

## Betwixt and between being healthy and ill: the stigma experienced by young adults with phenylketonuria

Plata Sofie Diesen<sup>a\*</sup>, Ingrid Wiig<sup>a</sup>, Lisbet Grut<sup>b</sup> and Bengt Frode Kase<sup>a</sup>

<sup>a</sup>Centre for Rare Disorders, Oslo University Hospital, Oslo, Norway; <sup>b</sup>Department of Health Research, Sintef Technology and Society, Oslo, Norway

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Today most adults with phenylketonuria (PKU) have followed a protein restricted diet from early infancy. This makes their disorder an invisible impairment which becomes visible only when food is served. When adhering to medical advice in public, adults with PKU find themselves in a liminal space, betwixt and between being medically ill and socially healthy. The affected adults may pass as being 'normal' if they ignore medical advice and eat ordinary food. By doing this, they risk severe neurological health consequences. Due to the rarity of this illness and limited knowledge regarding the health consequences of dietary intervention in adulthood, implications of adult PKU can be difficult to explain to others. Conscious stigma-handling strategies are therefore required. This article, which is based on qualitative interviews, discusses stigma and management strategies in the lives of eleven adults aged 20–30 with PKU.

**Keywords:** stigma; liminal space; phenylketonuria; rare disorders; lived experience

### Introduction and background

Almost all newborns in the Western world are tested for phenylketonuria (PKU), or Følling's disease, within the first week of life. For most people, a newborn screening test has no consequences. To some individuals, however, testing positive has a high impact.

This article addresses life experiences from 11 young adults who tested positive for PKU as newborns. We wish to elaborate the discussion on how PKU becomes visible to others, and how this requires stigma-handling strategies for managing the condition. We will argue that individuals with successfully treated PKU find themselves in a lifelong liminal space, as they are defined as medically ill, yet appear socially healthy. This article also relates the social challenges faced by Norwegian adults with PKU to the common Norwegian food culture. Few reports on this topic are available, and no Norwegian study has focused on this aspect.

PKU is an inborn metabolic disease which necessitates treatment with a protein restricted diet from early infancy. If left untreated, PKU results in severe, irreversible brain damage. All kinds of meat, fish, eggs, nuts, dairy products, ordinary bread and pasta are essentially eliminated. Potatoes, legumes, rice, cereals and protein containing fruits such as avocado and banana, are monitored and calculated into individually adjusted daily rations. Most vegetables, fruits, berries, sugar, spices, pure fat and special low protein products are

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\*Corresponding author. Email: [sofdie@ous-hf.no](mailto:sofdie@ous-hf.no)

allowed in free amounts. In addition, the diet includes a pharmaceutically produced protein substitute, which provides the remaining biologically necessary protein.

In addition to satisfying biological needs, meals serve to bring people together and strengthen relational ties (Quandt 1999; Kaufmann 2010; Polland 2013). Sharing a meal often involves social intimacy and thus implies strong normative rules. The patterns that establish not only what, but also how, why and under what circumstances we eat, are known as foodways (Edge 2007; Guptill, Copelton, and Lucal 2013). In order to understand a dietary deviance, it is necessary to understand what is considered normal, by describing the majority's foodways. The types of foods we eat can function as status symbols; how and when we prepare and consume them is argued to be part of our identity, similar to the choices we make in clothes and cars (Guptill, Copelton, and Lucal 2013, 18).

Dietary deviance may disqualify the affected individual from full social acceptance. A study of adolescents with coeliac disease described eating special food in public as a way of making an invisible condition visible (Olsson et al. 2009). A study of adults with coeliac disease revealed emotional dilemmas such as shame and fear of being a bother in food-related contexts (Sverker, Hensing, and Hallert 2005), and a study of young adults with type 1 diabetes described perceptions of having a stigmatizing condition which could easily undermine their identities as young and healthy individuals (Balfe et al. 2013). Our assumption was that the PKU diet had similar implications in the affected individuals' lives.

