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### The psychological impact of having a parent with a progressive neurodegenerative condition

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**The Psychological Impact of Having a Parent with a  
Progressive Neurodegenerative Condition**

Helen J Aslett

A thesis submitted in partial fulfilment of the regulations for the

**Doctorate in Clinical Psychology**

at

**Bangor University**

**Prifysgol Bangor**

## **Abstract**

This thesis examined the experiences of having a parent with a progressive neurodegenerative condition. An integrative literature review was conducted to assess the experiences of growing up in a family affected by Huntington's Disease (HD), and a qualitative research study was undertaken to examine the experiences of young adults living with a parent with young-onset dementia (YOD), dementia diagnosed before the age of 65. Five young adults, between 25-36 years of age, participated in the study and semi-structured interviews were analysed using Interpretative Phenomenological Analysis (IPA).

The integrative literature review found that young people growing up in families affected by HD experienced uncertainty, isolation, and unique challenges transitioning to adulthood as they balanced caregiving demands with their own potential risk of developing HD. Levels of family disruption and the age of the child at the time of the parental HD diagnosis were also related to later psychological outcomes.

Findings from the research study indicated that young adults with a parent with YOD experienced feelings of loss and guilt as they managed relationship changes with their parent, role shifts, and caregiving responsibilities. Concern for their non-affected parent was also apparent. Participants reported experiencing isolation from others due to lack of shared experience. The need to feel understood by both peers and health care professionals, was clear.

The integrative literature review and empirical study both suggest that individuals with parents with HD or YOD have significant needs and face considerable challenges. In both circumstances it is important that health care professionals are aware of these needs, offer

appropriate support and develop timely interventions to manage feelings of distress and isolation. Clinical and research implications are discussed.

**This thesis is dedicated to my parents.**

To my mum, Margaret Aslett there are no words to describe how grateful I am for all the love, help and support you have given me throughout my life, and in particular the last two years when my world fell apart. You picked up the pieces, nursed me through gruelling and debilitating treatment and supported me in my return to studies.

To my dad, Donald Aslett. I wish you were here to see me achieve this. I always bemused you with my thirst for studying and never being able to do things the easy way. I carry you with me every day and miss you always. This is for you.

**In memory of Donald John Augustus Aslett 13<sup>th</sup> September 1927- 2<sup>nd</sup> May 2013**

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*“A journey of a thousand miles begins with a single step”*

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Thank you all.



## Declarations

This work has not been previously accepted in substance for any degree and is not being concurrently submitted in candidature for any degree.

**Signed** .....

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This thesis is the result of my own investigations, except where otherwise stated. Other sources are acknowledged by footnotes giving explicit references. A list of references is appended.

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**SECTION 1: LITERATURE REVIEW**

**The experiences of young people growing up in families affected by Huntington's  
Disease: an integrative review**

Helen J Aslett<sup>1</sup>, Joanne Kelly-Rhind<sup>2</sup>, Jaci C Huws<sup>3</sup>, & Robert T Woods<sup>4</sup>

<sup>1</sup> North Wales Clinical Psychology Programme

School of Psychology

Bangor University

Bangor

Gwynedd

LL57 2DG

UK

<sup>2</sup> Betsi Cadwaladr University Health Board

Hergest Unit

Ysbyty Gwynedd

Bangor

Gwynedd

LL57 2PW

UK

<sup>3</sup> School of Health Care Sciences

Bangor University

Bangor

Gwynedd

LL57 2EF

UK

<sup>4</sup> Dementia Services Development Centre Wales

Bangor University

Bangor

Gwynedd

LL57 2DG

UK

**Suggested running head: A review of the experiences of young people growing up in families affected by Huntington's Disease**

Corresponding author: Helen J Aslett, North Wales Clinical Psychology Programme,  
School of Psychology, Bangor University, Bangor, Gwynedd, LL57 2DG, UK. Tel:  
+44 1248 382205, email: pspcb5@bangor.ac.uk



### Aims and Scope

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The *Journal of Genetic Counseling*, published for the National Society of Genetic Counselors, is a timely, international forum addressing all aspects of genetic counseling. The journal focuses on the critical questions and problems that arise at the interface between rapidly advancing technical developments and the concerns of individuals at genetic risk. The publication provides genetic counselors, medical social workers, medical and laboratory geneticists, and other health educators with a premier resource.

- Instructions for Authors

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Bennett, R. L., Steinhaus French, K., Resta, R. G., & Lochner Doyle, D. (2008). Standardized Human Pedigree Nomenclature: Update and Assessment of the Recommendations of the National Society of Genetic Counselors. *Journal of Genetic Counseling*, 17, 424-433.

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**Abstract**

This integrative review investigated the experiences of young people growing up in families affected by Huntington's Disease (HD). Twelve papers (six quantitative and six qualitative) were identified. The findings of this review suggest that younger age at the time of a parental HD diagnosis may increase vulnerability to poorer psychological outcomes in adulthood. This was principally noticeable where family dynamics were disrupted. Dominant themes which emerged from the review related to the impact, uncertainty and isolation that learning of HD and growing up in a family affected by HD had on young persons' lives. How young people managed care-giving duties for their parent with HD in the presence of their own potential risk of developing the disease was also prominent. Both adaptive and maladaptive coping strategies were identified in the papers reviewed. Methodological shortcomings of the included studies are discussed and recommendations for clinical practice are made to reduce the distress and uncertainty experienced by this population.

**Key words:** children, experiences, Huntington's Disease, integrative review, psychological implications, young people

## **Introduction**

Huntington's Disease (HD) is a progressive inherited neurodegenerative disease for which there is no known cure. Its presentation is characterised by motor dysfunction, cognitive deterioration and affective disturbances (Surrock & Leavitt, 2010).

Estimates vary as to how many individuals in the UK have a HD diagnosis. A recent study suggests that the number of symptomatic HD individuals has been underestimated and that the UK prevalence rate is 12.3 HD symptomatic people per 100,000 (Evans, Douglas, Rawlins et al., 2013). This contrasts with earlier findings which estimated the prevalence rates as between 4-7 people per 100,000 of the population (Williams, Skirton, Barnette & Paulsen, 2012). These higher estimates may be due to improvements in diagnosis and therapies and a willingness of individuals to be placed on HD registers.

### ***HD Symptoms and Disease Progression***

HD progression cannot be slowed or reversed, and there are individual differences in the rate of its progression. The mean time from disease onset to death is typically 15-18 years (Surrock & Leavitt, 2010). Recently a pro-dromal period typified by subtle cognitive difficulties has been identified that can be present for a number of years prior to a formal diagnosis (Paulsen, 2010; Surrock & Lavitt, 2010). Whilst medication is available for HD it is used solely for symptom control (e.g. involuntary movements, irritability and mood changes).

The pattern of symptoms may vary between individuals living with HD. However, common symptoms include: emotional lability, lack of motivation, impaired learning, reduced insight, low mood, obsessive-compulsive type behaviours, attention and

memory deficits, impaired spatial awareness; involuntary movements and co-ordination problems, choreic movements (e.g. fidgeting, jerking, uncontrolled facial expressions, muscle rigidity), feeding, swallowing, communication difficulties and executive dysfunction (e.g. inability to plan, lack of cognitive flexibility, perseveration). The majority of individuals with HD eventually develop dementia (Bates, Harper & Jones, 2002).

### *Causes of HD, genetic implications and pre-symptomatic testing*

An inherited mutation in the Huntingtin gene on the 4<sup>th</sup> chromosome is responsible for 97% of cases of HD, but in 3% of cases there is no obvious previous family history of the disease (e.g. premature death from other causes). HD is “autosomal dominant” thus, there is a 50:50 chance of the gene being passed from parent to child and a 50:50 chance that a child with HD will pass it to their offspring. However, if the defective gene is not inherited, then HD will not appear in later generations. Men and women are equally affected and the mean age of onset is between 30-50 years of age (Bates, Harper & Jones, 2002), although 5-10% of those affected develop HD before the age of 20 (Kremer, 2002; Quarrell, 2008). Approximately 25% of cases are in those over 50 years of age, the eldest reported case being observed in a 90 year old (Dennhardt & LeDoux, 2010).

HD is one of the few neurodegenerative diseases for which a pre-symptomatic genetic test is available. Since 1993 a simple blood test has been available to those aged 18 years or above, with a parent with HD, to detect the presence of the HD mutation (Huntington’s Disease Collaborative Research Group, 1993). It has an accuracy

range of 96-99%. In the UK uptake of such tests is low at less than 15% of eligible offspring (Morrison, 2010).

The impact, implications and potential ethical dilemmas of pre-symptomatic HD testing are great both for the individuals who undergo testing and their families and partners. A prominent philosophical concern in the literature is what may be gained from taking a test for a disease for which there is no known cure (Taylor, 2004; Wexler, 1977). A positive genetic result means that unless other life events intervene an individual will definitely develop HD.

Motivation for undertaking HD pre-symptomatic testing appears to be the reduction of uncertainty (Baum, Friedman, Zakoswski, 1997; Evers-Kiebooms & Decruyenaere, 1998), and clarifying the risk, or lack of, for other family members (Smith, 2013). It may also serve to inform future reproductive choices (Baum, Friedman, & Zakoswski, 1997). The reasons identified for not taking the test include fear of employment discrimination (Bombard et al, 2012; Penziner et al., 2008) and the fear of the inability to live with the consequences of a positive result (Tibben et al, 1992). Testing positive for the HD gene may heighten HD-related distress and hyper-vigilance to HD symptoms (Smith, 2013) and increase feelings of parental guilt in respect of their own children (Wahlin, 2007). A negative test may also increase feelings of survivor guilt if other siblings and family members test positive (Kessler, 1994; Wahlin, 2007).

***Psychological Impact of HD***

The psychological impact of HD is multifaceted both for the individual living with it and their families (Aubeeluck, Buchanan & Stupple, 2012; Hans & Koeppen, 1980; Kaptein et al., 2007; Lowit & van Teijlingen, 2005; Tyler, 1983; Williams et al., 2009). Navigating the emotional, behavioural and cognitive changes a HD diagnosis brings, and coming to terms with the genetic implications, may place significant burden and stress on family members and affect their quality of life (QoL; Aubeeluck & Buchanan, 2007; Aubeeluck et al., 2012). In the early stages of the disease, behavioural changes such as lack of motivation and apathy brought on by cognitive decline may prove difficult for relatives to manage. In the later stages, caregiver stress may be as a result of having to provide increased physical care and witnessing the physical deterioration of their loved one (Roscoe et al., 2009). HD caregivers have reported increased isolation, increased financial burden, lower levels of life satisfaction and poorer social relationships (Aubeeluck et al., 2012; Aubeeluck & Buchanan, 2007; McCabe, Firth & O'Connor, 2009)

The majority of research which examines the impact of HD on families has focused on spousal and partner distress and has primarily employed quantitative methodology (Hans & Koeppen, 1980; Lowit & van Teijlingen, 2005). Where other family members have been included the results of their experience have not been analysed separately so it has not been possible to examine the effect of HD on children and young adults growing up within a family where a parent has been diagnosed with HD.

As the mean onset of HD is between 30-50 years of age, children within HD families are likely to first observe symptoms in their parents from mid-childhood/early

adolescence through to emerging adulthood. Adolescence is often described as a period of “storm and stress” (Erikson, 1968) as it is a time of transition where a young person moves from childhood to young adulthood and seeks increased autonomy in terms of decision making related to life choices and in critical thinking. It is a period characterised sociologically by identity formation and changing familial and peer relationships whilst biologically it is marked by hormonal changes (Smith, Cowie & Blades, 2003). To be an adolescent within a HD family, seeing a parent develop and deteriorate with the condition and being aware of their own potential genetic risk may present unique challenges in addition to those already being navigated as they move into adulthood.

HD research which has focused on the experience of adolescents has predominantly focused on attitudes towards pre-symptomatic testing and the psychological harm or benefits of predictive testing in those under 18 years of age (Richards, 2006; Timman, Roos, Maat-Kievit, & Tibben, 2004).). More recently it has been suggested that HD predictive testing prior to the age of 18 would reduce potential psychological distress by resolving uncertainty (Duncan et al., 2005, 2008; Duncan & Delatycki, 2006; Richards, 2006).

### ***Purpose of Review***

The aim of this integrative review is to synthesise and summarise the qualitative and quantitative research which expressly examines the experiences and impact of growing up in a family living with HD with the underlying threat of potentially developing HD. The findings of this review will inform health care providers of the specific experiences and issues associated with growing up in a HD family; the

psychological sequelae and the unmet needs and unique challenges faced by this population. This review will also illustrate the methodological strengths and shortcomings of the studies and provide suggestions for future research

### **Methods**

This review employed an integrative design. An integrative review employs a rigorous methodology synthesizing diverse research (e.g. quantitative and qualitative, experimental and non-experimental) so that conclusions about a particular topic may be drawn. Its aim is to enhance understanding, identify gaps in current research and to inform policy, practice, and theory development (Cooper, 1998; Whittemore & Knafl, 2005). To ensure rigour, the 5 step integrative review framework developed by Cooper (1998) and adapted by Whittemore and Knafl (2005) was followed:

- (1) Problem identification – the purpose of the review.
- (2) Literature search - including both computerised database and reference searches for articles with clear search parameters.
- (3) Data evaluation – Development of quality standards by which published research is evaluated and decisions made as to its inclusion within the integrative review.
- (4) Data analysis- Extraction of relevant data from each published article using a common metric and finding commonalities and discontinuities within the data.
- (5) Presentation of findings – Tabulated data of studies included within the review.

