

Webinar 28.11.23

Insights into growing up in families affected by Huntington's disease

Siri Kjoelaas

PhD, psychology, counselor

Centre for Rare Disorders, Oslo University Hospital, Norway

Email: hagsir@ous-hf.no

Q1 What is your professional background?

(Single choice)

- a) Neurologist
- b) Neuropediatrician
- c) Neurology resident
- d) Psychiatrist
- e) Nurse
- f) Physiotherapist
- g) Geneticist
- h) Psychologist
- i) Patient or patient representative
- j) Other

Outline

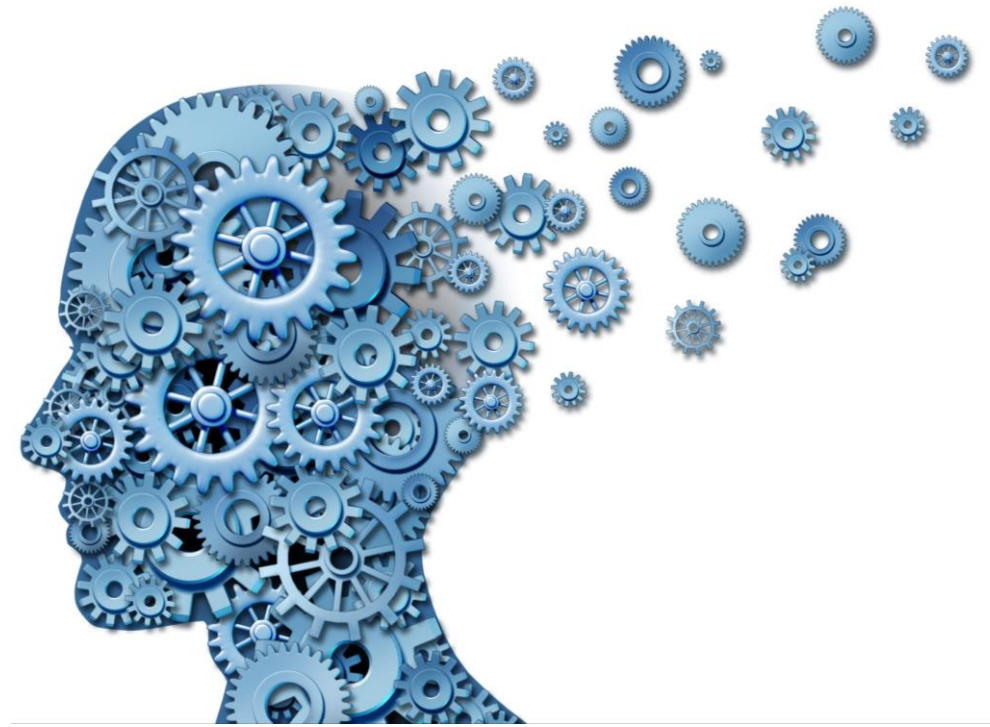
1. Topic introductions
2. Insights from the research
3. Placing the findings into context and frameworks of understanding
4. Key points
5. Questions and comments

Learning objectives

1. Demonstrate awareness and understanding of psychological and social impact of Huntington's disease on children and young people with parents who are affected
2. Connecting patients and families to appropriate psychological and social support if needed
3. Communicate information about psychological risk factors for children with parents with HD and ways to protect against these
4. Provide guidance on specific issues related to how families are affected by HD

HD - a rare disorder that affects the family

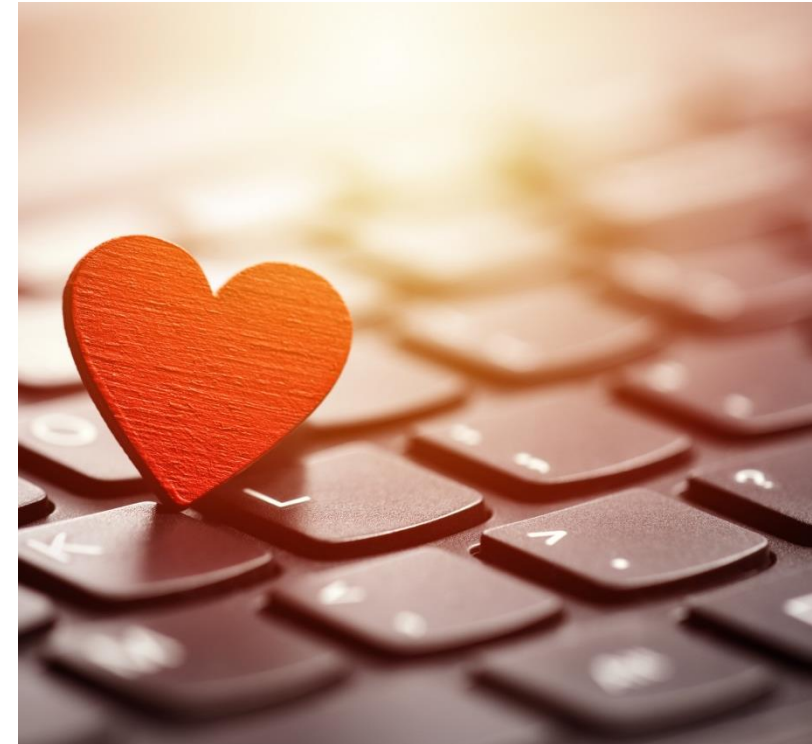
1. Adds genetic risk
2. Affect all functions of the brain
 - Changes in motor functioning
 - Changes in cognitive functioning
 - Changes in neuropsychiatric functioning
3. Symptoms usually start mid-life (30-50 years of age)
4. Long disease trajectory (17-20 years)
5. Large individual variability