Foodways reflect the cultural norms and expectations of individuals in the dominant group, and violating them may lead to stigmatization. Stigma is defined as 'the situation of the individual who is disqualified from full social acceptance' (Goffman 1963, 9). Stigma theory distinguishes between individuals with obvious stigmas, the discredited, and individuals with hidden disabilities, the discreditable. Discreditable individuals strive to control the information that may disclose their discreditable attribute (Barnes, Mercer, and Shakespeare 1999; Goffman 1963). Whether the negative asset disqualifies the individual from social acceptance may depend upon the nature of the stigma, the perceptions and reactions of the social surroundings, and the degree of acceptance towards the deviation within the stigmatized individual (Goffman 1963). For early treated adults with PKU, their stigma remains hidden until the food is served. When the stigma manifests itself, the individual is in opposition to the non-deviants, further referred to as 'others'. The disclosing situations require stigma-handling strategies, here understood as conscious behaviour to mitigate the situation (Goffman 1963).

Qualitative studies on living with PKU has revealed stigmatization as one possible reason for the varying level of adherence to medical advice and individual decision-making amongst adults with PKU (Frank, Fitzgerald, and Legge 2007; Vegni et al. 2009; Di Ciommo, Forcella, and Cotugno 2012). A key message from one study concluded: 'From the patients' perspective, the main topic in being affected by PKU is not the illness itself nor the diet, but the continuous and difficult balance between being different but socially included and being healthy and alone' (Vegni et al. 2009, 547).

Being in a state betwixt and between having good or bad health has been referred to as a liminal space (Turner 1964; Jackson 2005; Felde 2010). Turner (1964, 49) described the liminal space as a transitional stage between two relatively fixed stages: 'that which is neither this nor that, and yet is both'. The liminal space applies to adults with PKU because they are considered medically ill, yet they appear healthy. Liminality requires conscious and continuous thought and strategy, and for adults with PKU, the situation is connected to food and sharing meals with others.

Research on PKU and other rare disorders has mostly been medical. Interest for social research has however expanded. The rarity of a disease may become a stigmatizing aspect in its own right (Berglund, Mattiasson, and Randers 2010; Grut and Kvam 2013). Also, in a modern society, knowledge is often based on what is visible to the eye, and sight is assumed to be objective (Lingsom 2008). Having an invisible impairment may thus imply that the seriousness of the disease might be doubted by others.

There are approximately 50,000 people worldwide with PKU (ten Hoedt et al. 2011). Approximately three to five newborns are diagnosed yearly in Norway. Newborns in Norway have been tested for PKU since the 1970s. Consequently, the first successfully treated individuals with PKU are still relatively young, and there is little knowledge available on health consequences of dietary discontinuance in adulthood. However, outcome severity from dietary disruption does decrease with age. Without methods to predict who's risking what and when, the standard recommendation is to follow the diet throughout life, known as *diet for life* (Ahring et al. 2009). Low adherence is eminent in adult PKU (Burton and Leviton 2010; MacDonald et al. 2010; Berry et al. 2013), professional follow-up is no longer mandatory, and they tend to define their own degree of protein restriction (Das et al. 2014). Possible symptoms from low adherence in early treated adults are irritation, anxiety, depression, loss of concentration, headaches, unpleasant body and urine odour, osteoporosis and vitamin deficiency (Hoeks, den Heijer, and Janssen 2009; ten Hoedt et al. 2011; Bilder et al. 2013). Mental and emotional problems are hidden disabilities; they may go unnoticed for years, and may result in social difficulties. Some early treated adults with PKU have significant problems in forming interpersonal relationships, gaining autonomy and reaching educational goals, compared to the general population (Simon et al. 2008; Gentile, Ten Hoedt, and Bosch 2010).

### Research method and design

The aim of this study was to develop knowledge regarding everyday life for young adults with PKU. In order to describe the experience of living with PKU, a qualitative method approach was chosen. The interviews and further research process were open, inductive and inspired by grounded theory. The intention was to form hypotheses that emerged from the collected data, rather than defining them in advance (Glaser and Strauss 1967; Glaser 1998). The interviewer encouraged the participants to relate their story, asking *how* instead of *why*, in order to avoid placement of responsibility or guilt (Becker 1998). After interviewing, typical experiences were formulated into themes in an open coding process, and later categorized into core themes, in an axial coding process. A selective coding of the transcripts was performed, to locate quotes and variables to highlight the core themes of the emerging theory.