#### ***Literature search***

An electronic search of Psycinfo, Web of Science and Medline was conducted of all English language peer-reviewed articles published between January 1980 - October 2013.

A comprehensive set of terms related to Huntington's Disease, its psychological impact and genetic testing on family members of Huntington's patients was carried out using combinations of the following terms: Huntington Disease; and psychological impact (psych\* impact, experience, adjustment, adaptation, attitude); family (famil\*, son, daughter, child\*, adolescen\*, young adult, young person teen\*, parent, care\*); and risk (genetic testing, pre-symptomatic testing). Reference lists from papers deemed potentially relevant were also scrutinised for suitable papers.

Only English language primary research papers (qualitative, quantitative or mixed methodology), which included the experience of growing up in a HD family were included in the review. This included retrospective reports of the experience. Papers with a focus on caregiver stress and burden were included if the focus was on offspring providing care, or where the experience of caring for a parent with HD was analysed separately. Papers were excluded if the emphasis was on spousal or partner carer distress or if they focused on pre-symptomatic testing and genetic risk without reference to young people and the impact such testing had on their family.

### ***Data Evaluation***

Seven hundred and eleven papers were identified through the database searches. A further eleven papers were found through reference list searches. One hundred and fifty nine papers were excluded on the basis of being duplicates and a further four hundred and ninety-three papers were excluded on the basis of title analysis either for not meeting the basic review criteria in terms of subject area, or for being a review, commentary or guideline paper. Seventy papers remained for title and abstract analysis by the first author. Fifty were excluded on the basis of focusing on genetic



testing or spousal caregivers. Full text analysis was carried out with the remaining twenty records. Of these, twelve were identified as relevant. See Figure 1 for a flow diagram of how relevant studies were selected.

\*Insert Figure 1 here\*

A slightly modified version of the quality criteria framework devised by Caldwell, Henshaw and Taylor (2005) which was designed to assess the quality of both qualitative and quantitative studies, was used to evaluate the twelve selected studies (see Appendix 1). Studies were marked as to whether they fully or partially met the criteria. As all the studies were from peer-reviewed journals it was decided that it was unnecessary to assess the authors' credibility so this was omitted from the quality framework. There was some variation in study quality amongst the twelve studies, in particular with the older qualitative and quantitative studies, but all were deemed to be of a satisfactory standard for inclusion within the review by both the first and second author.

### ***Data analysis***

Data were extracted for each study included in the integrative review using a data extraction sheet. This included the study publication year, country of origin, study aim, design, sample, outcome measures (where applicable), findings and limitations of the study (see Appendix 2). Commonalities and discontinuities within the data were identified. These were then categorised under themes whose headings were representative of the commonalities.

## Results

### *Description of included studies*

Twelve papers were selected for this review based on ten separate studies published between 1983 and 2013 (Decruyenaere et al., 1999; Duncan et al., 2007; Folstein, Jensen, Chase & Folstein, 1983; Forrest-Keenan, Miedzybrodzka, van Teijlingen, McKee & Simpson, 2007; Forrest-Keenan, van Teijlingen, McKee, Miedzybrodzka & Simpson, 2009; Korner & Fitzsimmons, 1987; Sparbel et al., 2008; Vamos, Hambridge, Edwards & Conaghan, 2007; Van der Meer et al., 2006; Van der Meer, van Duijn, Wolterbeek & Tibben, 2012; Williams, Ayres, Specht, Sparbel & Klimek, 2009; Williams et al., 2013). Publications which used the same samples were the Forrest-Keenan et al., (2007) and Forrest-Keenan et al., (2009) studies; and the Sparbel et al., (2008) and Williams et al., (2009) studies. The studies were carried out in five different countries (Australia = 2, Netherlands = 3, UK = 3, USA/CANADA = 4). Participants for the studies were recruited via HD centres, genetic clinics, GPs and HD organisations and newspaper adverts.

There were six quantitative papers (Decruyenaere et al., 1999; Folstein et al. 1983; Vamos et al, 2007; Van der Meer et al., 2006, 2012; Williams et al., 2013) which included sample sizes ranging from 32 to 112. Five of the six studies employed questionnaire methodologies using standardised measures; one study utilised a newly devised questionnaire aimed at establishing distress of young people in HD families. The standardised measures included the Adult Attachment Interview (AAI; Main, Kaplan & Cassidy, 1985); Beck Depression Inventory (BDI; Beck, 1961); Family Relationship Index (FRI; Hoge, Andrews, Faulkner, & Robinson, 1989); K10 (Andrew & Slade, 2001); Measure of Parental Style (MOPs; Parker et al., 1997);

Minnesota Multiphasic Personality Inventory (MMPI; Hathaway & McKinley, 1940); Negative Life Events Scale (NLES; Cohen, Tyrell & Smith, 1993); NIMH Diagnostic Interview Schedule (DIS; Hendricks et al., 1983), and the State Trait Anxiety Scale (STAI; Spielberger 1983).

The sample size of the six qualitative papers (Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009; Koror & Fitzsimmons, 1987; Sparbel et al., 2008; Williams et al., 2009) ranged from 8 to 50. Four of the qualitative studies employed semi-structured interviews (Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009; Koror & Fitzsimmons, 1987) and two used focus groups (Sparbel et al., 2008; Williams et al., 2009). Three of the studies analysed the data using content analysis (Koror & Fitzsimmons, 1987; Sparbel et al., 2008; Williams et al., 2009) and three studies used thematic analysis (Forrest-Keenan et al., 2007, 2009; Duncan et al., 2007). Two of these studies (Forrest-Keenan et al., 2007, 2009) were in part informed by grounded theory (Glaser & Strauss, 1967).

### ***The HD family environment: Impact of age***

Five quantitative studies reported the impact of the age of the parent or child at the time of the HD diagnosis on later psychological well-being in their children (Folstein et al., 1983; Decruyenaere et al., 1999; Vamos et al., 2007; Van der Meer et al., 2006, 2012). In four of the quantitative studies, younger age of the child or parent at the time of the HD diagnosis was associated with increased negative experiences and outcomes for the child during adolescence and adulthood (Folstein et al., 1983; Decruyenaere et al., 1999; Vamos et al., 2007; Van der Meer et al., 2012).

Folstein et al. (1983) reported that 48% of their sample who had grown up within a HD household had psychological difficulties (anxiety, depression or conduct disorder/anti-social personality disorder). These difficulties were more prevalent in families where HD onset occurred at a younger age, suggesting that their children may also be young. Younger age of parental onset of HD was also associated with higher levels of familial disorganisation. Folstein et al (1983) defined such disorganisation as the child having to be raised away from the family home (e.g. in care or by other relatives) or abuse within the family setting.

The negative effects of younger age of a child at the time of a parental HD diagnosis were also observed by Decruyenaere et al. (1999). In 69 adults at risk of HD it was observed that those under 5 years of age at the time of their parent's diagnosis, experienced significantly higher levels of anxiety and depression and lower ego strength as adults when attending for pre-symptomatic genetic testing than those who were over 5 years of age at the time of diagnosis.

More recently Van der Meer et al. (2012) compared reports of adverse childhood experiences before the age of 16 in adults who had grown up in HD families, to those who had grown up in families affected by BRCA1 or BRCA2 breast cancer genes, and a control group. Van der Meer et al. (2012) found that children who had grown up in HD or BRCA families were more likely to have experienced negative childhood events and that younger age at the time of either a parental HD or breast cancer diagnosis was associated with an increase in negative childhood experiences and outcomes compared to controls.

In contrast Vamos et al. (2007) found no effect of a child's age at the time of their parent's HD diagnosis on later psychological well-being. This was despite 80% of the sample of forty 18-40 year olds reporting difficulty in growing up in a HD family, 40% reporting that HD had split their family up and 27.5% stating that HD was the biggest single issue facing them. However, these apparently discrepant findings may be an artefact of the low response rate to their questionnaire-based study, and that respondents may have had higher levels of psychological adjustment and greater resilience than non-respondents. This in turn acted as a barrier to participating in the study. If this is correct then their findings related to family disruption may be an underestimate of their prevalence. Alternatively the observations may be due to the study relying upon retrospective reports.

Despite the findings of Vamos et al. (2007), an increased vulnerability to psychological difficulties, as a result of younger age of children at the time of the parental HD diagnosis, is suggested. Early childhood is when key attachments are being formed between children and their primary caregivers, and the quality of such early attachments can have significant implications for later psychological functioning (Bowlby, 1989).

Adult attachment style in the children who had grown up in HD families was examined by Van der Meer et al. (2006) in 32 participants who were at 50% risk of developing HD and who had been under 18 years of age when their parent was diagnosed with HD. Only 38.7% of the HD sample had secure attachment representations in adulthood compared to 59.2% in a non HD reference group. Moreover, 45% of the HD sample had a preoccupied attachment style compared to

17.6% in the reference group. A preoccupied attachment style is related to negative childhood memories and events and insecure childhood attachments.

When participants were considered in terms of unresolved/disorganised attachment style, characterised by unresolved loss or trauma, 53.1% of the HD sample could be categorized into this typology compared to 18.4% of the reference group.

Van der Meer et al. (2006) found that those with insecure or unresolved/disorganised attachment representations were of a younger age when their parent was first diagnosed/displayed symptoms of HD. Age at which own risk of developing HD was learned of and death of the HD parent at a younger age were also associated with insecure attachment styles. Parents being diagnosed with HD during late childhood/adolescence seemed to have less effect on young people's later adult attachment representations. This may be related to there being a "critical period" for developing secure attachments.

### ***The HD family environment: family dynamics***

Three of the quantitative studies examined the role of family dynamics within the HD home and the impact this had for children growing up in HD families (Folstein et al., 1983; Vamos et al., 2007; Van der Meer et al., 2012). Folstein et al. (1983) found that psychological difficulties were greatest in families classified as "disorganised". This was related to early onset of HD diagnosis. Folstein et al. (1983) also reported that onset of major affective disorder within the children was related to the presence of it in the parent with HD. However, within a HD household the psychological well-being of the non-affected parent had implications for the child. Folstein et al. (1983) observed that antisocial personality disorder in young people within the HD families

was associated with a psychiatric diagnosis in the non-affected parent. It is unclear whether such diagnoses were a consequence of their partner's HD diagnosis and the strain placed on the family by HD or they preceded onset of symptoms in the affected partner.

Similarly Vamos et al. (2007) reported that participants' scores on the family relationship index (FRI), a measure of family cohesiveness, conflict and expressiveness, revealed significant levels of parental difficulty on all subscales, within the clinical range. Despite no evidence of affective disorder in their sample, such scores have been associated with adverse parenting experiences and increased psychological morbidity in children. Vamos et al. (2007) also observed high levels of parental difficulty from scores on the Measure of Parenting Style (MOPS) questionnaire which assesses parental indifference, over-control and abuse. This was found to be higher within HD participants than within a student reference sample. MOPS scores revealed that both the affected and non-affected HD parent displayed parental difficulties at similar levels, scoring highest on the over-control subscale. Interestingly, Vamos et al. (2007) observed that over-control scores related to fathers were significantly lower than a clinically depressed reference group, but mothers' scores were not. The reason for this finding is unclear, but suggests mothers within HD families may be more vulnerable than fathers to the increase in strains and demands. More recently Van der Meer et al. (2012) reported that significantly higher levels of parental difficulties were observed in HD families compared to both BRCA and control participants, and there were more likely to have been incidents of domestic violence and parental suicide attempts.

Themes of family stress, conflict and adverse childhood experiences were also evident in four qualitative papers included in this review (Forrest-Keenan, 2007, 2009; Sparbel et al., 2008; Williams et al., 2009). Sparbel et al. (2008) reported that levels of conflict were greatest where the HD parent was symptomatic and living at home, reflecting the behavioural unpredictability of HD. Participants spoke of parental conflict being borne of frustrations and misunderstandings between the affected and non-affected parent. However, there was also acknowledgement that conflict could extend to the wider family with detrimental effects to the young person placing extra pressure on them, impacting on their sense of autonomy and responsibility.

