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**Title: How young people find out about their family history of Huntington's disease**

**Abstract**

Family communication about adult-onset hereditary illness can be problematic, leaving some relatives inadequately informed or ignorant of their risk. Although studies have explored the barriers and facilitators in family communication about genetic risk, such disclosure continues to raise questions about when, what, how and indeed whether to tell relatives. The process of disclosure is also dependent upon the way in which genetic information is realized and understood by recipients, but little research has explored their views. Our paper explores one group of recipients' experiences of finding out about a family history of the hereditary disorder Huntington's disease (HD). In-depth interviews explored descriptive accounts of how and when young people found out, their reactions to different communication styles and any impact on family relations. We recruited young people through the North of Scotland regional genetics clinics and the Scottish Huntington's Association. Thirty-three young people (aged 9-28) were interviewed. A qualitative thematic analysis was undertaken. The analysis revealed four main themes: having always known about HD, learned gradually, it was kept a secret, or HD was a new diagnosis. In particular, the timing and style of disclosure from relatives, and an individual's stage of awareness, were fundamental in structuring participants' accounts. The article draws attention to questions of when, how and indeed whether to tell children about this hereditary illness. It sits within a broader set of research and practice issues about what professionals and families should tell children about parental illness and genetic risk.

## **Background**

Family communication about adult-onset hereditary disease may include discussion of individuals' risk of disease, the risk to future children, and may influence other important life decisions (Wilson, Forrest, van Teijlingen, McKee, Haites, Matthews et al., 2004; Gaff, Clarke, Atkinson, Sivelles, Elwyn, Iredale et al., 2007). In general, health professionals encourage the disclosure of genetic risk to relatives but the responsibility for passing on such information ultimately resides within families - only in exceptional circumstances is confidentiality broken (Petersen & Bunton, 2002; Clarke, Richards, Kerzin-Storarr, Halliday, Young, Simpson et al., 2005; Forrest, Delatycki, Skene, & Aitken, 2007). Yet relatives may be uninformed, misinformed or inadequately informed of their risk (Pilnick & Dingwall, 2001; Forrest, Simpson, Wilson, van Teijlingen, McKee, Haites et al., 2003; Featherstone, Atkinson, Bharadwaj, & Clarke, 2006). Barriers and facilitators in family communication about genetic risk are documented (Gaff et al., 2007), but little research has explored recipients' perspectives, particularly those of children and young people (Gallo, Angst, Knafl, Hadley, & Smith, 2005; Metcalfe, Coad, Plumridge, Gill, & Farndon, 2008). This paper focuses on young people's experiences of finding out about a family history of Huntington's disease (HD), and how different styles of family communication may impact on their experiences of growing up with HD.

### *What is Huntington's disease?*

HD is an incurable adult-onset hereditary neurodegenerative disorder affecting around 1/10,000 individuals in Scotland (Simpson & Johnston, 1989). Signs include involuntary movements, cognitive impairment, and personality change (Van der Meer, Timman, Trijsburg, Duisterhof, Erdman, van Eldern et al., 2006). Affected individuals are likely to become dependent on others and have usually had children before their diagnosis is made. Each child of an affected parent is at 50%

risk of developing the illness in adult life. Consequently, many children witness the physical and mental decline of a parent, whilst also finding out that they have a risk of developing the disease (Forrest Keenan, Miedzybrodzka, van Teijlingen, McKee, & Simpson, 2007). Genetic testing can establish whether an individual carries the mutated gene for HD or not.

### *How and when do children find out about HD?*

There is scant literature on children and young people's experiences of finding out about genetic illness, or HD in particular; although some studies have explored adults' reflections of discovering HD (Cox, 1999; Etchegary, 2006). Etchegary (2006) reports four different routes in which individuals may *initially* find out their family history: (1) 'Something is wrong' - HD is unknown and families search to find out what is wrong with their relative; (2) 'Out of the blue' - discovery is immediate and there is no protracted period of wondering what is wrong; (3) 'Knowing, but dismissing' - individuals have a vague knowledge that a distant relative has HD, but social and/or geographical distance does not expose them to it; and (4) 'Grown up with HD' - awareness is gradual and HD is not deliberately hidden. Several dimensions may affect each pathway into initial awareness: timing (whether told in childhood, adolescence or adulthood); process (gradual or sudden); family communication style (from openness to total secrecy); social and geographic proximity to an affected family member; and access to sources of information (e.g. genetics service or HD association) (Cox, 1999:230). For those living with HD, finding out is likely to be a process of awareness based on factual and experiential knowledge (Downing, 2001). Hence Downing (2001) suggests a five stage continuum moving from: (1) being *unaware*, where there is no conscious attention about a problem being in the family; to (2) *pre-awareness* where concerns begin to be expressed from experience of HD, but there is no factual knowledge; to (3) *limited awareness* when the term HD is known, but there is limited factual knowledge e.g. about the risk to one's self or others; to (4) *detailed awareness* where there is a fuller understanding of the illness and personal risk; to (5) *refined awareness* - where someone gains certainty from genetic testing or a diagnosis.

One of the few studies of young people's perspectives (Easton, 2003) demonstrated that many children do sense 'something is wrong' before they are told, even as young as age five. Some also realized that HD was a secret and learned not to ask any questions, or to talk about, the family illness - as a consequence they carried misinformation which caused anxiety and concern (Easton, 2003). Thus, being told in childhood or adolescence may help young people cope, and deal with their risk better than being told later in adulthood (van der Steenstraten, Tibben, Roos, van de Kemp, & Niermeijer, 1994; Skirton, 1998; Forrest Keenan et al., 2007; Holt, 2007). When information is withheld on purpose individuals often feel resentful, hurt, angry and disempowered (Skirton, 1998; Klitzman, 2007). There may even be a "critical age" when the protective effect (or 'psychological buffer') of early disclosure culminates i.e. disclosure is less beneficial than it could have been (Skirton, 1998). An optimal time for telling children about HD may be in the approach to adolescence (Holt, 2007), but some studies report that finding out around this age, or in mid adolescence was harmful (Van der Meer, Timman, Trijsburg, Duisterhof, Erdman, van Eldern et al., 2006; Klitzman, 2007).