The data consist of 11 semi-structured, qualitative interviews conducted with 20- to 30-year-old individuals with PKU. The Norwegian PKU population in this age group counted 44 individuals. As the screening programme and treatment is centralized, we could establish that none were excluded. The age group was targeted because it is the first to be expected to follow *diet for life*. Individuals typically gain autonomy in many life areas at this age (Persson et al. 2001; Vegni et al. 2009). Studying, finding a job, meeting new people, falling in love and building a family of one's own are all activities that will require sharing meals with others. These are social contexts in which the dietary treatment is visually manifested, which may lead to stigmatization.

To better understand life with PKU, the Norwegian PKU association's website ([pkuno.org](http://pkuno.org)) and membership magazine (PKU-bladet), as well as the open Norwegian

Facebook interest group (PKU Norge), were used as supplements during the research process. Ethical precautions were taken in order to recognize the topics that gained most attention, without focusing on identifying features. A presentation of the forthcoming study was printed in the membership magazine and on the Facebook 'wall'.

After acceptance from the 'Regional committee for medical and health research ethics' (REK), a postal invitation to participate in the study was distributed to all 44 registered patients. The invitations were sent from the national Centre for Rare Disorders at Oslo University Hospital. The letter of invitation pointed out that the study concentrated on life experiences, and no medical information would be obtained. Thirteen individuals replied to the invitation; however, for various reasons, two of them could not participate. The selection thereby consisted of 11 participants; seven women and four men. All participants had moved out of their parents' home and eight were currently in a relationship. All participants had completed the Norwegian equivalent of high school level. Higher education varied from none to high academic level. They were either employed or registered students. Seven adhered to a classical PKU diet. One was pregnant at the time of the interview and was therefore on a maternal PKU diet in order to avoid damage to the foetus. This dietary regimen is even stricter than the classical one. Three of the participants had ended their dietary treatment after reaching adult age, and were eating ordinary food.

The interviews included topics on the participants' current social life, their childhood, food preferences and risk perception (related to possible effects from non-adherence). Nine interviews were conducted face-to-face in a location chosen by the participant; in the participant's home, at quiet cafés, in confined rooms at the participant's campus site or at the Centre for Rare Disorders. Due to geographical distances, two interviews were conducted via video communication on Skype. Duration ranged from 45 minutes to two hours, depending on what and how much the participants wished to share. All interviews were recorded and transcribed. The study complies with the ethical principles set by the Declaration of Helsinki. Results of the study were presented to a larger group of adults with PKU in January 2012, at a national gathering at Oslo University Hospital, with a positive recognition feedback.

In the article the author has translated quotes from the original language of Norwegian, to English. Focus has been on translating the meaning of the quotes, rather than translating word for word.

### ***Limitations***

The specific age group may be a limitation as other cohorts may have other life experiences, but age-comparison was not in the scope of this study. Also, the participants' educational levels seem higher than expected from other studies. However, the aim of the Norwegian educational system is that all adolescents should finish 13 years of primary and secondary education, and facilitating arrangements are made where needed. The selection may be biased in having included only persons with good communication skills, as accepting an invitation to talk about one's social life and medical condition may require an outgoing personality and good conversation skills. Nevertheless, the selection included fully adherent participants, non-adherent participants and participants who juggled in between. This should secure a sufficient variety in life experiences within the selection.

## **Results**

When eating with close relatives and friends, none of the participants mentioned any stigma-related issues from following the diet. However, after reaching adult age, time spent with the close relatives and childhood friends became less frequent. PKU became an issue with semi-close relationships, such as new friends, friends of friends, working colleagues, fellow students, in-law family and elderly family. This situation has been described by Goffman (1963, 69): 'The area of stigma management, then, might be seen as something that pertains mainly to public life, to contact between strangers or mere acquaintances, to one end of a continuum whose other pole is intimacy'. Therefore, the following focus will be on semi- to non-intimate social situations that involve sharing a meal.