Williams et al. (2009) reported how young people kept their own emotions in check in order to reduce conflict with the HD parent. Yet within-family conflict and behavioural difficulties in the HD parent led to a sense of isolation in young people. This had a detrimental effect on their own social life, limiting opportunities for interactions with their peer group and leading to reluctance to have friends at the family home. In common with the quantitative findings, participants in Williams et al. (2009) spoke of the limitations a HD diagnosis placed upon the non-affected parent in terms of their ability to fulfil their parenting role in respect of availability, emotional and financial pressure. There appeared to be an acceptance of this amongst participants. However, some struggled with the lack of understanding the non-affected parent showed towards them, which could increase feelings of isolation (see also Forrest-Keenan, 2009).

Although three of the thirty-three participants in the study by Forrest-Keenan et al. (2007) reported abuse in childhood by the HD parent (see also Forrest-Keenan et al.,



2009), the majority of participants in this study appeared to be coping well and unlike the findings of Folstein et al. (1983) learning of their own HD risk at an early age appeared to facilitate this. The reason for this difference is unclear; however, it may be an artefact of the interrelationship between age of parent at HD onset, age of child, and levels of family disorganisation observed in Folstein et al. (1983).

### *Discovering HD risk*

Three qualitative studies investigated how children and young people find out about HD within their family and their own risk (Forrest-Keenan et al., 2007, 2009; Korner & Fitzsimmons, 1987). Korner and Fitzsimmons (1987) explored knowledge of HD in 50 13-25 year olds 29 of whom were at 50% risk of HD and 21 who were at 25% risk. Those at 50% risk were more likely to be aware of the term HD compared to those at 25% risk (96.55% v 80.9%). However, there was some confusion as to what the “genetic” nature of HD meant. Of their sample 59% of those at 50% risk learned about their risk from their parents compared to 43% of those at 25% risk. Interestingly three of the participants were only made aware of their own risk as a result of their parents being approached about the study.

Forrest-Keenan et al. (2007) found that openness in communication by parents in respect of both HD in general and the child’s risk enabled young people to cope, but being protected from knowledge of HD and its genetic risk until late teens /early twenties had a detrimental impact leading young people to become hyper-vigilant to their own HD risk in respect of physical symptoms. Those who discovered their own risk at a later age reported experiencing nightmares, panic attacks and a sense of dissociation. Exploring this further, Forrest-Keenan et al. (2009) investigated how

young people found out about HD and their own risk. Over 45% reported that they felt that they had always been aware of HD in the family. This was helpful to some, but others reported how being informed of their own risk in early childhood had caused a lot of fear and distress. Almost 25% of participants reported finding out gradually, knowing something was wrong, but being drip fed information by parents and colluding with this by not asking about their suspicions. This was highlighted by one participant who discovered his potential level of risk watching a TV documentary.

Forrest-Keenan et al. (2009) observed that parental gate-keeping was particularly evident in the 15% of participants who only learned about HD or their HD risk in early adulthood. Worry, anger, relief and fear were emotions most often expressed. Being able to see why their parents had kept the information from them in some cases improved relationships within the family. However, other young people were asked to conceal from younger and more distant family members, which increased pressure on them. Such gate-keeping was characterised by over-protection and authoritarian parenting styles as young people were growing up. Other parents who had less authoritarian parenting styles appeared to be waiting for the most appropriate time to tell their children. In this latter environment it was easier for young people to accept the information. Open communication was important also in those cases where the HD diagnosis was unexpected by all family members. Being open, supportive and being told in a relaxed informal setting, enabled the young person to have a better understanding of their own risk.

*Living with uncertainty*

Four qualitative papers addressed the issue of how young people growing up in HD families manage living with uncertainty and the risk of developing HD (Forrest-Keenan et al., 2009; Korner & Fitzsimmons, 1987; Sparbel et al., 2008; Williams et al. (2009). Williams et al. (2009) reported how young people struggle with constant reminders of HD within the family setting in light of their own potential genetic risk. Witnessing the decline of a parent or grandparent heightened young people's awareness of their own vulnerability and the restrictions HD placed on life, with future decisions having to be made in the context of potential risk.

Similar sentiments were expressed by participants in Sparbel et al. (2008) where all but one of the participants interviewed showed a reluctance to plan for the future or consider long-term goals. Despite this, a number of the participants were in romantic relationships, yet there was a general reluctance amongst this sample to consider having a family of their own. Forrest-Keenan et al. (2009) found similar reluctance in some of their participants. However, some were planning on having families irrespective of their genetic status in the hope of scientific innovations in the next 30 years. This was a theme echoed in Korner and Fitzsimmons (1987), a study which preceded the availability of pre-symptomatic testing.

Young people were found to keep their feelings of uncertainty to themselves. They reported struggling to discuss their own fears with the non-affected parent (Sparbel et al., 2008). Korner and Fitzsimmons (1987) found that 50% of those at 50% risk and 78.5% of those at 25% risk had not told their friends about their own risk of HD. This may potentially heighten feelings of isolation within the young person.

*Coping strategies*

Two quantitative studies considered coping strategies (Vamos et al., 2007; Williams et al., 2013). Vamos et al. (2007) reported that 72.5% of participants felt able to cope with HD in their families and their own risk. Similar findings were observed by Williams et al. (2013) who reported the results of a questionnaire-based study designed to identify the types of strategies employed by young people growing up in HD families, their degree of use and utility. Participants were 44 young people aged between 14-30 years of age who had a first or second degree relative with HD. They identified spending quality time with their HD parent, time with their friends, keeping busy with school and work, and actively researching HD related information as strategies that were most often employed and were of most benefit. Talking to Health Care Professionals (HCPs), attending support groups and conferences and limiting substance misuse in the HD parent were also identified as high utility strategies, but ones which were not often employed by young people. Participants reported that the most highly used but ineffective strategy employed was holding in their emotions. Maladaptive coping strategies such as substance misuse were reported as not being frequently used and were identified as being of low utility (cf. Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009).

Gender differences were observed by Williams et al. (2013) with females being significantly more likely to talk to others in a similar situation, make use of social support, psychological support and conduct research into HD and its implications. Age differences were also observed, with older participants being more likely to use humour as a coping strategy.

Four qualitative studies also examined coping strategies in young people growing up in HD families (Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009; Koror & Fitzsimmons, 1987). Forrest-Keenan et al. (2007) found that constructive coping strategies focussing on providing practical support and being positive about their parent's disabilities facilitated coping in young people. However, as previously noted, some young people particularly those who became aware of HD at an older age struggled to manage the uncertainty, becoming health anxious and hyper-vigilant to physical symptoms of HD (Forrest-Keenan, 2007; Koror & Fitzsimmons, 1987). Other participants had a fatalistic attitude towards their own risk of developing HD often based on supposition (Forrest-Keenan et al., 2007; Koror & Fitzsimmons, 1987). Use of avoidant coping strategies such as moving out of the family home at the earliest opportunity was also a theme in some of the participants in Forrest-Keenan et al. (2007).

Forrest-Keenan et al. (2009) suggested that coping with HD may progress through a series of stages of acceptance and non-acceptance similar to those who have experienced bereavement (c.f. Kubler-Ross, 1969). Resolution of uncertainty in respect of genetic status may also reduce maladaptive coping strategies. Duncan et al. (2007) found that prior to pre-symptomatic genetic testing a number of participants managed uncertainty in relation to risk by engaging in risk-taking behaviours such as underage drinking, drug use and criminal activity. This was echoed by participants in Forrest-Keenan et al. (2007, 2009).

Duncan et al. (2007) reported how attending genetic testing had enabled young people to manage the uncertainty of their HD status. Participants who had tested positive for

the Huntington gene expressed a sense of peace of mind (see also Forrest-Keenan et al., 2009) and were in time able to accept their positive HD gene status. Those who had received a negative result had as a result become more stable in their personal lives. However, other studies found that young people were uncertain as to whether or not they would pursue genetic testing (Sparbel et al., 2008) balancing the relief of finding out with the impact it may have on other family members and friends.

### *Young people as caregivers*

Three qualitative papers addressed the issue of young people caring for parents and relatives with HD and how this affected them (Forrest-Keenan et al., 2007; Sparbel et al., 2008; Williams et al., 2009). Forrest-Keenan et al. (2007) examined care-giving experiences in HD families in those under and over eighteen years of age. Those aged under eighteen years spoke of providing practical support within the house (e.g. housework, cooking, cleaning) whilst others were involved in providing personal care such as washing, and administering medication (see also Sparbel et al., 2008; Williams et al., 2009). Level of care provision impacted on normal teenage life, affecting their education, with time off school and becoming isolated from their friends and peer group. Those aged eighteen years and above who provided both practical and emotional support to their HD relative similarly reported how care-giving not only impacted upon their work and career, but also the demands of their own young families. Older participants also spoke of how they were perceived by other family members as being the person holding it all together and having to manage this responsibility.

Williams et al., (2009) detailed the forms of care-giving provided by 32 teenagers looking after a parent or grandparent with HD. A number were involved in providing practical support with feeding and bathing and supervising their relative so that they did not come to harm.

Sparbel et al. (2008) reported how young people felt overwhelmed by care-giving responsibilities that they assumed both in relation to the HD parent, and also supporting their non-affected parent both practically and emotionally. A sense of duty and responsibility hung over the young people as opportunities to move away from home (e.g. to attend college) arose. Williams et al. (2009) observed that fatigue and exhaustion in relation to care-giving demands was commonly reported by participants and this led to low mood and anxiety. Other participants spoke of the restrictions care-giving placed on friendships and socialising and how such meetings had to be carefully co-ordinated. Lack of support from statutory sources led to increased feelings of burden (Forrest-Keenan et al., 2007; Sparbel et al., 2008; Williams et al., 2009). Teenagers spoke with a resigned acceptance of growing up before their time and missing out on normal adolescent experiences.

## **Discussion**

### ***Summary of results***

From both the quantitative and qualitative evidence reviewed, it is apparent that the experiences of young people growing up in families affected by HD are distinct from their peer group. In addition to navigating the transition between child and adulthood, young people growing up in HD families have to manage the impact of watching a close family member live with the debilitating consequences of a HD diagnosis, and

the effect this has on their wider family, whilst personally discovering, processing and living with their own risk and potential vulnerability to HD.

From the data reviewed, there is evidence to suggest that early onset of HD and younger age of the child may be related to later negative psychological outcomes (Folstein et al., 1983; Decruyenaere et al., 1999; Van der Meer et al., 2012). This was associated with levels of familial discord. Findings by Decruyenaere et al. (1999) and Van der Meer et al. (2006) suggested that younger age at the time of a parental HD diagnosis may increase a child's vulnerability due to attachment formation in early childhood being disrupted.

Both quantitative and qualitative studies revealed significant levels of family difficulty within HD families which may affect young people as they grow up (Folstein et al., 1983; Sparbel et al., 2008; Vamos et al., 2007; Van der Meer et al., 2012; Williams et al., 2009). Although not formally assessed, the studies suggest that higher levels of emotional expression within HD families may be reflective of the cognitive and behavioural symptomology of HD and the stress placed on family members as a result of these. It may also be related to the strain and demands placed on the non-affected parent, which may limit the amount of time and ability to provide a parental and pastoral role for children and communication between parents and young people. This review also found evidence that mental health difficulties in the non-affected parent potentially affected young persons' well-being and family functioning (Folstein et al., 1983; Vamos et al., 2007). Together these findings highlight the potential impact HD has not only on the individual living with a HD



diagnosis but the wider family system and how it can lead to a sense of isolation in the young person which was revealed in the qualitative data.

The way in which young people learn of their genetic risk was found to have implications for their well-being (Forrest-Keenan et al., 2007, 2009). Open, sensitive, honest and timely communication by parents when informing young people of their risk was associated with more acceptance of the risk compared to parental gate-keeping (Forrest-Keenan et al., 2009). Where parents were more circumspect in the information, shared negative implications were observed as a consequence of parenting style. Parental-gate-keeping fell into two categories (i) to protect the young person from discovering their own potential risk and enabling them to enjoy their childhood, giving information in an informed and timely manner (ii) gate-keeping in combination with authoritarian parenting styles leading to suspicion and conflict and difficulties accepting potential risk.

This review suggests that the majority of young people appeared to adopt positive coping strategies to manage living with the uncertainty HD brings despite often constant reminders of their own risk by living with an HD parent. However, young people often experienced isolation from both family and peers. Providing care at a young age placed demands on young people in many ways. Some struggled in respect of education, work and social support and managing their own young families (Forrest-Keenan et al., 2007). For others care-giving for their parent and witnessing their deterioration, impacted on their ability to future plan and have romantic relationships (Sparbel et al., 2008).

Although there was some evidence of young people using maladaptive forms of coping (substance misuse and criminal activities) (Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009), they appeared to outgrow these as acceptance of their HD risk increased or they gained a definitive answer to their genetic risk whether positive or negative. It appears that the process of acceptance is for many preceded by feelings of loss, anger, fear and guilt.

