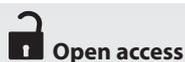


## ORIGINAL ARTICLE

# Walking with McArdle disease: alienation and solidarity in a rare disease journey

**Edmund Jessop**

Specialised services team, NHS England, London, UK



Open access

**Correspondence to**

Edmund Jessop  
Specialised services team  
NHS England  
80 London Road  
SE1 6LH London - UK  
Email [edmund.jessop@nhs.net](mailto:edmund.jessop@nhs.net)

**Competing interests**

None declared

Received: 30 October 2015

Accepted: 4 April 2016

©2015 Jessop. Open access  
article distributed under the  
terms of CC BY-NC-ND



The Rare Diseases and Orphan Drugs Journal has received funding from the European Union Seventh Framework Programme (FP7/2007-2013) under Grant Agreement n. 305690 RARE-Bestpractices project [www.rarebestpractices.eu](http://www.rarebestpractices.eu). Sole responsibility lies with the authors and the European Commission is not responsible for any use that may be made of the information contained therein.

**Abstract**

Narrative reveals what the author wants to share with the reader. First-hand written narratives are a particularly valuable resource for understanding the lived experience (as opposed to the medical facts) of disease. Stacey Reason is a person with McArdle disease, a very rare disorder of muscle, who wrote an account of her 32-day trek across the countryside and mountains of Wales. Her narrative is in the form of a daily diary detailing the events of each day's walking, together with her thoughts and feelings. I present an inductive and semantic analysis of this text. Prominent themes include the alienation of having a rare disease, but also a strong sense of solidarity in the journey, and the normal pleasures and tribulations of walking the British countryside.

**Key words**

Friends, McArdle disease, narrative medicine, social alienation.

**Introduction**

As doctors, we focus on the processes of disease. We try to understand molecular mechanisms to guide our choice of drugs; we investigate disordered anatomy to offer corrective surgery; we investigate abnormal physiology. We know that the patient in front of us is a person, not a disease, but the practice of medicine requires us to tackle problems system by system, one by one. Attending to the hopes, fears and experiences of the whole person in front of us is difficult.

In this situation, the first hand testimony of patients is an invaluable resource for understanding the lived experience of an illness. For chronic illness, this testimony typically takes the form of narrative, an account of events unfolding over a period of time. The narrative may be spoken or written, but full length spoken narratives are rare. Also, analysis of spoken narrative is complicated by the need to observe tone and gesture as well as the words. Full length written narratives provide stable material with form, content and words carefully constructed by the author.

We often speak of patients' journeys, meaning a metaphorical journey from symptom to diagnosis, from one state of health to another, or from even life to death. In this paper I analyse the written narrative of a literal, physical journey – a trek made by four people with McArdle's disease. McArdle is an inherited disorder of muscle metabolism which affects about 1 person in 100,000. It limits the capacity for sustained muscle activity, including walking, but also everyday tasks such as opening a new jam jar.

The narrative seemed to me worthy of analysis because even on a casual reading two themes stood out strongly, and in contrast to my earlier analysis [1] of (metaphorical) patient journeys. Specifically there were strong themes of solidarity, in contrast to the loneliness evident in the accounts by rare disease patients of their journey to diagnosis; there was also a strong emphasis on named individuals in the telling of the journey, in contrast to the almost entirely nameless narratives I analysed in the previous paper [1].

## Method

The material analysed here is text written by Stacey Reason in a book entitled 'One step at a time' [2]. The book is an account of a 32-day trek across Wales in the summer of 2010 by Reason and three others, all of whom have McArdle disease.

The book contains a considerable amount of other material, such as photographs and a daily blog, but I have chosen to focus on Reason's narrative. She writes a short introduction, followed by a page of 450-500 words for each of the 32 days of the journey, and an epilogue. Reason is a Canadian who writes in English.