### *Stigmatizing situations*

The PKU bread was a matter of concern for the participants, because of its' bright, white colour. It could, for instance, become an issue during lunch break. In Norway, bread is the main food ingredient in all meals except dinner. One of the participants had experienced the white bread to be an attention-trigger among others:

Now that I've started to work at a new place, people say: 'Wow, you have white bread', right? Because the slices are completely white.

In Norway, public nutrition recommendations are to eat wholemeal bread and, according to cultural foodways, most people choose brown bread. The Norwegian language distinguishes between bread (brød), being brown, grained and considered healthy, and white bread (loff), being light, fine and considered unhealthy. Thus, the all white bread in the PKU diet work as a stigma symbol, defined as 'signs which are especially effective in drawing attention to a debasing identity discrepancy, breaking up what would otherwise be a coherent overall picture, with a consequent reduction in our valuation of the individual' (Goffman 1963, 59).

The PKU bread also becomes a food anomaly, an irregularity that fails to fit with the socially expected. It catches people's attention. Such food anomalies can cause emotional reactions with others, varying from a slightly uncomfortable feeling to fear and disgust (Douglas 1966, 1970). Another participant described how PKU became visible during festive meals:

During a meal with people who eat normal food, you're always the underdog who's eating something different. You'll never become cooler or better than anybody else during a Christmas dinner, when the others are eating everything; meat and gravy and such, and you're sitting there eating cucumbers and sweet peppers.

The situation describes how the PKU diet makes it difficult to gain full social acceptance during semi-intimate festive and traditional meals. Eating food of a different appearance often led to eyebrows being raised and heads being shaken. Such social feedback could be experienced as negative and excluding, especially for young adults who are establishing new networks of friends and colleagues.

One of the main concerns of the participants was making others understand and accept the implications of PKU. The rarity of the disease added to the stigma, as the diet used differs from other, better known medical diets:

It's very strange because it's protein, right? Had it been sugar or gluten or lactose, it would have been more normal, if you know what I mean? But proteins, people have never heard of that. So they don't get it, I believe. It's really good for me to eat pure fat and sugar and stuff like that, and people say: 'What kind of diet is *that*', right? But it's like that, an absolute opposite to other diets.

The above statement points to two reasons why the PKU diet may be difficult to comprehend. First, it is the unfamiliarity, due to its rarity. Disorders like diabetes, coeliac disease and lactose intolerance are well known, and the public has a fair conception of the dietary treatments. However, protein restriction seems odd to most people. Second, as with white bread, pure fat and sugar do not fit in with public nutrition recommendations. Pure fat and sugar are considered unhealthy, and seem incompatible with the common idea of medical diets and dieting.

Persons with PKU obtain most of the protein they need from a protein substitute. However, small amount of food with natural protein is required. Dietitians adjust the daily prescribed amounts of natural protein for each individual. In contrast to people with allergy or intolerance, people with PKU can choose among all food items. This may appear as a dietary inconsistency to others. Knowing the confusion this would create, the dietary adherent participants were concerned with the signals they sent to others by eating small amounts of food containing protein. The following quote describes one participant's dilemma when dining in the university canteen:

If it's chicken, or whatever they have, now and then, it's not really a problem. The problem is the signal you send to others. That I'm eating chicken now, and that makes it ok. That's worse, because then they think I can eat chicken all the time.

The participant knew his choice of food would define his fellow students' perception of his condition in the future. If he had eaten chicken after revealing his condition, nothing noticeable would happen, and this could create a perceived inconsistency between his words and his actions. The seeming inconsistency could lead others to overlook the condition and to take it less seriously. It could also make PKU easy to forget:

It's typical with people that I've already explained it to, what it means to have PKU. After a while, I meet them again, and they ask: 'Do you want this and that', and it's clearly not food for a person with PKU. They don't understand, time goes by, and the next time, they ask if I want something that I can't eat again. It doesn't happen all the time, but it's often like that with peripheral friends.