### ***Methodological Issues***

Due to the dearth of research examining the experiences of young people growing up in HD families it was necessary to conduct an integrative review of both quantitative and qualitative research. This meant there was a degree of heterogeneity in the studies included. Also as a consequence of the paucity of research, two studies were included which predate the availability of pre-symptomatic testing in 1993 (Folstein et al., 1983; Korner & Fitzsimmons, 1987). In the latter study this may have influenced the observed results in relation to attitudes to risk.

The generalizability of the quantitative findings may be questioned due to small sample sizes. There is also a potential participant bias due to how participants were recruited in both the quantitative and qualitative studies. The majority of studies recruited via genetics clinics which may have biased findings, as only a minority of potential participants attend such clinics (Morrison, 2010). Three of the studies utilised HD organisations (Van der Meer et al., 2006; Forrest-Keenan et al., 2007, 2009). This may also have biased the observed findings as those in contact with HD organisations may be individuals who take an active interest in their HD risk.

Similarly, parental gate-keeping may have been a source of potential bias. In the studies by Korner and Fitzsimmons (1987), Forrest-Keenan, et al. (2007, 2009), Sparbel et al. (2008) and Williams et al. (2009) parents were approached about young people participating in the research studies due to parental consent being necessary for those under 16 years of age. Sparbel et al. (2008) reported that a number of parents declined to approach their children due to the potential stress it would cause (see also Williams et al., 2009). Korner and Fitzsimmons (1987) found that where the current HD risk in the child was 25% there was significant resistance by parents compared to where the risk was 50%. Both recruitment strategies and parental gate-keeping may therefore lead to biased samples which potentially led to an underreporting of difficulties as those most in need were hardest to reach.

In some of the studies, age may have biased the results as participants were asked to recount their experiences as teenagers growing up within HD families (Vamos et al., 2007; Van der Meer et al., 2006, 2012). For example, Van der Meer et al. (2012) required participants to recall adverse childhood experiences, yet participants ranged between 18-65 years of age. This meant that for some the events being recalled were recent, but for others the events occurred many years previously.

Whilst some of the quantitative studies used reference group norms, they lacked appropriate control groups (e.g. age or gender matched) which would have provided more specific comparison data. There were also particular methodological issues in the study by Folstein et al. (1983) where parental report was relied on in some cases. In the qualitative studies the use of focus group methodology by two of the studies

(Sparbel et al., 2008; Williams et al., 2009) may have impacted on the experiences disclosed as participants may have been reluctant to disclose in group settings.

### ***Practice Implications***

This review highlights the potential psychological difficulties faced by young people growing up in HD families, and the difficulties and conflicts that may arise. Clinicians need to understand the context of HD within the young person's life and provide timely interventions. The studies by Forrest-Keenan et al. (2007) and Williams et al. (2013) show that young people benefit from opportunities to have psychological support. However, this depends both on the ability of young people growing up in HD families to have access to psychological services via HD clinics, and the ability of HCPs to be able to access young people growing up in such families. Whilst the issue of early disclosure is one that is not fully resolved by the results of this review, psychologists and genetic counsellors can play a significant role facilitating dialogue within families and promoting open and honest communication about HD and HD risk.

HCPs need to involve the whole family in discussions about the potential impact of HD. They should enquire after children and young people in all HD appointments and make specific attempts to meet with young people growing up in families where they are at risk of HD. By doing this, care-giving responsibilities undertaken by the young person may be investigated and appropriate support such as referrals to young caregiver organisations made. Within-family conflicts may also be discussed and coping strategies assessed. The recently devised teen-HD measure, as employed by Williams et al. (2013), may be useful to assess the latter. HCPs also need to assess for

anxiety in young people at risk of HD and be mindful of hyper-vigilance and fatalistic beliefs.

Risk levels within a young person may change for a number of reasons and clinicians need to be aware of this. A young person may only be at 25% risk due to the disease being in a second-degree relative when they first become known to services.

However, if a parent then receives an HD diagnosis, their known risk increases to 50%. Pre-symptomatic testing may either increase the known risk to 100% or eradicate it. Clinicians need to consider their role in providing information to young people regarding pre-symptomatic testing. Such testing is only available to young adults over 18 years of age. By discussing the implications of genetic testing as a young person grows up this not only informs the young person, but also gives them a sense of future control in managing the uncertainty they may feel in relation to the potential risk.

To manage the loneliness and isolation experienced by some young people growing up in HD families, clinicians may also consider setting up HD support groups for young people within their clinics or encourage young people to join social networking private support groups (e.g. on Facebook and via the Huntington's Disease Association) where they can chat in confidence to others in similar positions to themselves. This peer support may provide an important source of social support to the young person.

***Research Recommendations***

This review highlights the dearth of research in this area and the need for high quality quantitative and qualitative research. Ethically it may always be difficult to interview younger children about HD and the effect it has on them whilst they are growing up, but studies are clearly needed where retrospective reporting is not relied on. Future quantitative research which has homogenous samples and appropriate control groups is also suggested as a result of the findings of this review.

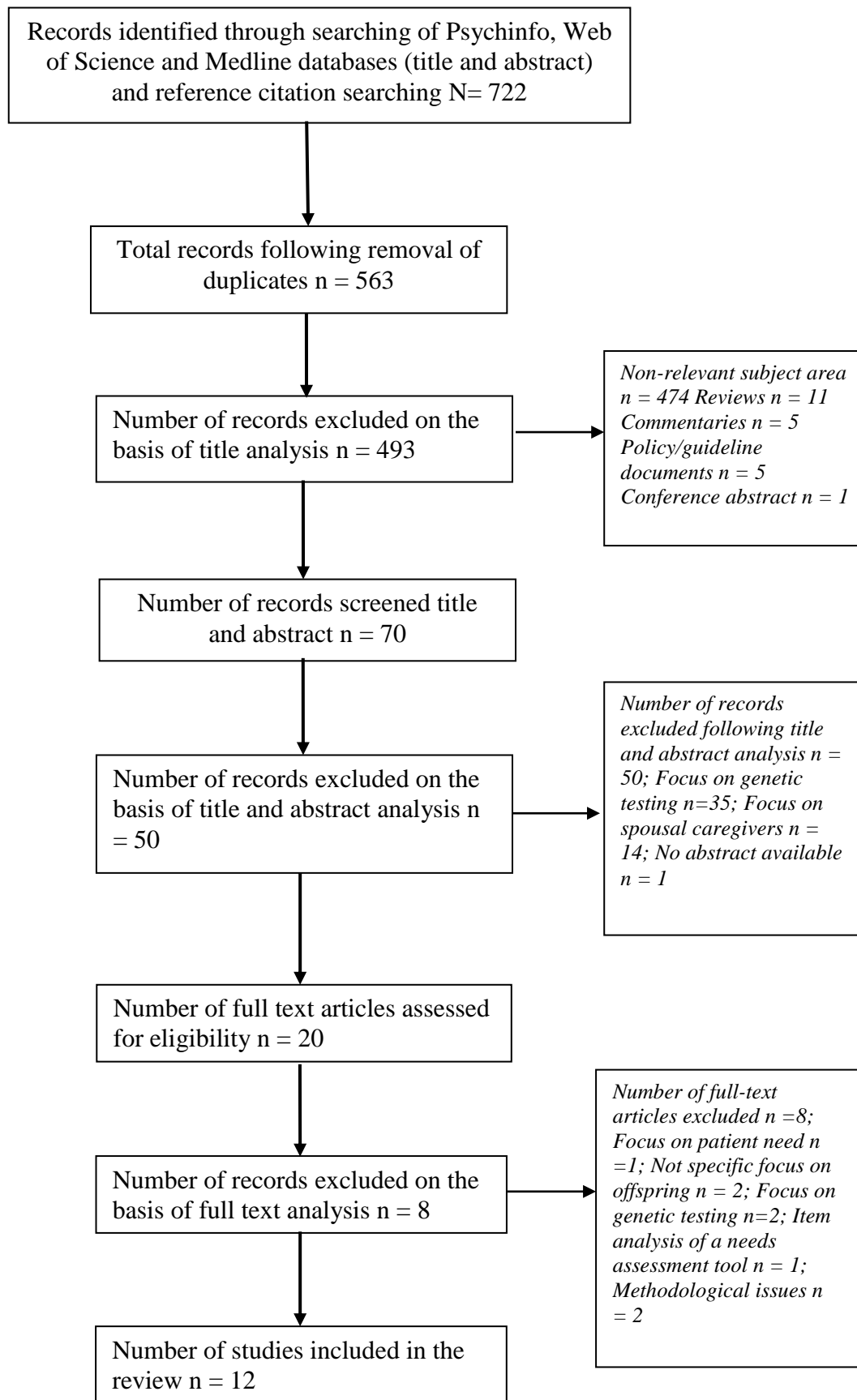
Extending qualitative research to focus on the individual meaning young people give to the presence of HD within their family lives, may serve to inform and develop HD specific outcome measures for this population. Exploring risk behaviours amongst young people growing up in HD families combining qualitative interviews with standardised measures of risk assessment may also inform targeted early interventions. Qualitative research is also needed to explore the decision making process regarding attending pre-symptomatic testing and the implications of a positive or negative test for a young person in respect of their own identity and their identity within their family.

***Conclusion***

Young people growing up in families affected by HD face unique challenges as they try to transition successfully to adulthood, whilst balancing care-giving demands, their own potential risk of developing HD, and their education and social lives. For many this can be an isolating experience. It is important that HCPs working with HD families are aware of these potential difficulties so that they are able to monitor their development, provide early interventions to reduce distress and feelings of burden,

and are able to counsel and support young people in respect of future decisions regarding pre-symptomatic testing.

Figure 1. Flow chart of study selection





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**SECTION 2: RESEARCH PAPER**

**“This is killing me inside”: the impact of having a parent with young-onset  
dementia** Helen J Aslett<sup>1</sup>, Jaci C Huws<sup>2</sup>, Robert T Woods<sup>3</sup>, & Joanne Kelly-Rhind<sup>4</sup>

<sup>1</sup>North Wales Clinical Psychology Programme

School of Psychology

Bangor University

Bangor

Gwynedd

LL57 2DG

UK

<sup>2</sup>School of Health Care Sciences

Bangor University

Bangor

Gwynedd

LL57 2EF

UK

<sup>3</sup>Dementia Services Development Centre Wales

Bangor University

Bangor

Gwynedd

LL57 2DG

UK

<sup>4</sup>Betsi Cadwaladr University Health Board

Hergest Unit

Ysbyty Gwynedd

Bangor

Gwynedd

LL57 2PW

UK

**Suggested running head: “This is killing me inside”: the impact of having a parent with young-onset dementia**

Corresponding author: Helen J Aslett, North Wales Clinical Psychology Programme,  
School of Psychology, Bangor University, Bangor, Gwynedd, LL57 2DG, UK. Tel:  
+44 1248 382205, email: [pspcb5@bangor.ac.uk](mailto:pspcb5@bangor.ac.uk)

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### **Abstract**

This study explored the experience of young adults having a parent with young-onset dementia. In-depth interviews were undertaken with five participants aged between 23-36 years of age and these were analysed using interpretative phenomenological analysis (IPA). Participants were found to experience a number of stresses in relation to their parent's illness, many of which were linked to loss and guilt. Five main themes were identified related to relationship changes, shifts in roles and responsibilities, support for the non-affected parent, support for self and the impact of living with their own potential risk of dementia. These findings are discussed in relation to the existing literature and suggest that individuals with a parent with young-onset dementia have needs which service providers should consider in the wider context of young-onset dementia care.

**Keywords: interpretative phenomenological analysis, parent, psychological impact, qualitative, young-onset dementia**

## Introduction

Estimates of the number of people in the UK having young-onset dementia (YOD), (defined as dementia diagnosed before age 65) vary from 17,000 to 64,000 (Alzheimer's Society, 2012; Alzheimer's Research Trust, 2010).

YOD has greater heterogeneity than dementia in individuals over 65, where the Alzheimer's type predominates (60% - Alzheimer's Society, 2012). The needs of people with YOD also differ both as a consequence of faster disease progression and socially as a result of being at a different life stage (Brown et al., 2012). Individuals may still be in employment, raising families and have financial commitments (e.g. mortgages). As dementia is perceived as a disease of old age, the impact on young-onset families may also be greater as it is "out of sync" with the normal life course (Harvey, Skelton-Robinson & Rosser, 2003).

Spouses and partners of those with YOD report higher burden, poorer emotional health and less social support than those providing informal care to older individuals with dementia (Arai, Matsumoto, Ikeda & Aria, 2007; Freyne, Kidd, Coen & Lawlor, 1999; Luscombe, Brodaty & Freeth, 1998; van Vliet, de Vugt, Bakker, Koopmans & Verhey, 2010). Spousal and partner distress in caregiving for someone with YOD may also impact on other members of the family, as individuals struggle to juggle the competing demands of caregiving for an ill spouse and raising a family (Gelman & Greer, 2011).