#### *Family communication about Huntington's disease and hereditary cancers*

Communication about HD may be particularly difficult as the disease can have a devastating impact on families and is associated with considerable stigma and taboo (Korer & Fitzsimmons, 1985; Vamos, Franz, Hambridge, Edwards, & Conaghan, 2007). Family rifts, secrecy and poor communication mean HD can appear unexpectedly 'out of the blue' when a relative is diagnosed (Cox, 1999) and children may still be living at home. Although some individuals actively choose to be open with their family (close and extended), others decide not to tell, or to put off telling (Forrest Keenan, Simpson, Wilson, van Teijlingen, McKee, Haites et al, 2005). Complex issues arise about when to tell, what to tell, who to tell and indeed whether relatives "need to know" (Sobel, 1997; Skirton, 1998; Hallowell, Foster, Eeles, Arden-Jones, Murday, & Watson, 2003; Klitzman, 2007).

Whilst some feel they are “doing right” by disclosing information, others feel burdened by this need and a strong desire to protect relatives from harm (d’Agincourt, 2001; Gaff et al., 2007).

Specific patterns of communication have been identified in families affected by hereditary breast/ovarian cancer (HBOC). Kenen, Ardern-Jones and Eeles (2004) observed five disclosure styles: (1) *Open and supportive*, where relatives spoke openly to each other about feelings/concerns, and used their family as a support system; (2) *Blocked (direct)* where family members would not talk about the disease and communicated this clearly to others e.g. by hanging up the phone; (3) *Blocked (indirect)* where individuals sent out signals that they did not want to discuss the illness and boundaries were more subtly drawn e.g. being uncomfortable when the issue was raised; (4) *Self-censored* where someone consciously limited what they said (this was *reactive or proactive*). Reactive self-censoring involved an individual sensing the other person’s unwillingness to talk and pulling back. Proactive self-censoring involved an individual choosing not to talk to certain family members because they didn’t want to cause them anxiety (e.g. children); and (5) *Use of Third Parties* where an intermediary was used to contact a relative for the individual. Hallowell, Ardern-Jones and Eeles et al. (2005) also identified three communication strategies in families affected by HBOC where parents chose between complete openness, limited disclosure or total secrecy.

#### *Family communication about genetic risk*

Overall, studies suggest that disclosure of genetic information is a process, rather than a one-off single event, in which several factors should be taken into account (Gaff et al., 2007). During this process individuals need to make sense of their own personal risk and to assess the receptivity of the recipient before disseminating this knowledge to relatives (Gaff et al., 2007). Individuals also reflect on how and what should be conveyed, and when may be the best time to tell (Gaff et al., 2007). Parents may have to make difficult decisions around the timing of disclosure - balancing a desire to protect children for as long as possible, versus the need to tell them in time to make

informed decisions (Gaff et al., 2007). Disclosure may also have two forms: *alerting* (where one's risk is announced); and *educating* (includes risk announcement and an explanation about the illness) (van den Nieuwenhoff et al., 2007). Differing styles of communication and disclosure practices have also been observed to vary within and between families, creating conflict and tensions (Forrest et al., 2003; Featherstone et al., 2006; Gaff et al., 2007). Whilst parents who 'selectively tell' may wait for their children to ask them specific questions (Gallo et al., 2005).

### *The Current Study; Focusing on How and When Young People found out about Their Family History of HD*

This paper focuses on the experiences of young people who live in a family affected by HD as little is understood about the appropriateness, timing and impact of disclosure or indeed about how risk can be or is conveyed within families. Although HD is a rare genetic disorder it offers a useful model to explore and compare psychosocial issues in other dominantly inherited disorders, and genetic illness in general (Brouer-Dudok deWit, Savenije, Zoetewij, Maat-Kievit, & Tibben, 2002).

## **Methods**

### *Theoretical and methodological framework*

The study is informed by a qualitative inductive method of data collection and analysis (Mason, 2002; Silverman, 2000) using elements of the grounded theory approach (Glaser & Strauss, 1967; Strauss & Corbin, 1990). This allowed us to explore the issues which emerged as most salient to participants, in contrast to imposing a predetermined framework upon their accounts (Glaser & Strauss, 1967). Nevertheless, the analysis was situated within a broader sociological framework drawing on theories about the social construction of childhood and family relations which highlight children's agency, rights, competence and reciprocity between adults and children (Finch & Mason, 1993; Alderson, 1999b; Lee, 2001). The accounts were also situated within recent models of family

communication about genetic risk (Cox, 1999; Downing 2001; Kenen et al., 2004; Etchegary, 2006) and research which has explored adolescents reactions to parents with a hereditary neurological illness (Power, 1977; Easton, 2003), particularly our main study which revealed four themes: Young People as carers, the worried well, those who cope, and those at risk/in need (Forrest-Keenan et al., 2007).

### *Recruitment and sampling*

Participants were drawn from the genetics service in Grampian (population 500,000), North East Scotland, and membership of the Scottish Huntington's Association (SHA). The clinical geneticists (SAS and ZM) and SHA advisors identified potential participants and invited them to take part by letter. If the young person was under 18 a letter was sent to their parent(s) asking them to pass on our study leaflet to their child. We did not invite participants who were considered by the clinical team and the SHA to be too vulnerable to participate (e.g. recent family bereavement). Participants were recruited between 2002 and 2005. Three sampling approaches were used: purposive sampling to include participants with as wide a range of demographic characteristics as possible (Mason, 2002) and theoretical sampling (Glaser & Strauss, 1967) to include individuals who were identified as experiencing communication difficulties in their families, as well as those families who were known to be more open. In the first year it was hard to access and recruit young people so we used a third method, snowball sampling (Atkinson & Flint, 2001), i.e. we asked participants and SHA advisors to pass on a study leaflet to other relatives. We also placed an advertisement on the SHA's website for young people (see [www.hdscotland.org/youth](http://www.hdscotland.org/youth)). We stopped recruiting when data saturation had been reached (Glaser & Strauss, 1967).

Fifty-one young people were invited to participate by the clinical geneticists (n = 16), an SHA Advisor (n = 23), or a family member (n = 12), of whom thirty-three agreed and were interviewed. Sixteen were recruited through the two clinicians, eleven through the SHA advisors, five through a

relative and one from the SHA website.

### *Data collection*

Semi-structured interviews were used to ask questions of interest but with flexibility to explore other emergent issues (Mason, 2002). The interview schedule was informed by a literature review, professional advice (from SAS, SHA Youth Advisor and Child Psychotherapist) and our previous research (Forrest et al., 2003; Forrest Keenan et al., 2005).