Having to repeat the information to the same person several times is tedious. Explaining the diet repeatedly was also identified as a source of frustration by adolescents with PKU (Sharman, Mulgrew, and Katsikitis 2013). With invisible and unobtrusive impairments, Lingsom (2008, 4) argued that knowing and forgetting go hand in hand. In this context, appearing to be 'normal' may be more of a challenge than a reward. Having experienced this several times could make revealing the condition less of an option:

It's recently, really, when I've moved away to study, that I'm getting tired of explaining over and over again. I don't feel that my friends here really understand. Because I don't weigh and calculate my food anymore, they are thinking: 'It doesn't matter, it's not important'. It's my friends at home who really know how it is. And now, I almost don't bother to explain it anymore. It's the first time in my life that I've felt that, and it's a little tiresome.

This participant struggled in her adult life, wanting to adhere to dietary advice whilst lacking social support in her new surroundings. Her new friends did not take her condition seriously, and she had started to anticipate being ignored. Among the participants, frustration from having to repeat information had increased in adult life. As children, they had weighed their food on portable kitchen scales and calculated accurately. They had friends and family around them who observed this and recognized the seriousness of PKU. As autonomous adults, they were no longer physically surrounded by the support of people who understood.

### ***Stigmatizing risks***

Receiving medical treatment is expected to prevent illness, and it is commonly presumed that the person knows what she is risking by non-adherence. The participants considered the risks from non-adherence in adult PKU to be vague:

It's the why-questions. They always come. For example, when I've brought another type of milk or something, they ask if its special food, 'those special things that you eat', and they want to know why. It's always the issue about standing out, you try to avoid it. And it's not easy, I'll say. Because with the why-questions, I don't really know what to say, nobody does. And people don't understand that.

The absence of a socially valid explanation to support dietary continuance in adulthood could lead to stigmatization in itself, or add to the stigma. During childhood, risks had been more distinct. For the adult person with PKU, it could still be shameful to reveal the facts about the condition, because it meant revealing an inherent link to mental illness:

I never like it when they ask me what would have happened, had I eaten this and that at a certain age. 'I'd be brain damaged', I say. And I just think it sounds terrible.

The participants had dieted all their lives to preserve their mental functioning, which reinforced the importance of not being associated with mental disability. From their point of view, telling the full story could give a worse impression than necessary. This result is supported by earlier research, describing a stigma hierarchy of disability groups, which place disabilities implying cognitive impairments and psychiatric illnesses amongst the very least accepted (Deal 2003; Westbrook, Legge, and Pennay 1993). These stigmatizing attitudes are not merely societal; they also exist within the stigmatized individual (Goffman 1963). Thus, in the search for a valid explanation for the PKU diet, revealing the link to mental disability became undesirable.

The participants all claimed to be aware of existing medical research. To the participants who had chosen to eat only ordinary food, the uncertain risk outcome had been decisive:

If someone had come to me and said: 'These are the consequences, when you're forty years old, you won't remember anything, because you've poisoned your brain with protein', then I'd restart the dietary treatment, but nobody knows. So I haven't got enough self-discipline to do it on my own.

Adhering to the dietary regime requires a strong will. Changes in treatment advice over time and absence of definite risks from dietary non-adherence in adulthood weighed heavily for those who had chosen to eat only ordinary food. While adherent participants

found it difficult to reason with others, the non-adherent found it difficult to reason with themselves. Reflection on this dilemma was stated in various ways by most of the participants. However, those following dietary advice had decided to trust the specialists:

I don't think I'd risk a lot from ending the diet. Well, I don't know. I probably wouldn't notice anything, but maybe other people would, from my conduct and behaviour and such. But I do think it's risk-free to stick with the diet and trust the people at the hospital when they say you should continue throughout life. I trust the people who know the most.

Some participants had experienced symptoms from eating ordinary food. The symptoms were lowered concentration, fatigue, headaches, impatience, social withdrawal and lack of initiative. However, it was difficult for them to decide if the mood-swings and headaches resulted from eating wrong food, or if they were just mood-swings and headaches. Symptoms were not decisive factors for the participants in regard to continuance of dietary treatment. The decisive factor for continuing the dietary treatment lay in trusting the professionals despite the lack of evidence, because, after all, it was better to be safe than sorry.