Adolescence and emerging adulthood is a stage in life where autonomy is sought, romantic relationships formed and aspirations followed. However, a parental

diagnosis of YOD may complicate these aspects of development. Recently a number of predominantly qualitative cross-sectional studies have been published investigating the impact of having a parent with a YOD diagnosis (Allen, Oyeboode & Allen, 2009; Barca, Thorsen, Engedal, Haugen, & Johannessen, 2014; Gelman & Greer, 2011; Millenaar et al., 2013; Nichols et al., 2013 Svanberg, Spector & Stott, 2010a; Svanberg, Stott & Spector, 2010b; van Vliet et al, 2010). Three studies have specifically considered the experiences of individuals aged under 18 years of age (Gelman & Greer, 2011; Nichols et al., 2013; Svanberg et al., 2010b), whilst three other studies have considered the experiences of adolescents and young adults (Allen et al., 2009; Barca et al., 2014; Millenaar et al., 2013). Despite wide age ranges in participants, similar themes have been reported across the studies suggesting that a parental diagnosis of YOD places significant stress upon children.

Svanberg et al. (2010b) in a mixed-method study of 11-18 year olds caregiving for a parent with YOD found that over 30% of their sample experienced clinically significant levels of low mood, and 50% were experiencing significant burden. This is related to children and young adults experiencing “anticipatory grief” where struggles are observed with the decline of the parental relationship, and the sense of who their parent was, even though their parent is still alive (Allen et al., 2009; Barca et al., 2014, Gelman & Greer, 2011; Millenaar et al., 2013; Nichols et al., 2013; Svanberg et al 2010b).

Role conflicts wherein children and young people take on roles and responsibilities which conflict with their age and stage of development (Allen et al., 2009; Barca et al., 2014; Millenaar et al, 2013; Nichols, 2013, Svanberg et al., 2010b), concern for

the well-being of the non-affected parent (Allen et al., 2009; Millenaar et al., 2013; Nichols et al., 2013), and levels of family conflict within the home due to behavioural changes in the parent with YOD (Allen et al., 2009; Barca et al., 2014; Millenaar et al., 2013) and the ability of the non-affected parent to manage this (Barca et al., 2014) have also been reported as leading to stress and distress. Children and young people may experience isolation as they struggle to find an outlet for their concerns both in and outside of the family (Allen et al., 2009; Barca et al. 2014; Gelman & Greer, 2011; Millenaar et al., 2013; Nichols et al., 2013).

Until now research has focused upon identifying themes of shared experience in this population. To date no published study has explored the subjective lived experience of having a parent with YOD. This is important as although individuals may be experiencing the same situation, their perceptions, experiences and processing can potentially vary considerably. This study addresses this by using Interpretative Phenomenological Analysis (IPA, Smith & Osborn, 2003), a qualitative method which aims to capture lived experiences and the meaning of such experiences to the individual.

By using IPA methodology this study aims to explore the personal meaning attached to having a parent with YOD; to consider how this impacts on relationships with other family members; and to consider positive as well as negative impact of having a parent diagnosed with YOD. The findings may be considered in relation to establishing the needs of this population and what interventions and services may best meet these.

## **Method**

### ***Design***

This qualitative study collected data via semi-structured face-to-face interviews with individuals who had a parent diagnosed with dementia before 65 years of age. The study utilised IPA methodology (Smith & Osborn, 2003). IPA is an idiographic approach which focuses on in-depth analyses of how small and homogenous groups of individuals make sense of their experiences and attach meaning to them. This is achieved through a process known as the “double hermeneutic” in which the researcher tries to make sense of the participant’s experience of making sense of their own experience (Smith, Flowers & Larkin, 2009).

### ***Ethics***

School of Psychology Ethics Review Committee at Bangor University (reference number 2012 -5162) and local NHS ethical and R&D approval (reference number 12/WA/055) was obtained for this study.

### ***Participants***

Participants were recruited by healthcare professionals (HCPs) working within dementia services across North Wales. Participants were aged 18 years of age or above with a parent with a diagnosis of dementia confirmed before the age of 65, who were between 6 months and 5 years of diagnosis and still alive. Participants needed to be aware of their parent’s diagnosis and be fluent in English.

Six participants were initially recruited; however this study only reports data from 5 of the participants aged between 23-36 years (2 males and 3 females) due to one



participant disclosing significant personal information unrelated to the research area. All participants had daily contact with their parent whether in person or by phone. One participant was the main caregiver for her parent with YOD and two of the participants had children of their own. Parents were aged between 51 and 63 years of age at the time of diagnosis and were between 1-5 years from diagnosis. Alzheimer's disease was the main type of YOD within this sample. Further patient characteristics are presented in Table 1. Pseudonyms are used to protect participants' identity.

\*insert Table 1 here\*

### ***Procedure***

At scheduled appointments prospective participants were either approached directly by HCPs if accompanying their parent, or an information pack was given to the attending partner or friend to pass on to them (see Appendix 3). Prospective participants then contacted the first author directly.

The first author conducted semi-structured interviews with participants in their home or work establishment. Written consent was obtained from participants prior to interviewing, this included consent for the audio-recording of the interview (Appendix 3).

A bespoke interview schedule was developed by the first author in consultation with the research team (Appendix 4). Interviews lasted between 32 and 90 minutes, and the mean length of interviews was 57 minutes. Each interview was transcribed verbatim. To protect participants' identity, participants' transcripts were assigned pseudonyms and potential identifiers were removed. Participants were offered a gift

voucher for participating and were telephoned 48 hours after each interview in case the interview had raised any difficulties for them.

### *Analysis*

In line with the IPA approach (Smith, Flowers and Larkin, 2009), each transcript was initially read whilst listening to the audio recording of the interview. Transcripts were then read and re-read several times on a case-by-case basis. Line-by-line analysis of each transcript was carried out, and three types of codes were identified: (i) descriptive codes – key words describing content (ii) linguistic codes – the words employed to describe the experience (iii) conceptual comments – where data was questioned for meaning to provide an interpretative context (see Appendix 5). Identification of these codes provided insight into the ways in which participants spoke of and thought about issues. These were used to develop emergent themes which captured the participant's individual experience. Emergent themes within each transcript were then collated and connections between themes were made. This process was repeated for each transcript. The themes that had arisen in each transcript were compared and the themes were integrated. Super-ordinate and sub-ordinate themes were identified. A summary table of themes with illustrative quotes from participants was then created (see Appendix 6). Yardley's (2000; 2008) four principles for validity in qualitative research were attended to throughout the study to ensure that the research was sensitive to context, rigorous, transparent and coherent.

### **Results**

The analysis revealed five overarching themes. They describe participants' experiences of having a parent with YOD diagnosis. The five themes are:

- (1) “Like I know them but I don’t know who they are”
- (2) “You just look up to them for all your life and then now they’re looking to you for help”
- (3) “I feel like she can get things off her chest”
- (4) “You are never going to understand until you are in my position”
- (5) “Hang on a minute, this could happen to ... to me”

***Theme 1: “Like I know them but I don’t know who they are”***

This theme addresses relationship changes between participants and their parent with YOD. This comprises two sub-themes: (i) “And they’ve just gone, that’s ... that’s the worst part”, considers relationship strength, and (ii) “You are sort of treading on eggshells”, examines communication within the relationship.

***“And they’ve just gone, that’s ... that’s the worst part”***: All participants struggled with loss of a meaningful relationship with their parent. Kate felt her father had “*just gone, that’s ... that’s the worst part about it*”. Kate still wanted her father to fulfill the parental role and struggled with this loss “*I’ve just wanted him to like ... hug me, or just tell me everything’s going to be okay*”. Kate felt she was left with “*nothing*”, even though her father was still alive.

*Last night I went to see him and he forgot who I was, he couldn’t even tell me my name, and I thought ... ah, it’s just ... that’s heartbreaking.*

Matthew spoke of a subtle loss and reflected on the conundrum of knowing, yet not knowing who his father was anymore:

*So you look at someone and you know who they are but ... you're not quite sure... [...] but the way they're talking is like ... completely different to how they would be.*

Quality of pre-diagnosis parental relationship impacted on relationship quality post-diagnosis for participants in different ways. Ben's relationship with his father had often felt uncertain and he felt that dementia had "robbed" him of a chance to reconcile this. Ben struggled with this, positioning his experience against those of his peers:

*I just kind of ... I think ... I think ... frustrated; frustrated and kind of ... robbed of a ... of one ... a big relationship that, you know, I see some people ... get to share with their dads, why they have ... they have a dad, they know that their dad was proud of them or something, [...] whereas I don't think ... I don't think I've ever really had...*

For Anne the irony of the situation was that it had enabled her to rekindle her relationship with her mother. This had been "a battle" but disease progression had led Anne to become more accepting of her mother in the presence of loss:

*...we didn't have an awful relationship, but we didn't have a ... brilliant one either [laughs]. [...] it was pretty horrific really, she ... you know, living in your Mum's home when she doesn't want you here is quite hard. [...] fortunately as ... as her disease has progressed I think she's just relaxed a lot more and ... actually its ... it's no problem, you know.*

**"You are sort of treading on eggshells":** Participants spoke of how they had become more aware of their communication style in interactions with the parent and how they

managed this. Diana felt a strong need to be compassionate in communication to what her parent was experiencing, yet this was sometimes difficult:

*You have to remember not to lose your temper with them because ... it's ... they can't help it, it's ... part of what's happening inside them and ... their mind. [...] not get frustrated with them and ... get angry or anything, because it is ... it's not helping you and it's not helping them.*

Matthew described the process as “treading on eggshells” which required a conscious effort to show greater sensitivity and patience in communication:

*You're sort of treading on eggshells, you're just ... things you say and that, you don't know if they know the ... quite get it.*

Participants struggled with their parent's changing thought processes, cognitive inflexibility, and one-sided communication which left them feeling unfulfilled. Diana grappled with this unpredictability:

*Some days you can get a really good conversation out of her, and then other days ... it's a bit ... mixed really.*

Anne clung on to her mother's residual language as the dementia progressed, but there was a cruel irony:

*Years into her Alzheimer's, she couldn't pick up a fork, she'd ... she'd quote Shakespeare whilst eating dinner.*

In trying to understand his father, Ben questioned what his father could understand. However such questioning did not provide Ben with answers and added to feelings of helplessness:

*...he's an enigma, we're just like 'I don't know ... I don't know what he's thinking'. You can make assumptions, like is he ... is he scared about it? Is*

*he worried about it? Is he upset? Does he really just think that there's nothing wrong with him, and he just gets up and he does his daily routine?*

**Theme 2: “You just look up to them for all your life and then now they're looking to you for help”**

This theme captures participants' adaptation to role changes since their parent's diagnosis. There are three subthemes (i) “I never thought I'd be doing for her” considers caregiving responsibilities (ii) “It's kind of on hold at the minute” relates to participants sacrifices to their own life, and (iii) “It's splitting me” examines responses to caring.

**“I never thought I'd be doing for her”**: Participants differed in the level and type of care they were providing, but they all seemed to experience similar feelings as they adjusted to their new role. For Kate the impact of her father's illness felt relentless, “*a nightmare*” and she struggled with the reversal of roles:

*It's just seeing someone that you love so much, like ... they are the ones that ... oh I don't know, you just look up to them for all your life and then now they're looking to you for help. [...] that's all I can say really, it's just a living nightmare. I hate it.*

Anne's life had been significantly disrupted by her mother's illness, she reflected on this and how her mother would be “*mortified*” of her daughter being responsible for her personal care:

*I never ever thought I'd be living back in my family home in my thirties [...] she needs full care, which I never thought I'd be doing for her, and I'm sure*

*she never ever dreamt I'd be doing it for her; she'd be mortified if she knew, you know, what was happening now. She was a very dignified lady in that sense, and very private.*

Role reversal was expressed by Diana as a sense of needing to protect her mother like a child yet experiencing helplessness:

*I suppose really ... you just want to be there for them, or you want to ... you know, I suppose it's like with a child, you want to protect them don't you? But then with this you don't know how you ... how you can do that.*

For Diana and Anne there was an additional sense of role reversal as their mothers could not perform grandmotherly duties. Anne reflected on this in terms of social comparison and a silent mourn over such a loss of role for her mother and resignation to this were clear:

*I often wonder what it would be like to have a family without my Mum [...] sometimes you see my friends and they've got their grandparents who look after their children, they go back to work and it's ... it's support rather than us supporting her. But it's just the way things are and I suppose you've just got to get on with it.*

**“It's kind of on hold at the moment”:** Some participants spoke about how they felt their own life had been placed “on hold”. Anne reflected on the scale of changes to her own life:

*You know, I changed what I thought was my career to having to come back to the UK, which was ... I never thought I'd be back in this country to be honest,*

*I was quite happy where I was. [...] I ... we've still got our lives ahead of us  
[...] But it's kind of on hold at the moment. [...] I didn't realise I was giving  
up everything then*

Anne was “quite happy” with her previous life and how the changes had been “huge”. Anne juxtaposes her own future with her mother’s. This aided acceptance of her life being “on hold”. Although Anne adopted a pragmatic approach to how her own life has been affected, the impact of the change is tinged with regret at not realizing how much she was “giving up”.