(e.g. McColgan & Tabrizi, 2018; Roos, 2010; Sparbel et al., 2008)

Insights into growing up in families affected by Huntington's disease

- National study from Norway (2018-2024)
- **Semi-structured interviews**
 - 36 teenagers and adults (13-65 years)
 - 14 parents with partners with HD
- **Qualitative research**
 - Analyses of texts and themes
- Keep in mind that there are methodological limitations...



Landsforeningen for Huntington sykdom





Psychology & Health

 Routledge
Taylor & Francis Group

ISSN: (Print) (Online) Journal homepage: <https://www.tandfonline.com/loi/gpsh20>

'I knew it wasn't normal, I just didn't know what to do about it': adversity and caregiver support when growing up in a family with Huntington's disease

Siri Kjoelaas, Tine K. Jensen & Kristin B. Feragen

To cite this article: Siri Kjoelaas, Tine K. Jensen & Kristin B. Feragen (2022) 'I knew it wasn't normal, I just didn't know what to do about it': adversity and caregiver support when growing up in a family with Huntington's disease, *Psychology & Health*, 37:2, 211-229, DOI: [10.1080/08870446.2021.1907387](https://doi.org/10.1080/08870446.2021.1907387)

To link to this article: <https://doi.org/10.1080/08870446.2021.1907387>

My childhood felt unsafe. We never knew what to expect at home. Huntington's disease made my father suspicious, always 'right', and very irritable. Us kids always had to be considerate of him: never saying what we really felt, trying not to get into arguments with him. If he didn't take it out on us, he took it out on our mum.

(Kjoelaas et al., 2022a)

Adverse Childhood Experiences and HD

- Unpredictability
- Grief
- Fear
- Emotional and physical neglect



(Kjoelaas et al., 2020)

Support provided by their caregivers were particularly important in how they coped and adapted

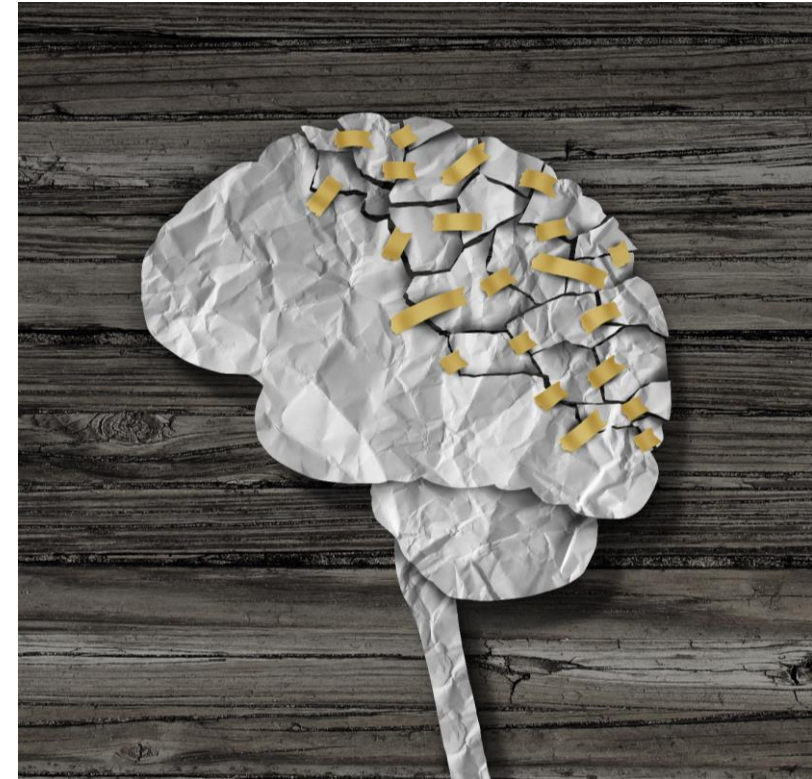
- Without support childhoods were described as overwhelmed by stress (toxic stress)
- With support participant's described having coped and adapted to adversity better