The interviews took place in participants' homes, a genetic clinic, or a neutral public venue. The majority were audiotaped with consent and fully transcribed. In three cases the tape recorder failed and notes were taken. In four cases the interview could not be taped as it was in a café and notes were taken instead. KFK (a sociologist and qualitative researcher) conducted all the interviews, explained the purpose of the study to participants, and ensured that they understood her role as a researcher was distinct from the genetic counselling services and the SHA. Background demographic data were collected using a short questionnaire at the time of interview.

### *Practical and ethical issues*

There are practical and ethical issues to consider when conducting research with children and young people (Alderson & Morrow, 2004) and particularly with those at risk of familial disease. This may be due to issues around access, informed consent and the sensitive nature of the research topic. In order to encourage young people to participate we offered to refund travel expenses and asked if they would like somebody else present for support e.g. a friend/relative (Gajraj et al., 1990). The participants were assured of confidentiality. An experienced Child Psychotherapist was consulted about how best to elicit the views and experiences of this group, and also how to protect them from any potential harm from taking part. Participants also had the opportunity to contact a genetic counsellor or Youth Advisor after each interview. Information about local health services was



offered to any young person as requested or perceived as necessary (e.g. NHS Grampian's 'Link Guide' and Glasgow's 'Youth Health Map').

The study was approved by the Grampian Research Ethics Committee.

### *Data analysis*

The analysis was conducted in two main phases. In phase one a thematic analysis of the transcripts and the interview notes was undertaken using the constant comparison method (Glaser & Strauss, 1967; Forrest Keenan, van Teijlingen & Pitchforth, 2005). KFK identified themes by reading and re-reading the transcripts and coded the main issues drawing upon aspects of Strauss and Corbin's (1990) approach to coding qualitative data by using a system of open, *in-vivo* and axial coding. Different segments of the data were grouped together into separate categories, which allowed reflection on the major themes and the relationships among them. The analysis was an iterative process, which was ongoing throughout the fieldwork. A second coder (EvT) read the transcripts in their entirety and coded a subset of the transcripts independently. Any differences in the coding were minor and reconciled through discussion. Both coders agreed on the main themes and sub themes which emerged. The second phase of analysis sought to situate participants' accounts within the theoretical framework outlined above.

### *The participants*

There were 21 females and 12 males; all were 'white.' There were four age ranges (with a mean age of 20.5): 12 and under (n=2), 13-18 (n=12), 19-25 (n=14) and 25-28 (n=5). We define young people as those aged 13-25 (Furlong & Cartmel, 1997). Children were defined as aged 12 years and under, and adolescents aged 13-18 years. Risk characteristics and domestic situation are detailed elsewhere (Forrest Keenan et al., 2007).

## **Results**

While wider findings from this study have been reported elsewhere (Forrest Keenan et al., 2007) the focus here is on the neglected aspect of young people's views and experiences of finding out about a family history of HD. The results are in two parts, focusing at first on when participants initially found out, who they were told by and any personal contact they had with an affected relative. We then move on to descriptive accounts of how young people found out, their reactions to different family communication styles and any impact on family relations. Six cases have been selected to reflect the main themes and subthemes identified (Table 1) and to provide an in-depth analysis of some individuals' experiences.

### **Initial finding out about HD**

Participants reported first learning about HD in childhood (n=21), adolescence (n=6) or as young adults (n=6). Table 2 summarises from whom each participant initially learned about a family history of HD, and when. For the purpose of this paper initially finding out means the participant knew the name Huntington's disease and that a relative had the illness. Table 3 summarises any personal contact participants had with an affected parent/grandparent when they initially found out.

### **How and when participants found out**

#### **Theme 1 - "Always known about Huntington's"**

Fifteen participants described having "always known" about HD since childhood. Five lived with an affected parent as a child, three lived with an affected grandparent, and seven had contact with an affected grandparent, but did not live with them. In this group the majority described being told initially by a female relative (mother, grandmother or sister), although two stressed they had found out by themselves, and one initiated communication with his father after being told by his older sister. The following account is from an older participant (Alison, age 28, 50% risk) who was purposely included in the study because of her in-depth reflection of what it was like to experience

a particularly traumatic childhood and adolescence, which she attributes to “Huntington's.”

### **Case 1 – Alison “I've always known about Huntington's ... but there are stages”**

When she was growing up Alison lived with two relatives who were affected by HD, her grandfather in childhood, and father in adolescence. She had “always known about Huntington's” being told about her grandfather's illness by her parents as a child, and of her own risk. Her grandfather (and grandmother) moved in with Alison's family when he “got really bad” to enable her gran to care for him with the support of her son and daughter-in-law. Thus, Alison described how:

“I've always known about Huntington's because we lived together ... all I can remember about my granddad is this grumpy man sitting in the big huge wheelchair ... he couldn't move and his speech was very poor ... And my gran used to cry for hours, with me sat on her lap giving me a cuddle ... and I would hear granddad screaming and shouting ... So you see things and you hear things ... But my gran believed I got told when I was too young, I was 6 or 7 ... I don't actually remember being sat down and told but I remember sitting on my gran's knee and me saying 'oh I'm going to die, I'm going to die'. It sounds really funny looking back on it, but my gran was saying 'don't be silly', she's always said 'I'll pray for you every night' ... That's why my gran was more of my hero to me. Because my gran looked after my granddad and went through a hell of a lot.”

Alison has no recollection of any disclosure “event” taking place in her childhood, as she has “always known about Huntington's” initially because of seeing (and hearing) how the illness affected her grandfather, and the impact this had on his carer, her gran. She believes she was told by her parents at the age of 6 or 7, and recalls saying to her gran as a child that she felt she was “going to die”, insinuating this was due to knowing about her risk of Huntington's. Whilst this period of her life was clearly distressing for Alison and her gran it seems to have initiated a huge closeness between them.

After her grandfather's death Alison's gran moved back to her home town leaving Alison with her mum and an increasingly affected father. Alison kept in close contact with her gran and went on to describe how:

. “Even though it was never spoken about, I know she knows some of the things that wasn’t spoken about ... It’s hard to explain because it was little things that she used to say ... that makes you think maybe she does know ... about the behaviour problems, the beatings and ... that side of it.”