Kate compared her situation to that of her peers, and felt that caring for her father had deprived her of “enjoying stuff”:

*I don't feel like a normal twenty-seven should do. I just feel like too much  
things have changed and ... you know, I ... I should be out there living my life,  
enjoying stuff, and I just don't feel like I've had that opportunity to. So it's ...  
yeah its bad.*

For Kate there was a sense not just of her life being on hold, but of being “held back”. This stemmed from a sense of duty:

*I want to do more stuff but I can't; I feel like my Dad holds me back...He's not  
holding me back, it sounds awful to say that as well, but I feel like, as if there's  
no-one here for him so I need to be here.*

**“It’s splitting me”:** All participants expressed difficulty managing their parent’s illness as well as other responsibilities, but the personal impact and reasons for the



burden varied. Anne describes this process as “*juggling*” and approached it with a degree of humour:

*I suppose it's a part of my life. It's not something I would want to do, but it's not particularly a huge issue, its more ... its more juggling the time with him [baby] and Mum; you can guarantee accidents always happen at the same time [laughs] [...] I think err ... sometimes I do feel quite guilty, because you ... sometimes you snap, or you say 'Come on, let's get going', and its pushing my Mum, moving her because ... I have him as well. So it's ... it's not always easy on her in a sense, it's splitting me.*

Anne perceived herself to have adapted to the situation, yet still says it is “*not easy*”, and felt “*split*” by her mother and young son physically and emotionally leading to guilt. Anne reflected on how her mother is metaphorically as demanding as a “*baby*”, drawing analogies to her son and how life now lacks spontaneity:

*I can't even just nip to the shops, for example, I can't leave my Mum here. So in that sense it's like having a baby [laughs]*

Ben describes the shock of responsibility and the sense of isolation and abandonment he experienced being put in charge of a family business:

*In the past few years I've been in tears in that workshop, practically having breakdowns standing there, just because ... I could ... I was not prepared for it at all.*

Kate struggled with feelings of failing her father as she was unable to support him at home:

*I felt like I'd failed him for that, because maybe I should have took the time out to look after him and give him a bit more attention, but ... I didn't. I couldn't do it.*

Sole decision-making responsibility and needing to ensure “*everything is right*” for her father highlights Kate’s sense of isolation and the enormity of the burden she feels over decisions she makes “*carrying that to my grave*”:

*So everything else is just ... my decision. [...] I've got nobody to make the decision with, [...] I've got a lot of guilt, I've got a lot of hate ... and I ... I've got to live with that for the rest of my life. But ... I'll obviously regret it when he's gone. I think maybe I should have done more with him, or ... yeah, but ... hard.*

### **Theme 3: “I feel like she can get things off her chest”**

This theme considers issues related to concern for the non-affected parent. For three of the participants this was predominant and was related to awareness of potential caregiver burden and the limits this placed on their non-affected parent’s life. Diana worried about her dad having time for himself, and taking a break from caregiving responsibilities:

*But my main concern is my Dad really more than anybody because, as I say, he's there day in, day out, and okay he goes out a little bit, but then ... you know, I worry about him, that I don't want him to feel that he can't ask somebody for help, you know. And that's why I've said to him ... and he's sort of said to me 'Oh no I'll be fine, I'll be fine'.*

Diana offers her father support, but her father appears to downplay the impact of his wife's diagnosis on him, this feeds into Diana's concerns for him.

Ben placed his concerns for his mother within the context of long-standing family relationship dynamics. Ben's concerns for his mother resulted from a recognition that his mother will always be a constant in his father's life:

*I just tend to find myself worrying more about ... about Mum. I just ... I think that's the thing, is that I think the lasting ... the lasting thing of my existing relationship with Dad is that ... I was always closer to Mum, and that now, in this ... at this point my main concern is that Mum's alright and supporting her [...] So ... I know that Mum will look out for Dad, and I'm ... my priority seems to be more well I'll look out for Mum.*

Ben spoke about the sorrow he felt for his mum and how she was now "doomed" by his father's diagnosis denied of a relaxed retirement. Ben attempts to see his father's dementia diagnosis through his mother's eyes and empathise with her experience and the impact this may have on her:

*I know she was looking forward to ... them being able to retire and do stuff together. [...] Dad's not the same ... person, he's changed; it's a subtle change, but it's enough of a change that if you've been married to someone for forty-odd years that's not the same person; looks the same, sounds the same, isn't ... isn't quite the man that she was in love with, you know.*

The powerlessness in the support participants were able to provide the non-affected parent was acknowledged by Matthew:

*I see her upset and it's like ... wishing you ... you wish you could do something, like if it was an organ and you can give it to someone, just so ... even if it just stopped the progression you'd ... you'd do that, I' feel like I'd want to do that straightaway, but ... I can't, got nothing to do about it is there?*

Matthew saw his role as being someone for his mother to talk to:

*I try and talk to her about it, but then it gets ... easier and easier. Well not, it doesn't get easier, but ... I just feel like she can get things off her chest and stuff like that.*

As a result Matthew assumed the role of confidante, by adopting this role he was happy for other family members to share their concerns with him and in return felt that this helped him to cope:

*I think I feel stronger because I know how bad everyone else is going to be, so I know ... I think to myself 'I have to be stronger for them', for when they need the help or whatever.*

#### **Theme 4: “You’re never going to understand until you are in my position”**

This theme examines both informal and formal support for participants. Most participants valued being able to talk to others about their situation, but limits in the utility of this were apparent. Ben preferred talking to family members rather than friends as he found that communication with them had become superficial: that he was “trotting” things out, retelling his story but without the emotional involvement:

*I find myself talking to them, and then you kind of ... I find myself saying the same thing. [...] is it ... am I really thinking about that or is that ... is that just an ... that's just how I've decided I feel about it and now that's just what I say.*

Others such as Kate preferred not to talk. Kate's reluctance was perpetuated by her perception that others could not understand her situation as they lacked shared experience. Even though the experience was "killing her inside", Kate rejected others' attempts at empathy choosing to isolate and distance herself, she also appeared to worry about what to say to others:

*People always ask me work 'Oh you must be going through a really bad time, I understand', well you're never going to understand until you're in my position, and that really gets my back up. But it ... it's difficult, I mean what do you say to people? [...] I'll sort of like distance myself, but I will never say like 'This is killing me inside'. And it is, but I ... I don't talk about it to my friends because I just feel like that's something that people don't need to know.*

Kate's reaction to the concern of others for her was not just limited to friends and colleagues, her sense of not being understood extended to the HCPs involved in her father's care:

*Every one of them in that hospital turned round and said 'I understand your position, it must be really difficult', and I thought 'You don't. You're never going to understand it because you're not me.*

Matthew and Ben found it easiest to talk to friends who, although did not have parents with YOD, had relatives with either a neurodegenerative or terminal illness. Ben reported how this was one of the most "helpful" experiences in identifying with the illusion of coping with a parent with a degenerative illness and the reality of it:

*It kind of just gives you a bit of ... perspective on it. And I think that's the hard, one of the hardest things to get.*

However, Ben reported that what would have helped most would have been to have spoken to others in the same situation:

*...it would probably help ... help more because, like I say, you might not ... necessarily have to go into the real ins and outs of it ...you can ... you can get a bit of insight into ... into how the situation looks from the outside right? Because it's really difficult when you're stuck in the middle of it.*

Participants suggested that formal services needed to be more proactive in addressing and attending to their needs. Anne felt that even though she was a health care professional navigating health care services was often difficult:

*I think if I hadn't been a nurse and I didn't understand the systems and how...how awkward they can be I wouldn't be able to access half of what I have been able to.*

Ben also felt that services needed to be more visible:

*We kind of felt like we needed ... the support givers to be a bit more proactive ... and come and tell us stuff.*

Ben acknowledged that the support would be available if he asked, but he felt strongly that the support available for those with a parent with YOD should be formally stated from the outset:

*My brother, myself and my sister don't ... have never really had anybody ... say 'actually I'm coming to see ... to see you guys, to see how you're getting on' or to get ... to provide sort of ... support in that respect. I ... I think that ... they would ... they would happily do it if we said 'Oh well actually can I*

*have a ...', you know, '... can we talk about it?', and they'd be more than happy to do it, but I don't ... it's not actually happened.*

### **Theme 5: “Hang on a minute, this could happen to ... to me”**

This theme looks at how participants' own lives were affected by their parent's diagnosis of YOD. It comprises 2 subthemes (i) “It just...knocks you about” addresses the threat of YOD to self (ii) “you only live once” considers how participants view their future.

**“It just...knocks you about”:** Participants spoke of “*shock*” upon discovering their parent's diagnosis. Kate described this in physical terms “*it just ... knocks you about. It's knocked me about, I still am as well*”. Even where family history was significant such as in Kate's, the diagnosis still challenged preconceptions “*And I think dementia is for someone who is old, not for fifty-three*”. Feelings of uncertainty led some participants such as Ben to question their own mortality. A dialectic is present between what is “inevitable” and what is “*possible*” in respect of his own dementia risk:

*I could find myself in Dad's situation, you know [...] well hang on, is this ... is this inevitably going to happen to me or is it possibly going to happen to me, have I got an increased likelihood?*

For Kate the dual uncertainty in respect of her father's health and her own potential genetic risk led to difficulties in planning ahead. Kate speaks of not being able to have “*any plans*” which imbues a sense of lacking direction. Kate defines her own risk of dementia as “*always a big worry*”, but despite this there is resistance to

genetic testing which would clarify her risk and reduce uncertainty; this appears to have come from her experience meeting a genetic counsellor:

*I think the worst thing that they said to me was 'If you've got it you would need to break the link', as in I couldn't progress in life to have children. And I just ... I can remember sitting there thinking 'You bitch'. Because I thought if my Dad would have known would I be here today?*

Kate's sensitivity to such comments led her to reflect on the trajectory of YOD. Kate expressed hopefulness that it can "*skip a generation*", yet shortly after this is contradicted by "*and I know more than likely one day I'm going to get it*". Reflecting the processing of uncertainty by Kate, there is an interesting dynamic between earlier comments regarding her risk being "*a big worry*" and her comments regarding the utility of ruminating on potential risk and how she considers the potential of other illness. Kate appears to be attempting to be pragmatic yet contradicts herself.

**"You only live once"**: All participants appeared to value life more as a result of their parent's dementia diagnosis and their own potential risk. For Ben "*living in the moment*" offered him a way of managing the uncertainty as well as giving himself time off from the burden of responsibilities he had taken on. This sentiment was echoed by Anne. Anne's approach to managing the uncertainty was informed by witnessing her mum "*scraping and worrying about her retirement*", but then not being able to enjoy it:

*She's got to retirement and actually ... she can't enjoy anything that she's saved or, you know, scraped and really worked hard for. [...]I've thought about it more than I would have done had this have not happened.*



Kate struggled more than any of the participants with her father's dementia and at times during the interview was overwhelmed about her future. There was defiance towards the dementia preventing her from living the life she wanted:

*I'm going to have children. I'm not going to let this stop me. And I feel like as if it is cruel ... if it is, but I'm not going to go ... my life, living a 50:50 chance that ... you know, I could get cancer tomorrow so what's the point in me worrying about dementia? If it happens it happens.*

### **Discussion**

This study investigated the subjective experiences of having a parent with YOD. Five main themes emerged from the analysis highlighting changes in relationships with the parent living with YOD, shifts in role and responsibilities, concern for the non-affected parent, the need for participants to be supported and the personal impact to self of the parental YOD diagnosis.

The experiences of participants in this present study showed some consistencies with previous published findings (Allen et al., 2009; Barca et al., 2014; Gelman & 2011 Millenaar et al., 2013; Nichols, 2013; Svanberg et al., 2010). However, this study also highlights important individual differences in the perception of, and response to, their circumstances. It provides unique insights into how young adults process the experience, navigate relationships and role changes, and how they manage the potential risks associated with a YOD diagnosis in terms of their own life stage when they are at an age of having their own family, relationship and career responsibilities.

The relationship between participants and their parent was defined by “anticipatory grief” and presence of an ongoing sense of loss. This had a significant impact on participants and how close they felt to their parent (Allen et al., 2009; Barca et al., 2014; Svanberg et al., 2010). The opportunity to review and renew parental relationships facilitated participants coping with the degenerative changes witnessed, but where denied, feelings of guilt, anger and frustration were exacerbated. The stress-process model of caregiving in dementia (Pearlin, Mullan, Semple & Skaff, 1990) suggests that relational deprivation and loss of closeness within a relationship can have a negative impact on caregivers and family members, with poor relationship quality affecting both emotional and physical well-being.

Participants adapted their own lives in light of their parent’s YOD. This took place in the context of role reversal. Increasing dependency of parents often magnified the ironies of dementia, particularly for those with families of their own. For some this evoked a sense of their own lives being interrupted and put on hold. This, combined with perceptions of their own life course being “out of sync” with that of their peers, illustrates how YOD impacted on participants’ own life stage development and the social norms associated with being a young adult (Harvey et al., 2003) and how it is likely to differ from offspring of those diagnosed with dementia over the age of 65 (Brown et al., 2012).