(Kjoelaas et al., 2022a)



Understanding toxic stress and how to buffer/protect from it

- **Fight or flight** response (Danese et al., 2012)
- **Toxic stress** = **excessive stress** accumulates over time (Danese et al., 2012; Shonkoff et al., 2012)
- Present **major risks** to current and future mental and physical **health** (Hughes et al., 2017; 2021)
- **Resources** that help **regulate stress** responses are promotive factors, **protective factors** or buffers (e.g. Fergus & Zimmerman, 2005; Fraser et al., 1999)
- One of the **most important resources** promoting healthy development in light of adversity is **social support** (Fergus & Zimmerman, 2005; Hughes et al., 2017; Shonkoff et al., 2012).



Central lessons on adverse childhood experiences and HD

- **Need for awareness of impact Huntington's disease can have on children and young people**
 - Understanding the range and severity of adverse experiences of family members
 - Understanding impacts of cognitive and neuropsychiatric symptoms
- **Offer appropriate psychological and social support to patients and families members**
 - Promoting early and sufficient help and support to caregivers with and without HD
 - Connecting children and other caregivers to support services to manage overwhelming stress (traumatic stress and post traumatic stress)
 - Connecting patients to appropriate support services to manage overwhelming stress (traumatic and post traumatic stress)

Q2: What adverse childhood experiences can patients and families affected by HD need help to prevent and/or cope with?

- a) Psychological abuse
- b) Physical abuse
- c) Neglect
- d) Grief
- e) All of the above

Children and young people in families with HD can potentially be supported through early management of cognitive and neuropsychiatric changes in their parents?

- a) True
- b) False

Dilemmas when talking about Huntington's disease: A qualitative study of offspring and caregiver experiences in Norway

Siri Kjoelaas^{1,2}  | Tine K. Jensen^{2,3}  | Kristin B. Feragen¹ 

Talking about HD from a child's perspective

In terms of learning about Huntington's disease I was left out. I felt like my parents figured I was too young, that they did not want to deal with it yet, and that they could talk with me when they thought I needed it, which would be later. So I figured it all out on my own.

Talking about HD from a parent perspective

It can't be all about Huntington's every day. I could see in my daughter how talking about it brought her down and all the time we spent trying to get her back up Many people in our family have committed suicide because of this, and her thoughts were there, too So you should tread carefully. That's my opinion.

Dilemmas when talking about HD

- Children need knowledge and conversation about HD with caregivers
- Still, a variety of dilemmas can hinder these conversations:
 - When?
 - What?
 - How often?
 - Do we share with others?



(Kjoelaas et al., 2022b)

Central lessons on parent-child conversations about HD

- **Offer appropriate psychological and social support to patients and families**
 - Approaching the topic of conversations about the disease with an understanding about the complex nature of these talks
- **Provide guidance on specific issues related to HD**
 - Guide or connect parents with HD to support services that can help them:
 - Balance the many dilemmas that arise in conversations and how to use dialogue to best help offspring adapt and cope with the many challenges that can come with HD.
 - Educate and prepare caregivers in families with HD by strengthen their ability to make conscious choices in current and future conversations with their children.

Q4 Informing and talking with children about neurogenetic disorders can feel difficult and come with many dilemmas for parents because of?

- a) Lack of words
- b) Unwillingness to address the subject
- c) Genetic risk and wanting to protect their children from information that can cause them harm
- d) I do not know



Health Psychology and Behavioral Medicine

An Open Access Journal



ISSN: (Print) (Online) Journal homepage: <https://www.tandfonline.com/loi/rhpb20>

Social support experiences when growing up with a parent with Huntington's disease

Siri Kjoelaas, Kristin B. Feragen & Tine K. Jensen

To cite this article: Siri Kjoelaas, Kristin B. Feragen & Tine K. Jensen (2022) Social support experiences when growing up with a parent with Huntington's disease, Health Psychology and Behavioral Medicine, 10:1, 655-675, DOI: [10.1080/21642850.2022.2104286](https://doi.org/10.1080/21642850.2022.2104286)

To link to this article: <https://doi.org/10.1080/21642850.2022.2104286>

I was lucky because I had my grandparents living in the house next-door to us. I really do not think I would have survived if it was not for them. Because there were a lot of fights at home, and it was not a good place to be most of the time.