The target audience for this narrative is stated on the book's cover to be 'patients, families and health professionals alike'. The author tells us that her purpose is as follows:

"... we wanted to share our experience with the world. We wanted to record our successes and challenges, what worked and what didn't. Most of all we wanted to demonstrate that people with McArdle disease can lead a normal, healthy productive life" (*day 9*)

My analytical method was inductive and semantic, following the 6 phase process outlined by Braun and Clarke [3]. An inductive process approaches the material with no preconceptions; a semantic analysis attends to the surface meaning of the narratives without seeking underlying hidden themes. First, I read the text several times to gain a thorough familiarity with the material. I then (phase 2) generated an initial list of ideas about the material. In phase 3, themes started to emerge, though in practice some themes emerged so strongly that this phase overlapped with phase 2 of Braun and Clarke's process. Ideas were grouped and sorted by grouping verbatim quotes, using standard word processing software. A particular text might appear under more than one theme. The themes were reviewed and refined (phase 4), and then (phase 5) named. Finally (phase 6) this report was written.

## Results

Two clusters of themes stand out immediately from Reason's narrative, which we can name as alienation (feeling different from other people) and solidarity (being with other people).

### *Alienation*

Reason opens her narrative thus:

"From early childhood you know there is something wrong. You try, but you cannot keep up with your friends... Every day, with everything you do, you fall behind... everyone. With no explanation at hand, you are left feeling embarrassed, humiliated, utterly defeated"

So from its very beginning, the narrative emphasizes the social consequence of the disease: embarrassment and hu-

miliation. Note also however that this is attributed to the lack of diagnosis – "with no explanation at hand".

Anxiety is also prominent. At first it seems that the anxiety is a performance anxiety-fear of failure.

"...my old familiar preoccupations returned – anxiety, fear and panic" (*day 2*)

"I had been a little nervous about our first big mountain day, who am I kidding – I was petrified" (*day 3*)

"My first thought is uh oh! How will I ever manage to keep up?" (*day 13*)

"What I had to work on though, was the pervasive feelings of inadequacy" (*day 26*)

"Having lived thirty six years without a diagnosis, I was perpetually embarrassed with the ambiguity of my seemingly poor level of fitness" (*day 1*)

No suggestion is made in the narrative that the embarrassment and humiliation come from the comments or teasing of other people: these are internal states of mind. And the telling of the story demonstrates how these problems become less as the walk proceeds: at day 18 we read:

"I no longer was afraid or embarrassed" (*day 18*)

"Perhaps this was part of my journey, to not only accept my diagnosis of McArdle disease, but a more global affirmation of self" (*day 7*)

There are frequent references in the narrative to the sense of achievement, which is situated within the bounds of what is possible for people with McArdle disease:

"I had learned my limits, and now was not the time to aggressively challenge them" (*day 12*)

"...I can do anything I set my mind to. McArdle disease is, and always will be part of my life; but it is not my life" (*day 18*)

"We were walking for everyone else with McArdle's. Our struggles and our successes were theirs too... We wanted to reach out to more people, to empower them in the same way we had empowered one another" (*day 22*)

"So today I happily trod up and down the hills of Wales with my newfound developing courage and acceptance of this rare metabolic disease. I hope I can hang onto this feeling forever. Could you?" (*day 18*)

The phrase "Could you?" in the last quote addresses the reader directly, engaging him or her in the author's story. This is the only place in the narrative where this happens, so the reference to courage and acceptance is especially important.

### *Solidarity*

The theme of alienation is matched by an equally strong theme of solidarity. It thus offers a counterpoint to the separation and alienation of rare disease.

Solidarity may consist in the immediate, physical presence of other people; or it may be the wider solidarity of our common experiences as ordinary people, human beings. Reason's narrative exhibits both types of solidarity.

Throughout the journey there are companions and helpers. In a previous analysis of rare disease narratives [1], I pointed out how rarely the actors were named. But in Reason's journey almost everyone she meets is identified by name. And in being named they are also honoured. For nowhere in this narrative do we find strangers who distance themselves from the McArdle walkers, none who taunt or tease.