Most participants were not responsible for primary caregiving duties. However, distress associated with caregiver burden was prominent and manifested itself in feelings of guilt. There was individual variation in how participants managed this. Where caregiving duties were tangible, participants appeared to employ problem

focused coping strategies. However, when participants experienced a lack of mastery or control of the caregiving situation, feelings of hopelessness were experienced. It may therefore be that perception of the caregiving role and ability to manage it, rather than actual level of role, may be the most influential feature in the level of burden experienced by individuals (Pearlin et al., 1990).

In accordance with other studies, concern for the non-affected parent was often the predominant worry (Allen et al., 2009; Barca et al., 2014; Millenaar et al., 2013). By providing space and support for the non-affected parent, participants assumed the role of protector as this was perceived to minimize burden for the non-affected parent. However, this concern could be constrained by existing relationship quality with the non-affected parent (Barca et al., 2014). The triadic relationship involving the child and both parents was strongly influenced by the previous relationship as well as relationship changes.

Participants' own care needs were many, but central to this was for others to understand, identify, and empathize with their situation. Lack of personal experience of YOD acted as a barrier to communication. Participants felt that they were better understood by friends who also had experience of illness within their families. However, the opportunity to meet with others in a similar situation to themselves was deemed invaluable. This adds to the findings of Barca et al (2014) that participants desired support groups which were personally meaningful to them.

Participants in this present study felt that their own needs were not specifically considered by HCPs. Although HCPs supported the non-affected parent, participants

felt that they lacked visibility and a presence in their own lives. Participants wanted HCPs to respond proactively to their needs to help them manage feelings of stress, burden and guilt and to enhance their understanding of YOD. Although recent policy drivers such as the “Dementia Triangle” (Royal College of Nursing, RCN, 2013) recommend that the whole family is considered within dementia, in practice focus is on the primary caregiver or partner (Roach, Keady & Bee, 2012; Svanberg et al., 2010a; van Vliet et al., 2010).

Participants experienced uncertainty about their own future. Most were able to manage their worry, but at times it overshadowed their lives. Fears over risk were managed by adopting mindful coping strategies which focused upon living in the moment rather than looking ahead and trying to live their lives before the possible onset of any dementia. This potentially helped participants cope with the day to day realities of YOD (Allen et al., 2009). Only one participant had been offered the option of genetic testing. Her reaction to such testing and implications in terms of her own future fertility was similar to findings reported in studies addressing attitudes to pre-symptomatic testing in those at risk from Huntington’s Disease (Duncan, 2008; Taylor, 2004; Wahlin, 2007)

### ***Study limitations***

There are several limitations to this study. Due to the rural location in which the research was carried out, the potential pool of participants to draw from was small; this created greater homogeneity, but meant that recruitment was limited. Potential participants were often not present at clinical appointments where information about the study was given out. Their awareness of the study was dependent upon parents

sharing the information. The interviews were carried out at a single point in time, which meant participants were often recalling events that had happened some time ago. The experience of having a parent with YOD is likely to vary as time passes from initial diagnosis to palliation. To map the YOD journey experience more coherently, future research could focus on obtaining longitudinal qualitative data sets either at different time points or disease phases within the dementia pathway.

### *Clinical Implications*

This study has shown how young adults who have a parent with YOD are a population who potentially go unnoticed by HCPs. Current service provision focuses on the primary caregiver and services are designed with that in mind (Roach, Keady & Bee, 2012; Svanberg et al., 2010a). Given the isolation experienced by participants in this present study, both from services as well as their peer group, it is vital that dementia services take account of the needs of the whole family. Although it may be easier to identify children that are still living in the family home, this should not take away from the impact a parental diagnosis of YOD may have on young adults living away. Young adults have differing needs, due to their specific life stage and personal responsibilities. It is important that HCPs take a full family history and that their availability to other family members is made clear. This should be undertaken in a proactive rather than responsive manner from an early stage.

Young adults with a parent with YOD may benefit from one-to-one support in understanding the diagnosis, disease trajectory and personal implications in terms of potential genetic risk. It is important that HCPS also acknowledge the role played by young adults in supporting their non-affected parent and how they are able to manage

their own lives in light of the YOD. Attempts should be made by HCPs to develop support groups for those who have a parent with YOD. This is something participants in this study identified as being valuable (see also Barca et al., 2014). Face-to-face groups or the development of an online forum, may suit this age group.

### ***Conclusions***

This study reveals the personal impact of a parental diagnosis of YOD for young adults in terms of relationship changes, impact on own life and threat to self.

Participants experienced isolation and stress as a consequence. HCPS working within dementia services need to be mindful of the needs of this population and provide wider family support to counteract feelings of isolation and lack of support.

**Table 1: Background participant information**

Pseudonym	Age at time of interview	Age at time of parent's diagnosis	Parent with YOD	Age of parent at time of diagnosis	Time since diagnosis*	Type of YOD	Family demographics
Kate	27	25	Father	53	1 year 5 months	Frontal lobe dementia	Only child parents divorced, lives alone Father in care home, has a partner
Anne	36	30	Mother	60	5 years	Alzheimer's Disease	One brother living abroad. Lives in mother's house with, mother, partner and young baby. Primary carer for her mother with assistance from carers.
Diana	35	33	Mother	63	1 year	Alzheimer's Disease	Lives with partner and young baby and daughter aged 4. Parents live 3 miles away. Brother nearby.
Ben	36	35	Father	63	2 years	Frontal lobe dementia	Lives next door to parents, brother nearby, sister has moved away.
Matthew	23	21	Stepfather	51	1 year 6 months	Alzheimer's Disease	Lives in family home with parents and 3 siblings aged 14, 17 and 24). Been brought up by mother and stepfather since 18 months old

\*confirmed by nursing team

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**SECTION 3: DISCUSSION PAPER**

### **Discussion Paper**

This section considers the main findings of the thesis and implications for clinical practice, and future research. It concludes with a reflective commentary on the process of data collection and analysis for the research study.

#### **Main findings of the thesis**

This thesis examined the psychological impact of having a parent with a progressive neurodegenerative condition with specific reference to Huntington's Disease (HD) in the literature review, and Young-Onset Dementia (YOD) in the research study. The findings from the integrative literature review suggest that growing up in a family affected by HD presents unique challenges. It was reported that younger age at parental onset of HD was associated with an increase in psychological difficulties and poorer psychological outcomes in adulthood (Folstein et al., 1983; Decruyenaere et al., 1999; Van der Meer et al., 2012). Family discord and difficulties were prominent in the studies reviewed. This was in part attributed to the wider consequences of the pronounced behavioural symptomology of HD (Folstein et al., Sparbel et al., 2008; Vamos et al., 2007; Van der Meer et al., 2012; Williams et al., 2009).

How young people learned of HD and their own potential risk of developing the disease was found to be related to coping and psychological well-being. Open communication within families was seen as facilitating this (Forrest-Keenan et al., 2007, 2009). Pre-symptomatic testing was reported to resolve feelings of uncertainty for individuals, irrespective of the test results (Duncan et al., 2007), and was found to promote adaptive coping. However, reflective of the general levels of genetic testing uptake, the majority of participants in the studies reviewed had not undergone pre-

symptomatic testing. Feelings of uncertainty were therefore prominent in the papers reviewed. This led to young people describing feelings of isolation from their peer group as they struggled with uncertainty over their own potential HD risk (Korer & Fitzsimmons, 1987; Sparbel et al., 2008). For some this uncertainty led to reluctance to consider long-term plans (Forrest-Keenan et al., 2009; Sparbel et al., 2008).

Assuming caregiving responsibilities magnified this as young people provided care in the presence of their own potential personal risk (Sparbel et al., 2008; Williams et al., 2009). Opportunities to meet and discuss experiences with others in a similar position appeared to be limited, but valued by young people (Forrest-Keenan et al., 2007; Williams et al., 2013).

In the research study participants reported the impact of a parental diagnosis of YOD. They described how pre diagnosis relationship quality impacted on their relationship with their parent with YOD. Participants detailed how they managed changes in the parental relationship in the presence of YOD and the sense of ongoing loss brought by a dementia diagnosis. They felt it was important to adapt communication to meet the changing cognitive status of their parent, but limits in understanding and communication changes could lead to participants experiencing frustration. In order to support their parent with YOD, they reported adjusting their own lives as they adopted roles and responsibilities which conflicted with their own life stage. Some participants described this as making them feel that their own lives were on hold. This was related to burden and guilt.

In terms of relationship triads between participants and their parents, some expressed a duty of care to their non-affected parent in order to enable that parent to fulfil

primary caregiving responsibilities for the parent with YOD. This was influenced by prior relationship quality. Participants expressed feelings of isolation from peers and a lack of understanding as they tried to manage the impact of the parental YOD and the uncertainty it brought to their own lives. This impacted on their outlook on life. The opportunity to be able to talk with others in a similar situation was highlighted by participants as being something they would value. Service provision which was more visible to family members other than the primary caregiver was also emphasized.

From both the review and research study, it is evident that participants growing up in families affected by a progressive neurodegenerative condition may experience significant distress as they try to manage their parent's declining health, as well as their own responsibilities, and potential personal risk of developing the condition. It is important therefore that health care professionals (HCPs) are aware of the impact a parental diagnosis of a progressive neurodegenerative condition may have and develop timely psychological interventions and support.

### **Implications for clinical practice**

There are a number of different implications arising from the literature review and the empirical research for clinical practice in relation to the needs of individuals with a parent with HD or YOD. These will be discussed under four subheadings: (i) Reducing barriers to services (ii) Support in relation to genetic risk (iii) Family interventions (iv) Support groups.

(i) *Reducing barriers to services and assessing need*

It is clear from this thesis that young people growing up in HD families and those with a parent with YOD are at risk of being invisible to services. The review paper identified that there may be several barriers which prevent young people in families affected by HD from seeking psychological help, and prevent HCPs from being able to access the young people. It is important that HCPs are mindful of these. The levels of family disruption reported within HD families (Folstein et al., Sparbel et al., 2008; Vamos et al., 2007; Van der Meer et al., 2012; Williams et al., 2009), and the reluctance by some families to discuss a young person's potential risk of HD, may lead to gate-keeping by parents in respect of service involvement. This may manifest itself in young people not attending family appointments (e.g. with genetics teams) and as a consequence not having the information to access services themselves.

Young people may also be worried that accessing services independently may impact on their parent's care or create family discord. Young people may also perceive that such services have no personal relevance if they are unaware of their own potential HD risk. It is therefore imperative that the whole family is identified and involved in HD services from an early stage, and that HCPs work with the whole family and make the service personally meaningful to young people. This requires HCPs being mindful of family systems and the dynamics therein.

A different set of barriers may face those with a parent with YOD. The National Institute for Health and Care Excellence guidelines for Dementia, states that:



*Younger people with dementia have special requirements, and specialist multidisciplinary services should be developed, allied to existing dementia services, to meet their needs for assessment, diagnosis and care.*

*(Section 1.1.2.1, NICE, CG42, 2006)*

Indeed in Wales there is specific drive to create young onset dementia services as a consequence of the Dementia Vision for Wales (2011). However, across Wales and the UK provision is still patchy. Specific reference to the psychological needs of families affected by YOD is absent from both NICE and other guidance (NICE, 2006; Roach, Keady & Bee, 2012). It is important that these needs are recognised. Recent policy drivers such as the Royal College of Nursing “Dementia Triangle” (RCN, 2013) recommend that the whole family is considered in dementia services generally. However, in practice the focus to date has been on the primary / spousal caregiver (Roach et al., 2012; Svanberg et al., 2010a; van Vliet et al., 2010).

This was evident in the research study where it was clear that some participants interviewed felt that HCPs lacked a presence in their own lives and needed to be more proactive in their approach. Anne, who herself was a nurse, highlighted in her interview how “*the systems aren’t always easy*” which suggests if she struggles to navigate health and social care systems for her parent, then others potentially encounter greater difficulties:

*I think if I hadn’t have been a nurse and I didn’t understand the systems and how ... how awkward they can be I wouldn’t have been able to access half of what I have been able to. I think for people who would want to do this who don’t necessarily have that insight, I think it would be incredibly difficult with regards to extra support.*

It is vital that services take a full family history. In an interview-based study of clinical practice in the care of individuals and families affected by YOD, Roach et al. (2012) found that full biographical information was not always collected by HCPs. Roach et al. (2012) made key recommendations that: (i) HCPs ask about the whole family and the roles of each member within the family; (ii) timelines are created to plot key biographical data to understand how families functioned pre and post-diagnosis; (iii) HCPs engage with all family members in care planning and provision, and (iv) clinical decision making of the care of the individual with YOD is informed by this knowledge. The findings of Roach et al. (2012) potentially have wider applicability to HD. In respect of HD, where young people may be more likely to be less than 18 years of age and living at home, it is important for HCPs to be aware of the Social Care Institute for Excellence guidance (SCIE, 2007) which states that there is a responsibility for adult health care providers to ask individuals about children at home.