### ***Managing stigmatizing situations***

People with social stigmas need strategies in order to manage or pass the possibly stigmatizing situations. Passing here refers to full acceptance in social interaction. With invisible impairments, the individual may choose to conceal the stigma and thus pass as being 'normal', or to disclose it, preferably in an unobtrusive manner (Goffman 1963; Lingsom 2008). Goffman (1963, 95) argued that 'because of the great rewards of being considered normal, almost all persons who are in a position to pass will do so on some occasion'. The participants in this study told of a variation of solutions for concealment and disclosure in managing stigmatizing situations.

The attitude of the participants who were openly adherent to the diet was to be open when people asked questions. They recognized that others could feel uncomfortable asking questions, due to a fear of saying something offensive. To Goffman (1963), such tense situations would oblige the lifelong stigmatized individual to have patience with the unknowing person, because to the unknowing, the situation is new and thus uncomfortable. To the stigmatized individual, however, this rather familiar situation should enable them to act nicely, to educate in a sympathetic manner and overlook untactful comments:

A lot of people wait for a while, and they just glance at your food. After a while, they say: 'I don't really want to ask you, you've probably had questions asked a thousand times before' and I say: 'You know what, it doesn't matter, just go on asking'.

This participant handled the attention she received in a positive manner. She knew by experience that people would notice her food, whether she wanted it or not. However, she also knew that others were probably more uncomfortable asking the questions than she was getting them. Her strategy was to be positive and allow all questions.

With mere acquaintances, the participants were often reluctant to reveal the medical explanation, knowing it could lead to more confusion and more questions. The aspiration to pass as being 'normal' and avoid negative attention was a priority for them.

Frank, Fitzgerald, and Legge (2007) described how their participants emphasized the feelings of others in situations that required denial of food-related intimacy. In the present study, participants recounted similar experiences. Adhering to the PKU diet was said to

be especially difficult when the hosts were unaware of the situation and proud of their cooking. In order to avoid spoiling the good atmosphere, such situations could lead to temporary non-adherence. One participant described the very first time she decided to accept protein-rich food. This happened shortly after she had moved to another city to attend college. At the end of an internship at a hospital, a colleague prepared her a farewell meal, without knowing about her dietary restrictions:

They were so pleased that I had been there, so they cooked. It's the first time in my life that I could not tell them what I had. She said: 'You should help yourself first'. And I sat there, and I hadn't said it yet, and I thought: 'I'll have a small bite'. [...]. It wasn't an option to reveal it, because as I hadn't said it before, I didn't dare to say anything now. She was so proud of her food.

To benefit the social context, she compromised her medical needs. Participants also stated that dining with the parents-in-law was a difficult situation to manage. It was important to impress the parents of the person they loved. One of the participants talked about this problem:

I tried really hard not to be too different. I was served dinner there, normal dinners, we got a lot of meat and they were good cooks, I'll give them that. But I felt that I couldn't be picky about what was on my plate and not eat it. I told them what I had and everything, and they asked polite questions, but I felt like I had to try to fit in the best I could.

Having to reject food made by his mother-in-law could be interpreted as a denial of her offered intimacy, and thus compromise his chances of being fully accepted as a part of the family. In the above situation, the overall goal was for him to fit in. He worried about being perceived as picky, a highly negative trait when seeking acceptance. Thus, he faced the problem of inconsistency; he revealed his condition, yet politely ate the protein-rich food. The strategy resulted in ignoring his diet.

To some of the participants, going public with PKU was not an option. One solution was to eat ordinary food in public and follow the diet at home. This strategy was planned and aimed for passing. To one participant, the idea of eating her own food in company with extended friends made her giggle during the interview, saying:

To eat my own food? No, I don't know. I don't like that; I want to be like everybody else. I don't want anything to be different about me. It's very easy to just eat what the others are eating.

Eating her own food was referred to as not being like everybody else. This participant showed that the stigma can become unbearable. She would rather fit in as a 'normal' person than adhere to medical advice in public. She believed her friends knew about her condition, but she did not think they considered it. And she wanted it to stay that way.