Here Alison joked nervously that she had a lot more to disclose, i.e. physical abuse by her dad during her teens and witnessing some highly inappropriate sexual behaviour (Forrest Keenan et al., 2007). Although Alison and her gran never openly discussed this abuse Alison felt she at least knew “some of the things” having previously cared for two affected relatives. She remained Alison's one constant source of support throughout her adolescence and into adulthood. Thus for Alison:

“Childhood wasn’t good because I used to get hit, and there was a lot of social workers in and out ... And because of the Huntington's my dad was in and out of the psychiatric hospital ... and there was other behaviour things that he used to do that I know was part of the disease ... Which is why I think I was so close to my gran because she understood, *and* she’d looked after granddad’s dad. But I’ve never gone into this with anyone before apart from a counsellor ...”

Alison went on to reflect that although she had “always known” it was only in recent years that she had been able to speak more openly about her experiences of growing up with HD, and to voice the word “Huntington's” itself out loud (from fieldnotes). She also felt she had been through some specific “stages”, suggesting this could be similar to other young people's experiences - stages of coping and not coping, and engagement with her own risk (McAllister, 2002).

“It has taken me until now to actually really talk about it and admit to people my dad had Huntington's. Before I wouldn't tell anyone ... Because there is stages ... To start with it was denial, I completely blanked it ... turned into a bit of a rebel ... alcohol, drugs and the Goth scene ... totally went into oblivious, literally. Not coming home 'til late. Hitting walls ... And then I got engaged, I was eighteen (laughs). And I got my big head on and phoned her up [the genetic counsellor] and demanded to see her. 'I want that test done, I'm engaged, I want to know!' Which I never, because I still haven't had the test done (laughs). And then it was ignore it again, and then it was like a couple of years, and it would creep up, usually around April because my dad died in April.”

Similar to work on bereavement (Kubler-Ross, 1969) Alison's account reflects some of the stages people experience when they suffer loss and trauma i.e. denial, anger, bargaining, depression and acceptance. Like others growing up with HD Alison became involved with a group outside of her family (Easton, 2003) following their lifestyle to escape from her home situation, and to possibly alert others to “some of the things that wasn't spoken about”. Alison intimated that some of her behaviour was a form of self harm, supporting clinical observations of other young people who have grown up with HD (by SAS). Alison's level of engagement with her own personal risk also changes during different points in her life, citing two particular triggers which cause it to “creep up” and “get her big head on”: getting engaged and the month of her father's death. Other younger participants described wanting to know more about HD, or “paying attention” at the age of 11 or 12, and even anger about “not being told enough” around this time, suggesting this age was another trigger. Alison went on to comment that:

“See I've never spoken to anyone about how *I* feel, what *I* went through ... The kids always seem to get ignored, I mean you were there but you're not listened to ”.

Like other participants Alison felt no one had sought to listen to her views of what it was like to live with such an affected father, even though there was professional involvement with her family (e.g. from social services, health, education) and she was clearly a child 'at risk'.

## **Theme 2 – Learning gradually**

A second subgroup (n=8) described a more gradual process of finding out about Huntington's. They didn't feel they had “always known” about the family disease nor their own risk. They described being initially told after a period of thinking “something was wrong” with a parent, or grandparent, but also stressed how they learned about HD “a bit at a time” as they were growing up. Whilst some experienced quite limited forms of parental disclosure during childhood and adolescence, others were told considerably more at an earlier age, but sometimes this “didn't mean anything” to them e.g. being told as a child about a grandparent having HD. The next accounts highlight the experience of learning gradually about HD and the considerable impact that this illness and different styles of family communication has had on interviewees' lives.

### **Case 2 – Nicola “They were always going on about little things”**

Nicola (age 16, 50% risk) was a young carer who lived alone with her affected mum and younger brother (aged 12). Nicola's close and extended family had a long history of contact with the SHA and the genetic clinic, but she had only known about her mum's illness and her personal risk for two years (also see Forrest-Keenan et al., 2007). Thus, Nicola described how:

“I've known about it for about two years. My granny told me ... Because they were always going on about little things ... but they never came out with a name. I didn't really notice it until my mum and dad split up ... She just told me that it was run from granddad to mum and then down to me, so your cousins and your brother and you might get it. But it's nothing to worry about just now, because it's when you're older ... I did understand it then, but I didn't really. It's not until now that

I feel like more, I don't know ... But it wasn't open in my family before. Nobody spoke about it until everybody noticed how bad my mum was getting [and she went for the test – from fieldnotes] ... And everybody just fights now, half of the family doesn't speak to one another because of all the pressures with having Huntington's. And see if you speak to somebody in the family that you're not supposed to, somebody else falls out with you ... It's horrible that it's breaking the family apart ... I hate it being like this. And I just get really angry ... But I didn't really want to speak about it before. There's some things you just can't say ... Today's the first time I've ever really spoken about it.”

Prior to being told about HD by her granny Nicola had sensed something was wrong because her relatives “were always going on about little things.” Nicola's family colluded in the denial of the onset of her mum's symptoms (Kessler, 1989), which may have enabled them to maintain a situation of 'closed awareness' from Nicola - where several relatives know but fail to openly acknowledge or discuss it with someone who may have a right to know (Downing, 2001). Nicola was finally told when her parents divorced, becoming more aware of the changes in her mum once she became her key carer. When her mum was eventually diagnosed Nicola's family became more open about the cause of her behaviour but family dynamics significantly deteriorated. Although Nicola came from a large family who all lived nearby she described feelings of helplessness, anger and isolation (Forrest-Keenan et al, 2007), and a belief that Huntington's “tears your family apart.” Like Alison she didn't feel she could speak to anyone in her family about her feelings, admitting this was the first time she had ever been so open - only now being able to ask for support with her caring role and for more information (particularly about testing). When asked about her views of having children Nicola asserted that:

“I've got a really strong opinion about this ... if I've got it I'm not having children at all ... it's pointless because you've just to put your children through what we're going through, like with somebody having it.”

For Nicola the experience of living with such an affected mother meant that if she finds out she is a gene carrier she doesn't want to have children i.e. she wasn't prepared to accept the parenting risk (Downing, 2001).

