

Supporting treatment adherence in young people with tyrosinaemia type 1: co-creation of a discussion guide for healthcare professionals (HCPs)

■ **Author:** Dr Sumaira Malik
■ **Contact:** sumaira.malik@atlantishealthcare.com

AIMS:

- ▶ To develop a brief HCP-led intervention to support treatment adherence discussions between HCPs and their patients.
- ▶ To explore the feasibility, acceptability and relevance of the intervention for HCPs.

1 Background

Hereditary Tyrosinaemia type 1 (HT-1) is a rare metabolic disorder caused by an enzymatic defect in the metabolism of the amino acid tyrosine.

HT-1 typically presents in infancy as failure to thrive and, if left untreated, can lead to progressive liver and kidney dysfunction. HT-1 has an estimated incidence of 1 case in every 100,000 births.¹ The primary treatment for HT-1 is lifelong adherence to nitisinone medication and a specialist low-tyrosine/ phenylalanine diet.

The appropriate use of medication and strict dietary adherence is thus central to successful management of the condition. However, research across various chronic conditions has shown that adherence to strict medication and dietary guidelines can be challenging for many young people and their families, particularly during teenage years.²

There is a consensus that young people with a chronic condition should be more involved in clinic consultations



from a young age to ensure they understand their condition/ treatment and develop self-management skills early on. However, HCPs are sometimes unsure how best to initiate discussions around adherence and involve younger patients in their treatment plans.³

A brief intervention was developed to support treatment adherence discussions between HCPs and young people with tyrosinaemia.

2 Methods

Intervention development and co-creation steps:

- Initially, a literature review of psychosocial and treatment challenges for young people living with rare disease was conducted
- Semi-structured qualitative interviews were then carried out with patients, caregivers and healthcare professionals in the UK and France
- Based on insights from the literature and qualitative research, an initial prototype for the discussion guide was developed
- A co-creation focus group with HCPs (n=5) involved in the care of tyrosinaemia patients was conducted to assess the feasibility, acceptability and relevance of the discussion guide for HCPs
- Feedback from the focus group was used to further refine and develop the discussion guide
- The updated version of the discussion guide will be piloted with a small number of patients in clinic
- The discussion guide will be amended in accordance with feedback from pilot
- A final version will be made available to HCPs

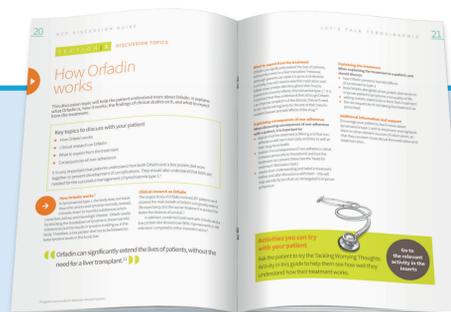
3 Results

The discussion guide focussed on six factors that were found to be important for adherence to treatment:

1. Practical barriers
2. Self-efficacy
3. Treatment necessity
4. Treatment concerns
5. Understanding of the condition
6. Understanding of treatment

The discussion guide included several key components:

- ▶ A summary of insights on adherence challenges in tyrosinaemia
- ▶ A screening questionnaire to help HCPs elicit a patient's barriers to adherence
- ▶ A corresponding discussion checklist for each barrier
- ▶ Interventional activities to help facilitate conversation around these barriers e.g. quizzes, CBT-based activities



The prototype of the discussion guide was positively received by the HCPs participating in the focus group. They felt that both themselves and their colleagues would find the booklet useful for structuring and facilitating treatment discussions with their patients.

Key suggestions for improvement included:

- ▶ Increased content around dietary non-adherence
- ▶ Tips and strategies for communicating specifically with young people and teenagers
- ▶ Providing additional information and guidance to describe how the guide could be used in practice

Discussion Guide for healthcare professionals

Supporting you to help tyrosinaemia type 1 patients overcome key challenges and achieve better treatment outcomes

ACTIVITIES | TYROSINAEMIA QUIZ

Quiz: How well do you understand tyrosinaemia?

How well your doctor or nurse has gone through the important things to be aware of with tyrosinaemia, take our quiz to find out how well you understand your condition and your treatment plan. The more knowledge you have about tyrosinaemia, the easier you will find it to manage.

- For each question simply circle your chosen answer. Good luck!
1. Tyrosinaemia is an inherited condition, which means you are born with it. What causes the symptoms of tyrosinaemia?
 - a. High levels of all proteins build up and cause problems in the kidneys, liver and brain
 - b. High levels of tyrosine build up and cause problems in the kidneys, liver and brain
 - c. Low levels of all proteins cause problems in the kidneys, liver and brain
 - d. Levels of tyrosine cause problems in the liver and brain
 2. Which of the following treatment plans will best control tyrosinaemia to prevent problems occurring?
 - a. Orfadin
 - b. Orfadin + a protein supplement
 - c. Orfadin + a protein supplement + a low protein diet
 - d. Orfadin + a protein supplement + a high protein diet
 3. You have eaten a large beef burger and don't feel unwell. Is it safe to continue eating such foods?
 - a. Yes - the diet guide says you can eat what you like
 - b. No - you should avoid eating such foods
 - c. Not sure
 - d. Not treated, what does it matter?

4 Summary/Next steps

▶ The co-creation focus group with HCPs was effective for informing the design and development of the HCP discussion guide. Initial feedback suggests that the tool has the potential to help structure and support discussions around adherence to medication. Further piloting will be carried out to examine the feasibility of adopting the tool in clinical practice and explore impact on clinical outcomes.

¹ Sniderman King L, Trahms C, Scott C (2006) Tyrosinemia type 1. SourceGeneReviews™[Internet]. Seattle (WA): University of Washington, Seattle.

² Dean AJ, Walters J, & Hall A (2010) A systematic review of interventions to enhance medication adherence in children and adolescents with chronic illness. Archives of Disease in Childhood, 95(9), 717-723.

³ Malik S et al (2015) Treatment Adherence in Type 1 Hereditary Tyrosinaemia (HT1): A Mixed-Method Investigation into the Beliefs, Attitudes and Behaviour of Adolescent Patients, Their Families and Their Health-Care Team. JIMD Reports, 19, 13-22.