For those who did follow dietary advice in public, a strategic way of teaching others about the importance of their condition and make them remember it was to consistently exaggerate the dietary rules. One participant said:

If you eat a little bit, they remember it the next time, like: 'She ate that; I'll serve it again this time'. It's better to just say: 'I can't eat it'.

Another participant said:

Even though the diet allows it, you can't eat a small slice of the chicken in the canteen. It will only send the wrong signal. And people don't understand that, so you might as well give up explaining it to them.

The solution of not eating food containing natural protein was a way of telling by showing, they placed PKU into a category with clear limitations, evident enough to make people remember.

In meals shared with people they did not expect to meet again, the participants could choose to conceal the real nature of PKU and avoid extra attention. According to Goffman (1963), those who are stigmatized and unable to pass as being 'normal' might sometimes try to pass as having another, less significant stigma. With invisible impairments, such passing can involve telling less than the whole story (Lingsom 2008). In a study on adolescents with PKU, participants used veganism as an alternative explanation for the PKU diet (Sharman, Mulgrew, and Katsikitis 2013). In the present study, the participants gave two alternative explanations, namely having coeliac disease or being vegetarian. It did not remove the stigma; but it removed the parts of the stigma that involved rarity, and it became easier to explain:

If I say I can't eat proteins, I know it starts a conversation, right? So I say I have coeliac disease. I just say I'm allergic to flour. And I feel like I'm not really lying.

Claiming to have coeliac disease would apply to meals with baked products or pasta. The stigma was reduced to something less deviant than PKU. Another stigma-reducing option was to pass as a vegetarian:

Lots of people think I'm a vegetarian, but I say: 'No, I'm really not, I can't eat meat, and vegetarians can actually eat meat.' But sometimes I just don't bother, and I just don't say anything, and I let them believe that I'm a vegetarian.

By eating only vegetables, one could let others believe vegetarianism to be the reason, and easily get away with it. The differences between PKU and vegetarianism, however, were evident to the participants themselves. Firstly, contrary to people with PKU, vegetarians get plenty of natural protein from protein-rich vegetables and legumes. Secondly, and this was most important for the participants, vegetarianism is a choice; PKU is not.

To make the symbol of their stigmatizing condition less evident, the participants could make the food look more similar to ordinary food. Recipes for low protein food that looks and tastes similar to ordinary food is a frequently shared topic on the Norwegian PKU interest group on Facebook. Among the participants, making the bread appear darker than its original, white colour was a priority. Low protein bakery products are usually baked at home, leaving the baker free to experiment. One participant told a narrative about adding gluten-free flour in the dough:

Our bread rolls weren't like everybody else's, to put it like that. They were bright white, and people said: 'What? Are you eating *that*?' because they didn't look very appealing, and it wasn't cool to have a disease that made you stand out. But now, you can add a small amount of dark, gluten-free flour, so that it looks darker. It looks more normal. It has some protein in it, but I just add enough to give it a little colour. Which means that the food is not like: 'Wow, you eat different food'.

Finding a solution was a relief, as it meant being able to eat bread among others without constant stares. Another option mentioned was to add brown food colouring to the dough.

## **Discussion**

Foodways influence the vast majority of social meals. They are culturally determined by repetitive social interactions, which produce and reproduce internalized truths about the right types of food and the right ways to prepare them (Guptill, Copelton, and Lucal 2013; Douglas 1966, 1970). By deviating visibly, PKU food violates the norms of expectations for a meal. However, cultural beliefs do not only exist within those who stigmatize, they are also internalized by the stigmatized individual (Goffman 1963). Having accepted the majority's foodways, the early treated adult with PKU may come to expect and accept stigmatization in various food-related contexts, which may serve as an intrinsic barrier to dietary adherence.