### **Case 3 – Robert “It was a bit secret”**

Robert felt HD had been kept “a bit secret” by his mum until he was nearly an adult. When interviewed Robert was 18 and living with his father (affected with late stage HD), his mother (full-time carer), and older sister. Since leaving school Robert had become a young adult carer, helping his mother to look after his father, whilst also caring for his elder sister who had learning difficulties. Robert appeared to be a linchpin for the family and without his presence there was an underlying sense of fear that it may breakdown (Forrest-Keenan et al., 2007). Robert described how his mum had told him a limited amount when he asked her in childhood, but deferred fully *alerting* and *educating* (van den Nieuwenhoff et al., 2007) him about the severity of his father's HD and his own high risk until he was 17. Thus, after initially questioning his mum when he was younger Robert didn't seem to pursue the subject with either of his parents, nor through any other means.

“My mum wasn't sure whether to tell me when I was about 10, I think she kept it a bit secret ... So it wasn't until I asked mum later why dad couldn't drive that she began to say. I didn't fully understand at the time when she explained it, but she said it was an illness and it means that he won't be able to do certain things and you might notice some changes ... I began to notice how his movements and speech were affected and how going outdoors he would say people were looking at him in a funny way, and things like that. But the only time I really found out a lot was from a programme less than a year ago ... My mum was watching it too ... I was quite shocked there was a 50% chance of having it or not ... Before I just knew the name, but I didn't really know too much about it ... I had just asked a few questions from my mum and it was really finding out little bits of



information a bit at a time ... I guess if I was to learn so much very quickly it would have come as a shock ... but before seeing the programme or meeting Annie I hadn't really checked any books or anything for information on it.... I'd never really put much thought into it about the Huntington's disease and I think Annie's made me think a lot more about it, think about myself a bit more and how it does affect me ... because there's all these kind of tensions."

Although shocked at the news of his own risk Robert seems to support his mum's more lengthy disclosure style, and to believe he had been told in time. Nevertheless, he had only recently begun to reflect upon the considerable impact of HD on his life, mainly because of his recent contact with a professional (Annie) in the field.

From these accounts it appears that some young people do not seek out information or support from sources other than a parent (or grandparent) for what can be a considerable amount of their adolescence - even if they suspect their parent is selectively telling (Gallo et al., 2005), or where it means they have no one else to talk to. This is consistent with psychosocial work on child development and socialisation which suggests that although some children may sense parental 'procrastination,' 'half-truths' and distortions they may still conform to parent's wishes that they 'do not know' (Bowlby, 1988) accepting parental authority and children's dependence (Alderson, 1999b). This appears to have been a protective factor for Robert (e.g. limiting the burden of knowing his risk), but a considerable risk for Nicola (e.g. by increasing her isolation).

### **Theme 3 – Kept a secret by parents**

A third theme concerned participants who described their family history of HD being kept a total secret from them by their parents (n=5). Whilst three young people sensed "there was something wrong" when they were teenagers because they lived with an affected parent, or went to visit an affected grandparent, another two had grown up having no contact with any affected relatives and

were in complete ignorance about the family disease until their early twenties.

#### **Case 4 - Kate (and sister Claire) “I knew there was something”**

Kate (age 24, 50% risk) and her sister Claire (age 19, 50% risk) were not told about their family history of HD until they were young adults'. Their circumstances differed from previous accounts because they did not grow up with a parent who was affected with the disease, but they had visited their affected grandfather since childhood. Kate's mum knew she had the gene mutation herself, but had kept this hidden from her children using a proactive self-censoring style (Kenen, Ardern-Jones and Eeles, 2004). Thus Kate described how:

“I didn’t know until I was 19 or 20 ... by that time grandpa was quite ill and I had been at the stage of what is wrong? What is it? And there was fudging ... I knew there was something ... I had asked my dad ‘what’s wrong with grandpa?’ but he never said anything, my mum had decided that she wanted to be the one to say ... I was very unhappy at home. Mum and I were arguing all the time. Just arguing about anything, about being irresponsible, about lots of stuff.”

When she was growing up Kate had a very difficult relationship with her mother. She vividly outlined her mum's controlling and non-trusting style of parenting, but until she was 19 there was no explanation for her mother’s behaviour (primarily fear that her daughter may get pregnant). Thus, when Kate was eventually told she experienced ambivalent feelings of relief, worry (about her mother becoming ill) and fear (of her own risk). Kate became so anxious that she experienced severe insomnia and suicidal thoughts (see Forrest-Keenan et al., 2007). At the same time she also felt a sense of relief because she now understood her mother’s disclosure burden. From that point on their relationship significantly improved, but the relationship with her dad became more distant.

“When I found out I remember feeling my life was considerably shorter than it had been in the

morning when I woke up. I felt like something had been taken away ... but everything fell into place ... why she was pressing this responsibility issue on me ... And from that day everything was much better. It was like two new different people ... It was such a relief ... *we just fought so much before* ... Previously I would have been closer to my dad, but I have kind of drifted away from him ... the way he approaches it sometimes irritates me and he doesn't have - although he is involved in it he doesn't *know* what it feels like."

Kate and her mum also talked about whether to tell her younger sister, Kate agreed with her mum's decision not to tell Claire, colluding in her parent's secret keeping. But as her sister went through her teens Kate described watching her mum use the same oppressive parenting style with Claire, as Kate had experienced:

"She said she wasn't going to tell my sister at the moment because she felt she was too young – and I agreed ... she shouldn't have to worry about that ... And then I saw her put pressure on her, almost test her, almost testing to see if she was ready ... Wanting her to take responsibility .... Sort of just wanting her to think for herself and just being an adult. She was looking for some signs of an adult nature before she told her. And in me as well."

Kate's account draws attention to her mum's anxiety about telling her children, and how this may impact on parenting styles (e.g. controlling and oppressive) and forms of communication (e.g. secretive and testing for maturity). Six months later Kate agreed to a second interview, and brought Claire along (who was now 19). Kate's feelings about her own risk had changed: "I've not been obsessing about it very much lately, I've been quite relaxed." Whilst Claire - who had only known about their family history for a year - described feeling angry, hurt and disappointed at her mum and sister for keeping HD a secret from her, and the lack of trust she felt this embodied.

“I was quite angry at one point because I hadn’t been told ... and almost disappointed they thought I wouldn’t be able to deal with it ... I felt I wasn’t trusted ... But I understood why they didn’t tell me ... I try not to let it get me down ... I’m busy with work all the time ... It’s just some days, normally at the weekend, those are the sort of days when you start thinking about other things and that’s when it sort of creeps up.”