"The group maintained contact with one another – forever connected" (*intro*)

"our kind hearted support driver" – this phrase appears in the second paragraph of the whole journey (*day 1*)

"Heather... fed us, cleaned up after us and nurtured us" (*day 8*)

"Meri shows us a different way of being in the world; of living in the moment, and being one with the world around her. I feel truly blessed to have met such a wonderful, creative, strong woman" (*day 15*)

"Dan, Andy and I were thoroughly enjoying the hospitality of the Wakelin siblings"

"The day culminated with a lovely dinner – all thirteen of us, together celebrating" (*day 26*)

Part of solidarity is to do what other people do – to be normal:

"We talked, we laughed, we ate – all pretty normal stuff" (*day 1*)

"...feeling normal is not familiar to us – so it felt great!" (*day 27*)

This solidarity of "feeling normal" and doing "pretty normal stuff" touches on our common experiences as ordinary people. Also normal, in travelogues from the British countryside, are accounts of getting lost, getting wet, and enjoying natural beauty. This conveys a sense of shared experience: enjoying the common feelings of trekking in the countryside – beauty, good weather, bad weather, getting lost and so on. So the narrative is full of the normal experiences, good and not so good, of walking in the countryside. The narrative is not wholly determined by illness.

Beauty is a prominent theme:

"The views... were breath-taking" (*day 11*)

"...the mountains and their magnificent proportions" (*day 6*)

"For now, I will enjoy the beauty that surrounds me..." (*day 8*)

"The west-east route... can only be described as breath-takingly majestic and gracefully alluring" (*day 19*)

So also is the weather:

"the weather was dismal... the rain fell heavy, and the wind carried it sideways – there was no escaping it" (*day 3*)

"The weather had turned for the worse – the winds were strong, the air was cold and I was certain it was snowing – okay, maybe they were just really cold raindrops" (*day 5*)

"the wet weather was relentless..." (*day 8*)

"The sun was shining and our mood was relaxed. It really was a perfect day" (*day 11*)

And getting lost:

"The map was telling us to go in one direction, the trail – another" (*day 14*)

"Lunch was followed by a second wrong turn" (*day 14*)

"But seriously, how many times can you get lost in the rain?" (*day 17*)

## Discussion

Reason has provided first hand testimony of her experience over 32 days of walking in Wales. She comments on the daily events and reflects on present and past. Analysis of her narrative shows that themes of alienation and solidarity are prominent.

Alienation is the sense of being separate or different. Any illness makes us different from our fellow human beings, marks us out as alien. Sometimes this separation is temporary, as when a bout of sickness leaves us unable to share the family meal. But for genetic diseases, the separation is lifelong. It is from birth, and it may be profound:

"I thought I was the only one in the world, you see. In fact for a long time I used to comfort myself with the thought that actually I was an alien child. And I spent most of my childhood waiting for the mother ship to come back for me, to take me away" [4]

For common conditions such as diabetes and asthma, alienation may be overcome by finding fellow sufferers. But people with rare disorders typically know of no-one who shares their particular pattern of disability and dysfunction. For those with McArdle disease, which is primarily a limitation of motion, difference and alienation may be more obvious in the energetic days of youth than later on when a sedate pace is the norm. Spoiled identity [5] becomes obvious only when there is a requirement to keep up, to match the pace of people who do not have McArdle disease – as Reason comments "You try, but you cannot keep up with your friends". Even in adulthood, however, the sense of difference remains, with frequent reference in the narrative to fear of failure, embarrassment and anxiety.

Solidarity comes from the presence of fellow sufferers, people who share our particular same pattern of ability and disability, engagement and limitation. The comfort may be emotional, or it may be practical – tips and hints on how to manage particular actions and situations. In one sense, the whole journey narrated by Reason is a sharing of advice on how to manage a long distance walk across hill country. The walking group also clearly developed emotional solidarity – “forever connected” as Reason puts it.