In order to identify socially constructed barriers, which consist of negative attitudes and social exclusion, contributing to disabling the individual (Oliver 1990), the social model of disability has been used. This model essentially blames society for failing to include people who are biologically different. While the medical model defines disability as the outcome of bodily impairment, the social model conceives disability as an outcome of socially processed and constructed categories that discriminate people with impairments (Shakespeare 1996). Even though the social model is debated (Shakespeare 2006; Edwards 2008), we have found the concept of socially created barriers suitable for the purpose of describing our results. For people on medical diets, cultural foodways become social barriers and function as real obstacles. For adults with PKU, acceptance of these barriers may in fact lead to an increased impairment. Thus, without balancing the information they release to others, social and biological health may paradoxically be mutually excluding.

The sociological concept of identity may be approached as a biographical chain of events, and the identity core is argued to lie in the individual's capacity to continue a particular narrative (Giddens 1991, 54). Individuals place their identity in a collective context, and may experience their disability as a highly negative trait in contexts of social interaction. While doing this, they fail to challenge the socially established structures, and aim for assimilation instead (Shakespeare 1996). Previously, the PKU identity was one of severe mental disability. In contrast, the modern PKU identity is defined by normality. Having received early treatment, adults with PKU may feel the need to emphasize their normality. They do this in order to highlight the contrast to what might have been the result, had they not been diagnosed early and treated. By gambling with their future health, they appear to maintain their acquired identity as healthy and 'normal' individuals.

As a positive trait to their identity, adults with PKU may choose to emphasize having defeated the cognitive impairment outcome, by maintaining the challenging and highly stringent dietary regime. However, the combination of being rare, being invisible, and lack of immediate and defined consequences from eating the wrong food, may make it difficult for others to understand the extent of having PKU – and in remembering it on later occasions, as previously illustrated in the results. After revealing something as personal as one's medical condition, being ignored must be seen as a challenge to the impaired individual's need for social support. Individuals need people around them to act supportively in order to uphold motivation for dietary adherence. Thus, by relaxing the diet in some situations, it may make the diet easier and more practical to manage. However, in social interaction, seemingly dietary inconsistencies could make it more

demanding to live with, as the dietary principles become even more difficult to comprehend for others.

According to the participants in this study, the most difficult aspect was the lack of evident risks. Following the tradition of medical sociology, the common expectations of the patient in a society where the science of medicine holds the power of definition, is that the patient is socially expected to follow doctor's orders (Parsons 1951; James and Hockey 2007). However, disease is observed through symptoms (Foucault 1989), and medical reasoning is expected to imply definable and immediate consequences. Early treated adults with PKU may struggle in this no man's land. Explaining and acting understandably when the invisible condition becomes visible may be challenging, as the medical advice can be perceived as a precaution rather than preventive. To manage a medical condition in a liminal space requires releasing and withholding information and adherence in order to be considered both socially and medically healthy. The alternative could be, to be considered both medically and socially ill, a highly undesirable outcome. Increased knowledge, openness and acceptance within the general population would always help to ease the problem, but it would not remove the challenges of living with PKU.

In regards to future research on PKU and other dietary treatable diseases, correlations between diet integration and the majority's foodways should be considered. With PKU, in societies and social situations where people traditionally eat large amounts of protein rich foods and less fruits and vegetables, food anomalies become more visible and level of stigmatization might be higher. Conversely, in societies that accept heterogenic foodways, the level of stigmatization might be lower, and adhering to medical advice might be easier for adults with PKU.

## **Conclusion**

Stigmatization and ignorance are barriers of considerable importance for young and early treated adults with PKU. Our results show that the challenges of living with PKU and following a strict diet seem to have more social than practical implications for young adults. Having to eat different food causes unwanted attention and such situations need to be managed. The PKU diet differs significantly from other, more commonly known diets in its rarity, cause and character. This is difficult to explain to others. Thus, the rarity should in itself be recognized as a stigma. To undergo medical treatment without risking any evident and immediate symptoms, places the individual in a lifelong liminal space, betwixt and between being healthy and ill. With PKU, the liminal space might force the individual to practice a stricter diet than necessary and alter the truth about the condition, in order to make the diet socially acceptable. Choosing to eat only ordinary food, either permanently or temporarily, can be experienced as a relief for individuals who are unwilling to burden the extra attention, despite the possible health consequences. This study may be a step towards providing a wider understanding to the varying level of adherence amongst adults with PKU and other rare, dietary treatable medical conditions.

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