Assessing the role of Materials and Technology among the families of children and young people with Metabolic Rare Diseases

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Rare Diseases 360
Collaborative Strategies to leave no-one behind
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Abstract

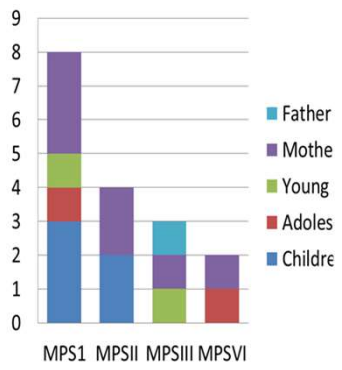
It has been reported that significant numbers of children with rare diseases, life-limiting conditions, and other chronic illness are living longer due advances in medical care and technology. Therefore, it is important to understand how technology and materials (things) subsequently affect the families experiences of life with a rare disease where their world is now transformed by materials and things (for example hoist, wheel chair, hospital appointment letter, feeding pump, insulin pump) and social media like face book, twitter etc.

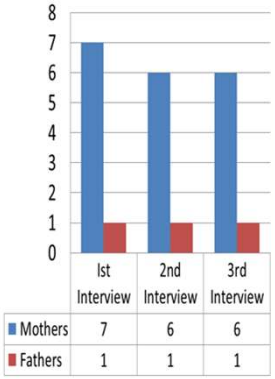
This poster will provide an overview on how materials and technology emerged and played an active role in the everyday life in the context of families of children, adolescents and young adults with MPS I syndromes (Hurler syndrome, Scheie syndrome), MPS II (Hunter syndrome), MPS III (Sanfilipo syndrome) and MPSVI (Maroteaux-Lamy syndrome).

Mucopolysaccharidosis (MPS)

- MPS is a group of rare inherited metabolic disorders.
- Category 3 of life limiting conditions.
- The highest world recorded incidence of MPS-type 1(Hurler Syndrome) in Ireland.
- Bring the living experiences of parents through their own voices, and in doing so make their lives more understandable to the wider audience.

Sample Selection





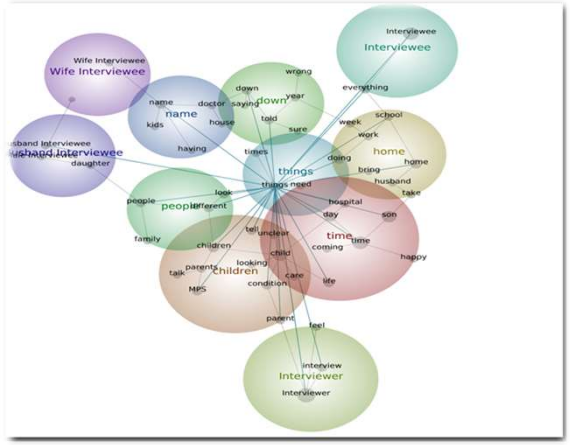
	Ist Interview	2nd Interview	3rd Interview
Mothers	7	6	6
Fathers	1	1	1

Data Analysis

An automated content analysis was carried out using Leximancer computer-aided text analysis program (Smith and Humphreys 2006), to identify the main concepts embedded in "Things" from the 19 interview transcripts.

Van Manen's (2007) six-stage hermeneutics phenomenological research method is used as a framework to guide through the process of research, providing a structure by which to explore the meaning of the lived experiences of parents of a child, adolescent or young adult with MPS

Content Analysis



Selected Concept: **things**

Related Name-Like	Count	Likelihood
MPS	16	09%
Wife Interviewee	13	05%
Interviewee	40	05%
Female Interviewee	4	04%
Husband Interviewee	10	04%
Interviewer	32	03%
Male Interviewee	2	02%

Related Word-Like	Count	Likelihood
bring	17	15%
take	18	12%
times	11	12%
house	7	12%
doing	22	11%
kids	12	11%
different	22	11%
life	22	10%
work	14	10%
child	37	09%
told	17	09%
everything	20	09%
parent	13	09%
look	13	09%
coming	10	08%
down	18	08%
interview	14	08%
parents	11	08%
care	9	08%
talk	12	08%
saying	8	08%

Things in their day to day life

Hoists

"The hoists are great, but it still involves a bit of lifting and what have you. But that is where we are at the moment, she is just physically challenging more so, and it requires teamwork. Even going on a trip in the car, it is like packing to go on a holiday sometimes with nappies, wipes, feed, emergency stuff and then getting her into the wheelchair van, up the ramp, securing her. It is just ... When you are going out for a day, you make the most of it." (FA04, pp. 5) (F4 1 st Interview)

Bilevel Positive Airway Pressure (BiPAP)

"...Shock because it is getting worse, they are after finding another problem with it. We just thought it was a routine sleep study; then you find that he is gone on the BiPAP now for the rest of his life. So that is another new thing for us. It was a bit of a shock, but she said it was better to go on it now instead of waiting. You'd be waiting another year to go on the machine and stuff so start if off now before it gets any worse before it does any more damage." (FC03, pp. 23)' (F3, 3rd Interview)

Telephone

"Over the phone, it was horrendous for the reason being I had no support around me, I was at home, the children were at school or crèche or wherever they were, and the name xxx was way beyond my comprehension of a word I had ever heard of before. It sounded big, and it sounded serious, and it was just an awful way to be told. Automatically I said, 'I have to get my husband to ring you.' Because I couldn't take it in, even though I didn't know what it meant, I knew it didn't mean something good." (FA01, pp. 6) (F1, 1st Interview)

Social Media and Internet

"But now if you put in the word, Sanfilippo, hundreds and thousands of things come up. There are images of pictures; actually, our daughter is even on it as an image of a Sanfilippo person, so you can see straight away what they look like … I think the internet has changed the whole world, but by pressing the button, you can be in touch with families now and online support." (FC04, pp. 12-13) (F4, 3rd Interview)

Conclusions

This study recommends improving the quality and quantity of information available to parents of children with rare diseases, and it is crucial to clarify whose role it is to provide the required information. Also, the need for understanding how material and things can gain power in their own right in the healthcare systems and how we can promote the tactful sensitivities in healthcare practices by listening to parents and children's experience and, thereby improving the quality of healthcare delivery from service users' perspectives.

Acknowledgement

The families who so generously shared their stories, experiences, and insights with this study, without their input this thesis would not have been possible.Should you have any queries or require any further information please do not hesitate to contact us:
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