***(ii) Support in relation to potential genetic risk***

For young people growing up within HD families, consideration of own potential risk of developing HD may become a predominant concern during adolescence. Although pre-symptomatic HD testing is not available to individuals until 18 years of age, it is important that dialogue about potential risk is encouraged in families by HCPs from an early stage. HCPs should work closely with families and target interventions to promote open communication, and use psycho-educational interventions to enhance understanding of HD. HCPs, in particular genetic counsellors, should provide support to young people as they transition to an age where they are eligible for testing. It is

therefore vital for services to continue supporting the young person through the process of genetic testing should they choose to pursue this.

Sensitive communication surrounding the issue of genetic testing is vital irrespective of disease type. It was evident in the experience of one of the participants in the research study how communication in such settings could have a significant impact on an individual's experience of, and engagement with, genetics services. The genetic links are not so clearly defined with YOD, given the heterogeneity of dementias within this classification. It is nevertheless important, where potential genetic links are identified, for individuals to be given the opportunity to be referred to genetic services in line with NICE guidance (NICE, 2006).

*(iii) Working with families*

The literature review and research study both reported how individuals with a parent with HD or YOD may experience difficulties within families as a result of the respective conditions. In HD families, discord and disruption was prominent in the literature (Folstein et al., 1983; Sparbel et al., 2008; Vamos et al., 2007; Van der Meer et al., 2012; Williams et al., 2009). In addition the research study highlighted how the relationship triad between the (adult) child and both parents was influenced both by previous relationship quality and relationship changes as a consequence of the YOD.

In both circumstances family focused interventions may be valuable for young people experiencing significant distress and disruption. Family systems therapy (Bowen, 1978) offers an appropriate theoretical framework for therapists to work in with such families. This posits that families change dynamically as they are required to adapt to

situational changes. However, a single family and family members will all respond differently to such changes despite inter-dependence. When major life events occur and problems arise that do not follow the typical life-stage of development (e.g. HD or YOD), then families are forced to readjust in order to manage these atypical circumstances. Family systems therapy acknowledges that individuals may need to redefine their sense of self as a result of the life event and this will impact on other family members. A key feature of the therapy is for individuals to recognise such shifts, both within themselves and others, in order for family members to work through the difficult emotions and circumstances experienced. There is preliminary evidence of the effectiveness of family systems therapy in YOD (Gelman & Greer, 2011).

*(iv) Support groups*

Both the literature review and the research study highlighted how participants felt isolated by their respective situations. This was related to relationship changes, role shifts, uncertainty, and a sense of peers not being able to understand them. It was clear from both papers that participants would value the opportunity to talk to others in a similar situation. HCPs should encourage peer support and mentoring schemes whereby individuals are put in contact with others in a similar position to share experiences and potentially reduce feelings of isolation. At a local level this may be achieved through the setting up of support groups linked to the clinics responsible for the parent's care, or by establishing private online groups via social networking sites such as Facebook where individuals can share their experiences.

Signposting individuals to national organisations who have their own established support groups may also help. For individuals growing up in families affected by HD, groups such as the UK based Huntington's Disease Association (HDA), and the international Huntington's Disease Youth Organisation (HDYO) may be beneficial. Both the HDA and HDYO offer moderated online forums and age specific advice. In particular the HDA website is well developed and offers age specific resources and telephone support. It also runs summer camps for young people up to 16 years of age in the UK providing individuals with the opportunity to meet with other young people in a similar situation. However, for individuals with a parent with YOD such support is less well developed. The Alzheimer's Society has a thread on its "talking point" forum, but it is not private and the majority of posts are from partners and spouses. This may therefore be off-putting for individuals who seek to share their experiences of having a parent with YOD only with those in a similar situation to themselves. For example one participant in the research study (Kate) reported that:

*I think somebody who's ... who's older, I just ... there's no point because I feel like ... you know, they've got dementia and they're seventy or eighty, that technically doesn't bother me so [...] Yeah I think if there was somebody with ... you know, a parent who was at the same age as my Dad, that would be ... quite interesting.*

HCPs should also signpost individuals to young carer organisations. In England these are available to children aged 18 years of age and under, but in Wales young people may access young carer services until the age of 25 (Welsh Assembly Government, 2013).

**Implications for theory and future research*****Individuals with a parent with HD***

It is clear from the review paper that there is a dearth of quality research into the experiences of having a parent with HD. The review paper revealed how extant quantitative research into young people growing up in HD families has methodological shortcomings related to recruitment, lack of appropriate control groups, and sample selection (Decruyenaere et al., 1999; Folstein et al. 1983; Vamos et al, 2007; Van der Meer et al., 2006, 2012; Williams et al., 2013). Future quantitative research into the experiences of having a parent with HD therefore needs to be methodologically robust. The qualitative research into the experiences of growing up with a parent with HD also had methodological weaknesses, focusing on thematic or content analysis with only two of the studies (Forrest-Keenan et al., 2007,2009) being informed by a particular approach such as Grounded Theory (Glaser & Strauss, 1967).

Extant qualitative studies into the experiences of growing up in a HD family have been cross-sectional in nature (Duncan et al., 2007; Forrest-Keenan et al., 2007, 2009; Korner & Fitzsimmons, 1987; Sparbel et al., 2008; Williams et al., 2009). There are still opportunities for cross sectional qualitative research in respect of IPA studies examining the lived experiences of adolescents and young people within HD families. However, given the reported levels and experiences of family discord within HD families (Folstein et al., 1983; Sparbel et al., 2008; Vamos et al., 2007; Van der Meer et al., 2012; Williams et al., 2009), it would also be interesting to qualitatively explore relationship dyads between children and the parent with HD. Potentially the triadic relationship between the child and both parents could be explored, where appropriate,

in order to consider how family dynamics and circumstances impact on the individual. There is also a clear need for well-designed longitudinal qualitative interview-based research. By adopting a prospective approach following young people within HD families over time, it would be possible to map the individual experience not only of growing up in HD families, but also, in separate studies, the individual experiences and processes of pre and post genetic testing and counselling.

Future quantitative research may also consider the development of additional measures to screen for distress and burden in young people within HD families. This would extend the development of measures such as the HD-teen inventory (Williams et al., 2013) which may then inform HCPs and help them to identify and target those in need of tailored support.

### ***Individuals with a parent with YOD***

In the past five years there has been an increase in research into the experiences of having a parent with young onset dementia. The evidence base, however, is still small and underdeveloped with studies being cross-sectional in design (Allen, Oyebode & Allen, 2009; Barca, Thorsen, Engedal, Haugen, & Johannessen, 2014; Gelman & Greer, 2011; Millenaar et al., 2013; Nichols et al., 2013 Svanberg, Spector & Stott, 2010a; Svanberg, Stott & Spector, 2010b; van Vliet et al, 2010). The research study in this thesis addressed the issues facing young adults with a parent with YOD using IPA methodology. Although as far as possible a homogenous sample was sought, there were differences in time since parental diagnosis and type of YOD. Future IPA research could therefore consider investigating the experience of individuals at a particular point since diagnosis, or whose parent has a specific type of YOD (e.g.

Alzheimer's, frontal lobe, vascular) as there can be differences in the behavioural and cognitive presentations of the different types. In the research study participants either lived at home or within 3 miles of the family home. It would be valuable to understand how a parental diagnosis of YOD may impact on young adults living and working away from home, as their experiences may differ.

Only the experiences of adults aged over 18 years of age were explored in the research study (see also Barca et al., 2014). Other studies have explored the experiences of individuals either in those under 18 years of age (Gelman & Greer, 2011; Nichols et al., 2013; Svanberg et al., 2010b) or in adolescents and young adults (Allen et al., 2009; Millenaar et al., 2013). Given the different life stages and experiences of this age range, it would be valuable to undertake prospective longitudinal qualitative studies to map the experience of having a parent with YOD more coherently from initial diagnosis to palliation with both children and young adults. This would enable insights to be gained into how changes in a parent's health over time may impact on a young person's psychological development. Research could potentially focus on obtaining qualitative data either at different time points or disease phases within the YOD pathway. Such research may also be useful in identifying continuity of care, and times when children and young adults may feel in most need of support. Exploring relationship dyads between child and YOD parents is also an area that future research needs to address. In the research study it was observed how pre-YOD diagnosis relationship quality impacted on relationship strength post diagnosis. It would be interesting to see if there were any differences in this as a result of gender or age. Moreover, the findings highlight the need for future research to explore relationship triads between individuals and both of their parents.



**Personal reflection**

It is important for me to personally reflect on the process of conducting research into the experience of having a parent with YOD, and the influence my own position may have had on the research and interview process. I also feel it is vital to consider the impact of participants' experiences on myself.

I came to the interviews as a trainee clinical psychologist, who was a similar age to the participants, and who had a background in health services research of patient and caregiver experiences in chronic and life-threatening illnesses. I was drawn to the research area by the experiences of a close friend whose mother, years previously had been diagnosed with YOD in her 40s. More personally I was conducting these interviews after receiving treatment for a life-threatening illness "out of sync" for my age and within six months of losing my father. Given the research area I feel that all these experiences, particularly those of serious illness and loss, may have potentially impacted on how I approached the research.

Having previously been involved in qualitative research, primarily employing thematic analysis, I approached the interviews with confidence and curiosity, for three reasons: (i) I always loved being actively involved in research; (ii) I was interested to see how approaching interviews from an IPA perspective differed from other forms of qualitative research; (iii) I was keen to see if and how clinical training informed and had changed my approach to research interviewing. From the very first interview with Kate I felt an urge at times to drop the researcher role of active listener and to slip into the therapist role seeking to work with difficult thoughts and feelings when the participant became distressed. This reminded me of the powerlessness I used to

feel as a researcher, and of why this had served as a motivation for me to move out of research into clinical training. This was something I remained mindful of throughout each interview.

The interviews heightened my awareness of clinical and research interviews in other ways, both in terms of similarities and differences. Similarities between clinical interviews and the IPA paradigm were evident when participants were probed about their thoughts and feelings in order to understand their subjective experience and their underlying cognitive and emotional processing. However, there were notable differences. Depending upon the client group within an initial clinical session, one may not always gain a full history or be able to clearly formulate the client's difficulties as rapport needs to be established and client's may not be willing to disclose. However, within this research setting I needed to build rapport and enable participants to feel comfortable in disclosing their experiences within the space of a single visit, with no benefit of therapeutic intervention from myself, the researcher. The fact that participants did talk and spoke in so much depth within the confines of a one hour interview, and some talked for the first time about their experiences, was truly humbling for me as a researcher and something which I may prior to clinical training have taken for granted when carrying out interviews.

During the course of the interviews I became acutely aware of my own subconscious sensitivities that I brought to the situation. I was surprised at how often "cancer" was mentioned in interviews and how thrown I was by it. In the first interview, from the outset Kate compared YOD to cancer. I could feel emotions stirring in me as she spoke, but knew I needed to keep both them and my words in check. I thought that by

the second interview, with Diana, I would be more prepared to manage this, but yet again towards the start of the interview cancer was mentioned and again I felt momentarily pulled away from the research. Discussions of loss regarding their parent also impacted on me. When Ben spoke of feeling robbed of a relationship with his father, feelings and memories of my own father were evoked as in the latter stages of his non-dementia related illness I had experienced this, although not in the context of relationship strain. Being mindful of these feelings within the research paradigm highlighted for me the “double hermeneutic” of IPA and how researchers, working within an IPA framework, are engaging in analysis “on-line” even during the interview process, responding to what participants say, reflecting on it and considering personal responses.

Looking back I feel that I had maybe always been a little tentative, perhaps even cynical about conducting IPA research, feeling it was a dark art and that there was nothing to actually discriminate it from thematic analysis. However, there was a point in the analysis where it suddenly clicked just how different it was from the qualitative studies I had previously been involved in. I could see how it offered me the chance to explore the process of the individual experience and individual nuances in a way I had not done before. This led me to review my prior conceptions and emerge myself in the data and analysis at a different level.

The quality of the interview data meant that writing up became a bit of a battle as I attempted to do justice to the participants’ experiences using IPA analysis within the confines of a 7000 word limit. It was very important to me that I did not compromise on this richness. Thus it became a labour of love for me. I also became mindful of the

flourishing research into the impact of having a parent with YOD. When I had planned the study and submitted my NHS ethics application in early 2012 there was a dearth of literature in the area. I felt ahead of the game, but suspending my studies for a year left me with the feeling that I had to some extent missed the boat. I am hoping that the experiences that participants' shared with me show that I had not and that their insights have a unique place within the emerging evidence base.

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**SECTION 4: UNIVERSITY AND NHS ETHICS AND R&D**