Claire described how she “went off the rails” for a few months after she was initially told – binge drinking and staying out late - but within a year she began “caring about” and “planning for things” again. Family relationships between her mum and the two sisters also improved, and ultimately Claire felt she understood why they had kept it a secret.

### ***Grew up in ignorance***

A further two participants had grown up in complete ignorance about their family history of HD until adulthood because it had been kept a secret. One didn't see an affected relative until she was a young adult and the other had still never met a relative with HD.

### **Case 5 - Stacey**

Stacey (age 20, no longer at risk) described having only known about her family history of HD for a year and very much supported her parents decision to keep her grandma's illness and dad's 50% risk a secret from her and her sister whilst they were growing up. Stacey's dad had recently been for the test and discovered he did not carry the gene. Although Stacey was relieved she remained anxious about her extended family, agreeing to collude with her mum in keeping HD a secret from her cousins. Thus, Stacey described how her sister went:

“onto the internet and she put two and two together with my grandma and asked mum and dad ... I think mum and dad were scared ... they had known for ten years that my grandma had it before we

found out ... and I think when they did find out I was still quite young [age 10]. So I think they were just wanting to find the best time to tell us. It didn't bother me because ... I knew they had our best interest at heart ... and I'm a worrier as well. I take after my mum ... Once I found out, it was still there - thinking about my cousins and my uncles and that - but I was just relieved that dad didn't have it ... but that was one thing my mum said 'don't tell your cousin'. And I said, no I wouldn't."

Again, Stacey's account highlights parents' anxiety about finding the 'right time' to tell their children, in this case choosing to wait until they were young adults, although their eldest daughter deduced the cause of her grandmother's symptoms before their parents found the 'right' opportunity. Interestingly, Stacey's parents had known about her dad's family history for a decade before their children found out, but this secrecy does not seem to have had a detrimental impact on family relations either before or post awareness. This suggests her parents used a different parenting style to Kate's mum, and/or were perhaps better able to contain their own anxiety from having any impact on their children.

#### **Theme 4 – HD is a new diagnosis**

The last theme concerns participants who experienced HD being a new diagnosis (n=5), where their family had known a parent or grandparent was ill but not what was wrong, the diagnosis of HD being unexpected and 'out of the blue'. In a similar way to participants from whom HD had been kept a secret, this group described two quite different reactions to finding out about their family history.

#### ***“It was a huge shock”***

Two young adults described HD being a new diagnosis, both of whom who had contact with their affected parent when they found out, but did not live in the family home. They experienced feelings

of shock, panic and immense anxiety about their own risk (Forrest Keenan et al., 2007), as did Kate (see Theme 3), and all had wanted to get tested straightaway. Like Kate one had been to speak with a genetic counsellor and the other was considering it, but none had taken the test as yet.

Two young people (aged 15 and 16) also described knowing nothing about their family history until they were 12 when a parent went for a diagnostic test and discovered they had early signs of Huntington's. Both were informed by their parent about the diagnosis soon afterwards, and they reflected that finding out was a "huge shock" with which they were still trying to come to terms. There was a sense that these parents had perhaps 'blurted out' the news about HD, in a shocking manner (Forrest Keenan et al., 2007; Klitzman, 2007). In contrast to other participants these two accounts mainly focused on the life-changing events which took place *after* being told e.g. a parental divorce and becoming a young carer. Daniel (age 16, 50% risk) was particularly assertive about how his life had changed since finding out about HD - he was now the sole carer for his affected father who was estranged from his mum, admitting that this new role could be extremely stressful (see Forrest Keenan et al., 2007).

#### **Case 6 - Clara "*It wasn't a huge thing*"**

Clara's account differs from the others in this group as she (now 22 and a gene carrier) appears to have coped well with finding out about her family history of HD in her mid teens, and with her own carrier status since then (also see Forrest-Keenan et al, 2007). Whilst she was growing up Clara and her family had known there was something wrong with her grandfather, but it wasn't until she was 15 that he was diagnosed with HD. That year she also found out that her mum was a gene carrier and about her own risk. As Clara's grandparent's lived nearby she witnessed her grandfather's deterioration, as well as her grandmother's burden of caring for him before he moved into a nursing home. However, she reflected that:

“It wasn’t a huge thing - we knew grandpa was ill, it was just a name to what was happening ... We didn’t know then my mum was going for the test ... But once she had the test and come to terms with it herself they got the two of us together and sat us down one night at tea and said this is what it is and this is what’s happening. A strange tea conversation ... I understood it wasn’t going to affect mum at the time and you’re so young yourself, you don’t really think about the consequences for you ... My little brother would have been about 8 and I think it’s been different for him than it has been for me because he’s been brought up with it. I found out about it when I was a bit older, but he was only young when he found out. But in a way I think that’s a good thing, because it’s not such a, it’s something he’s been brought up with, it’s something he knows there’s a possibility of in life. It’s not such a shock as it were.”

Clara's account suggests her parents chose an open style of communication (Kenen et al., 2004) in order to tell their children about their grandfather's diagnosis. Then once their mum had “come to terms” with her result she also told them about her status and their personal risk, supporting other studies where parents needed time to make sense of their own risk and to assess the receptivity of their children (Forrest et al., 2003; Gaff et al., 2007). Clara seems to have been told in a calm and supportive manner, in an everyday family context (i.e. having tea) with both parents and children present. Thus, although Clara found out the same information as Kate the circumstances were considerably different. For Clara there was a more open and supportive form of communication, no controlling or 'testing' parenting style in the pre-awareness stage, and she was told at a younger age, all of which appear to have contributed towards her ability to cope. Clara went on to describe her views about living with her gene-carrier status, and the possibility of having children in the future:

“When I got the result ... it’s one of those things where you think about it all the time, and then one day it just goes to the back of your mind ... and we will definitely have family. We will have our own ... that’s what I wanted and that’s what my husband wanted ... We did talk about adopting at

one point. But, I don't see the need to. By the time they've grown up, hopefully there will be some kind of cure ... So I don't see it being that much of a problem really."

Clara's experience of HD was considerably different from Kate, she was able to assimilate the news about her grandfather's illness and mum's result into her daily life, and even knowing her own carrier status she had gone on to get married, begun a successful career and was planning to have children, accepting the parenting risk (Downing, 2001). She was optimistic about the future, believing in a discourse of hope (Novas, 2005).

