practice', implementing research-based interventions and developing contemporary practices based on evidence.
- Actively contributing to research, the body of evidence based practice and development.
- Ensuring effectiveness and efficiency through provision of cost effective services that are responsive to community and individual need.
- Improving the health outcomes and community tenure of people affected by HD.
- Providing education to health and welfare agencies and the general public
- Identifying service gaps that impact on people affected by HD and advocating on their behalf
- Considering holistic, bio-psychosocial needs through assertive case management and individual service planning
- Increasing capacity within primary care and Community Sector Organisations (CSO's) support services through collaboration, education, consultation and liaison
- Reducing the stigma associated with HD through the mainstreaming of our services within the community and general health services
- Supporting ongoing workforce skills and development by:
- Developing and implementing a workforce development and innovation plan consistent with the Model of Care
- Encouraging clinicians to publish in recognised forums.

**Triage/Intake by HDS**

Clients of HDS can access service through the Mental Health Helpline or directly to the HDS clinicians. Triage is the process of determining the need and urgency for service. Triage involves the delivery of clinically relevant information, screening and referral.

Access via the Helpline is provided on a 24 hour basis by phoning 1800 332 388.

To accept referrals from the triage service, determine the required response and coordinate that response. Intake will be conducted on an agreed format and will include:

- Reason for referral
- A comprehensive assessment including bio-psychosocial needs
- A collateral history



**Assertive Case Management**

Case management is an essential element for continuity of care for consumers within an integrated service delivery system and ensures each consumer is able to access the services they need, when they need them, with one clinician accountable for ensuring (though not necessarily providing) ongoing assessment, treatment and review. Assertive case management is a proactive, collaborative process that facilitates the engagement of a range of external community services where appropriate to meet the needs of the consumer.

**Individual Service Plan (ISP)**

Every consumer in the mental health service will have an Individual Service Plan (ISP) and current risk assessment developed collaboratively, at the point of entry to the service. The ISP will contain strategies for relapse prevention and service discharge and will be evaluated by the case manager. A complete ISP will include a discharge plan collaboratively determined with the consumer, and where appropriate, carer or family member. Some consumers will move in and out of HDS several times during their lifetime, and each episode of care will require a comprehensive assessment process to identify current needs and put in place appropriate services and/or therapies that support recovery, minimise relapse and achieve positive outcomes.

**The Individual Service Plan should detail**

- The current situation and definition of problems
- The goals to improve the situation and indicators of their achievement
- The strategies for achieving the goals
- The person responsible for implementing strategies
- The date for review

**Consultation and Liaison**

Consultation and liaison is an inherent component of HDS. Partnerships, with internal and external stakeholders are improved upon with the provision of consultation, liaison, support, undertaking community development and participating in activities to increase community awareness and understanding of Huntington's.

**Performance and Outcome Monitoring**

The development of formal partnerships with stakeholders ensures clarity of roles and responsibilities and cooperation to develop and implement a shared evaluation strategy.

- All community mental health teams will be required to collect the following essential data
- OARS data (or the future electronic replacement service collection data)
- Agreed consumer/carer outcome measures
- Agreed suite of performance indicators
- Nationally and locally agreed Quality and Safety information
- Agreed evaluation data for monitoring effectiveness of the Strategic Plan 2006 – 2011

<b>Client Registration</b>	<ul style="list-style-type: none"> <li>• Clients requiring ongoing psychiatric intervention will be registered in the Mental Health system, via completion of OARS Admission</li> <li>• Clients requiring ongoing care for Huntington's (without Psychiatric intervention).</li> </ul>
<h2 style="background-color: #003366; color: white; padding: 5px;">Client Service Pathway</h2>	
<b>Entry to Service</b>	<p>Entry to the service can occur through a number of channels both family, community and professional.</p>
<b>Assessment</b>	<p>There is a standardised Huntington's Disease Service Assessment tool.</p> <p>Specialist assessments as clinically indicated.</p>
<b>Ongoing Care/Shared Care</b>	<p>On acceptance and service entry, the individual consumer will be allocated a case manager. The case manager works as part of the multidisciplinary team to undertake formal assessments to develop an individual Service Plan (ISP), for consumers requiring both short term and long term care.</p> <p>The ISP will specify the level of service to be provided to the consumer and significant others by the CMHS. The case manager will coordinate access to specialist treatments, groups and support services and ensure continuity of care if a consumer is admitted for inpatient care</p>
<b>Clinical Review</b>	<p>The ISP Plan is reviewed regularly with consumers and the multidisciplinary team and others service providers involved in the consumers care.</p> <p>Registered clients will be reviewed on a three monthly basis. Non registered clients will be reviewed not less than six monthly.</p>
<b>Separation from Service</b>	<p>Discharge occurs when the ISP goals have been met, and/or the consumer no longer needs the Huntington's Service.</p> <p>A discharge summary is documented with comprehensive discharge information indicating current treatment and support and relapse prevention plans, provided to Primary Health Providers and Support service on transfer of care. Formal Transfer of care occurs at the time of active engagement by the primary health Services/ support service. Clients are also made aware of how they can access the service as and when required.</p>

## Service Evaluation

### Client Outcome Measurement

- Health of the Nation Outcome Scales for registered clients
- Client Satisfaction Survey for all clients.  
A client satisfaction survey will be conducted within six months of intake. When a client indicates dissatisfaction with the service, or component therein, the ISP will be reviewed and the issue of dissatisfaction discussed with the client with a view to remediating the dissatisfaction and another Client Satisfaction Survey conducted within six months. Otherwise the Client Satisfaction Survey will be conducted annually.

Draft for Consultation