The wider solidarity of common humanity can be illustrated by comparing Reason’s narrative with two other accounts of long distance walks.

In the very same month as Reason was walking 338 kilometres across Wales, Simon Armitage, a professional poet, walked the 429 kilometres of the Pennine Way, a long distance footpath in England. Like Reason, Armitage also told the story of his journey a day at a time [6]. He mentions no specific locomotor disability, though he doubted his physical fitness: ‘Physically, I’d assumed I wasn’t up to it’ (p 278). His text shows several points of solidarity with Reason in describing the common experiences of hillwalkers in Britain.

Armitage walked solo but an ever changing cast of companions accompanied him on various sections of the walk. Like Reason, he names them and celebrates them: “...my sincere thanks to the following, for their kindness and encouragement, for their time and energy, but mostly for their company...” (p 282). Armitage also shares with Reason the familiar anxieties of a hillwalker about getting lost in bad weather: “Fear is what is stopping me. I don’t mind the wet and the cold, but I don’t want to get lost” (pp 268-9).

In the telling of a narrative, the author selects what to include and what to leave out. A contrast to Reason and Armitage is provided by Ranulph Fiennes. Fiennes is a professional explorer whose book *Mind over Matter* [7] describes an attempt to walk unsupported across Antarctica. His companion in the 65-day journey was Mike Stroud, a doctor. Fiennes is an interesting character whose strengths and weaknesses have been revealed in his many books. His narrative is full of dry wit but is devoid of enjoyment; it is all about the struggle to survive and to keep moving. There is no mention of beauty in the ice fields. Although Stroud has saved Fiennes’ life after a fall through the ice on a previous expedition (and in this journey Fiennes later does likewise for Stroud), there is little discussion of

the emotional warmth that comes from companionship; references to his travelling companion by Fiennes focus almost entirely on whether Stroud will have the physical strength to complete the journey. Here is a typical passage:

“The next day, after two hours of steep yard-by-yard progress, Mike was forced by another diarrhoea attack to stop... A few minutes later he told me he could not continue. He must rest. I was furious. We erected the tent and made tea. After Mike had rested for an hour or two I told him we must get going. He was angry and said I was boorish and graceless; in short a prick of the first order” (p 139)

Antarctica is a harsh environment where life hangs by a thread; so perhaps the lack of enjoyment is dictated by the life-threatening dangers of the trek. But note that for people with McArdle’s the same can be true of the north Wales hills: Reason’s book is dedicated to a young woman, Jessica Binder, who died from McArdle’s disease at the age of 31, and Reason herself was hospitalized on day 5 of her trek. Like Fiennes in Antarctica, Reason must make her walk step by step. So her frequent references to beauty and enjoyment, solidarity and kindness, rather than struggle and hardship, are worthy of note. Reason has an illness, but she is not defined by, or not fully defined by it: she still enjoys the normal experiences of common humanity.

MacIntyre [8] has drawn attention to our status as “dependent rational animals”, all in our own way vulnerable and all in our own way dependent on help and support from our fellow human beings. Reason’s narrative illustrates well this thesis.

## References

1. Jessop E. The quest for diagnosis – a narrative analysis of patient journeys. *Rare dis orphan drugs* 2014;1:109-115.
2. Reason SL. One step at a time. *Walking with McArdle disease*. Droxford: Association for Glycogen Storage Disease (UK) 2013.
3. Braun V, Clarke V. Using thematic analysis in psychology. *Qual Res Psychol* 2006;3:77-101.
4. Cornwall S. Telling stories about intersex and Christianity: saying too much or not saying enough? *Theology* 2014;117(1):17-33.
5. Goffman E. *Stigma: notes on the management of spoiled identity*. Englewood Cliffs NJ: Prentice Hall 1963.
6. Armitage S. *Walking home*. London: Faber and Faber 2013.
7. Fiennes R. *Mind over matter*. London: Sinclair-Stevenson 1993.
8. MacIntyre A. *Dependent rational animals*. London: Duckworth 2009