## **Discussion**

This paper explores one issue that emerged in a study of young people's experiences of living in a family with HD, that is, how young people find out about their family history. A qualitative thematic analysis was undertaken which revealed four main themes: having always known about HD, learned gradually, HD was kept a secret, or it was a new diagnosis. In particular, the timing and style of disclosure from relatives, and an individual's stage of awareness, were fundamental in structuring participants' accounts.

The majority of participants were told about HD in childhood (n=21) and many (n=15) described having "always known". This suggests some families choose an open style of communication with their children. Those who had "always known" either lived with, or were in close contact with an affected relative, which may have encouraged disclosure. Our data highlight how children can be active agents in their own learning about HD, for example, through "watching," "hearing," "asking," or "searching" (Alderson, 1999b; Easton, 2003). Nevertheless, some chose not to ask questions because they wanted to protect their parents (Skirton, 1998), or had learned not to break 'family rules' of silence (Easton, 2003). Three participants also described being told as a child about a grandparent having HD, but that it "didn't mean anything to them" until they were teenagers.



Thus, children may have 'limited awareness' (Downing, 2001), where the term Huntington's becomes known, but it means very little to them at the time. Nevertheless, whilst some did not remember "or really understand" the first information they were given they did recall feelings e.g. of shock, upset, fear – or ambivalence because this 'telling time' brought them closer to a relative, affirming (or initiating) trust (Klitzman, 2007). Other participants wanted to know more about HD around the age of 11/12, probably reflecting a different stage of cognitive development (Easton 2003; Gallo et al., 2005).

Participants who were told gradually about HD gave more complex accounts, similar to Downing's (2001) five stage continuum. For some disclosure was limited even until late adolescence, whilst others were told more - "bit by bit" - from an earlier age. Our findings highlight a period of initial awareness which could be distressing, particularly where a lone parent was diagnosed with little family or professional support (see also Forrest-Keenan et al., 2007).

Like other studies we found participants whose parents had chosen to keep HD a secret, disclosing in late adolescence or adulthood. The first subgroup had contact with an affected parent/grandparent in childhood and adolescence and described feelings of confusion about their family dynamics, and a sense of "knowing something was wrong" but not what. Our results bring attention to a 'testing' form of parenting disclosure, supporting other work on families with HD (Downing, 2001). The second subgroup had no contact with an affected relative and grew up in complete ignorance about HD (although Stacey unknowingly lived with an at-risk father). Participants in this small subgroup not only understood but supported their parent's decisions to defer telling them, demonstrating that nondisclosure may be supported by participants even into adulthood.

Our results also draw attention to participants for whom HD was a new family diagnosis. For most in this group finding out was a "huge shock" triggering anxiety (Forrest-Keenan et al., 2007). The

two younger interviewees primarily focused upon the traumatic events after their parents' diagnoses such as divorce, unemployment, moving house and becoming a young carer, again suggesting the initial period post diagnosis can be stressful. Participants who find out in adulthood (whether it was kept a secret or a new diagnosis) are likely to have little knowledge about HD; consequently their support needs may be different from those who have grown up knowing (Etchegary, 2006). This may be particularly important if they have made major life decisions which they may not have chosen if they had known, and/or they react by needing to regain a sense of control e.g. through predictive testing (van der Steenstraten et al., 1994). In contrast Clara's account highlights that some younger people do cope well with being told about Huntington's and their own personal risk (see also Cox and McKellin, 1999).

Our data supports a small body of work which suggests that those children and young people who have close contact with a relative affected by HD want to know what is wrong and can cope with knowing (something) from an early age (van der Steenstraten, Tibben, Roos, van de Kemp, & Niermeijer, 1994; Skirton, 1998; Forrest Keenan et al., 2007; Holt, 2007). When children are not told, but sense something untoward, they feel anxious and confused and have little understanding of an affected person's behaviour or overprotective parenting (Easton, 2003). This supports studies of nondisclosure in other areas e.g. parental cancer, or parental AIDS/HIV, negating the protective element of secrecy and silence about illness (Slavin, O'Malley, Koocher, & Foster, 1982; Katz & Jay 1984; Skirton, 1998; Waugh 2003).

This study lends some support to there being a 'critical age' for finding out, that is, a time where the benefits of telling outweigh the benefits of not telling. The approach to adolescence was an important time for some to know (more), whilst others coped with finding out earlier, or with limited forms of disclosure even up to late adolescence/early adulthood (Gallo et al., 2005:272). For those who learned gradually there was an important distinction between being told about a parent's

illness first, then later about one's own risk (Klitzman, 2007). Thus, telling children about Huntington's is not a one-off event but a potentially lengthy process of disclosure (Forrest et al., 2003) which should be negotiated at the child's pace (Gallo et al., 2005).

Our findings draw particular attention to children's experiences of parental anxiety about disclosure of genetic risk demonstrating how this may impact on family relationships, parenting and styles of communication, similar to studies exploring telling children in other hereditary disorders (Featherstone et al., 2006; Metcalfe et al., 2008). Participants revealed a range of attitudes towards their family's style of communication - from anger at "everybody fighting" and distress about things being kept hidden, to acquiescence about, and even an understanding of parents 'disclosure dilemmas' (Forrest et al., 2003). Where HD had been more open in a family some participants were supportive of this, whilst others had possibly been told too much, in a distressing manner. Thus, we observed five disclosure styles similar to Kenen et al. (2004), adding two new categories from our results: 'open and overloading' and 'proactive testing,' both of which could have a detrimental impact on participants' well being and family relationships. There may be more open styles of communication in families affected by familial cancer (Forrest et al., 2003; Gaff et al., 2007) indicating some styles of communication may be more or less prevalent than others depending on the nature of the disease and availability of treatment (Wilson et al., 2004).

Our findings also support previous studies which show that authority rules based on social constructions of family and kinship may influence genetic disclosure (Forrest et al., 2003; Featherstone et al., 2006). Like Featherstone et al. (2006) we found collusion between generations to protect others and secret keeping across generations (vertical blocking), but also within generations (horizontal blocking) because participants lacked the 'authority' to tell certain relatives.