How support helped in childhood

- Relationships were experienced as supportive when they provided:
 - Love, care, or belonging
 - Coping skills
 - Reduced or alleviated stressors at home



(Kjoelaas et al., 2022c)

How support can be hindered

- Parent's lack of acknowledgement and insight
- Others lack of knowledge
- Familial aspects of the disease
- Protecting oneself



(Kjoelaas et al., 2022c)

Central lessons on social support for children with parents with HD

- **Provide guidance on specific issues related to HD**
 - Educate patients and other family members that social support outside the family help and can compensate for support that is compromised in caregiving relationships by disease.
- **Offer appropriate psychological and social support to patients and families affected by HD**
 - Educate patients and make appropriate referrals to patient organisations
 - E.g. Huntingtons Disease Youth Organisation (HDYO), European Huntington Association (EHA)
 - Include information from family members when evaluating their support needs

Q5: Which of the following provide social support and consequently help children in families with HD cope with adversity?

- a) Close adults who can provide love and care
- b) Others in similar situations
- c) Activities that give them a break from responsibilities at home
- d) Others who provide information and act as role models
- e) All of the above

Key points/ conclusions

- Neurogenetic disorders like Huntingtons disease can lead to a **range of adverse impacts on** children and other family members
 - Double risk (genetic and psychological) with adverse childhood experiences
 - Risk of lacking support from caregiving and other relationships
- Many **impacts can be understood** and helped **within frameworks of how we understand and help any child** that grow up in families that are vulnerable for any reason
- Evidence of **positive outcomes and coping in light of these impact** should be used to make appropriate guidance and/or referrals to psychological or social support
- However, **unique aspects of neurogenetic disorders** like HD potentially **exacerbates** and complicates **impacts on children and other family members**



References

- Danese, A., McEwen, B. S. (2012). Adverse childhood experiences, allostasis, allostatic load, and age-related disease. *Physiology & Behavior*, 106(1), 29–39. <https://doi.org/10.1016/j.physbeh.2011.08.019>
- Fergus, S., & Zimmerman, M. A. (2005). Adolescent resilience: A framework for understanding healthy development in face of risk. *Annual Review of Public Health*, 26(1), 399–419. <https://doi.org/10.1146/annurev.publhealth.26.021304.144357>
- Fraser, M. W., Galinsky, M. J., & Richman, J. M. (1999). Risk, protection, and resilience: Toward a conceptual framework for social work practice. *Social Work Research*, 23(3), 131–143. <https://doi.org/10.1093/swr/23.3.131>
- Hughes, K., Bellis, M. A., Hardcastle, K. A., Sethi, D., Butchart, A., Mikton, C., Jones, L., & Dunne, M. P. (2017). The effect of multiple adverse childhood experiences on health: A systematic review and meta-analysis. *The Lancet Public Health*, 2(8), 356–366. [https://doi.org/10.1016/S2468-2667\(17\)30118-4](https://doi.org/10.1016/S2468-2667(17)30118-4)
- Hughes, K., Ford, K., Bellis, M. A., Glendinning, F., Harrison, E., & Passmore, J. (2021). Health and financial costs of adverse childhood experiences in 28 European countries: a systematic review and meta-analysis. *The Lancet Public Health*, 6(11), 848–857. [https://doi.org/10.1016/S2468-2667\(21\)00232-2](https://doi.org/10.1016/S2468-2667(21)00232-2)
- Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2022a). ‘I knew it wasn’t normal, I just didn’t know what to do about it’: adversity and caregiver support when growing up in a family with Huntington’s disease. *Psychology & health*, 37(2), 211–229.
- Kjoelaas, S., Jensen, T. K., & Feragen, K. B. (2022b). Dilemmas when talking about Huntington's disease: A qualitative study of offspring and caregiver experiences in Norway. *Journal of Genetic Counseling*, 31(6), 1349–1362.
- Kjoelaas, S., Feragen, K. B., & Jensen, T. K. (2022c). Social support experiences when growing up with a parent with Huntington’s disease. *Health Psychology and Behavioral Medicine*, 10(1), 655–675.
- McColgan, P., & Tabrizi, S. J. (2018). Huntington’s disease: A clinical review. *European Journal of Neurology*, 25(1), 24–34. <https://doi.org/10.1111/ene.13413>
- Roos, R. A. (2010). Huntington’s disease: A clinical review. *Orphanet Journal of Rare Diseases*, 5(1), 1–8. <https://doi.org/10.1186/1750-1172-5-40>
- Shonkoff, J. P., Garner, A. S., Siegel, B. S., Dobbins, M. I., Earls, M. F., McGuinn, L., Pascoe, J. Wood, D. L. (2012). The lifelong effects of early childhood adversity and toxic stress. *Pediatrics*, 129(1), 232–246. <https://doi.org/10.1542/peds.2011-2663>

**Thank you for your
attention**