Our analysis reinforces the role of 'pivotal' relatives in disclosing or withholding information

(Forrest Keenan et al., 2005), and the extent to which their influence may be protective for some young people whilst increasing the vulnerability of others (e.g. by keeping silent about suspected abuse). Disclosure may also be highly gendered (D'agincourt-Canning, 2001; Forrest et al., 2003; Featherstone et al., 2006), as women are often the 'gatekeepers' of genetic information (Richards, 1993). At the same time some fathers do take responsibility for telling relatives (Forrest-Keenan et al., 2005) and may also influence women's disclosure decisions (Gaff et al., 2007).

Young people use various defense mechanisms to cope with any knowledge and/or experience of HD such as: denial, avoidance, minimization or assimilation (Power, 1977; Forrest Keenan et al., 2007), drawing upon personal, interpersonal and group styles of coping (Easton, 2003). In addition, some young people's sense of self-belief may protect them from anxiety (Skirton, 1998), whilst others may be experiencing loss or grief (Skirton, 1998; Sobel & Cowan, 2003). These accounts demonstrate that young people's views and experiences of HD are varied, with some coping well and others experiencing considerable anxiety, or even physical and/or emotional harm (Forrest-Keenan et al., 2007).

Similar to studies of other sensitive issues e.g. sexual health and disclosure of abuse (CRFR, 2007), we found that young people may value the opportunity to talk about HD with a parent/relative, or someone outside the family (Forrest-Keenan et al., 2007) at their own pace and when they are emotionally and cognitively ready. We observed various triggers for more information and support, which brought Huntington's and one's personal risk to the forefront of participants' attention e.g. a relative's death, parental divorce, career/exam choices, first relationships and having 'free time', what McAllister (2002) refers to as changes through time and illness-related events.

### *Study strengths and limits*

This study breaks new ground through exploring young people's experiences of growing up at risk

of adult-onset hereditary illness. While the sample size is arguably small, the emphasis has been on gaining depth insights into a typically invisible hard-to-reach population. Although we have explored young people's experiences of finding out solely about HD, our findings will have relevance for family communication about other genetic conditions and parental diseases.

We chose a qualitative thematic analysis using some elements of a grounded theory approach in order to give a voice to participant's views (Opie, 1992) and to enhance validity, but acknowledge that a different theoretical perspective, such as that used by a health psychologist or family therapist, might illuminate different issues (Brouer-DudokdeWit et al., 2002; Gooding et al., 2006). The use of more varied research tools to elicit young people's experiences (particularly about their own risk) may also have been beneficial (Alderson & Morrow, 2004; Coad, 2007). We are aware that the small sample size and mean age of 20.5 make generalisability difficult to other populations (Forrest Keenan et al., 2007).

There were also limits in accessing young people under 18 through a parent. We know little about those young people whose parents chose not to inform them about our study. Use of snowball sampling also means that some members of the same family were recruited which could have reduced variation, but this was addressed to some extent by using purposeful and theoretical sampling. A further possible limit is that everyone who was invited by the two clinicians took part, which may be because of an established rapport suggesting a "gratitude bias" (Øvretveit, 1992), and again reflects the selective nature of the sample. Some caution may also be needed in relation to the accounts of those who 'grew up in ignorance' as both were no longer at risk and had no experiential knowledge of HD which is likely to have lessened any fear about the illness. Also, we did not include individuals considered by the professionals as too vulnerable to take part, and families who do not come into contact with any SHA or genetics services. Nor did we interview anyone who grew up living with an affected parent, but from whom HD was kept a secret until adulthood.

Future research would benefit from interviewing individuals in these circumstances.

## **Conclusion**

This paper adds to a growing body of work which suggests that those children and young people who live with, or who have close contact with, a parent or grandparent affected by HD want to know 'what is wrong?' and can cope with knowing - at least something about the illness - from an early age. Although knowing about Huntington's cannot remove the emotional trauma of living with an ill parent, or the burden of finding out about one's personal risk, it can ease anxiety from inaccurate information (e.g. fear of a parent's imminent death), and may ultimately improve some family relationships. At the same time there were circumstances where limited or even nondisclosure was supported by participants into late adolescence or early adulthood. Those who learned gradually made an important distinction between learning about a parent's illness, then later about their own risk. Our findings highlighted particular triggers which may bring personal risk to the forefront of a young person's attention, and a need for further information and support at these times. We also observed two additional categories to Kenen et al's model, namely 'open and overloading' and 'proactive testing', both of which could have a detrimental impact on young people's well being. Where HD was a new diagnosis for a family young adults were also likely to have different support and informational needs from those who had grown up knowing. Thus, the timing and style of disclosure from relatives, and an individual's stage of awareness, were fundamental in structuring young people's experiences of learning about HD.

Future research is needed to explore more about the impact of different parenting styles and forms of family communication upon children's experiences of growing up with hereditary illness. Further research is also needed to explore the extent to which female relatives are gatekeepers of genetic information, and what support fathers may need about telling. Given that this generation of young people is likely to grow up knowing about new genetic technologies such as predictive and prenatal

testing it would also be interesting to explore what actual decisions they make with regards to finding out about their own carrier status and having children, particularly as studies indicate low uptake rates (Simpson & Harper, 2001). As people are enabled to find out what hereditary illnesses exist in their family, professionals will continue to be asked about how, what and when to tell children about genetic risk, highlighting the need for training and awareness on family communication issues.

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Table 1. Main themes and sub themes arising from interviews

Always known about HD	<p>“Always known ... but there are stages”</p> <p>When told “it meant nothing”</p>
Learned gradually	<p>“They were always going on about little things”</p> <p>“It was kept a bit secret”</p>
HD was kept a secret	<p>“I knew there was something”</p> <p>Grew up in ignorance</p>
New diagnosis for family	<p>It was a “huge shock”</p> <p>“It was just a name”</p>

Table 2. Initial informant about HD

When told:	Childhood	Adolescence	Adulthood
<i>Initial informant</i>			
Parent: Mother	11	3	3
Father	0	1	2
Both	4	0	
Grandparent	3	1	
Sibling	1	1	1
Self	2	0	
Total	21	6	6



Table 3. Personal contact with affected relative

When told:	Childhood	Adolescence	Adulthood
<i>Contact with affected relative</i>			
Living with affected parent	6	4	
Living with affected grandparent	3	0	
Not living with affected grandparent, but have contact [parent at risk or carrier]		10	2 3
Not living in family home with affected parent, but have contact (not known others with HD)			2
Never known relative with HD, parent carrier	2		
Never known relative with HD, parent at risk			1
Total	21	6	6

